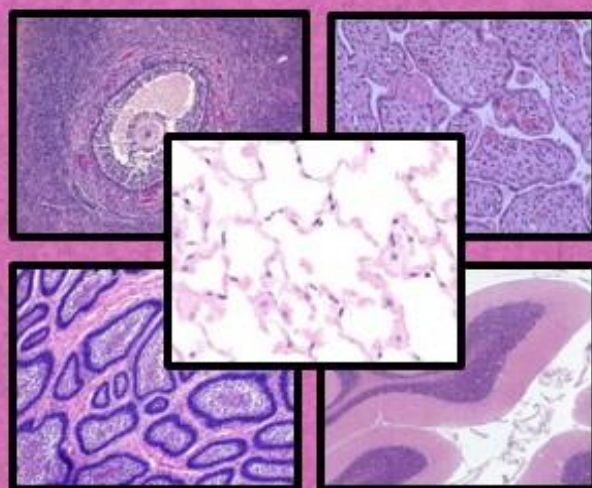




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**Annual Academic Sessions 2023**



**Updates on**  
**Lung, CNS, Gynaecological, Oral and Placental Pathology**

**20<sup>th</sup> to 21<sup>st</sup> October 2023**

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**MESSAGE FROM THE PRESIDENT - DR. SRIYANI NANAYAKKARA**  
**THE COLLEGE OF PATHOLOGISTS OF SRI LANKA**



As the president of the College of Pathologists of Sri Lanka, 2023, I am honoured and privileged to share this message with all of you on behalf of the members of the council. We are having a long-awaited physical Annual academic session this year after two years of the virtual platform, at a time of economic slowdown and with the decline of the COVID-19 pandemic.

My deep appreciation is extended to Dr Jayasundara Bandara Project Director, Primary Health Care System strengthening Project (World Bank) and former Director General of Health Services for accepting my invitation to be the Chief Guest at short notice and the Guest of Honour at the Inauguration of the Sessions Dr Sanjiv Manek. I am extremely grateful to all the resource persons, overseas resource persons, Dr Sanjiv Manek, Prof Geeta Chacko, Prof Deepali Jain and Prof Jane Dahlstrom. Local resource persons Major General Dr Sanjeeva Munasinghe, Dr Sunil Perera, Dr N Jeyakumaran, Prof. Vajira Dissanayake and Brigadier Dr Geethika Jayaweera for sharing their knowledge and experience.

I wish to extend my gratitude to our Platinum, Gold, and Silver sponsors and all the other sponsors for their financial support for this conference at a time of economic slowdown. Your financial support was invaluable to us at this difficult time, and I wish all the sponsors the very best for the future.

I extend my deep appreciation for the commitment made by my two secretaries of the college Dr Avanthi Rajapakse and Dr Charisma Shahi Fernando to make this event a success. I would like to extend my sincere thanks to Council members, IT personnel, College administrative assistant Ms Asuntha Thamel and all who contributed to make this event a success.

Finally, I welcome all participants from near and far who will join us. We are sure you will greatly benefit from the wide variety of topics that will be discussed. I believe Annual Academic Sessions 2023 will be a platform for all the participants and the resource persons to share their experience and update their knowledge which will further upgrade the current practice of Histopathology in the country.

I wish the Annual Academic Sessions 2023 all success.

**MESSAGE FROM JOINT SECRETARIES (CONFERENCE COORDINATORS)**

**DR. AVANTHI RAJAPAKSE**

**DR. CHARISMA SHAHI FERNANDO**



At the outset, we are indeed greatly honored, humbled and extremely privileged to be able to share this message of felicitation, as secretaries cum coordinators of this year's Annual Academic Sessions.

As we have come out of the pandemic but immersed in the current economic crisis, we have tried our utmost not to let the current situation impede us in any way in our quest for knowledge and further learning.

From its very inception in 1975, The College of Pathologists of Sri Lanka has played a significant role in the continuous medical education of its members and other medical professionals. This year has been no exception and this sessions once again will reiterate the absolute dedication and commitment of the college.

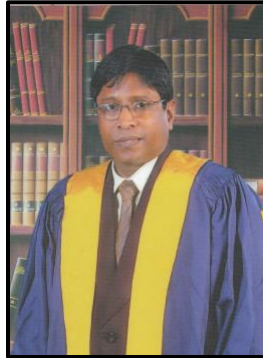
This is our first year hosting a physical conference after three years of virtual sessions, through which we bring to you all, a very esteemed and learned group of resource persons from Australia, UK and India, including our very own home bred speakers from the neurospeciality who will be imparting their knowledge on updates on Lung, CNS, Gynaecological, Oral and Placental Pathology.

The objective of this year's program is to achieve high standards and get updated in the field of Lung, CNS, Gynaecological, Oral and Placental Pathology, through sharing of knowledge, expertise and special inputs from other clinical fields including oncology, neurosurgery, radiology and molecular genetics.

Finally, we take this opportunity to thank our dynamic President, other members of the organizing committee and the council of the College of Pathologists of Sri Lanka, for the immense faith entrusted on us and the endless co-operation extended to us in true team spirit, to ensure the success of the session. We also extend our heartfelt gratitude to all the sponsors for their generous contributions. We sincerely hope that this will be a memorable event, meeting the expectations and high standards of all the participants.



**MESSAGE FROM CHIEF GUEST**  
**DR. J.M.W. JAYASUNDARA BANDARA**



It is with great pleasure I am placing this message as a medical administrator because of the fact that I was the first Director Laboratory services in the Ministry of Health. I am happy that recognition given to the laboratory sector today is at a very high level compared to what it was a 30n years ago. Every one has to congratulate the college of pathologists for the untiring effects it has taken during the past few decades to uplift the potential of their members and in turn raising the professional, intellectual and influential capacity of pathologists in the country with regard to the quality of tertiary health care services

Pathology, by it's name, the study of suffering has been a very important branch of Medicine from ancient days. In the past, the pathologist was basically considered to be the decisive character to give the diagnosis of a disease as he was called upon to render the final diagnosis most of the time after the patient is dead specially when the cause of death was unknown and when a specific treatment or management did not work resulting in death. In short the pathologist's main job was to submit the final verdict after a post mortem examination.

However, due to improvement in clinical diagnosis linked with advancements that have been made in the field of imaging and other disciplines, requirement of the pathologists for planning, monitoring and evaluation of a course of treatment has become important rather than post mortem wordlists.

I sincerely hope the pathologists in the future will search for the universal knowledge and improve the discipline further at the cellular and nuclear level so that pathologist will be a predictor to help clinicians to de-route the pathological process to prevent a disease or complications. I wish this academic sessions a great success!

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Dr Sanjiv Manek  
Consultant Gynaecological Pathologist  
Department of Cellular Pathology  
John Radcliffe Hospital  
Oxford  
United Kingdom.



Professor Geeta Chacko  
Head of Department of Pathology  
Christian Medical College  
Vellore  
India



Professor Deepali Jain  
Department of Pathology  
All India Institute of Medical Sciences  
New Delhi  
India



Dr Sunil Perera  
Consultant Neurosurgeon  
Asiri Central Hospital  
Sri Lanka



Major General  
Dr Sanjewa Munasinghe  
Consultant Radiologist  
Army Hospital  
Sri Lanka



Dr N Jeyakumaran  
Consultant Oncologist  
Apeksha Hospital  
Sri Lanka



Brigadier Dr Geethika Jayaweera  
Consultant Histopathologist  
Army Hospital  
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Professor Vajira H.W. Dissanayake  
Senior Professor  
Department of Anatomy, Genetics, and Biomedical Informatics  
Dean of the Faculty of Medicine, University of Colombo  
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Professor Jane Dahlstrom OAM  
Acting Head, Canberra Clinical School,  
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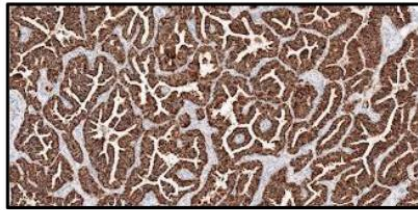
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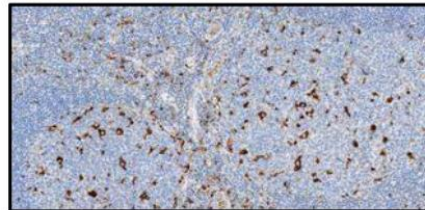
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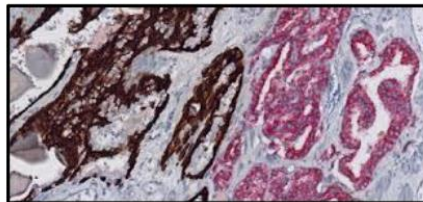
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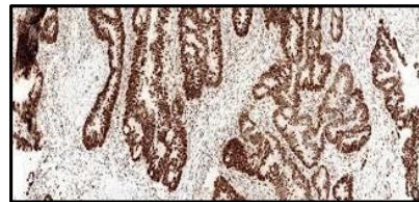
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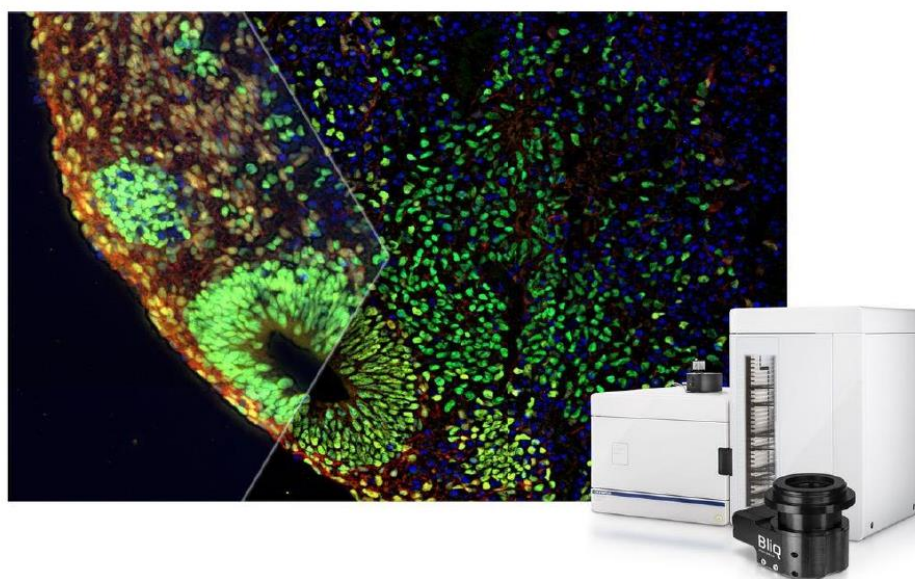
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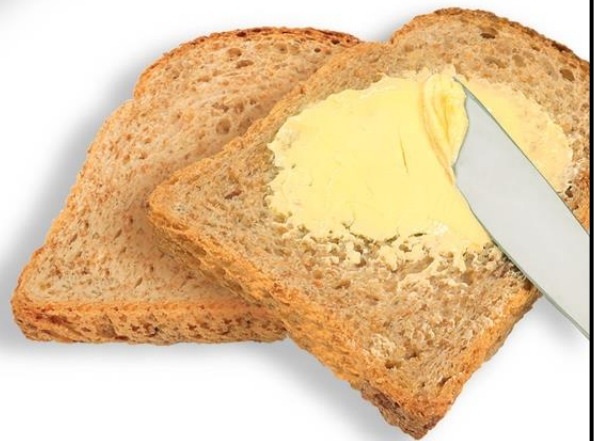
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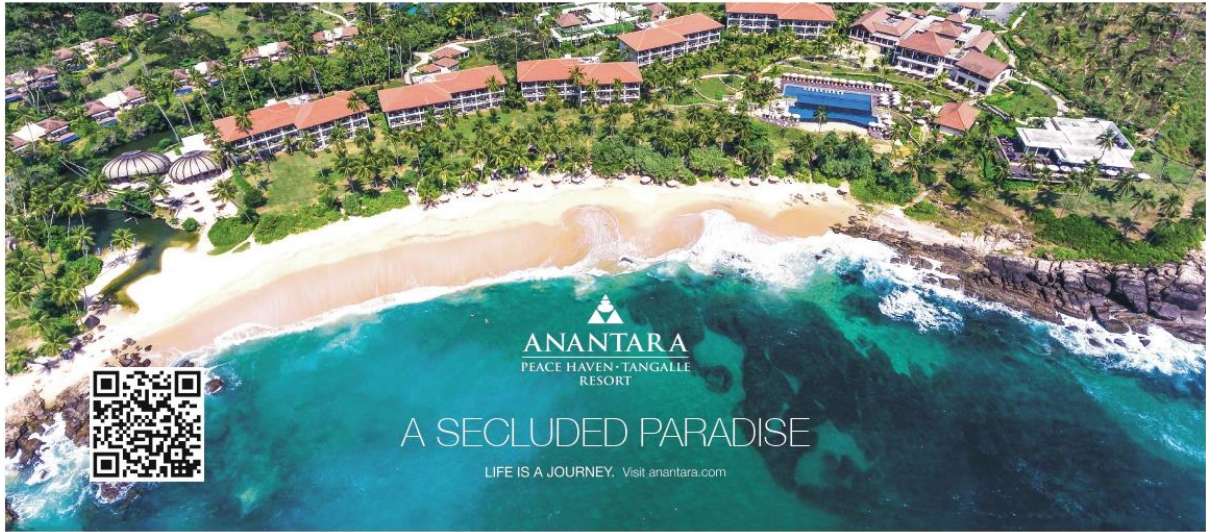
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**PROGRAMME – PRE CONGRESS SESSION**

19 <sup>th</sup> October 2023	
TIME	
7:30 am - 8:00 am	Registration
<b>CHAIRPERSONS : Professor Priyani Amarathunga Dr Sriyani Nanayakkara</b>	
8:00 am - 8:45 am	Approach to Pigmented Lesions in the Oral Cavity <i>Professor Jane Dahlstrom OAM</i>
8:45 am - 9:30 am	Human Papilloma Virus (HPV) infection and head and neck tumours <i>Professor Jane Dahlstrom OAM</i>
9:30 am - 10:30 am	Oral Presentations
<b>10:30 am - 11:00 am</b>	<b>TEA</b>
11:00 am - 12:30 pm	Slide Seminar
12:30 am - 1:00 pm	Measuring your impact: Impact factor, H-index and citation-based indicators <i>Professor Jane Dahlstrom OAM</i>

**PROGRAMME AAS 2023**

<b>DAY 1</b> <b>20<sup>th</sup> October 2023</b>		
7:30 am - 8:00 am	Registration and introduction to the programme	
8:00 am - 9:00 am	Advances in endometrial pathology	Dr Sanjiv Manek
9:00 am - 10:00 am	Immunohistochemistry and molecular studies in gynaecological pathology	Dr Sanjiv Manek
<b>10:00 am - 10:30 am</b>	<b>TEA</b>	
10:30 am - 11:30 am	New everyday entities in gynaecological pathology	Dr Sanjiv Manek
11:30 am - 12:30 pm	Diagnostic approach to non-neoplastic lung disease	Professor Deepali Jain
12:30 pm - 1:15 pm	Diabetes mellitus, obesity, and the placenta: Defender or offender	Professor Jane Dahlstrom OAM
<b>1.15 pm - 2.00 pm</b>	<b>LUNCH</b>	
2.00 pm - 3.00 pm	EBUS TBNA / Liquid biopsy	Professor Deepali Jain
3.00 pm - 4.00 pm	Interpretation of small biopsies and cytology of lung WHO recommendations	Professor Deepali Jain
4.00 pm - 5.00 pm	Slide seminar – Case based discussion	Professor Deepali Jain
<b>5.00 pm - 5.15 pm</b>	<b>TEA</b>	

<b>DAY 2</b>		
<b>21<sup>st</sup> October 2023</b>		
7.30 am – 8.00 am	Registration	
8.00 am - 8.45 am	WHO classification of CNS tumours in the Asian context	Professor Geeta Chacko
8.45 am - 9.45 am	Case based approach using the 5 <sup>th</sup> WHO classification of CNS tumours	Professor Geeta Chacko
9.45 am – 10.30 am	Smears in the intraoperative diagnosis of CNS lesions	Professor Geeta Chacko
<b>10.30 am -11:00 am</b>	<b>TEA</b>	
11.00 am - 12.30 pm	Slide seminar	Dr Sanjiv Manek
12:30 pm – 1.00 pm	Lessons from litigations in gynaecological pathology	Dr Sanjiv Manek
<b>1.00 pm – 2.00 pm</b>	<b>LUNCH</b>	
2.00 pm - 3.00 pm	Case based approach to dural based neoplasms	Professor Geeta Chacko
3.00 pm - 4.00 pm	Case based approach paediatric type diffuse gliomas	Professor Geeta Chacko
4.00 pm – 5.00 pm	CNS symposium	Dr Sunil Perera Major General Dr. Sanjewa Munasinghe Dr N. Jeyakumaran Brigadier Dr. Geethika Jayaweera Professor Vajira Dissanayake
5.00 pm – 5.30 pm	Closing remarks	
<b>5.30 pm</b>	<b>TEA</b>	



**ABSTRACTS  
OF  
RESEARCH PAPERS**

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RP 07	Audit of documentation of quality control parameters in histopathology	<u>E. E. Harold</u> C. S. P. Sosai
RP 08	Second opinion in diagnosis of cancers: experience of a cancer referral centre	<u>B. Y. Hettiarachchi</u> C. M. Baranasuriya K. K. S. Lakshika H. D. Wijesinghe M. V. C. de Silva
RP 09	Undergraduate student perception of an online course for clinical clerkship in histopathology	<u>D. R. Jayasinghe</u> K. A. G. De Abrew M. D. S. Lokuhetty M. V. C. de Silva

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RP 19	Serum concentration of soluble apoptosis-associated molecules in coeliac disease	<u>K. K. Prasad</u> A.K. Sharma U. Debi P.K. Gupta S.K. Sinha

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RP 22	Prevalence of sarcomatoid morphology in different types of renal cell carcinoma and its association with adverse prognostic factors	<u>P. D. K. Thiranagama</u> A. Abeygunasekera C. S. P. Sosai
RP 23	Testing for IDH mutation in astrocytoma by immunohistochemistry: an analysis of potential misdiagnosis in the Sri –Lankan setting	<u>I. L. Wickramanayake</u> C.M. Baranasuriya S. Abeygunawardhane B.Y. Hettiarachchi K.K.S. Lakshika H.D. Wijesinghe M.V.C. de Silva



**RP 01**

**Is there an association between papillary thyroid carcinoma and chronic autoimmune thyroiditis: a retrospective study**

N.G.M.M. Amarasinghe<sup>1\*</sup>, S.J.de S. Hewavisenthi<sup>2</sup>, S.M. Fernandopulle<sup>1</sup>

<sup>1</sup>North Colombo Teaching Hospital, Ragama, Sri Lanka

<sup>2</sup>Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka

malikaamarasinghe@yahoo.com

**Introduction and objectives:** The link between papillary thyroid carcinoma (PTC) and chronic autoimmune thyroiditis (AIT) was first described by Dailey et al in 1955, but the association between these two diseases is still controversial. The objective of this study was to evaluate the association between PTC and chronic AIT and to compare this with the association between benign thyroid disease and AIT.

**Methodology:** The data of 1014 patients who had undergone thyroidectomy from 2017 to 2021 at North Colombo Teaching Hospital (NCTH) was retrospectively analyzed. Chronic AIT was diagnosed based on the presence of diffuse lymphoid aggregates with at least focal germinal center formation, follicular destruction and Hurthle cell change. Patients who were found to have malignancies other than PTC were not included in the study. The patients were divided into two groups, those having PTC and those having benign thyroid disease. The odds ratio was used in the statistical analysis to determine the difference between these two groups.

**Results:** The first group consisted of 282 patients who were diagnosed with PTC. Among these 282 patients, 106 patients were found to have AIT in the background thyroid tissue (37.59%). In the second group of 832 patients who did not have PTC (i.e.: benign thyroid disease), 106 patients were found to have AIT (12.74%). The odds ratio was 4.125 (95% confidence interval 3.01-5.65).

**Conclusion:** 37.59 % of PTC demonstrated AIT in the background thyroid tissue. The odds of PTC in the presence of chronic AITs was four times that of benign thyroid disease and the difference was statistically significant.

**Keywords-** autoimmune thyroiditis, papillary thyroid carcinoma, association

**RP 02**

**The association between the biological characteristics of papillary thyroid carcinoma and the severity of background inflammation: a histological analysis of 282 patients**

N.G.M.M. Amarasinghe<sup>1\*</sup>, S. J. de S. Hewavisenthi<sup>2</sup>, S.M. Fernandopulle<sup>1</sup>

<sup>1</sup>North Colombo Teaching Hospital, Ragama, Sri Lanka

<sup>2</sup>Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka

[malikaamarasinghe@yahoo.com](mailto:malikaamarasinghe@yahoo.com)

**Introduction and objectives:** Chronic inflammation is associated with the development and progression of some cancers. Studies carried out to determine the relationship between background inflammation and papillary thyroid carcinoma (PTC), have shown varying results. The objective was to describe and assess the degree of background inflammation in PTC and assess its relationship with some tumour characteristics.

**Methodology:** Haematoxylin and eosin-stained slides and reports of 282 patients diagnosed with PTC at North Colombo Teaching Hospital from 2017 to 2021 were re-examined. The degree of background inflammation was categorized into four semi-quantitative grades; 0 - no inflammation, 1- minimal inflammation with rare formation of lymphoid follicles, 2-scattered follicle formation with minor Hurthle cell changes and focal follicular destruction, 3-diffuse thyroiditis with marked follicular destruction and Hurthle cell change. The association between the grade of inflammation and the tumour characteristics were assessed using Fisher's exact test.

**Results:** Among the 282 cases of PTC, 2.48%, 24.11%, 35.82% and 37.59% of cases showed Grade 0, 1, 2, and 3 background inflammation respectively. The degree of inflammation showed no significant differences in association with sex ( $p=0.516$ ), mean age of patients ( $p=0.923$ ), multifocality versus unifocality of PTC ( $p=0.711$ ), tumour size ( $p=0.149$ ), extra thyroid extension ( $p=0.840$ ), lymphovascular invasion ( $p=0.840$ ) or nodal metastases (0.426).

**Conclusion:** The degree of inflammation in the background thyroid tissue in patients with PTC showed no relationship with the biological features of the tumour assessed such as tumour size, focality, extrathyroidal extension, lymphovascular invasion and nodal metastases.

**Keywords:** papillary thyroid carcinoma, chronic inflammation, biological characteristics

**RP 03**

**Comparison of dynamic perfusion CT parameters of biopsy proven benign and malignant lung lesions at a tertiary care centre**

U. Debi\*, M. Garrg, N. Prabhakar, N. Singh, A. Bal

*Postgraduate Institute of Medical Science, Chandigarh, India*

Debi\_uma@yahoo.co.in

**Introduction and objectives:** At present non-contrast CT(NCCT) is the initial modality of choice for diagnosis in patients with suspicious lung lesions and requires confirmation by biopsy. It is invasive and requires high expertise. Dynamic perfusion CT (dVPCT) may be used as a non-invasive diagnostic modality to differentiate benign lesions from malignant lesions. The objective of this study was to compare various dVPCT parameters of biopsy-proven benign and malignant lung lesions.

**Methodology:** Ten patients with suspicious lung masses/nodules underwent a routine NCCT chest, followed by dVPCT on a 384-row multi-slice CT scanner system after injecting 50 ml Iohexol 300mg/ml. A subsequent tru cut biopsy was performed and the perfusion parameters of benign and malignant lesions were compared.

**Results:** The mean age was  $58 \pm 10.87$  years. All patients underwent biopsy to confirm the diagnosis; five were diagnosed with lung carcinoma, four were diagnosed to have a non-malignant lesion, one report was non-confirmatory, and this patient was excluded from the study. Dynamic perfusion parameters were compared between benign and malignant groups. Measurements of blood flow (malignant:  $53.29 \pm 18.43$  ml/dl/min; benign:  $9.94 \pm 7.1$  ml/dl/min,  $p=0.032$ ) and flow extraction product (malignant:  $12.18 \pm 1.9$  ml/dl/min; benign:  $3.86 \pm 3.7$  ml/dl/min,  $p= 0.035$ ) revealed a statistically significant difference between the two groups. Measurements of blood volume did not reveal a statistically significant difference between the groups (malignant:  $5.222 \pm 1.11$  ml/dl; benign:  $1.61 \pm 1.5$  ml/dl,  $p >0.05$ ).

**Conclusion:** A statistically significant difference in blood flow and flow extraction product was found between the benign and malignant groups. Blood volume did not show a statistically significant difference between the groups. The findings suggest that dVPCT can be added to routine NCCT to facilitate diagnosis. However since, this is a preliminary pilot study, further prospective multicentric studies are required to determine the usefulness of dVPCT in the diagnosis of lung lesions.

**Keywords:** lung, malignant, benign, biopsy



**RP 04**

**The association between tumour infiltrating lymphocytes and known prognostic factors of breast carcinoma in a cohort of Sri Lankan women**

K. S. Maheshika<sup>1\*</sup>, S. M. Fernandopulle<sup>2</sup>, S. J. de S. Hewavisenthi<sup>1</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

<sup>2</sup>*Department of Pathology, Colombo North Teaching Hospital, Ragama, Sri Lanka*

[saramaheshika@gmail.com](mailto:saramaheshika@gmail.com)

**Introduction and objectives:** The balance between tolerance of self-antigens and elimination of non-self-antigens plays a major role in breast carcinoma progression. Tumour infiltrating lymphocytes (TIL) in the tumour microenvironment play a crucial part in this surveillance. The objective of the study was to determine the association between TIL and known prognostic factors in a cohort of Sri Lankan women.

**Methodology:** Two hundred and ninety-four patients were included in the study conducted during a period from 2018 January to December 2021. Histological reports were used to gather data on age, tumour grade, tumour stage, extent and grade of DCIS, presence of lymphovascular involvement and the nodal stage of breast carcinoma. All histological types of breast carcinoma were included in the study and 85% (250/294) were of no special type (NST). The haematoxylin and eosin slides were examined, and stromal TILs were graded according to the recommendations given by an International Tumor Infiltrating Lymphocytes Working Group 2014. A Pearson's chi-square test was used to assess the relationship between categorical variables. The Spearman rank correlation was used to determine the correlation between stromal TILs and age and tumour size.

**Results:** The size of the tumour and Nottingham grade demonstrated a statistically significant correlation with stromal TIL ( $p < 0.05$ ). Age, lympho-vascular invasion, DCIS and nodal stage showed no significant correlation with TIL.

**Discussion and conclusion:** This study demonstrated a correlation between TIL and tumour grade and size. Results of previous studies on the association between TIL and prognostic factors have shown mixed results. It is suggested that these differences may be due to genetic influences resulting in different interactions with the host immune system and further studies are recommended to validate these findings.

**Keywords:** breast carcinoma, tumour infiltrating lymphocytes

RP 05

**Association between stromal tumour infiltrating lymphocytes and hormone receptor status and Her2/neu in a cohort of Sri Lankan women with breast carcinoma**

S. M. Fernandopulle<sup>1</sup>, K. S. Maheshika<sup>1</sup>, S. J. de S. Hewavisenthi<sup>2</sup>

<sup>1</sup>*Department of Pathology, Colombo North Teaching Hospital, Ragama, Sri Lanka*

<sup>2</sup>*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[saramaheshika@gmail.com](mailto:saramaheshika@gmail.com)

**Introduction and objectives:** Stromal tumour infiltrating lymphocytes (TIL) has become an area of interest in discovering new therapies because it is thought to play a role in tumour progression. The aim of this study was to document the association between stromal TIL, ER, PR and Her 2/neu status in breast carcinoma.

**Methodology:** Stromal TIL was assessed and graded as no/minimal, intermediate and high based on the recommendations of the International Tumor Infiltrating Lymphocytes Working Group 2014 in a cohort of breast carcinoma patients who underwent wide local excision (WLE) or mastectomy. The ER and PR status were assessed using Allred score and Her2/neu results were extracted from the pathology reports. The Chi-squared test was used to determine the relationship between these parameters.

**Results:** In the ER-negative cases, 17.8% (13/73) had no/minimal, 43.8% (32/73) had intermediate and 38.4% (28/73) had high TIL. In the PR-negative group, 19.5% (16/82) had no/minimal, 42.7% (35/82) had intermediate and 37.8% (31/82) had high TIL. When the intermediate and high TIL groups were combined and considered the high-TIL group, 82.2% (60/73) of the ER-negative group and 80.5% (66/82) of the PR-negative group showed high TIL. Out of ER-positive cases, 47.7% (62/130) had no/minimal immune cells, 32.3% (42/130) had intermediate and 20% (26/130) had high immune TIL. Out of PR-positive cases, 49.2% (59/120) had no/minimal immune cells, 34.2% (41/120) had intermediate and 16.7% (20/120) had high TIL. The majority were HER2-negative (79.1%;159/201). The distribution of TIL was, 55/159 (34.8%) no/minimal, 67/159(42.4%) intermediate and 36/159(22.8%) high TIL.

**Discussion and conclusion:** When the intermediate and high TIL groups were combined, high stromal TIL was associated with negative ER and PR ( $p < 0.001$ ), raising the possibility of different immune-tumour interactions based on the ER/PR status. There was no significant association between stromal TIL and Her2/neu in this study.

**Keywords:** breast, tumour infiltrating lymphocytes, hormone receptors, Her2/neu

**RP 06**

**Diagnostic accuracy of rapid stain technique in touch imprint cytology of axillary sentinel lymph nodes**

*E. E Harold<sup>1</sup>, D. M..T. Chandramali<sup>1</sup>, P. D. K. Thiranagama<sup>1</sup>, K. Wijesinghe<sup>2</sup> and C. S. P. Sosai<sup>1\*</sup>*

<sup>1</sup>*Department of Histopathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

<sup>2</sup>*Department of Surgery, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

[cherinesosai@yahoo.com](mailto:cherinesosai@yahoo.com)

**Introduction and objective:** Touch imprint cytology (TIC) is a well-recognized technique in the intraoperative assessment of axillary sentinel lymph nodes (ASLN) for metastases. Rapid stain technique (RST) can reduce TIC turnaround time (TAT). The objective is to determine the diagnostic accuracy of rapid stain TIC.

**Methodology:** Data was retrieved and analysed from the departmental database. ASLN reports from January 2019 to November 2022 were reviewed. ASLN received for TIC was subjected to a validated RST, in which, the TIC smear was fixed and stained for one minute in 95% ethyl alcohol and haematoxylin. The same slides were also routinely fixed and stained with Papanicolaou stain (PAP). The remaining ASLN was fixed and routinely processed. Following both methods, the slides were assessed microscopically. The sample size was calculated by the recommended formula. Sensitivity, specificity, positive and negative predictive values were calculated using the Chi square test.

**Results:** For 55 ASLNs, the sensitivity, specificity, and positive and negative predictive values for RST was 100%, 90.9 %, 73.3%, and 100% in diagnosing metastasis. Sensitivity, specificity, positive and negative predictive value for routine PAP stain was 100 %, 95.5%, 84.6% and 100% in diagnosing metastasis. The false positive results were, 4 (7.3%) in RST and 2 (3.6%) in PAP stain.

**Discussion and conclusion:** The specificity is reduced in both methods due to the false positive results thus the diagnostic accuracy of RST is less compared to PAP stain. The histiocytes which mimicked malignant cells when in clusters and due to unstained cytoplasm gave the false positive results. Hence RST should be used together with routine stains to acquire satisfactory competence as RST will reduce TAT for intraoperative TIC.

**Keywords:** intraoperative cytology, rapid stain, sentinel lymph node, diagnostic accuracy



## RP 07

### Audit of documentation of quality control parameters in histopathology

E. E Harold, C. S. P. Sosai\*

*Department of Histopathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

[cherinesosai@yahoo.com](mailto:cherinesosai@yahoo.com)

**Introduction and objectives:** Provision of safe and quality laboratory reports requires stringent quality control measures. Assessment and documentation of tissue section quality at reporting is mandatory to prevent pitfalls related to poor quality. The objective is to audit documentation of quality parameters against a 100% benchmark and to identify reasons for any unsatisfactory quality.

**Methodology:** Request forms from January to December 2022 were analysed for documentation of the quality of fixation, processing, microtomy, staining, mounting and labelling as satisfactory or unsatisfactory for reporting. If unsatisfactory, the reason indicated was recorded. Data were analysed using SPSS software.

**Results:** There was a total of 4344 requisition forms. Quality parameters were documented in 3881 (89.3%) and not documented in 463 (10.7%). Fixation was recorded as satisfactory in 3855 (99.3%), processing in 3773 (97.2%), microtomy in 3748 (96.6%), staining in 3860 (99.5%), mounting in 3873 (99.8%) and labelling in 3875 (99.8%). Fixation was unsatisfactory in 26 (0.7%), processing in 108 (2.8%), microtomy in 133 (3.4%), staining in 21 (0.5%), mounting in 8 (0.2%) and labelling in 6 (0.2%). The reasons for unsatisfactory sections were due to fragmentation resulting from poor processing, retention of water and inadequate removal of wax, over or under-staining with eosin, incorrect embedding plane, thick sections, tissue loss, folds, rolled up tissues, crush artefacts, holes, scores, chatters, carryovers and air bubbles or combinations of these factors.

**Discussion and conclusion:** The rate of documentation of the quality parameters was high but not on par with the benchmark level. Unsatisfactory status was higher in areas of processing and microtomy. Hence, stringent quality control measures for processing and microtomy followed by reaudit are due.

**Keywords:** quality parameters, histopathology, tissue processing, microtomy

## RP 08

### Second opinion in diagnosis of cancers: experience of a cancer referral centre

B.Y. Hettiarachchi<sup>1\*</sup>, C.M. Baranasuriya<sup>1,2</sup>, K.K.S. Lakshika<sup>1</sup>, H.D. Wijesinghe<sup>1,2</sup>, M.V.C. de Silva<sup>1,2</sup>

<sup>1</sup>Centre for Diagnosis and Research in Cancer, <sup>2</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

buddhinihettiarachchi21@gmail.com

**Introduction and objectives:** The Centre for Diagnosis and Research in Cancer (CeDARC) provides second opinions to Sri Lankan pathologists on diagnostically difficult cases. This audit analyses the type of cases received at CeDARC, and whether it was possible to provide a clinically relevant comprehensive diagnosis with the addition of immunohistochemical tests.

**Methodology:** All cases referred to CeDARC from January 2022 to June 2023 were reviewed. The initial diagnoses of the referring pathologist (RP) and CeDARC were categorized as being concordant or having major or minor discordance. Cases with major discordance (MD) were further analyzed.

**Results:** There were 259 referred cases; skin (35;14%), haematolymphoid (33;13%), soft tissue (32;12%), gynaecology (26;10%), head and neck (HN) (26;10%), neuropathology (23;9%), bone (23;9%). The RP had provided a diagnosis in 204 cases (78.8%). The majority (144/204;70.6%) were concordant with the final diagnosis of CeDARC and included in the differential diagnosis of the RP. The overall discordance rate (DR) was 23.1% (major-57/204;27.9%, minor-3/204;1.5%). Soft tissue (10/57;17.5%), bone (7/57;12.3%), haematolymphoid (10/57;17.5%), skin (10/57;17.5%), HN (6/57;10.5%) and gynaecology (5/57;8.8%) had the highest DR. The cases with MD were categorized as a change in diagnosis from malignant to benign (10/204;4.9%), benign to malignant (3/204;1.5%), different entity but still benign (3/204;1.5%), different entity but still malignant (20/204;9.8%) and change in soft tissue/bone tumour category (21/204;10.3%). The final diagnoses were based on further immunohistochemical testing in 59.6% (34/57) of cases that showed MD, and each of these required an average of four immunohistochemical markers.

**Discussion and conclusion:** The soft tissue, haematolymphoid, bone, skin, HN and gynaecology cases showed the highest rates of MD. Most of these cases required extra immunohistochemical markers, highlighting the need to establish more IHC facilities around the country. Moreover, these areas with the highest rates of MD require expert panels, workshops in the local setting, and specialized training in local and foreign centres.

**Keywords:** discordance rate, cancer, referral, immunohistochemistry

**RP 09**

**Undergraduate student perception of an online course for clinical clerkship in histopathology**

D.R. Jayasinghe<sup>1,2\*</sup>, K.A.G. de Abrew<sup>3</sup>, M.D.S. Lokuhetty<sup>1</sup>, M.V.C. de Silva<sup>1</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>2</sup>*University Hospital Lewisham, Lewisham and Greenwich NHS Trust, United Kingdom*

<sup>3</sup>*Department of Medical Education, Faculty of Medicine, University of Colombo, Sri Lanka.*

[dineshajayasinghe\\_01@yahoo.com](mailto:dineshajayasinghe_01@yahoo.com)

**Introduction and objectives:** The COVID-19 pandemic instigated a boom in blended learning, compelling academics to employ innovative methods of teaching/learning. This study evaluates the perceptions of medical undergraduates on a one-day self-directed online course in clinical histopathology.

**Methodology:** The clerkship in clinical pathology in the MBBS course spans two weeks in the 3<sup>rd</sup> year of the course. During COVID-19, a one-week online course was developed as an adjunct to clinical work, which included a one-day ‘Moodle lesson’ on clinical histopathology. Excerpts from lectures, videos and images were used to teach a range of topics including specimen collection and transport, grossing, processing, embedding, sectioning, staining, reporting and laboratory errors. An online quiz with real-time feedback assessed learning. Students’ perceptions of structure, content, appropriateness, attractiveness and usefulness of the lesson were obtained using a 10-item Google form. Free comments were encouraged. Results were summarised using descriptive statistics.

**Results:** 245 students who completed the lesson submitted the Google form. All 10 items (“attracted interest”, “stated objectives”, “content appropriate”, “well-organized”, “relatable to practise”, “tools enhanced understanding”, “quiz was useful”, “feedback in quiz useful”, “useful for self-learning” and “would you like to have this type of activity during system-based modules?”) received positive feedback (4-5/5 in 5-point Likert scale) from >90% of students. Overall mean score for the lesson was 4.49/5 ( $\pm 0.61$ ). Free comments were positive, with suggestions for improvement.

**Discussion and conclusion:** Online platforms such as Moodle can be used effectively for teaching not only theoretical concepts, but also practical aspects of medical education, and bring positive perceptions among students, especially when used as an adjunct to clinical teaching.

**Keywords:** blended learning, clinical pathology, medical education, online teaching, histopathology laboratory

## RP 10

### Tumour budding and its association with the TNM stage in neoadjuvant treated and untreated colorectal cancers

E.W. Kosgallana<sup>1</sup>, S. Wijetunge<sup>2</sup>, S.L. Malaviarachchi<sup>3</sup>, G.S.S. Hegoda<sup>2</sup>, I.V. Prematilleke<sup>4\*</sup>

<sup>1</sup>Department of Anatomy, <sup>2</sup>Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka

<sup>3</sup>Department of Oncology, University Hospital, Plymouth NHS Trust, United Kingdom

<sup>4</sup>Department of Pathology/Centre for Cancer Research, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka

[ishaprem@sjp.ac.lk](mailto:ishaprem@sjp.ac.lk)

**Introduction and objectives:** Tumour budding (TB) is recognised as an independent adverse prognostic factor in colorectal cancer (CRC). However, its significance after neoadjuvant treatment is still unclear. The aim of this study was to assess the association between TB and TNM stage in neoadjuvant-treated and untreated CRCs.

**Methodology:** The histology slides of neoadjuvant-treated and untreated CRC from four tertiary care centres were evaluated for PTB (peritumoral TB), ITB (intratumoral TB) and TNM stage. The presence of TB was assessed according to the International Tumour Budding Consensus Conference 2016 recommendations. Chi-square, Fisher's exact and rank biserial correlation statistical tests were used to assess the associations between PTB and ITB with TNM stage.

**Results:** Ninety-two CRCs were included in this study, and 27.17% (25/92) were neoadjuvant-treated tumours. TB was present in 56% (14/25) of treated and 73.13% (49/67) of untreated cases (p=0.116). In the untreated CRC, PTB showed a significant association with the increasing T stage (p=0.025) but not with the N stage (p=0.203) or M stage (p=0.392). ITB showed a significant relationship with the increasing N stage (p=0.036) but not with the T stage (p=0.288) or M stage (p=0.631) of the tumour. In the neoadjuvant-treated cases, PTB was not significantly associated with the T stage (p=0.197), N stage (p=0.952) or M stage (p=1.000). Similarly, ITB was not significantly associated with the T stage (p=0.778), N stage (p=0.456) or M stage (p=1.000).

**Discussion and conclusion:** In the neoadjuvant untreated CRCs, PTB was associated with the T stage and ITB with the N stage. In treated cases, neither PTB nor ITB was associated with the TNM stage. Neoadjuvant therapy may alter the predictive value of TB in CRC.

**Keywords:** colorectal cancer, neoadjuvant treatment, tumour budding

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## RP 11

### Analysis of immuno-histochemistry markers and costs incurred at a tertiary referral centre for precise cancer diagnosis

K. K. S. Lakshika<sup>1\*</sup>, B. Y. Hettiarachchi<sup>1</sup>, H. D. Wijesinghe<sup>1,2</sup>, M. V. C. de Silva<sup>1,2</sup>

<sup>1</sup>Centre for Diagnosis and Research in Cancer, Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

[kkslakshika96@gmail.com](mailto:kkslakshika96@gmail.com)

**Introduction and objective:** Immunohistochemistry (IHC) though essential for diagnosis of difficult cases, incurs a heavy cost. This is an audit of utility and cost of IHC at a tertiary centre for cancer diagnosis.

**Methodology:** All cases referred to the Centre for Diagnosis and Research in Cancer (CeDARC), Faculty of Medicine, University of Colombo from January 2022 to May 2023 were reviewed. Details of IHC tests were obtained from laboratory records.

**Results:** From a total 265 cases received at CeDARC, 181(68.03%) required 941 IHC tests. Seventy-nine cases were received with initial IHC done at referring hospital (345 tests). Fifty-five required further IHC testing, whilst 102 had all IHC tests done at CeDARC (total=157 cases). An average of five tests were done per case (range 1-23). Estimated price per marker ranged from LKR8,140–25,900 with an estimated total cost of LKR10,103,540(181 cases) and an average cost of LKR55,820 per case(range=LKR8,620-251,300). The most often used IHC markers were Ki67(n=71), S100(n=42), PanCK(n=38), CD20(n=35), SMA(n=33), CD3(n=32), CD34(n=27), Desmin(n=26), CD10(n=23), and CK7(n=22). Of the tests done at CeDARC(n=596), 37.24%(n=222) were haematopathology, 14.93%(n=89) basic immunohistochemical, 13.75%(n=82) soft tissue, and 11.57%(n=69) breast and gynaepathology markers. Markers available only at CeDARC and utilized included IDH-1(n=18), IgG4(n=9), PAX-8(n=7), INI-1(n=7), SOX10(n=6), Glypican3, TLE-1(n=5 each), MDM-2, kappa, lambda(n=4 each), HBME-1, STAT-6, myeloperoxidase(n=3 each), DOG-1, BerEP4, Granzyme-B, SAL-4, ERG(n=2 each), c-myc and PD1(n=1 each).

**Discussion and conclusion:** The most used markers at CeDARC were haematopathology, basic immunohistochemical and soft tissue markers. Costs incurred for IHC testing in the diagnosis of the referred cases was considerable with an average cost of LKR55,820 per case.

**Keywords:** immunohistochemistry, referral centre, cancer diagnosis, cost.

## RP 12

### Lymphoid cell infiltrate and its association with coexistent pathologies in the thyroid gland

L. I. Madaluwage\*<sup>1</sup>, L. Jayasinghe<sup>2</sup>, T. P. M. Bopagoda<sup>1</sup>, S. J. De. S. Hewavisenthi<sup>1</sup>

<sup>1</sup>Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka

<sup>2</sup>Military Hospital, Colombo, Sri Lanka

[imanilakshika@gmail.com](mailto:imanilakshika@gmail.com)

**Introduction and objectives:** Lymphoid cell infiltrate (LTI) can co-exist with benign and malignant thyroid pathologies and their relationship with LTI is complex and is not completely understood. The objective of this study was to describe the significance of LTI and its relevance to co-existing pathologies.

**Methodology :** This was a descriptive cross-sectional study of all thyroidectomies reported from 2018 to 2022 at Faculty of Medicine, University of Kelaniya. Prevalence, severity of LTI and coexisting benign and malignant pathologies were assessed. The severity of LTI was graded according to number of lymphocytic foci based on criteria by Williams and Doniach, defined as grade 0->0-1focus, Grade 1->2-8 foci, Grade 2->9-40 foci, Grade 3->>40 foci, Grade 4->more than half of parenchyma replaced by lymphocytic foci. Grades 0 and 1 were categorized as mild, grade 2 as moderate and grade 3 and 4 as severe LTI.

**Results :** Of the 203 cases examined, 128(63%) showed LTI of which 41/128(32.0%) was mild,43/128 (33.6%) moderate and 44/128( 34.4%) severe. In cases with LTI, benign pathologies were seen in 81/128(63.4%) and malignant pathologies were seen in 47/128(36.7%) of which papillary thyroid carcinoma was the commonest(85.1%,40/47). In 75 cases without LTI, 53(70.6%) cases showed a benign pathology and 22(29.3%)cases were malignant. Severity of LTI in benign lesions was mild in 32/81(39.5%) moderate in 22/81(27.2% )and severe in 27/81,( 33.3%) while in malignant lesions it was mild in 9/47(19.2%) moderate in 21/47(44.7%) and severe in 17/47(36.1%). A greater proportion of malignant lesions(47/69,68.12%)were associated with LTI than benign(81/134,60.4%), although there was no statistically significant difference.

**Discussion and conclusion :** Majority of malignant pathologies were associated with LTI of moderate to severe degree. In benign pathologies it was of mild degree. Though the presence or absence of LTI do not relate with an underlying pathology, the cautious assessment of thyroidectomies for a possible coexisting malignant pathology would be beneficial when moderate to severe LTI is encountered.

**Keywords:** lymphocytic thyroid infiltrates, severity, co-existing pathologies

**RP 13**

**A comparison of two audits on liver biopsies carried out in a tertiary care setting**

L. I. Madaluwage\*, S. J. de S. Hewavisenthi

Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka

imanilakshika@gmail.com

**Introduction and objectives:** The emergence of less invasive, more sophisticated techniques to evaluate the liver has led to a belief that liver biopsies (LB) will be performed in lesser numbers in the future. However, in our setting with limited advanced technology, liver biopsies are still performed in considerable numbers. The aim of this study was to carry out an audit of LBs performed during a three-year period and compare this with a similar audit done previously.

**Methodology:** A retrospective descriptive study was done on all LBs reported from 2019 to 2022 at the Department of Pathology, Faculty of Medicine, University of Kelaniya. Data were collected regarding the number, demographic factors of patients undergoing LB and diagnoses, and these were compared with a similar audit done in the same unit from 2009 to 2012.

**Results:** A total of 327 LBs were performed, in comparison to 216 in the previous audit, an increase of 51.3%. The age range was two months to 78 years, which included 165 males (50.4%) and 162 (49.6%) females. Most LBs (21%;69/327) were done in the 51-60-year age group compared to the previous audit where 21% of LBs were done in the 61-70-year age group. The common histological diagnoses were focal lesions (24%;77/327), cirrhosis (57/327,17%), fatty liver disease (50/327,15%) and chronic hepatitis (39/327,12%). Diagnoses were similar, except that transplant biopsies accounted for 6.1% (20/327) and cholestasis for 9.1% (30/327), in comparison to none and 6.1% (13/216) in the earlier audit.

**Discussion and conclusion:** The number of LBs performed has increased during the 10-year period with some variations in demographic factors and diagnostic entities and the addition of transplant biopsies. These findings support the view that liver biopsies are still performed in considerable numbers even though there are more sophisticated techniques.

**Keywords:** audit, liver biopsy, comparison

## RP 14

### Comparison of ER and HER2 expression in screen-detected and symptomatic invasive breast carcinomas: an audit

H. Maddumage\*, S.D. Palma, A. Gamage

*Department of Cellular Pathology, Royal Surrey Hospital-NHS-Foundation Trust, United Kingdom*

[hansikamaddumage@gmail.com](mailto:hansikamaddumage@gmail.com)

**Introduction and objectives:** Invasive breast cancer (IBC) is the most common cancer among women, and the majority overexpress ER while a minority expresses HER2. Reliable receptor assessment is crucial to minimize false-negative and false-positive results that may lead to suboptimal or overtreatment, respectively. The objectives of this study were to assess the distribution of ER and HER2 expression in screen-detected and symptomatic IBC and to compare the results within these two groups and with the national data and data of the South-East region of the UK.

**Methodology:** The ER and HER2 status of the IBCs, screen-detected and symptomatic, reported on breast core biopsies at the Department of Cellular Pathology Jarvis breast screening center and Royal Surrey Hospital-NHS Foundation trust in one-year period from 01 June 2021 to 31 May 2022 were retrospectively reviewed. The Allred score of 0-1 for ER was considered negative and a score of 2 and above was considered positive. A HER2 score of 3+ was considered positive and 0 - 1+ was considered negative. A score of 2+ was considered following further investigations.

**Results:** There was a total of 606 IBCs [60% (n=362) were screen-detected and 40% (n=244) were symptomatic from 574 females between the ages of 28-100 years. Positive ER expression was seen in 94.5% (n=342) of screen-detected IBCs and 86% (n=209) of symptomatic IBCs. HER2 was positive in 7% (n=26) of screen-detected IBCs and 13% (n=31) of symptomatic IBCs.

**Discussion and conclusion:** ER expression was high in screen-detected IBC, while HER2 expression was high in the symptomatic group. The pattern of ER expression in this study was similar to that of the national and regional data but the HER2 expression was slightly lower. As the pattern of ER expression was similar to national and regional data, dissemination of results among clinical breast teams and re-auditing in 12 months was recommended. With the lower percentage of HER2 positivity, monitoring the EQA process, paying attention to the scoring system and re-auditing in 12 months was recommended.

**Keywords:** ER, HER 2, screen-detected, symptomatic, breast carcinoma



## RP 15

### The spectrum of adult renal cell carcinoma, including possible molecular types, presenting to a tertiary care hospital in Sri Lanka

I. W. G. A. L. Malhasi<sup>1\*</sup>, M. M. A. Jayawickrama<sup>1</sup>, M. D. S. Lokuhetty<sup>2</sup>

<sup>1</sup>*Department of Histopathology, National Hospital of Sri Lanka, Sri Lanka*

<sup>2</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[malhasig@gmail.com](mailto:malhasig@gmail.com)

**Introduction and objective:** Renal cell carcinoma-(RCC) is the commonest renal malignancy, for which histomorphology remains important in determining the type. The aim of this study was to describe the RCC types encountered at a tertiary care delivery setting based on routine histomorphology and to identify potential cases requiring molecular analysis for further typing, as per the 5<sup>th</sup> WHO urinary and male genital tumours classification (Uro5).

**Methodology:** Microscopy of all RCC cases reported at the Histopathology Departments of the National Hospital and Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka from 2016 to 2020 were reviewed. Based on the most representative routine haematoxylin and eosin histomorphology, these were typed according to the WHO-Uro5. The tumours which need further molecular testing for typing were separated out.

**Results:** A total of 228 RCC cases were reviewed. This included 189 clear cell RCC (82.9%), 24 papillary RCC (10.5%) and six chromophobe carcinomas (2.6%). One eosinophilic solid and cystic RCC (0.3%) and one multilocular cystic renal neoplasm of low-malignant potential (0.3%) were identified. Seven cases which possibly required molecular testing for tumour typing in addition to histomorphology were separated out (3.1%).

**Discussion and conclusion:** With the introduction of targeted therapy, molecular profiling has significantly impacted the classification of adult renal malignancies, albeit histomorphology being the foundation of tumour taxonomy. The latest WHO-Uro5 has introduced a group of molecularly defined RCC entities. Therefore, it is worth facilitating molecular analysis to determine the RCC type as evidenced by this study (3.1%). The actual numbers of molecularly defined RCC types could even be higher, as some may mimic histomorphologically defined RCC types.

**Keywords:** renal cell carcinoma, histomorphology, classification, molecular testing

## RP 16

### A comparison of intraobserver variability of WHO/ISUP and Fuhrman nuclear grading systems for clear cell renal cell carcinoma

I. W. G. A. L. Malhasi<sup>1\*</sup>, M. M. A. Jayawickrama<sup>1</sup>, M. D. S. Lokuhetty<sup>2</sup>

<sup>1</sup>*Department of Histopathology, National Hospital of Sri Lanka*

<sup>2</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[malhasig@gmail.com](mailto:malhasig@gmail.com)

**Introduction and objectives:** The nuclear grade (NG) is an indicator of prognosis for certain renal cell carcinoma (RCC) types. The WHO/ISUP NG system replaced the previously used Fuhrman system (FS) in 2012. The objective of this study was to assess the intraobserver variability within these two systems.

**Methodology:** All clear cell RCCs (CC-RCC) reported in the Histopathology Departments of the National Hospital and the University of Colombo, Sri Lanka, from 2016 to 2020 were selected. One routinely stained slide with a representative tumour section was selected from each case. Two rounds of NG assessment were performed using each system with two-week wash-out periods. The slide sequence was changed between each round, and the findings were recorded in four separate data-entry sheets to prevent bias. The intraobserver agreement for each system was calculated using the kappa index (SPSS version 25.0). The slides with different NGs within the same system were re-examined to identify reasons for the discrepancy.

**Results:** There were 189 CC-RCCs. The kappa values for the WHO/ISUP NG system and FS were 0.953 and 0.856, respectively (95% CI = 0.916-0.99 and 0.794-0.918, respectively). The complexity of criteria, artefacts and poor slide quality hampered the accurate assessment of nuclear details and affected NG.

**Discussion and conclusion:** The WHO/ISUP system showed an almost perfect level of intraobserver agreement. Complexity/multiplicity of nuclear criteria would have resulted in higher intraobserver variability in FS. Although discordant with prior documentation, FS also demonstrated a strong level of intraobserver agreement in this study.

**Keywords:** renal cell carcinoma, WHO/ISUP, Fuhrman, intraobserver variability, kappa

**RP 17**

**Frequency and patterns of direct immunofluorescence positivity in glomerular diseases: a descriptive analysis**

M.A.D.N Munasinghe<sup>1</sup>, E.E. Harold<sup>1</sup>, K.R. Leelaratne<sup>1</sup>, S. Rodrigo<sup>2</sup>, R. Champika<sup>3</sup>, C.S.P Sosai<sup>1\*</sup>

<sup>1</sup>*Department of Histopathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

<sup>2</sup>*Department of Histopathology, Sri Jayawardenepura General Hospital, Sri Lanka*

<sup>3</sup>*Department of Histopathology, Medical Research Institute, Borella, Sri Lanka*

[cherinesosai@yahoo.com](mailto:cherinesosai@yahoo.com)

**Introduction and objectives:** Direct immunofluorescence (DIF) is the gold standard investigation, adjunct to morphological diagnosis, vital to establish the aetiology of glomerular diseases, which enables initiation of treatment to mitigate disease progression. DIF patterns may vary due to geographic variation in renal diseases. The objective of this study was to determine the frequency and pattern of DIF positivity in glomerular diseases.

**Methodology:** This was a retrospective analysis of renal biopsies with glomerular pathology reported at Colombo South Teaching Hospital in 2020. Biopsies with less than five glomeruli and transplant biopsies were excluded. Morphological diagnoses and frequency and pattern of DIF positivity were documented. Analysis was done using SPSS.

**Results:** DIF results were available in 392 renal biopsies, and 63.9% (212/392) showed positive DIF results. The most common morphological pattern of renal disease was FSGS, noted in 21.9% (73/392), followed by acute diffuse proliferative glomerulonephritis (ADPGN) in 19.6% (65/392), IgA nephropathy in 12.7% (42), minimal change disease (MCD) in 11.1% (37/392), and lupus nephritis in 10.6% (15/392). In FSGS, IgM was the most common immune deposit present in varying intensity and present in combination with C3 (46.6%;34/73). In MCD, 78.3% (29/37) had no immune deposits. ADPGN had variable immune deposits with IgG predominance in 60% (39/65) and C3 in 83% (54/65). IgA nephropathy had IgA in 90.4% (38/42) and C3 in 78.6% (33/42). Lupus nephritis had all immune reactants in 85.7% (30/35) and variable in 14% (5/35).

**Discussion and conclusion:** As the electron microscopy facility is lacking in the local setting, awareness of the different immune reactant patterns associated with glomerular diseases is helpful in differentiating glomerular diseases of similar morphology, such as FSGS and MCD. DIF-positive MCD cases were likely to be FSGS. DIF in lupus nephritis is likely to have all immunoreactants.

**Keywords:** direct immunofluorescence, FSGS, MCD, IgA nephropathy, lupus nephritis

**RP 18**

**A comparison of lymph node yield in left colectomy specimens, with and without neoadjuvant therapy: an audit in a tertiary care centre**

M. K. Piyasinghe<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, K. Ratnatunga<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Department of Surgery, Faculty of Medicine, University of Peradeniya, Sri Lanka*

mekupi@gmail.com

**Introduction and objectives:** The lymph node (LN) count and node positivity are important prognostic indicators in primary colorectal adenocarcinomas. The LN count shows significant variations with adjuvant therapy (AT). This study compares the LN status of AT-given (AT-G) and AT-not-given (AT-NG) left colectomy specimens.

**Methodology:** This retrospective study included all left colectomy specimens reported at the Department of Pathology, University of Peradeniya, for one year from October 2021. Clinicopathological data were extracted from the archives. The data were analyzed with demographic factors and neoadjuvant status.

**Results:** There were twenty-five (25) left colectomy specimens; 48% (n=12) were AT-G and 52% (n=13) were AT-NG. The male-to-female ratio was 2.3:1 (n=9:4) and 1:1 (n=5:5) in the AT-G and AT-NG groups, respectively. Patients' age range was 42-83 years, and the majority were between 60-69 years in both groups. The AT-G group had >12 LNs in 46% (n=6) of cases and a mean LN count of 12. The AT-NG group had >12 LNs in 83% (n=10) of specimens, and a mean LN count of 19. LN positivity was 0% in 84% of the AT-G category and 53% in the AT-NG category.

**Discussion and conclusion:** The total cases were relatively similar in both groups, but more men received AT than women. The mean LN count and the LN positivity in the AT-G group were lower than that of the AT-NG group. The majority of the cases in the AT-G group were classified under the ypN0 category.

**Keywords:** neoadjuvant therapy, lymph node count, left colectomy specimens



## RP 19

### The serum concentration of soluble apoptosis-associated molecules in coeliac disease

K.K. Prasad<sup>1\*</sup>, A.K. Sharma<sup>1</sup>, U. Debi<sup>2</sup>, P.K. Gupta<sup>3</sup>, S.K. Sinha<sup>1</sup>

<sup>1</sup>Departments of Gastroenterology, <sup>2</sup>Radiodiagnosis and <sup>3</sup>Biostatistics

Postgraduate Institute of Medical Education and Research, Chandigarh, India

[kaushalkp10@hotmail.com](mailto:kaushalkp10@hotmail.com)

**Introduction and objectives:** The exact role of soluble apoptosis-related mediators in celiac disease is still not fully understood. Serum sFas-sFasL may play a role in the pathogenesis, and the aim of this study was to assess the circulating soluble Fas and Fas ligand levels in celiac disease and compare that with the normal individuals.

**Methodology:** This prospective study included 50 adults with newly diagnosed celiac disease (CD-group/CDG) and 25 non-celiac adults (N-CD group/N-CDG) as controls. The circulating Fas and sFasL levels were assessed by ELISA kits. All duodenal histological parameters were evaluated, classified and histological lesions were graded according to criteria proposed by our group. Results were expressed as the mean±standard error (SE), range and percentage. The serum sFas and sFasL levels and duodenal histological results were analyzed using statistical methods, Mann-Whitney U test, One-way ANOVA and Linear regression using SPSS packed program (version 22 software, SPSS Inc. Chicago, Illinois, USA). The *P* values<0.05 were considered statistically significant.

**Results:** The mean age of CDG was 31.28±12.83 (15-62 years) years, with a male: female of 1:1. In the CDG, mean serum sFas was significantly lower (*p*<0.001) than N-CDG. However, the mean serum sFasL was significantly higher (*p*=0.018) than N-CDG. One-way ANOVA indicated no relationship between the grade of duodenal mucosal damage and concentration of serum sFas (*p*=0.35). On the other hand, one-way ANOVA indicated a statistically significant relationship between different grades of duodenal mucosal damage and sFasL (*p*=0.04).

**Discussion and conclusion:** The concentration of serum soluble Fas and FasL are significantly altered in CD. Altered serum sFas does not correlate with histological grades of duodenal mucosal lesions. Further studies are required to see any relation between membrane-bound Fas or FasL and the grading of duodenal mucosal lesions, which may help better understand the pathogenesis of celiac disease.

**Keywords:** celiac disease, soluble Fas, Fas ligand, apoptosis, serum

**RP 20**

**Critical appraisal of histological features of autoimmune hepatitis: an experience in the local setting**

H.R.S.D. Sumanasekara\*, D. Walisinghe, P. Rathnayake, A. Vithanage, N. Rathnayake, C.L. Piyarathne  
*National Hospital Kandy, Sri Lanka*  
[sachinhanasumanasekara@gmail.com](mailto:sachinhanasumanasekara@gmail.com)

**Introduction and objectives:** Autoimmune hepatitis (AIH) is diagnosed on the basis of clinical, biochemical, immunological and histological features, using scoring systems and by exclusion of other causes. Therefore, a liver biopsy is mandatory in the diagnostic workup. The objective of this study was to critically evaluate the histological features of AIH in a Sri Lankan cohort.

**Methodology:** This was a retrospective descriptive study. All the initial liver biopsies of patients with AIH (selected following clinical, serological and histological correlation) at the National Hospital Kandy from January 2022 to June 2023 were selected. The composition of the inflammatory infiltrate, the presence of interface hepatitis (IF), emperipolesis(E), hepatocyte rosettes(R), sinusoidal lymphocytic infiltration and the stage of chronic hepatitis were evaluated.

**Results:** Twenty-five liver biopsies that were stained routinely were analysed. All biopsies showed portal tract inflammation and interface hepatitis with lymphocytes (100%). Definitive plasma cells were present in 20 % (n=5). 84% showed hepatocyte rosette formation and emperipolesis. Sinusoidal lymphocytes were seen in 100%. Established cirrhosis/stage 4 disease was seen in 96%(n=24).

**Discussion and conclusion:** The main histological features (IF, E and R) in international scoring systems (The revised International Autoimmune Hepatitis Group (IAHG) modified scoring system-1999 and simplified scoring system of the IAHG-2008) were seen in most of our biopsies. Plasma cells were not prominent in the infiltrate. Lymphocytes were seen in the sinusoids in all biopsies and may be a morphological feature that favours AIH. The features highlighted in our study may have value in diagnosing AIH, although these should be validated by a case-control study with a larger sample.

**Keywords:** autoimmune hepatitis, emperipolesis, interface hepatitis

## RP 21

### **An audit on completeness of the core data in histopathology reports of peripheral neuroblastic tumours**

H. G. D. Thilini\*, S.A. Guneratne

*Teaching Hospital, Karapitiya, Sri Lanka*

*Lady Ridgeway Hospital for Children, Colombo 8, Sri Lanka*

[t.gamage1983@gmail.com](mailto:t.gamage1983@gmail.com)

**Introduction and objectives:** Neuroblastoma is the fourth most common childhood tumour. Histopathology plays a major role in the diagnosis of peripheral neuroblastic tumours (PNBT), a family of tumours arising in embryonal remnants of sympathetic nervous system, accounting for 7-10% of tumours in children. The aim of this study was to assess the completeness of histopathology reporting of PNBT according to Royal College of Pathologists (RC PATH) guidelines and set out a standard reporting proforma for our local setting.

**Methodology:** Histopathology reports of all PNBT reported at the Lady Ridgeway Hospital from 2017 to 2023 were included. Clinical, macroscopic and microscopic core data items were assessed.

**Results:** A total of 86 PNBTs were reported. The data criteria most often not reported was the pre- or post-treatment status (14%) followed by the sites of separate lymph nodes (8%), number of nodules present and nodular variant subtype (11%), grade of tumour differentiation (28%), necrosis (67%), calcification (67%) and immunohistochemistry utilized two neuroblastoma antibodies (77%). All other criteria were included in over 90% of reports.

**Discussion and conclusion:** Separate lymph nodes were usually not received in our setting. The absence of tumour necrosis and calcification was not mentioned. Macroscopic identification of nodules is important in the nodular variant subtype of neuroblastoma. Owing to low economic status, performing two neuroblastoma antibodies is impossible. Clinicians must be promoted to provide the treatment status. Guidelines are to be set in order to include important macroscopic findings such as the presence of nodules. Documenting the negative findings must also be encouraged.

**Keywords:** Peripheral neuroblastic tumours, PNBT, neuroblastoma, histopathology

**RP 22**

**Prevalence of sarcomatoid morphology in different types of renal cell carcinoma and its association with adverse prognostic factors**

P. D. K Thiranagama<sup>1</sup>, A. Abeygunasekera<sup>2</sup>, C. S. P. Sosai<sup>1</sup>

<sup>1</sup>*Department of Histopathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

<sup>2</sup>*Department of Urology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

prasangasiriwardena@gmail.com

**Introduction and objectives:** Sarcomatoid morphology (SM) in renal cell carcinoma (RCC) is associated with poor outcomes. The objective was to evaluate the prevalence of sarcomatoid morphology in different types of RCC and determine its association with other adverse prognostic factors.

**Methodology:** Data was retrieved and analysed from the departmental database from January 2018 to December 2021. Presence or absence of sarcomatoid morphology in different tumours and its association with tumour size, WHO/ISUP grade, necrosis, renal sinus invasion, extracapsular invasion, renal vein invasion and tumour stage were documented. SPSS was used for data analysis.

**Results:** Of the 94 RCCs clear cell RCC was the commonest 75/94(79.8%), followed by papillary RCC 16/94(17%) and chromophobe RCC 3/94(3.2%). SM was present in 23/94 (24.5%) RCCs and the highest prevalence was in clear cell RCC 19/75(25.3%) followed by papillary RCC 4/16(25%). SM was not seen in chromophobe RCC. Of the RCC with SM, 11/23(47.8%) had tumour size>7 cm, 23/23(100%) were WHO/ISUP grade 4, 19/23(82.6%) had tumour necrosis, 13/23 (56.5%) showed renal sinus invasion and 11/23(42.8%) had lymphovascular invasion. Of the RCC without SM, 22/71(31%) had tumour size>7 cm, 1/71(1.4%) was WHO/ISUP grade 4, 28/71(39.4%) had tumour necrosis, 14/71 (19.7%) had renal sinus invasion and 23/71(32.4%) had lymphovascular invasion. Chi square test showed a significant difference between the tumours with and without SM in relation to WHO/ISUP grade ( $p<0.001$ ), renal sinus invasion ( $p=0.003$ ) and tumour necrosis ( $p=0.001$ ). A significant difference was not seen in relation to tumour size ( $p=0.303$ ), renal vein invasion ( $p=0.179$ ), TNM stage ( $p=0.150$ ) and lymphovascular invasion( $p=0.181$ ).

**Discussion and conclusion:** SM is more prevalent in clear cell RCC compared to other renal tumours which indicates that extensive sampling is warranted for clear cell RCC. Renal tumours with SM show a significant association with several poor prognostic factors.

**Key words:** clear cell renal cell carcinoma, sarcomatoid differentiation, WHO/ISUP grade



**RP 23**

**Testing for IDH mutation in astrocytoma by immunohistochemistry: an analysis of potential misdiagnosis in the Sri Lankan setting**

I. L. Wickramanayake<sup>1,2\*</sup>, C.M. Baranasuriya<sup>1,2</sup>, S. Abeygunawardhane<sup>1,2</sup>, B.Y. Hettiarachchi<sup>2</sup>, K.K.S. Lakshika<sup>2</sup>  
H.D. Wijesinghe<sup>1,2</sup>, M.V.C. de Silva<sup>1,2</sup>

<sup>1</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup>Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka

[Indikalakmali507@gmail.com](mailto:Indikalakmali507@gmail.com)

**Introduction and objectives:** The new World Health Organization Classification bases the diagnosis of astrocytoma on the presence of IDH mutation. Immunohistochemistry (IHC) for IDH mutation was introduced for the first time in our setup. This audit analyses the results of IDH testing by IHC and the potential for misdiagnosis.

**Methodology:** Twenty-two glial tumours were referred for analysis of IDH mutational status by IDH1 R132H IHC from January 2022 to July 2023. The histology was evaluated by two pathologists to confirm the diagnosis, grade and IDH status.

**Results:** Twenty tumours were high-grade (90.91%;20/22) and two were low-grade (9.09%;2/22). Among the high-grade tumours, IDH mutation was present in seven tumours including one with areas of oligodendroglial morphology. These were diagnosed as CNS WHO grade 4 astrocytoma with recommendations for IDH sequencing and 1p/19q codeletion in the tumour with oligodendroglial morphology. One of the tumours with low-grade histology showed IDH mutation and was reported as astrocytoma CNS WHO grade 2 with recommendations for IDH sequencing. The remaining thirteen high-grade tumours were IDH wild type and diagnosed as glioblastoma, WHO grade 4. One case of IDH wild type showed low-grade morphology.

**Discussion:** The lack of resources for molecular testing has led to several limitations in the diagnosis. IDH mutation should be tested in all glial tumours irrespective of grade, however, it is requested mainly in high-grade tumours. There are no facilities for testing of IDH wild-type low-grade tumours on IHC with IDH sequencing, to exclude false negatives. Facilities for testing for TERT promoter mutation, EGFR amplification, and combined chromosome 7 gain and chromosome 10 loss to exclude glioblastoma CNS WHO grade 4, in low-grade IDH wild-type tumours lacking microvascular proliferation or necrosis are also lacking. These limitations may result in the misdiagnosis of low-grade glial tumours.

**Keywords:** glioblastoma, IDH sequencing, oligodendroglia

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**CR 01**

**Mediastinal histiocytic sarcoma: the diagnostic dilemma of a rare tumour**

A. A. S. D. Abeygunawardhane\*, C. M. Baranasuriya, A. A. H. Priyani, H.D. Wijesinghe,

M. V. C. de Silva

*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*

[sakuraabeygunawardhane@gmail.com](mailto:sakuraabeygunawardhane@gmail.com)

**Introduction:** Histiocytic sarcoma (HS) is a rare malignant neoplasm, that mostly occurs in adults, with an incidence of 0.17 per 1000,000 people. Although the involvement of extranodal sites is more common than nodal involvement, the mediastinal origin is extremely rare.

**Case report:** A previously well 37-year-old man presented with dysphagia and night sweats for one month. PET-CT scans showed posterior mediastinal lymph node masses with hepatosplenomegaly and possible bone marrow involvement, suspicious for lymphoma. FNAC and biopsy revealed a predominant population of large pleomorphic cells with abundant eosinophilic cytoplasm and pleomorphic nuclei, some containing prominent nucleoli. Bizarre tumour cells were present. There was marked neutrophil phagocytosis and focal extravasated mucin. The background contained mixed inflammatory cells. There was no associated germ cell tumour component. The tumour cells showed diffuse and strong positivity for CD68, weak membranous staining for CD15 and negative staining with PanCK, EMA, CAM5.2, TTF1, LCA, CD3, CD20, CD30, ALK1, S100, calretinin and desmin. INI1 stain was retained in tumour cells. The Ki-67 index was 50%. These histomorphological features were diagnostic of HS, and the patient is currently undergoing chemotherapy.

**Discussion and conclusion:** The differential diagnosis of anaplastic large cell lymphoma, germ cell tumour, mesothelioma, poorly differentiated metastatic carcinoma, malignant melanoma, extrarenal rhabdoid tumour and myxoinflammatory myofibroblastic tumour were excluded by negative stains of relevant immunohistochemical stains. HS can have associated germ cell tumour components. Surgical resection and chemoradiotherapy are the available treatment options for this aggressive tumour with poor treatment response. However, clinically localized and small tumours have more favourable outcomes. Though rare, the possibility of HS should be considered in a tumour with large cells showing phagocytosis.

**Keywords:** mediastinal histiocytic sarcoma, phagocytic tumours

**CR 02**

**Intraprostatic solitary fibrous tumour: a rare spindle cell lesion in the prostate**

A. A. S. D. Abeygunawardhane\*, I. L. Wickramanayake, H.D. Wijesinghe, M. V. C. de Silva  
*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*  
*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*  
[sakuraabeygunawardhane@gmail.com](mailto:sakuraabeygunawardhane@gmail.com)

**Introduction:** Solitary fibrous tumour (SFT) is a rare mesenchymal tumour in the prostate, with less than a hundred cases reported in the literature.

**Case report:** A 54-year-old man presented with lower urinary tract symptoms and an abdominally palpable mass. Radiology revealed gross prostatomegaly without any extraprostatic mass lesions. His serum PSA level was 3.05 ng/mL. Intraoperative findings revealed a mass lesion confined to the prostate. The resected specimen was composed of multiple, solid, white pieces of tissue measuring 100x120x50 mm. Microscopy showed a lesion with bland spindle cells admixed with benign prostatic tissue. The spindle cells were uniform with a vague fascicular arrangement and admixed with collagen bundles and proliferating vessels. There was no nuclear atypia, increased mitotic activity, myxoid change, necrosis or inflammatory infiltrate. The cells showed diffuse and strong nuclear staining for STAT6 stain, focal cytoplasmic positivity for SMA and membranous positivity for CD34. The cells were negative for desmin, S100, CD117, EMA, PanCK and ALK1. The Ki-67 proliferation index was less than 5%.

**Discussion:** Diagnosis of intraprostatic SFT needs exclusion of more common spindle cell lesions of the prostate, such as postoperative spindle cell nodule, prostatic stromal nodule of uncertain malignant potential, prostatic sarcoma, neural and spindle cell soft tissue tumours and myofibroblastic tumours.

**Conclusion:** Awareness of this rare site of occurrence of SFT is needed for the correct diagnosis and risk stratification.

**Keywords:** intraprostatic SFT, prostatic spindle cell lesions

**CR 03**

**Sertoli-Leydig cell tumour of ovary with benign and borderline heterologous elements: a rare presentation**

A. A. S. D. Abeygunawardhane<sup>1,2</sup> \*, K. K. S. Lakshika<sup>2</sup>, R.C.U. Priyadarshika<sup>3</sup>, G. Ranaweera<sup>1,2</sup>,  
M. V. C. de Silva<sup>1,2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>2</sup>*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*

<sup>3</sup>*Base Hospital, Wathupitiwala, Sri Lanka*

[sakuraabeygunawardhane@gmail.com](mailto:sakuraabeygunawardhane@gmail.com)

**Introduction:** Sertoli-Leydig cell tumours of the ovary account for < 0.5% of all ovarian neoplasms and heterologous elements, which are usually benign, occur in less than 20% of them. The presence of a borderline heterologous component is extremely rare.

**Case report:** A 39-year-old woman presented with abdominal pain and discomfort. Radiology revealed a large left ovarian mass and a normal right ovary. Macroscopically, the mass showed solid and cystic areas and measured 110x110x50 mm. Microscopy revealed a tumour with alternating hypo and hypercellularity with compressed and open tubules, cords and diffuse sheet-like architecture. The constituent cells showed mild to moderate atypia and occasional mitotic figures. The hypocellular stroma was fibrous with psammomatous calcifications. The Leydig cell component was inconspicuous. The cells stained positively with calretinin and inhibin with focal staining for PR. PAX8 was negative. The tumour contained glands lined by intestinal-type mucinous epithelium showing benign and borderline morphology. There was no evidence of retiform, sarcomatous or malignant heterologous elements.

**Discussion and conclusion:** The mixed morphology gave rise to the differential diagnosis of an ovarian endometrioid tumour with Sertoli-Leydig-like differentiation, which was excluded with immunostains. Moderate differentiation and heterologous elements are adverse prognostic factors. Identification of all the elements within the tumour is needed for the correct diagnosis and determination of better patient outcomes.

**Keywords:** Sertoli-Leydig cell tumour, heterologous elements

## CR 04

### A primary diffuse large B cell lymphoma of the central nervous system mimicking a metastatic carcinoma and a glioblastoma

B. N. Adhikari \*, P. Abeygunasekara

*Apeksha Hospital, Maharagama, Sri Lanka*

[buddhiadhikari2590@gmail.com](mailto:buddhiadhikari2590@gmail.com)

**Introduction:** Primary central nervous system lymphomas (PCNSL) are rare extranodal non-Hodgkin lymphomas confined to the central nervous system (CNS) at presentation, with no evidence of systemic involvement. Most PCNSL are diffuse large B Cell lymphomas (DLBCL). They typically occur in older people and have a poorer prognosis than systemic DLBCL.

**Case report:** A 69-year-old previously healthy man presented with sudden loss of consciousness. He had no history of immunodeficiency. Magnetic resonance imaging of the brain showed multiple supratentorial brain lesions, suggestive of metastasis. The right vocal cord asymmetry was seen radiologically. No lymphadenopathy, hepatosplenomegaly or mediastinal masses were detected. Excision of a left parietal brain lesion revealed closely packed sheets of polygonal cells with pleomorphic, hyperchromatic nuclei and moderate to abundant cytoplasm. Nucleoli were inconspicuous. A fibrillary background was not seen. Focal necrosis and microvascular proliferation were present. The main differential diagnoses were metastatic carcinoma and glioblastoma. The initial panel of immunomarkers for GFAP, pan-cytokeratin and EMA were negative. Subsequent immunohistochemistry panel showed strong, diffuse positivity for CD45 and CD20 and focal positivity for CD30. CD138 was negative. The Ki-67 proliferation index was 70%.

**Discussion and conclusion:** The atypical radiological and histological features led to a delay in diagnosis in this case. CD10 is useful to exclude CNS involvement by a systemic DLBCL, as fewer than 10% of all DLBCL-CNS express CD10. The majority of DLBCL-CNS express BCL2 and MYC, which would also help to confirm the diagnosis. EBV positivity would indicate an underlying immune deficiency. Some systemic DLBCL and anaplastic large cell lymphomas show diffuse positivity for CD30, which would help to exclude systemic lymphoma. Due to economic constraints, these markers were not performed in this case. PCNSL is on the rise and a high index of suspicion should be maintained in elderly patients presenting with neurological symptoms. Systemic lymphomas must be excluded.

**Keywords:** primary central nervous system lymphoma, primary diffuse large B cell lymphoma, central nervous system

## CR 05

### Classic papillary thyroid carcinoma arising in a mature cystic teratoma of the ovary

B. N Adhikari\*, P. Amarabandu

*Apeksha Hospital, Maharagama, Sri Lanka*

buddhiadhikari2590@gmail.com

**Introduction:** Mature cystic teratoma is the commonest ovarian germ cell tumour seen in young women. Malignant transformation is a rare occurrence.

**Case report:** A 33-year-old woman underwent surgical excision of a cystic right ovarian mass. Serum tumour markers were within normal limits. Macroscopic examination showed a multilocular cyst with a capsular breach. The cyst contained hair, sebaceous material and papillary structures accounting for 40% of the mass. Microscopically, papillary structures lined by cuboidal cells displaying nuclear features of papillary thyroid carcinoma and a few interspersed normal thyroid follicles were seen. The rest of the cyst showed mature stratified squamous epithelium, skin adnexa and respiratory epithelium. Immature elements were not seen. Imaging of the thyroid showed a solitary nodule in the left lobe. A total thyroidectomy was performed, and histology showed a colloid nodule with no evidence of papillary thyroid carcinoma. Staging laparotomy and radical hysterectomy were done. The patient underwent radioiodine therapy.

**Discussion and conclusions:** Malignant transformation of mature cystic teratomas is rare and typically occurs in postmenopausal women, and squamous cell carcinoma and adenocarcinoma are the most common malignancies. Although differentiated thyroid carcinomas are known to arise in association with struma ovarii, their occurrence within a mature cystic teratoma is exceptional. There are no optimal treatment protocols for this entity due to its rarity, and only a few studies have been published. Some authors recommend thyroidectomy to allow monitoring for recurrence with serum thyroglobulin levels and radioiodine ablation. Long-term follow-up studies are needed to prevent unnecessary therapeutic interventions.

**Keywords:** classic papillary thyroid carcinoma, mature cystic teratoma of the ovary, ovarian tumour



CR 06

**Retroperitoneal bronchogenic cyst masquerading as a pancreatic cyst: a rare case**

A.A.T.I. Adhikari<sup>1</sup>, N.J Asanthi<sup>1\*</sup>, V. Bandaranayake<sup>2</sup>, P. Nellihela<sup>2</sup>, S. Thalaspitiya<sup>3</sup>

<sup>1</sup>*Department of Histopathology, Teaching Hospital Anuradhapura, Sri Lanka*

<sup>2</sup>*Department of Onco-Surgery, Teaching Hospital, Anuradhapura, Sri Lanka*

<sup>3</sup>*Department of Surgery, Faculty of Medicine, University of Rajarata, Sri Lanka*

[jayanjana\\_asanthi@yahoo.com](mailto:jayanjana_asanthi@yahoo.com)

**Introduction:** Bronchogenic cysts are typically formed because of abnormal budding of the tracheobronchial tree during embryonic development. They are commonly found in the thorax, specifically in the mediastinum. Although rare, cases of bronchogenic cysts occurring within the retroperitoneum have been reported.

**Case report:** A 48-year-old previously healthy, non-alcoholic, man presented with left-side abdominal pain of three weeks duration. Physical examination revealed mild tenderness over the left hypochondrium. Radiological investigations were suggestive of a pancreatic cyst. Surgical excision of the lesion was performed. The gross specimen comprised a multilocular cyst measuring 60 mm in diameter, with focal whitish, solid areas. Histological sections revealed cysts lined by respiratory epithelium and mucinous epithelium with smooth muscle tissue, seromucinous glands and cartilage within the cyst wall. There was no necrosis, mitoses or nuclear pleomorphism indicative of malignancy.

**Discussion:** Retroperitoneal bronchogenic cysts are extremely rare lesions that are believed to occur either from trapping of the abnormal buds of the tracheobronchial tree or aberrant budding from the primitive foregut. The left adrenal gland and pancreatic region are the frequent sites where they occur in the retroperitoneum. Therefore, these lesions might be diagnosed radiologically as pancreatic cysts, as in this case, or as adrenal lesions. The presence of cartilage, seromucinous glands and smooth muscle helps to differentiate this from the differential diagnoses of intraabdominal cystic teratoma, bronchopulmonary sequestration and foregut cysts.

**Conclusion:** Despite their rarity, retroperitoneal bronchogenic cysts should be considered in the differential diagnosis of retroperitoneal cystic lesions to ensure accurate diagnosis and appropriate management.

**Keywords:** retroperitoneal bronchogenic cyst

## CR 07

### **Anaplastic large cell lymphoma presenting as multiple polyps within the upper gastrointestinal tract: a rare presentation**

A.A.T.I. Adikari<sup>1</sup>, N.J. Asanthi<sup>1\*</sup>, C De Silva<sup>2</sup>, P. Nellihela<sup>3</sup>

<sup>1</sup>Department of Histopathology, <sup>2</sup>Gastroenterology Unit, <sup>3</sup>Department of Onco-Surgery,

Teaching Hospital Anuradhapura, Sri Lanka

[jayanjana\\_asanthi@yahoo.com](mailto:jayanjana_asanthi@yahoo.com)

**Introduction:** Anaplastic large cell lymphoma (ALCL) is a heterogeneous T-cell lymphoma involving both nodal and extranodal sites. However, involvement of the gastrointestinal tract (GIT) is rare.

**Case report:** A 28-year-old man was investigated for vomiting and dyspeptic symptoms of one-month duration. His physical examination and laboratory investigations, including the FBC and blood picture were within normal limits. LDH was not performed as lymphoma was not suspected initially. Upper GI endoscopy revealed multiple polypoidal lesions extending from the lower oesophagus up to the second part of the duodenum. A representative gastric antral polyp was biopsied, and microscopy revealed non-specialized gastric mucosa infiltrated predominantly by diffuse sheets of small to medium-sized lymphoid cells with irregular nuclei. There were scattered large cells with irregular nuclei with binucleate forms, and amphophilic moderate to abundant cytoplasm. These cells were positive for immunohistochemical stains LCA, CD4, CD30, and ALK1 and CD3 were positive in occasional cells. CD20 and PCK were negative. The Ki-67 index was 80%. These features were compatible with ALK+ALCL. Subsequent radiological assessment excluded any nodal involvement, confirming this as a primary GIT disease.

**Discussion:** ALCL is most frequent in childhood and young adults, with male predominance. Among the extranodal sites, skin, bone, soft tissue, lung and liver are frequently involved. GIT involvement is very rare, and ulceration, thickening and mass-forming lesions are the usual presentations. Polyposis is typically seen in B cell lymphomas, especially in mantle cell lymphoma. ALCL presenting with polyposis, as in this case, is unusual. There is a broad spectrum of microscopic appearances in ALCL, and some variants are associated with unfavourable prognosis.

**Conclusion:** Despite the rarity, ALCL should be considered when there are multiple polypoid lesions involving the GI mucosa.

**Keywords:** anaplastic large cell lymphoma, gastrointestinal tract

**CR 08**

**Cutaneous lymphadenoma: a rare skin adnexal tumour in a young Sri Lankan woman**

B. D. B. M. Baduraliyage\*, T. G. Liyanage

*Department of Pathology, Faculty of Medicine, University of Ruhuna, Sri Lanka*

buddimanimadavi@gmail.com

**Introduction:** Cutaneous lymphadenoma, a distinctive variant of trichoblastoma, is an exceedingly rare benign skin adnexal tumour that is also referred to as lymphotropic adamantinoid trichoblastoma. It primarily occurs in adults as a small nodule on the face, back or legs.

**Case report:** A 24-year-old woman presented with a painless left eyebrow lump for two years. An excisional skin biopsy measuring 10×5×2 mm was received. Microscopic evaluation revealed an intact epidermis and a circumscribed, unencapsulated organoid lesion in the dermis. The lesion comprised multiple lobules of basaloid cells with peripheral palisading nuclei in a desmoplastic stroma. The centres of lobules contained pale, large cells with vesicular nuclei and abundant eosinophilic cytoplasm admixed with mature lymphocytes. Lymphocytes spilled over into the stroma. There were no brisk mitoses, atypical cells or retraction artefacts.

**Discussion and conclusion:** Cutaneous lymphadenoma poses diagnostic challenges due to its rarity and overlapping features with other skin adnexal neoplasms and basal cell carcinoma (BCC). Cutaneous lymphadenoma is characterised by irregular basaloid islands with lymphocytic infiltration and focal follicular differentiation, distinguishing it from other skin adnexal tumours. Differentiating it from BCC is supported by the lack of mitoses, the presence of papillary mesenchymal bodies, and the absence of retraction artefacts. Accurate diagnosis of cutaneous lymphadenoma requires careful histopathological evaluation to avoid misdiagnosis as BCC or other skin adnexal neoplasms.

**Keywords:** cutaneous lymphadenoma, adnexal tumour, adamantinoid trichoblastoma

**CR 09**

**Primary ALK-negative anaplastic large cell lymphoma of ischium: a rare encounter**

C. M. Baranasuriya<sup>1\*</sup>, H. D. Wijesinghe<sup>1</sup>, B.Y. Hettiarachchi<sup>1</sup>, M. Jayawickrama<sup>2</sup>,  
W. Rathnayake<sup>3</sup>, M. V. C. de Silva<sup>1</sup>

<sup>1</sup>Centre for Diagnosis and Research in Cancer (CeDARC), Department of Pathology, Faculty of Medicine,  
University of Colombo, Sri Lanka

<sup>2</sup>Department of Pathology, National Hospital of Sri Lanka

<sup>3</sup>National Cancer Institute, Sri Lanka

[chamalib@gmail.com](mailto:chamalib@gmail.com)

**Introduction:** Primary bone lymphomas (PBL) account for <1% of all non-Hodgkin lymphomas (NHL). Anaplastic large cell lymphoma (ALCL) of bone is rare. This is a case of ALK (-) ALCL affecting the ischium.

**Case Report:** A 17-year-old girl presented with a pelvic mass. MRI revealed an expansile lytic bony lesion involving the right ischium measuring 9.8x5.8x4.2 cm, with the possibility of Ewing sarcoma or soft tissue sarcoma. Biopsies showed aggregates and singly scattered medium to large, atypical cells containing irregular vesicular nuclei with coarse chromatin and small nucleoli, cells resembling hallmark cells and wreath-like giant cells in an inflamed oedematous background. Neoplastic cells expressed diffuse, strong positivity for CD4, CD5, EMA, CD30, Granzyme B, and weak positivity for CD2 and CD99. ALK, SMA, CD3, PAX-5, LCA, AE1/AE3, PLAP, Desmin, MyoD1 and WT-1 were negative. A diagnosis of ALK (-) ALCL was made. There was no systemic involvement. Bone marrow biopsy was normal. The patient responded to treatment well.

**Discussion:** The negativity for desmin/MyoD1 and PLAP/PanCK excluded alveolar rhabdomyosarcoma and embryonal carcinoma, respectively. Negativity for WT-1 ruled out CIC rearranged sarcoma. Negativity for ALK, positivity for EMA, CD30 and T cell markers excluded epithelioid inflammatory myofibroblastic sarcoma. The presence of atypical large cells, including hallmark cells, and diffuse strong expression of CD30, EMA, Granzyme B, CD4, and CD5 helped confirm the diagnosis of ALK-ALCL. If facilities are available, cytogenetics will be useful to confirm the absence of ALK gene rearrangement. Irrespective of the ALK status, primary bone ALCLs have poorer outcomes compared to their nodal counterparts.

**Conclusion:** A higher degree of suspicion and appropriate selection of immunohistochemical stains is essential to make a diagnosis of ALCL at uncommon sites like bone.

**Keywords:** ALK, anaplastic large cell lymphoma, bone, Ewing sarcoma

## CR 10

### Myoepithelial carcinoma of the sinonasal tract: a rare tumour with potential for misdiagnosis

C. M. Baranasuriya<sup>1\*</sup>, H. D. Wijesinghe<sup>1</sup>, B.Y. Hettiarachchi<sup>1</sup>, I. Kumarasinghe<sup>2</sup>, M. V. C. de Silva<sup>1</sup>

<sup>1</sup>Centre for Diagnosis and Research in Cancer (CeDARC), University of Colombo, Sri Lanka

<sup>1</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup>Department of Pathology, University Hospital KDU - Kotelawala Defence University, Sri Lanka

[chamalib@gmail.com](mailto:chamalib@gmail.com)

**Introduction:** Myoepithelial carcinoma (MC) of the head and neck region is very rare, with a high rate of recurrence and metastasis, accounting for less than 1% of malignant salivary gland tumours in this region.

**Case report:** A 37-year-old man presented with change of voice and nasal mass for three months. MRI scan showed a mass lesion filling the right nasal cavity with extension into the right maxillary sinus, nasopharynx and right orbit. Endoscopic excision of the tumour was done. Microscopically, the lesion showed an infiltrating lobular tumour composed of anastomosing cords, nests, short fascicles of tumour cells intermingled with pseudovascular spaces and hyalinized bands throughout the lesion. The epithelioid to spindled tumour cells showed mildly pleomorphic hyperchromatic nuclei and moderate eosinophilic cytoplasm. The mitotic count was 16/2 mm<sup>2</sup>. Focal tumour necrosis was evident. The tumour cells showed diffuse and strong positivity for SMA, S100 and PanCK and stained negatively with CD34 and CD31. The Ki-67 proliferative index was 60%. A diagnosis of MC was made.

**Discussion:** The presence of pseudovascular spaces raised the differential diagnosis of malignant vascular tumours such as epithelioid haemangioendothelioma (EHE) and angiosarcoma (AS). Due to the nested architecture, clear cell sarcoma (CCS) needed to be excluded. The negativity for CD31 and CD34 excluded EHE and AS. Strong and diffuse positivity for SMA and PanCK excluded CCS. The neoplastic cells can show variable expression of calponin, GFAP, SOX10, p63, p40 and PLAG1. EWSR1 gene rearrangement is identified in clear cell and rhabdoid variants of MC in the salivary gland.

**Conclusion:** Awareness of the presence of different architectural patterns, including pseudovascular spaces in MC, as observed in our case, is important to avoid misdiagnosis of this rare tumour, especially at an unusual site like the nasal cavity.

**Keywords:** myoepithelial carcinoma, sinonasal tract, pseudovascular space

## CR 11

### Scirrhus hepatocellular carcinoma: diagnostic dilemma of a CK7 positive, HepPar1 negative hepatoid-looking carcinoma

C. M. Baranasuriya<sup>1,2\*</sup>, H. D. Wijesinghe<sup>1,2</sup>, K.K.S. Lakshika<sup>2</sup>, L.A.H.A. Albert<sup>3</sup>, M.V.C. de Silva<sup>1,2</sup>

<sup>1</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup>Centre for Diagnosis and Research in Cancer (CeDARC), University of Colombo, Sri Lanka

<sup>3</sup>Department of Pathology, Teaching Hospital Jaffna, Sri Lanka

[chamalib@gmail.com](mailto:chamalib@gmail.com)

**Introduction:** Scirrhus hepatocellular carcinoma (s-HCC) is a rare subtype of hepatocellular carcinoma (HCC), accounting for approximately 4% of all HCCs.

**Case report:** A 51-year-old woman presented with upper abdominal pain, loss of weight and appetite. Contrast-enhanced CT abdomen showed a hypodense mass measuring 10.5 cm in a non-cirrhotic background, suggestive of an intrahepatic cholangiocarcinoma (IC). Serum tumour markers were unavailable. Core needle biopsies revealed thick trabeculae, cords and small clusters of hepatoid-like large polygonal cells with mildly pleomorphic round nuclei and abundant eosinophilic cytoplasm. The surrounding stroma showed dense fibrosis. The neoplastic cells showed moderate to strong positivity for CK7, strong diffuse cell membranous positivity for Glypican-3, and HepPar1 was negative. A diagnosis of s-HCC was made. The patient was referred to an oncology unit for further management.

**Discussion and conclusion:** The differential diagnosis for the above clinical and immunomorphological features included IC and the fibrolamellar variant of HCC (F-HCC). Although the radiological appearance and immunophenotype (CK7+/ HepPar1) favoured cholangiocarcinoma, the absence of glands, mucin and positivity for glypican-3 excluded IC. F-HCC occurs primarily in the young and shows uniform positivity for HepPar1. 90% of s-HCC show arginase-1 positivity. Potential diagnostic pitfalls of s-HCC include radiological appearance similar to cholangiocarcinoma, normal serum AFP levels in 30%, occurrence in cirrhotic and non-cirrhotic liver, showing CK7, CK19 and EpCAM positivity and negativity for HepPar1. The prognosis of s-HCC is variable compared to conventional HCC. Awareness of this rare HCC subtype and knowledge that it is CK7 positive and HepPar1 negative should prompt staining with glypican-3 to avoid misdiagnosis as an IC.

**Keywords:** scirrhus hepatocellular carcinoma, cholangiocarcinoma, CK7, glypican-3



**CR 12**

**Acute lymphoblastic lymphoma with unusual microscopic morphology in small biopsies: a potential pitfall**

C. M. Baranasuriya<sup>1,2\*</sup>, S. A. Abeygunawardhane<sup>1,2</sup>, H. D. Wijesinghe<sup>1,2</sup>, K.K.S. Lakshika<sup>2</sup>,  
S. Nanayakkara<sup>3</sup>, V. Gupta<sup>4</sup>, M. V. C. de Silva<sup>1,2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>2</sup>*Centre for Diagnosis and Research in Cancer (CeDARC), University of Colombo, Sri Lanka*

<sup>3</sup>*Department of Pathology, National Eye Hospital, Sri Lanka*

<sup>4</sup>*Asiri Surgical Hospital, Sri Lanka*

[chamalib@gmail.com](mailto:chamalib@gmail.com)

**Introduction:** Acute lymphoblastic lymphoma/leukaemia (ALL), the most common childhood malignancy, is known to have a characteristic morphological appearance. We report two cases of ALL with unusual morphologies, with a high potential for misdiagnosis.

**Case report:**

**Patient A:** A 24-year-old man presented with worsening chest pain and shortness of breath. The contrast-enhanced CT scan of the chest showed a large anterior mediastinal mass encasing the mediastinal vessels. Biopsies showed sheets of small lymphocytes with hyperchromatic nuclei and scanty cytoplasm. Mitotic figures were inconspicuous. Tingible-body macrophages (TBM) were not seen. The neoplastic cells showed diffuse, strong positivity for CD3 and TdT. The Ki-67 proliferative index was 90%. PanCK-staining epithelial cells were absent. A diagnosis of T-ALL was made.

**Patient B:** A one and half month-old-baby boy presented with left eye proptosis. The MRI scan showed thickening of the left globe suggestive of lymphoma/orbital neoplasm. Biopsies showed sheets of monomorphic, small round cells containing hyperchromatic nuclei and scant cytoplasm. Mitoses were indistinct. TBM were not seen. The neoplastic cells showed diffuse, strong positivity for LCA and TdT. CD3 and CD20 showed positivity in scattered cells. The Ki-67 proliferative index was 50%. A diagnosis of ALL was made.

**Discussion and conclusion:** The usual morphology of ALL includes the presence of medium-sized lymphoid cells containing round to irregular nuclei with stippled chromatin and nuclear convolutions, frequent mitoses and TBM. These two cases showed an unusual morphology comprising small cells resembling mature lymphocytes without stippled chromatin and nuclear convolutions, inconspicuous mitoses and absent TBM. Knowledge of unusual morphologies of ALL and prompt confirmation with TdT will enable life-saving early treatment.

**Keywords:** acute lymphoblastic lymphoma, mature lymphocytes, TdT

## CR 13

### Focal nodular hyperplasia: an uncommon liver lesion in a child

D.M.T Chandramali\*<sup>1</sup>, D. H. Beneragama<sup>1</sup>, M. Nandasena<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

<sup>2</sup>*Department of Surgery, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

[tharanga.dissanayake88@gmail.com](mailto:tharanga.dissanayake88@gmail.com)

**Introduction:** Focal nodular hyperplasia (FNH) is a mass-forming, benign liver lesion commonly seen in adults, with female preponderance. FNH of the liver in the paediatric age group is extremely uncommon and rarely reported.

**Case report:** A nine-year-old girl presented with a right hypochondrial mass. The ultrasound scan revealed a tumour arising from the liver. The contrast-enhanced CT scan of the abdomen showed an exophytic liver mass. The radiological differential diagnoses included FNH, giant haemangioma, fibrolamellar carcinoma and hepatoblastoma. A non-anatomical liver resection was performed. Macroscopically, there was a well-delineated, lobulated, pale lesion with a maximum diameter of 10 cm with a central stellate scar. Histologically, it was a well-defined, non-encapsulated, subcapsular lesion composed of nodules of hepatocytes arranged in plates of 1-2 cell thickness. The central scar and fibrous septa were traversed by dystrophic, arterial-type vessels unaccompanied by portal veins. The reticulin framework was intact within the lesion. Glypican-3, performed to exclude a well-differentiated hepatocellular carcinoma, was negative throughout the lesion. Glutamine synthetase was not available in the local setting.

**Discussion and conclusion:** FNH comprises only 2% of paediatric hepatic tumours, and it is essential to differentiate this from more common hepatic tumours in this population. Map-like glutamine synthetase staining is characteristic of FNH and helps differentiate it from hepatocellular adenoma. Careful clinical, macroscopic and microscopic assessment, along with immunohistochemistry, is crucial to arrive at the correct diagnosis. Most cases of FNH are managed conservatively, but surgery is indicated for large and symptomatic lesions, as in this case.

**Keywords:** focal nodular hyperplasia, hepatic tumours

## CR 14

### Duodenal teratoma: a rare case

D.M.T. Chandramali<sup>1\*</sup>, D. H. Beneragama<sup>1</sup>, P. Kumarasinghe<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura,*

<sup>2</sup>*Hepatobiliary Surgical Unit, National Hospital of Sri Lanka*

[tharanga.dissanayake88@gmail.com](mailto:tharanga.dissanayake88@gmail.com)

**Introduction:** Teratoma is a benign neoplasm composed of mature tissue representing at least two embryonic layers: ectoderm, mesoderm or endoderm. Teratomas usually arise from germ cells in the gonads and midline structures of children and young adults. The gastrointestinal tract is a rare site for extragonadal teratoma, and duodenal teratoma is extremely rare.

**Case report:** A 41-year-old woman presented with bloating and upper abdominal discomfort. On examination, she was found to have an abdominal mass, and the CECT scan revealed a cystic lesion arising from the third part of the duodenum with the possibility of a teratoma. Resection of duodenal teratoma, ROUX-EN-Y duodenojejunostomy and gastrojejunostomy was performed. The resected small bowel segment was 110 mm in length. One end of the bowel was yellow and thickened, with part of the cyst wall measuring 50x30x20 mm attached to the bowel. The separate part of the cystic mass measured 80x70x20 mm and contained sebaceous material, hair and bony tissue. Microscopically, the cyst was lined by a stratified squamous epithelium, and the wall contained adnexal structures, mature adipose tissue, glandular structures, smooth muscles, bone and bone marrow. Thickened areas of the bowel wall showed features of fat necrosis. There were no immature elements or evidence of somatic malignancy.

**Discussion:** Duodenal teratoma is an extremely rare tumour in adults, with only a few reported cases in the literature. The treatment of choice is complete surgical excision.

**Keywords:** duodenal teratoma, extragonadal teratoma

## CR 15

### **Widespread haemorrhage and plasma leakage into Bowman's space: a rare renal manifestation in dengue hemorrhagic fever**

L.D.S. De Silva<sup>1\*</sup>, M.V.S. Amarangani<sup>1</sup>, S. Wijetunge<sup>1</sup>, D.P.P. Senasinghe<sup>2</sup>, T. Jayamaha<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Department of Pathology, Teaching Hospital, Peradeniya, Sri Lanka*

[shashids84@gmail.com](mailto:shashids84@gmail.com)

**Introduction:** Dengue haemorrhagic fever (DHF) and related deaths have significantly increased in Sri Lanka over the last few years. Most renal manifestations in DHF are due to “expanded dengue syndrome”, described by the WHO. Primary renal haemorrhages and plasma leakage (HPL) are rare complications, with less than 100 published case reports. This is a postmortem case report of DHF with widespread HPL into Bowman's space.

**Case report:** A 60-year-old man was admitted to the ward with high-grade fever and managed as DHF following positive serological results. Despite intensive treatments, he developed multiorgan failure with sepsis and died after three days. The postmortem examination revealed haemorrhages in multiple organs, including the lungs, subendocardium, and peritoneal and pleural cavities. The microscopy revealed extensive intra-alveolar haemorrhage and focal hepatocytic necrosis associated with steatosis and significant lymphocytic infiltration, consistent with DHF-related changes. Macroscopically, the kidneys were pale with no obvious haemorrhagic manifestations. However, on microscopic examination, there was widespread HPL in Bowman's space with red cell casts in tubules. There were no other renal pathologies.

**Discussion and conclusion:** In DHF, acute kidney injury is most commonly due to acute tubular injury secondary to hypotension, myoglobinuria or haemolysis. Limited data is available on primary renal involvement, and most of these were glomerulonephritides with mesangial proliferation due to dengue immunocomplex deposition. Even though there is an increased risk of HPL in DHF, it is site-selective and transient, and widespread HPL in Bowman's space is described rarely in the literature. Reporting unusual findings in DHF is important for future management decisions and the development of novel treatment modalities.

**Keywords:** DHF, Bowman space, plasma leakage, expanded dengue syndrome, glomerulonephritis

## CR 16

### Carcinosarcoma, a rare malignant biphasic tumour in the pancreas

M. M. N. Darshani\*, T. G. Liyanage

*Department of Pathology, Faculty of Medicine, University of Ruhuna, Karapitiya, Sri Lanka*

[darshanimalwatta@gmail.com](mailto:darshanimalwatta@gmail.com)

**Introduction:** Carcinosarcoma is a rare malignant biphasic neoplasm containing carcinomatous and sarcomatous components with a predilection for the female genital tract, especially the uterus. It is an extremely rare tumour in the pancreas.

**Case report:** A 65-year-old woman presented with epigastric pain for one month. USS revealed a mass in the tail of the pancreas. USS-guided fine needle aspiration cytology was reported as adenocarcinoma. USS-guided core biopsy of the mass was reported as a malignant spindle cell lesion. A distal pancreatectomy was performed. Macroscopy revealed a partially circumscribed mass in the tail of the pancreas measuring 10x8x6 cm. It showed solid and cystic cut surfaces with necrotic and haemorrhagic areas. Microscopic examination revealed a malignant biphasic tumour with a predominant sarcomatous component comprising atypical spindle cells arranged in vague fascicles. The carcinomatous component showed neoplastic glands and multilayered papillary structures. Atypical mitoses and necrosis were noted within both components. Carcinomatous and sarcomatous components showed positive cytoplasmic staining with pan-cytokeratin and vimentin, respectively.

**Discussion and conclusion:** Carcinosarcoma of the pancreas is a rare neoplasm having a dismal prognosis. Morphological assessment and immunohistochemical evaluation are necessary to differentiate it from poorly differentiated or undifferentiated carcinomas, including sarcomatoid carcinomas. Biphasic synovial sarcoma and other malignant spindle cell lesions should also be excluded.

**Keywords:** carcinosarcoma, biphasic, carcinomatous, sarcomatous

## CR 17

### Hepatic amyloidosis: a case report

M. M. N. Darshani\*, H. A. S. Perera

*Department of Pathology, Teaching Hospital Karapitiya, Galle, Sri Lanka*

[darshanimalwatta@gmail.com](mailto:darshanimalwatta@gmail.com)

**Introduction:** Amyloidosis is a rare disorder characterized by extracellular deposition of non-branching fibrillary proteins. It can involve any organ in isolation or in conjunction with other organs. Hepatic involvement in amyloidosis is common in patients with systemic amyloidosis, but isolated hepatic involvement is rare.

**Case report:** A 41-year-old man presented with lower limb swelling and ascites of two weeks duration. Ultrasound scan revealed hepatomegaly. Alkaline phosphatase was mildly elevated, but liver transaminases were normal. Hepatitis viral serological tests were negative. Microscopic examination of the liver core biopsy revealed distorted lobular architecture with vague nodule formation. Perisinusoidal deposition of extracellular amorphous acellular eosinophilic material was identified. Atrophy of liver cell plates and sinusoidal narrowing were present. Some of the vessels in the portal tracts showed thickened walls with evidence of deposition of similar material. Hepatic amyloidosis was diagnosed as this material-stained orange-red with Congo red stain and showed apple green birefringence on polarized microscopy. The clinical impression was of isolated hepatic amyloidosis as there was no evidence of other organ involvement or evidence of secondary amyloidosis due to chronic inflammatory conditions. Serum electrophoresis and bone marrow biopsy were planned to rule out primary amyloidosis, but the patient defaulted to follow-up.

**Discussion:** Congo red negative amyloid-like material within the liver can be seen in Waldenstrom macroglobulinemia and light chain deposition disease. Congo red special stain performed on 10 µm thick sections is the gold standard for diagnosis of amyloidosis. It is important to classify amyloidosis as primary or secondary and as localized or systemic as it affects the management.

**Keywords:** hepatic, amyloidosis, Congo red



## CR 18

### Warthin-like papillary thyroid carcinoma: a rare tumour

M. M. N. Darshani\*, T. W. Wijesiri

*Department of Pathology, Faculty of Medicine, University of Ruhuna, Karapitiya, Sri Lanka*

[darshanimalwatta@gmail.com](mailto:darshanimalwatta@gmail.com)

**Introduction:** Papillary thyroid carcinoma (PTC) is the most common endocrine malignancy in both the adult and paediatric populations. Warthin-like variant of papillary thyroid carcinoma (WL-PTC) is a rare subtype which shares morphologic similarities with the Warthin tumour of the salivary gland.

**Case report:** A 57-year-old woman underwent a completion right thyroid lobectomy as she was diagnosed to have a PTC of conventional type in the left lobe. Macroscopic examination of the right thyroid lobe showed a whitish lesion measuring 4x4x3 mm. Microscopic examination revealed a tumour composed of papillae with fibrovascular cores lined by oncocyctic cells. The nuclei showed enlargement, nuclear clearing, nuclear grooves and intranuclear pseudoinclusions. The papillary cores were filled with a dense lymphoid cell infiltrate. The background thyroid showed patchy chronic autoimmune thyroiditis. Lymphovascular emboli, extrathyroidal extension or nodal metastasis were not identified. It was concluded as Warthin-like papillary microcarcinoma (WLPMC) as it showed the typical histological features of this variant.

**Discussion and conclusion:** WL-PTC is considered to have an excellent long-term prognosis similar to low-risk PTC. There is limited published data on the prognosis of the various histological subtypes of papillary microcarcinoma. However, a recent study showed that WLPMC falls into the non-aggressive category of papillary microcarcinoma which also includes conventional, follicular, oncocyctic and cystic subtypes. It showed WLPMC has a better prognosis than the aggressive subtypes of papillary microcarcinoma which include diffuse sclerosing, solid, tall cell, columnar cell and hobnail variants, as it shows less tendency for extrathyroidal extension and nodal metastasis.

**Keywords:** Warthin-like PTC, oncocyctic cells, lymphoid stroma

## CR 19

### Peptic duodenitis with gastric foveolar metaplasia presenting as duodenal polyps

K. Dharani\*, T.P.M. Bopagoda

*Faculty of Medicine, University of Kelaniya, Sri Lanka*

[dhara\\_ni\\_ghs@yahoo.com](mailto:dhara_ni_ghs@yahoo.com)

**Introduction:** Gastric foveolar metaplasia (GFM) of the duodenum is associated with mucosal healing in peptic duodenitis (PD) caused by *Helicobacter pylori* infection or NSAID use. It is known to be associated with GNAS and KRAS mutations that are associated with different tumours. These usually present as mucosal swelling, erythema, erosions and ulcerations. Rarely, as in this case, PD with GFM presents as endoscopically recognizable duodenal polyps.

**Case report:** A 71-year-old man presented with gastroesophageal reflux disease and jaundice for one-week duration. Ultrasound scan showed chronic calculous cholecystitis. Endoscopy revealed multiple polyps in the duodenum (D1), and the biopsy showed a polypoidal duodenal mucosa with intact crypt-villous architecture and marked foveolar metaplasia of the surface epithelium highlighted with PAS stain. The lamina propria was expanded with increased chronic inflammatory cells and scattered neutrophils. There was marked Brunner gland hyperplasia with glands extending above the muscularis mucosa. No *Helicobacter pylori*, dysplasia or malignancy was seen. This was concluded as PD.

**Discussion and conclusion:** The presentation of PD with GFM as a duodenal polyp is a recently recognized, poorly understood entity with limited literature. It has been proposed that Brunner gland hyperplasia associated with GFM is the cause for the polypoid nature of these lesions, thus indirectly relating to GFM. The endoscopic findings of these polyps are heterogeneous, with no specific features to distinguish them from other lesions, especially adenomatous polyps. This case emphasizes the importance of considering PD with FGM as a differential diagnosis in patients presenting with duodenal polyps.

**Keywords:** peptic duodenitis, gastric foveolar metaplasia, duodenal polyp, histopathology, endoscopy

**CR 20**

**Carcinosarcoma of the ovary associated with bilateral serous cystadenofibromas: an unusual presentation**

K. Dharani\*, T.P.M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[dharani\\_ghs@yahoo.com](mailto:dharani_ghs@yahoo.com)

**Introduction:** Carcinosarcoma, a biphasic malignancy with both high-grade carcinomatous and sarcomatous components, accounts for only 2% of all ovarian malignancies. This is a unique case of carcinosarcoma associated with bilateral serous cystadenofibromas.

**Case report:** A 48-year-old woman presented with recurrent lower abdominal pain of three years and was found to have a left adnexal mass on an ultrasound scan. She underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy and infracolic omentectomy. Macroscopy of the left adnexal mass revealed irregular tissue pieces with solid, cystic, friable, white and gelatinous areas that collectively measured 150x120x70mm. The right ovary showed a unilocular cyst measuring 20 mm in diameter. Microscopy of the left adnexal mass revealed a carcinosarcoma comprising highly atypical cells forming tubular and cribriform glands with occasional papillae and atypical spindle cells. Brisk mitotic activity and foci of tumour necrosis were present in both components. Heterologous malignant elements were not seen. The cystic areas showed features of serous cystadenofibroma with intervening focal areas of borderline serous tumour (BST). The right ovary showed a serous cystadenofibroma with focal epithelial proliferation. The uterus, right fallopian tube and omentum were free of tumour. The peritoneal washing was negative for malignant cells.

**Discussion and conclusion:** Primary ovarian carcinosarcoma is an aggressive tumour with a dismal prognosis and sparse literature due to its rarity. In this case, the presence of bilateral serous cystadenofibromas and transition into carcinosarcoma with intervening areas of BST raised the possibility of carcinosarcoma arising from a serous cystadenofibroma. No such cases have been reported previously, and the aetiology or precursor lesions for carcinosarcoma have not yet been identified. Therefore, further research and meticulous recording of such cases are warranted for a better understanding of this rare tumour.

**Keywords:** carcinosarcoma, serous cystadenofibroma, ovary, histopathology

CR 21

**Xanthogranulomatous cholecystitis mimicking carcinoma of the gallbladder: a case report**

K. Dharani\*, L. I. Madaluwage, S.K. Liyanage

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[ddharani\\_ghs@yahoo.com](mailto:ddharani_ghs@yahoo.com)

**Introduction:** Xanthogranulomatous cholecystitis (XGC) is an uncommon form of chronic cholecystitis with an incidence of 0.7-1.8% of excised gallbladders. It has been reported that around 20-30% of pathologically diagnosed XGC can be misdiagnosed as gallbladder carcinoma (GBC) based on preoperative radiographs and intraoperative findings, as the inflammation infiltrates the adjacent organs.

**Case report:** A 62-year-old man with diabetes mellitus presented with right hypochondriac pain for three weeks. CECT appearance favoured gallbladder carcinoma with evidence of infiltration into the adjacent liver tissue and regional lymph node metastasis. The CA 19-9 level was 6.5 U/mL. The radical cholecystectomy specimen included the gallbladder with an attached liver segment and regional lymph nodes. Macroscopically, the gallbladder wall of the fundus showed a yellowish-brown lesion with a central cystic area extending into the hepatic bed, measuring 28x15x15 mm. There were no intraluminal lesions. Microscopy of the lesion showed sheets of foamy histiocytes admixed with lymphocytes and granulation tissue with fibrosis that extended into the adjacent fatty tissue and the liver parenchyma with focal hepatocellular damage. These histomorphological features were in keeping with XGC. The background gallbladder showed evidence of chronic cholecystitis and adenomyomatosis. There was no dysplasia or malignancy. The lymph nodes showed no significant pathology.

**Discussion and conclusion:** XGC is possibly a reaction to bile within the gallbladder wall. Histological assessment is mandatory for the correct diagnosis. Surgical treatment for XGC is challenging, and complete cholecystectomy is preferred because of the rare risk of coexisting carcinoma.

**Keywords:** gallbladder, xanthogranulomatous cholecystitis, carcinoma

CR 22

### **Sinonasal undifferentiated carcinoma: a rare case**

A. W. K. Dilrukshi\*, H. A. S. Perera

*Department of Pathology, Teaching Hospital, Karapitiya, Sri Lanka*

[kaushikadilrukshia@gmail.com](mailto:kaushikadilrukshia@gmail.com)

**Introduction:** Sinonasal undifferentiated carcinoma (SNUC) is a rare tumour accounting for 3-5% of all sinonasal carcinomas. SNUC commonly affects males, and the median age of presentation is in the sixth decade. These tumours frequently arise in the nasal cavity and paranasal sinuses, and large masses involve multiple sites. This is a rare case of SNUC in an elderly woman.

**Case report:** A 73-year-old woman presented with a growth in the right nasal cavity. CT scan revealed a large, polypoidal mass in the right nasal cavity extending into the right ethmoid and sphenoid sinuses with destruction of the right orbit. The biopsy from the nasal growth showed a poorly differentiated malignancy composed of sheets of tumour cells having enlarged, pleomorphic, vesicular nuclei with prominent nucleoli, scanty cytoplasm and brisk mitotic activity. Large areas of tumour necrosis were noted. There was no evidence of glandular or squamous differentiation, rosette formation or a fibrillary matrix. The tumour cells showed strong positivity for immunohistochemical stains AE1/AE3 and CK7 and negative for CD3, CD20, S100, synaptophysin and CK5/6.

**Discussion and conclusion:** The differential diagnosis of an undifferentiated tumour of the sinonasal region includes SNUC, nasopharyngeal undifferentiated carcinoma (NPUC), neuroendocrine carcinoma, olfactory neuroblastoma and lymphoma. NPUC often has a prominent lymphoplasmacytic infiltrate and is positive for CK5/6. Neuroendocrine carcinoma and olfactory neuroblastoma show consistent positivity for neuroendocrine markers. Negative staining for CD3 and CD20 excludes lymphoma. Diagnosis of SNUC by morphological features alone is difficult and correlation with clinical and immunohistochemical findings is necessary to differentiate from other poorly differentiated tumours.

**Keywords:** sinonasal undifferentiated carcinoma, poorly differentiated malignancy, nasal cavity

**CR 23**

**A hybrid tumour with schwannoma and meningioma in a patient with neurofibromatosis type 2**

M. K. S. Amarawickrama\*, K. Dissanayake

*National Hospital of Sri Lanka*

[ksandamaliama@gmail.com](mailto:ksandamaliama@gmail.com)

**Introduction:** Neurofibromatosis type 2 (NFT-2) is an autosomal dominant disease, and these patients present with multiple intracranial and intraspinal neoplasms. Around 50% of NFT-2 patients develop meningiomas, and around 30% develop schwannomas. However, the coexistence of meningioma and schwannoma is extremely rare, with around ten documented cases in the literature.

**Case report:** A 45-year-old woman diagnosed with NFT-2 presented with right-side hearing impairment and left-side facial numbness. MRI shows multiple dural-based extra-axial lesions in the posterior fossa along the falx cerebri and over cerebral convexities. During surgery, a hybrid tumour was suspected as the tumour was infiltrating the brain tissue. The resected CP angle tumour, measuring 55x50x10 mm, was sent as multiple fragments for histological assessment. The tumour was composed of two histological components. One area contained whorls, fascicles and bundles of meningotheial cells with multiple psammoma bodies. Constituent cells showed positivity for EMA and S100, compatible with transitional meningioma (WHO grade 1). The other part of the tumour contained hypercellular and hypocellular areas: Antoni A areas with Verocay bodies and Antoni B areas. The constituent cells showed strong positivity for S100 and negative for EMA. These features were compatible with schwannoma.

**Discussion and conclusion:** NFT-2 is characterized by acoustic schwannomas, meningiomas, astrocytomas and extracranial schwannomas. Transitional meningiomas share the features of frequent 22q deletions and NF2 mutations. Also, NF2 mutation is a precursor neoplastic lesion for Schwann cells, commonly presenting with plexiform schwannoma. Surgical excision is the treatment for both tumours. When surgical excision is impossible, radiosurgery is the preferred treatment modality.

**Keywords:** schwannoma, neurofibromatosis type 2, meningioma



## CR 24

### Adrenal cortical oncocytic tumour with uncertain malignant potential

T. S. Dissanayake<sup>1</sup>, A. S. Rodrigo<sup>1</sup>, N. Senevirathne<sup>2</sup>

<sup>1</sup>*Department of Pathology, Sri Jayewardenepura General Hospital, Sri Lanka*

<sup>2</sup>*Department of Surgery, Sri Jayewardenepura General Hospital, Sri Lanka*

[sonalirodrigo@yahoo.com](mailto:sonalirodrigo@yahoo.com)

**Introduction:** Adrenal cortical tumours are rare. They are mostly non-functioning adenomas.

**Case report:** A 47-year-old woman presented with left loin pain for two weeks with no symptoms to suggest hormonal imbalance. CT showed a heterogeneously enhancing well-defined lesion in the left adrenal gland, displacing the kidney inferiorly. Adrenal hormone levels were normal. The tumour was excised with the entire adrenal gland. The entire specimen weighed 0.085kg. The tumour was round with a smooth outer surface, measuring 6.5x6.2x4 cm. Sectioning showed a tan-to-brown heterogeneous cut surface. Microscopy revealed a circumscribed, encapsulated tumour comprising clusters and nests of polygonal cells with mildly atypical nuclei and oncocytic cytoplasm. Capsular invasion was identified. Adjacent adrenal tissue was normal. Reticulin stain highlighted the preserved nested architecture of tumour cells. Calretinin was diffusely positive in tumour cells, confirming adrenal origin and excluding a renal oncocytic tumour. The Ki-67 proliferation index was 6%. The diagnosis of an adrenal cortical oncocytic tumour of uncertain malignant potential was made.

**Discussion:** Patients with adrenal cortical tumours present with abdominal mass, hormonal imbalances or incidentally. Weiss criteria are applied to differentiate adenoma from carcinoma. Specific Lin-Weiss-Bisceglia criteria are applied to subtype tumours with oncocytic morphology. Identifying the adrenal origin is important. The presence of capsular invasion and the borderline Ki-67 index were considered for this diagnosis.

**Conclusion:** Adrenal cortical oncocytic tumours are rare tumours needing strict histological criteria with clinical and endocrinological evaluation for diagnosis. Regular follow-up for the identification of potential malignant behaviour is mandatory.

**Keywords:** adrenal cortical tumour, oncocytic adenoma

**CR 25**

**Clear cell renal cell carcinoma with hemangioblastoma like features: a rare presentation**

T. S. Dissanayake<sup>1</sup>, N. Senevirathne<sup>2</sup>, A.S. Rodrigo<sup>1</sup>

<sup>1</sup>*Department of Pathology, Sri Jayewardenepura General Hospital, Sri Lanka*

<sup>2</sup>*Department of Urology, Sri Jayewardenepura General Hospital, Sri Lanka*

[sonalirodrigo@yahoo.com](mailto:sonalirodrigo@yahoo.com)

**Introduction:** Clear cell renal cell carcinoma (CCRCC) is the most common renal carcinoma with a range of histomorphological features.

**Case report:** A 64-year-old woman presented with gross haematuria. Imaging showed a heterogenous mass in the left kidney. A left radical nephrectomy was performed. Sectioning showed two well-circumscribed tumours abutting each other. The larger tumour had a variegated appearance. The smaller tumour was homogenous and tan-coloured. Microscopy of the larger tumour showed a CCRCC of ISUP grade 2 comprising alveolar-like areas and solid nests with cuboidal cells, pleomorphic nuclei and clear to granular cytoplasm. The smaller tumour showed a haemangioblastoma-like morphology with proliferation of vessels composed of thin intervening capillaries lined by small endothelial cells. The stroma was densely cellular with large, irregular-shaped cells with variable-size nuclei and prominent nucleoli. Pancytokeratin positivity was observed in both tumours, excluding the possibility of a coexisting hemangioblastoma. CD10 positivity in both tumours confirmed renal origin. A diagnosis of CCRCC with hemangioblastoma-like features was made.

**Discussion and conclusion:** The morphological diversity of CCRCC is well known, and a haemangioblastoma-like variant is described. CCRCC should be differentiated when coexisting with other renal tumours, especially if the other tumour is benign. Although macroscopic and microscopic features indicated coexisting tumours of different origins, immunohistochemistry was needed to confirm the diagnosis in this case. Immunohistochemistry is mandatory in the diagnosis of renal tumours of different morphology, which is important for staging, prognostication and management.

**Keywords:** clear cell renal cell carcinoma, hemangioblastoma, pancytokeratin, CD10

## CR 26

### **Rare occurrence of a cellular angiofibroma in an elderly female mimicking a vaginal vault prolapse**

R. M. M. L. Ekanayake\*, R. Waduge

*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[manoeka249@gmail.com](mailto:manoeka249@gmail.com)

**Introduction:** Cellular angiofibroma is a rare benign fibroblastic neoplasm, usually found in the vulvovaginal region of middle-aged women. Its occurrence at the vaginal vault in an elderly female with post-vaginal hysterectomy is an extremely rare presentation.

**Case report:** A 79-year-old woman presented with a painless lump at vulva for four months. She had a history of vaginal hysterectomy 30 years back. Speculum examination and imaging revealed a posterior vaginal wall cyst measuring 5x3 cm, which was excised. The specimen was received as multiple whitish tissue fragments together measuring 2x1x1 cm. Microscopy revealed a spindle cell neoplasm comprising randomly distributed uniform, plump spindle cells in an oedematous and collagenous stroma. There were small to medium-sized thick-walled proliferating blood vessels, scattered mast cells and small aggregates of adipocytes. Mitoses were sparse. Tumour cells showed diffuse positivity for ER and PR. Desmin was negative. The Ki-67 index was 8%. These features were compatible with angiofibroma.

**Discussion and conclusion:** The clinicopathological differential diagnoses for vaginal vault lump following hysterectomy include vaginal vault prolapse, inclusion cysts, other vulval mesenchymal neoplasms and vulva carcinomas. Histomorphological and immunohistochemical evaluation is necessary for diagnosing and distinguishing cellular angiofibroma from other mesenchymal lesions of the vulva. Local recurrence is infrequent in cellular angiofibroma and sarcomatous transformation has been described rarely.

**Keywords:** angiofibroma, desmin, hysterectomy, vulvovaginal

## CR 27

### Mixed epithelial and stromal tumour of the kidney

R. M. M. L. Ekanayake\*, A. Vithanage, R.M.P. Rathnayake

*Department of Histopathology, National Hospital, Kandy, Sri Lanka*

manoeka249@gmail.com

**Introduction:** Mixed epithelial and stromal tumour is a rare biphasic renal neoplasm comprising both epithelial and mesenchymal elements. It occurs predominantly in perimenopausal women with a history of long-term hormone therapy.

**Case report:** A 42-year-old woman presented with left abdominal pain of two months duration. She denied any history of hormonal therapy. Clinical examination was unremarkable. The CT scan revealed a complex, cystic left renal lesion, classified as Bosniak type III. She underwent a partial nephrectomy. Macroscopic examination of the specimen showed a multilocular, complex cystic lesion with thin and thick septa measuring 50x30x20 mm. These cysts contained clear fluid. Microscopy revealed variable-sized cysts lined by cuboidal to columnar epithelial cells with focal hobnail configuration. The stroma was moderately cellular and showed a proliferation of bland, plump spindle cells and thick-walled blood vessels. The adjacent renal tissue was unremarkable. The epithelial cells were positive for pancytokeratin. The stromal cells were positive for ER, PR, SMA and desmin. HMB45 and melan A were negative. Accordingly, a diagnosis of mixed epithelial and stromal tumour was made.

**Discussion and conclusion:** Differential diagnoses of renal mixed epithelial and stromal tumour include adult cystic nephroma, multilocular cystic renal neoplasm of low malignant potential and angiomyolipoma with epithelial cysts. The presence of a solid cellular stroma, positivity for ER and PR and negativity for HMB45 and melan A are helpful immunomorphological features to distinguish it from the other differential diagnoses. The majority of mixed epithelial and stromal tumours are benign and have a good prognosis. Rare cases of malignant transformation have been reported.

**Keywords:** Bosniak, kidney, tumour, mixed epithelial and stromal

## CR 28

### **Cribriform morular thyroid carcinoma in association with familial adenomatous polyposis: a rare occurrence**

A. Ekanayaka\*

*Department of Pathology, Countess of Chester Hospital, NHS Foundation Trust, United Kingdom*

[anju.inesha@gmail.com](mailto:anju.inesha@gmail.com)

**Introduction:** Cribriform morular thyroid carcinoma (CMTC) is a rare malignant thyroid neoplasm which occurs predominantly in women. It is frequently associated with familial adenomatous polyposis (FAP) but can occur as a sporadic neoplasm.

**Case report:** An 18-year-old girl presented with an anterior neck swelling. Ultrasound scan showed a nodular goitre. Her family history was thought to be unremarkable at the time. Fine needle aspiration cytology revealed squamoid cells arranged singly and in small groups admixed with scattered thyroid epithelial cells. The differential diagnosis on cytology was a primary thyroid carcinoma or a secondary deposit of squamous carcinoma. Histology of the total thyroidectomy specimen revealed a multifocal tumour comprising well-formed papillae with widespread squamoid morules. Nuclear features of papillary thyroid carcinoma, neuroendocrine differentiation or high-grade nuclear features were not seen. A diagnosis of multifocal CMTC of both lobes was made. She was referred to the oncology clinic, where a subsequent colonoscopy showed multiple polyps. Three polyps were excised; two were tubular adenomas with high-grade dysplasia, and the third was a tubulovillous adenoma with a microinvasive carcinoma. Prophylactic colectomy was performed, and histology confirmed a moderately differentiated adenocarcinoma arising in a background of FAP. Colonoscopies performed in her mother and a maternal uncle also showed features of FAP. The patient was followed-up at the oncology clinic.

**Discussion:** CMTCs which are associated with FAP are multifocal and bilateral. Sporadic tumours are usually solitary. CMTC has an indolent clinical behaviour like classic papillary thyroid carcinoma. However, high-grade CMTC behaves aggressively with distant metastasis. CMTC occurs secondary to activation of the WNT/Beta-catenin pathway due to germline or sporadic mutations.

**Conclusion:** The diagnosis of CMTC should alert the clinician to the possibility of FAP and the need for family screening.

**Keywords:** cribriform-morular thyroid carcinoma, familial adenomatous polyposis

## CR 29

### Pleomorphic leiomyosarcoma of the tibia: a rare case

A. Ekanayaka<sup>1\*</sup>, L. Wijetunge<sup>2</sup>, R. Laxsan<sup>3</sup>

<sup>1</sup>Department of Pathology, Countess of Chester Hospital NHS Foundation Trust, Liverpool Road, Chester, UK

<sup>2</sup>Department of Pathology, District General Hospital, Chilaw, Sri Lanka

<sup>3</sup>Postgraduate Institute of Medicine, University of Colombo, Sri Lanka

[anju.inesha@gmail.com](mailto:anju.inesha@gmail.com)

**Introduction:** Pleomorphic leiomyosarcoma (PL) is a rare malignant tumour derived from smooth muscle cells, mainly in the uterus, gastrointestinal tract and soft tissue. Primary leiomyosarcoma of bone is exceedingly rare.

**Case report:** A 27-year-old man presented with a tibial fracture. Radiology revealed an aggressive cortical expansile lesion in the right proximal tibial diaphysis, raising the suspicion of adamantinoma. An excisional biopsy of the tibial tumour was performed, and the microscopy revealed a malignant spindle cell tumour infiltrating the surrounding soft tissue and skeletal muscle. Tumour cells exhibited marked pleomorphism with a brisk mitotic activity accounting for 37/10 high-power fields. No osteoid formation or epithelial component was present. The differential diagnoses were malignant peripheral nerve sheath tumour, undifferentiated pleomorphic sarcoma and PL. The immunohistochemistry demonstrated positivity for SMA and h-caldesmon and negativity for CD99, CK7, CD34 and S100. Desmin was repeatedly negative, which is a known finding in 20-30% of leiomyosarcomas. Following the full-body PET scan, the diagnosis of primary tibial PL was confirmed.

**Discussion:** Primary bone leiomyosarcoma is a challenging diagnosis and requires careful immunohistopathological analysis. Due to the rarity of this entity and the possibility of metastatic leiomyosarcoma from another primary site, it is crucial to consider a comprehensive diagnostic workup, including a full-body PET scan, to exclude other potential sources. The prognosis depends on the histological grade. High-grade lesions are highly aggressive with distant metastasis with a 5-year survival rate of <50%.

**Conclusion:** This case highlights the importance of distinguishing PL from other mimicking tumours, as their management and prognosis differ significantly.

**Keywords:** Pleomorphic leiomyosarcoma, bone tumour, immunohistochemistry, pathological fracture



**CR 30**

**Vacuolated cells on FNA in salivary neoplasms: a potential pitfall**

P. N. Managoda\*, S. Fernandopulle

*Department of Pathology, Colombo North Teaching Hospital, Ragama, Sri Lanka*

[piumi628@gmail.com](mailto:piumi628@gmail.com)

**Introduction:** Secretory carcinoma is a rare low-grade salivary gland carcinoma. Tumours with a prominent papillary cystic component have a possibility of being misdiagnosed as cyst contents during the initial cytological assessment.

**Case report:** A 50-year-old man presented with a painless, slow-growing right parotid mass for six months. The ultrasound scan was suggestive of pleomorphic adenoma. A 10 ml of fluid was aspirated during FNAC, which showed cyst macrophages and haemosiderin-laden macrophages. The macroscopy of the right side parotidectomy specimen showed a cystic lesion filled with gelatinous material. Solid projections were seen extending into the cyst cavity. Histology showed a cyst lined by a flat to cuboidal ciliated epithelium. The solid areas showed papillae, microcysts and tubules lined by hobnail cells. An extracellular and intracytoplasmic PAS-positive, PASD-resistant material was identified. Immunohistochemistry showed diffuse and strong positivity for mammaglobin and S100 in the lining cells, confirming the diagnosis of secretory carcinoma. The patient has been followed up in the clinic for five months and is well with no recurrences.

**Discussion and conclusion:** On review of the initial cytology, vacuolated tumour cells that were reported as macrophages in the initial assessment were identified. The tumour cells showed a larger nucleus with a conspicuous nucleolus and a denser cytoplasm, compared to macrophages. Acinic cell carcinoma with a papillary cystic component can give a similar appearance in cytology. Careful surveillance is crucial in reporting salivary gland lesions with cyst contents in cytology as neoplasms can be missed, including rare tumours like secretory carcinoma.

**Keywords:** secretory carcinoma, parotid gland

## CR 31

### Epithelioid leiomyosarcoma arising in a leiomyoma: a rare case

G. M. D. D. Galagedara\*, T. P. M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[dilushi.da@gmail.com](mailto:dilushi.da@gmail.com)

**Introduction:** Leiomyoma is the most prevalent tumour in the female genital tract, with a remote possibility of malignant transformation. A few cases of such transformation have been reported in the literature. This is an exceptional case of epithelioid leiomyosarcoma (ELS) arising from a leiomyoma.

**Case report:** A 60-year-old woman was found to have a fibroid uterus on an ultrasound scan while investigating for nonspecific urinary symptoms. She underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. Macroscopy of the uterus showed a well-circumscribed lesion completely filling the uterine cavity and measuring 100x80x80 mm. It showed a predominantly uniform, white and whorled cut surface. There was a friable area with haemorrhage adjacent to the uterine wall that measured 60x20x30 mm. Microscopy showed predominantly intersecting bundles of smooth muscle fibres in a collagenous stroma with no significant atypia or mitosis. The sections from the friable areas showed sheets of epithelioid cells with abundant clear cytoplasm in a stroma rich in thin curving blood vessels. These cells showed moderate nuclear pleomorphism and brisk mitoses of 12/10 HPF, including abnormal forms. In many foci, these areas of the tumour were juxtaposed with the benign spindle cell areas showing abrupt transition, while occasional foci showed spindle cells with cellular atypia, increased mitosis and occasional foci of coagulative tumour necrosis intermingled with these cellular epithelioid cell areas. Occasional foci showed a transition from benign spindle cells to atypical spindle cells to frank malignant epithelioid cells. The lesional cells diffusely expressed SMA and caldesmon in both the spindle cell and epithelioid areas and were negative for HMB45, EMA, and AE1/AE3. In the presence of all three diagnostic criteria for leiomyosarcoma and with immunohistochemical confirmation, the diagnosis of epithelioid leiomyosarcoma was established.

**Discussion and conclusion:** This case report highlights the extremely rare transformation of leiomyoma into epithelioid leiomyosarcoma and emphasizes the challenges associated with preoperative diagnosis. A comprehensive histopathological examination with careful attention to morphological variations is essential for accurate diagnosis and appropriate management.

**Keywords:** epithelioid, leiomyosarcoma, leiomyoma, transition

**CR 32**

**Pure, non-hilar, benign Leydig cell tumour of the ovary: a rare case presenting with virilization**

G. M. D. D. Galagedara\*, T. P. M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[dilushi.da@gmail.com](mailto:dilushi.da@gmail.com)

**Introduction:** Leydig cell tumour (LCT) is a rare, unilateral, benign steroid cell tumour primarily located in the ovarian hilum. It represents <0.5% of all ovarian tumours and typically presents with symptoms related to excess androgen secretion. This is a rare case of a pure, non-hilar, benign LCT leading to virilization.

**Case report:** A 54-year-old woman presented with voice change, alopecia, increased facial and genital hair growth, and increased libido for six years. Her elevated serum testosterone level was 12.9 ng/ml. CT scan of the abdomen revealed a solid left ovarian neoplasm. A bilateral salpingo-oophorectomy was performed, which showed a well-defined tumour measuring 40x30x25 mm. The cut surface was solid, white-grey, and yellow. The capsule was intact. Microscopy revealed a well-circumscribed tumour comprising lobules and sheets of cells separated by fibrous bands. The tumour cells were polygonal to round with abundant eosinophilic granular cytoplasm, uniform, round nuclei, prominent central nucleoli and more than one nucleolus. Few clusters and scattered cells with clear/foamy cytoplasm were seen. There was moderate nuclear pleomorphism. Scattered cells with enlarged, hyperchromatic bizarre nuclei were present. Many intracytoplasmic and rare extracellular elongated, eosinophilic Reinke crystals and scattered tumour cells with lipofuscin pigment were seen. Mitoses, necrosis or vascular invasion were not identified. A rim of normal ovarian tissue was present surrounding the tumour. The diagnosis of a non-hilar, benign, LCT was made based on the histology and clinical picture. Her symptoms declined over the ten months of the post-operative period.

**Discussion and conclusion:** Diagnosis of ovarian LCT relies on clinical history, hormone levels and histological characteristics. In this case, the presence of virilizing symptoms, elevated testosterone levels and distinctive histopathological features were helpful.

**Keywords:** Leydig cell tumour, virilizing symptoms

## CR 33

### Juvenile hyaline fibromatosis: a rare case

G. M. D. D. Galagedara\*, K. S. Maheshika, S. M. Fernandopulle

*Colombo North Teaching Hospital, Ragama, Sri Lanka*

[dilushi.da@gmail.com](mailto:dilushi.da@gmail.com)

**Introduction:** Juvenile hyaline fibromatosis (JHF) is a rare, autosomal recessive, inherited disorder that occurs mainly in children. It is characterized by papulonodular skin lesions, gingival hyperplasia, joint contractures and bone lesions. Less than 70 cases have been reported worldwide.

**Case report:** An eight-year-old girl born to nonconsanguineous parents presented with multiple, gradually enlarging, papulonodular skin lesions on her face, scalp, back and distal extremities since the age of 1 year. Gingival hypertrophy was present on examination. She had an elder sister with the same clinical features. The nodules from different sites were excised and ranged from 10 mm to 60 mm in diameter. The cut surfaces were yellow, homogenous and gelatinous. Microscopy showed dermal nodules with ill-defined margins, exhibiting hypercellular and hypocellular areas composed of cords of uniform fibroblasts simulating vascular channels, embedded in an abundant amorphous, eosinophilic, hyaline matrix compatible with juvenile hyaline fibromatosis. No nuclear atypia, necrosis or increased mitoses were seen. Periodic acid-Schiff (PAS) stain highlighted the matrix, which was diastase resistant.

**Discussion and conclusion:** JHF is a rare autosomal recessive genetic disorder associated with mutation of the capillary morphogenesis gene (CMG2) on 4q21. Age and the site of missense mutation are the main prognostic factors. In this case, as both members of the same generation were affected by the disease, and in the absence of any consanguinity or family history, molecular genetic studies will be helpful for confirmation, genetic counselling and prediction of prognosis.

**Keywords:** juvenile hyaline fibromatosis.

## CR 34

### Myopericytoma of the renal artery: a rare entity presenting as young hypertension

G. M. D. D. Galagedara\*, S. M. Fernandopulle

Colombo North Teaching Hospital, Ragama, Sri Lanka

[dilushi.da@gmail.com](mailto:dilushi.da@gmail.com)

**Introduction:** Renal artery stenosis is commonly caused by atherosclerosis and fibromuscular dysplasia. The latter is common in young. An intravascular neoplasm is a rare cause of renal artery stenosis. Myopericytoma is a myoid neoplasm primarily found in dermis or subcutis, but it can occur in deep tissues. It usually affects extremities, but intravascular cases exist. We present a unique case of myopericytoma in the renal artery, leading to young-onset hypertension.

**Case report:** An 18-year-old girl presented with a recent onset severe headache and significantly elevated blood pressure of >200 mmHg. Initial investigations, including serum creatinine, were within normal limits. A renal angiogram and diethylenetriamine pentaacetate (DTPA) scan revealed a normal left kidney and a small-sized right kidney with compromised function and significant renal arterial stenosis. A nephrectomy was performed, and the excised kidney measured 70x42x25mm. The renal parenchyma was unremarkable. However, the renal artery had a solid, white, homogenous cut surface and showed complete obliteration of the lumen at the hilum, up to a length of 10 mm. Microscopy revealed a well-circumscribed, intravascular lesion composed of varying-sized, haemangiopericytoma-like vascular spaces lined by uniform endothelium with perivascular proliferation of uniform, myoid, spindle cells. No significant atypia, necrosis or increased mitoses were seen. The endothelial cells were highlighted by CD34 and the myoid cells showed diffuse cytoplasmic positivity for SMA. Intravascular angioleiomyoma was excluded by the absence of desmin positivity, confirming a myopericytoma. Van Gieson stain highlighted the absence of medial thickening and a preserved internal elastic lamina, with no fragmentation and duplication, excluding fibromuscular dysplasia. In the absence of thrombi, fibrosis, subendothelial foamy cells or significant inflammation, atherosclerosis was excluded.

**Discussion and conclusion:** This case highlights the importance of thorough evaluation and consideration of unusual causes of renal artery stenosis in young patients, especially in cases with unilateral stenosis.

**Keywords:** myopericytoma, renal artery, hypertension

## CR 35

### Undifferentiated pleomorphic sarcoma of paratesticular soft tissue: a case report

N.H.S.M. Gunasekara\*, H.A.S. Perera

*Department of Histopathology, Teaching Hospital, Karapitiya, Galle, Sri Lanka*

[sammanigunasekara@gmail.com](mailto:sammanigunasekara@gmail.com)

**Introduction:** Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma (MFH), is an aggressive, high-grade soft tissue sarcoma of unknown origin. UPS is a rare tumour in the paratesticular region.

**Case report:** A 48-year-old man presented with non-tender left scrotal swelling for one year. MRI revealed a well-defined, extra testicular mass lesion at the posterior aspect of the left scrotal sac. The gross examination of the excised specimen revealed a partially encapsulated paratesticular tumour measuring 43x40x35 mm with a pale tan, solid cut surface. The tumour was separated from the testis, epididymis and spermatic cord. Microscopic examination revealed partially encapsulated malignant neoplasm with focal infiltrative areas. There were many bizarre tumour giant cells with multilobated nuclei, prominent nucleoli and scanty pale eosinophilic cytoplasm. Frequent mitotic figures were noted. Focal spindle cell areas were noted. The background contained a heavy infiltration of plasma cells, lymphocytes and neutrophils. The tumour focally infiltrated the hilar soft tissue. There was no invasion into the testis, epididymis or spermatic cord. Lymphoma and pleomorphic sarcoma were the initial differential diagnoses. Neoplastic cells revealed strong, diffuse cytoplasmic positivity for vimentin while negative for CD3, CD20, CD30, CD15, S100, SMA, desmin and CD34, which excluded lymphoma and pleomorphic sarcomas with differentiation such as pleomorphic leiomyosarcoma, pleomorphic rhabdomyosarcoma, pleomorphic liposarcoma, high-grade malignant peripheral nerve sheath tumour and poorly differentiated angiosarcoma. The final diagnosis was undifferentiated pleomorphic sarcoma.

**Discussion and conclusion:** Common sarcomas in the paratesticular region are liposarcoma, leiomyosarcoma and rhabdomyosarcoma which need to be excluded prior to making a diagnosis of UPS. As recurrence and metastasis are common, the prognosis of UPS is often poor.

**Keywords:** undifferentiated pleomorphic sarcoma, paratesticular



## CR 36

### **Sinonasal teratocarcinoma: a rare entity**

N. Hajani\*, M. Jayawickrama

*Department of Histopathology, National Hospital of Sri Lanka*

[hajaninn@gmail.com](mailto:hajaninn@gmail.com)

**Introduction:** Sinonasal teratocarcinoma is an aggressive malignant sinonasal neoplasm with combined histological features of teratoma and carcinosarcoma. The median age of presentation is 60 years, with a male predilection. It commonly involves the nasal cavity followed by the ethmoid sinus.

**Case report:** A 67-year-old man presented with periodic epistaxis of five months duration. Magnetic resonance imaging revealed a solid lesion with a small cystic component arising from the posterior ethmoid sinus and extending into the sphenoid sinus. Excision was done and the specimen was received as multiple friable fragments. Microscopically, the tumour was composed of carcinomatous, sarcomatoid, and neuroepithelial elements. The carcinomatous component included glandular structures lined by tall columnar cells and squamous nests. Immature squamous nests with foetal-appearing clear cells and a stroma containing rhabdomyoblasts and fibroblasts were present. MyoD1, NSE and CK7 were positive in the rhabdomyoblasts, neuroepithelial cells and glandular elements, respectively. CK20 and CD117 were negative.

**Discussion and conclusion:** The average survival for sinonasal teratocarcinoma is less than two years. Timely diagnosis and surgical excision combined with adjuvant therapy will improve prognosis. The differential diagnosis includes sinonasal adenocarcinoma, olfactory neuroblastoma, squamous cell carcinoma, adenosquamous carcinoma and salivary gland-type malignancies. Histomorphological features and immunohistochemical staining pattern are helpful to confirm the diagnosis. Adequate sampling is necessary to demonstrate all the components. Awareness of this condition will help to arrive at the correct diagnosis.

**Keywords:** sinonasal, ethmoid sinus, neuroepithelial

**CR 37**

**Intestinal perforation secondary to mucormycosis infection in postoperative patients: a report of two cases**

G.S.S. Hegoda<sup>1\*</sup>, S. Sumanasekara<sup>1</sup>, S. Wijetunge<sup>1</sup>, K. Ratnatunge<sup>2</sup>, H.M.A.U. Jayasinghe<sup>3</sup>

<sup>1</sup>Department of Pathology, <sup>2</sup>Department of Surgery, Faculty of Medicine, University of Peradeniya, Sri Lanka  
[sanwimck@gmail.com](mailto:sanwimck@gmail.com)

**Introduction:** Mucormycosis of the gastrointestinal tract is a rare and often fatal condition, usually associated with immunocompromised states. The mode of spread to the intestine is via ingestion, as a part of systemic dissemination, or direct implantation. Diagnosis can only be confirmed by demonstration of the characteristic Mucormycosis morphology by histology.

**Case reports:**

Case 1: A 27-year-old previously healthy immunocompetent, primipara underwent an emergency LCSC at the POA of 38 weeks due to pre-eclampsia and fetal distress at a peripheral hospital. She was put on an abdominal drain due to bleeding and was transferred to a tertiary care unit. Two weeks later, while seemingly recovering at the hospital, she developed acute abdominal distension and pain. An emergency laparotomy was performed, which revealed perforated and necrotic small bowel segments.

Case 2: A 51-year-old woman with moderately controlled diabetes mellitus underwent an elective uncomplicated hysterectomy following dysfunctional uterine bleeding at a base hospital. On postoperative day 6, she presented with fever, surgical site discharge and signs of peritonitis. She was transferred to a tertiary care centre, and an emergency laparotomy was performed, which showed gangrenous small bowel segments.

Microscopy of both small bowel specimens showed necrotic bowel segments with angioinvasive ribbon-like pauci septate fungal hyphae with right-angled branching compatible with Mucormycosis fungal spp. Despite starting antifungal treatment, both patients passed away a few days following surgery. The autopsy of patient-1 showed disseminated Mucormycosis infection involving the pancreas, kidneys and spleen.

**Discussion and conclusion:** Although gastrointestinal mucormycosis is rare, it may occur in those without risk factors. Since the prognosis is poor, a high degree of clinical suspicion and urgent reporting are required. The source of infection in postoperative patients could be direct implantation during surgery or via abdominal drains.

**Keywords:** mucormycosis infection, gastrointestinal, intestinal perforation

## CR 38

### Light chain cast nephropathy with “intratubular amyloidosis”: a rare finding in monoclonal gammopathy

G. S. S. Hegoda<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, E.H.C.K. Bandara<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Nephrology and Transplant Unit, National Hospital, Kandy, Sri Lanka*

[sanwimck@gmail.com](mailto:sanwimck@gmail.com)

**Introduction:** Major renal manifestations in monoclonal gammopathy include cast nephropathy and AL amyloidosis. Isolated intratubular amyloidosis is rare, and pathogenesis is unclear. This reflects an amyloidogenic capacity of light chains that can manifest as systemic light chain amyloidosis.

**Case report:** A 65-year-old man with diabetes mellitus for ten years presented with features of acute kidney failure. The initial serum creatinine was 700  $\mu\text{mol/L}$ , which progressively increased and he was dialysed. The urine full report showed proteinuria (1+). There were no red cells or pus cells. Ultrasound scan showed features of chronic kidney disease. His renal biopsy showed 40 glomeruli: eleven were sclerosed and the others showed no significant pathology. There was tubular atrophy with many tubules showing crystalline casts with evidence of tubular injury and associated inflammatory reaction. Interstitial fibrosis was around 50% and showed moderate lymphocytic infiltration. Congo red stain was positive for amyloid in the tubular casts with apple green birefringence under polarized light. Amyloid was not detected in glomeruli, blood vessels or the interstitium. Features of diabetic nephropathy were not present. Immunofluorescence revealed lambda light-chain restriction in casts. Accordingly, a diagnosis of cast nephropathy with intratubular amyloidosis and severe chronic interstitial nephritis was made. Subsequent investigations revealed a monoclonal band in the gamma region of serum protein electrophoresis and positive urine Bence Jones protein. However, the bone marrow biopsy showed no significant increase in plasma cells. The final diagnosis was monoclonal gammopathy of renal significance.

**Discussion and conclusion:** Isolated intratubular amyloidosis is rare and can lead to amyloid tubulopathy. It could also be a risk factor for systemic amyloidosis.

**Keywords:** intratubular amyloidosis, cast nephropathy

**CR 39**

**Pulmonary artery pseudoaneurysm secondary to a lung abscess caused by pulmonary aspergillosis**

H.H.W.S.B. Herath<sup>1\*</sup>, L. D.S. De Silva<sup>1</sup>, S.B.P.S. Wijethunga<sup>2</sup>, G. Ganga<sup>2</sup>, W. Kudagammana<sup>3</sup>, D. Medagedara<sup>4</sup>, B. Dassanayake<sup>2</sup>, S. Wijetunge<sup>1</sup>

<sup>1</sup>*Department of Histopathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Department of Surgery, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>3</sup>*Department of Microbiology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>4</sup>*Respiratory Medicine Treatment Unit, National Hospital Kandy, Sri Lanka*

[wsbherath@gmail.com](mailto:wsbherath@gmail.com)

**Introduction:** Pulmonary artery pseudoaneurysm (PAP) is a rare vascular phenomenon with a high mortality rate since it can enlarge, rupture, and lead to asphyxiation. Pulmonary aspergillosis is an underdiagnosed but known cause of PAP. The infection is most commonly seen in immunosuppressed patients. We report a rare case diagnosed incidentally in an immunocompetent patient with no pre-existing risk factors.

**Case report:** A 57-year-old woman presented with massive haemoptysis for one month. She was initially managed for community-acquired pneumonia complicated by a right pulmonary abscess. Contrast-enhanced CT revealed a right lower lobe abscess with a right pulmonary artery pseudoaneurysm. She underwent a right-sided lower and middle lobectomy. The resected lung tissue demonstrated a mycotic pseudoaneurysm of the pulmonary artery and an adjacent abscess. Histology of the surrounding lung tissue showed an organizing pneumonia pattern. Both the aneurysm wall and the surrounding lung tissue showed numerous filamentous fungi. These showed positivity with Grocott methenamine silver stain, morphologically compatible with *Aspergillus*.

**Discussion and conclusion:** PAP is mostly acquired due to causes including infection, trauma, iatrogenic and as a complication of neoplasms. The pulmonary artery lacks an adventitial wall. Therefore, repeated endovascular seeding with septic emboli creates saccular dilations that are more likely to rupture than systemic arterial aneurysms. The most common clinical presentation of PAP is massive haemoptysis with resultant worsening hypoxaemia. The most common infective causative organism is tuberculosis, which is called Rasmussen aneurysm. Although rare, fungal infection should always be excluded as a cause.

**Keywords:** aspergillosis, pulmonary artery pseudoaneurysm, lung abscess

## CR 40

### **Pancreatic clear cell neuroendocrine tumour in a patient with Von Hippel-Lindau syndrome: A rare case associated with a cerebellar hemangioblastoma, pheochromocytoma and clear cell renal cell carcinoma**

H.H.W.S.B. Herath\*, T. Jayamaha

*Department of Histopathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[wsbherath@gmail.com](mailto:wsbherath@gmail.com)

**Introduction:** Clear cell neuroendocrine tumour (CC-NET) of the pancreas is rare and is found in von Hippel-Lindau syndrome, multiple endocrine neoplasia type I (MEN I) or in sporadic form. CC-NET is often misdiagnosed as metastatic carcinoma with clear cell differentiation.

**Case report:** A 34-year-old man presented with a headache and elevated blood pressure for two months. He had a history of bilateral pheochromocytoma, right cerebellar hemangioblastoma and clear cell renal cell carcinoma (CC-RCC) in the right kidney. The patient was clinically stable. His 24-hour urine metanephrine levels were normal. Serum chromogranin levels were elevated. Contrast-enhanced computed tomography of the abdomen revealed a pancreatic-enhancing lesion. The pancreaticoduodenectomy specimen showed a well-circumscribed, unencapsulated, tan-coloured, solid and homogenous lesion in the pancreatic head. Microscopically, the tumour showed nests of polygonal cells separated by fibrous septa. The constituent cells had mildly pleomorphic centrally located nuclei with salt and pepper chromatin pattern, inconspicuous nucleoli, and clear vacuolated cytoplasm. Lymph node metastasis was present in two superior pancreatic lymph nodes. Alcian blue and Periodic acid Schiff stains were negative for mucin. The tumour cells were positive for synaptophysin, chromogranin, CK19 and vimentin and negative for CK7, RCC Ag and CD10. The Ki-67 index was 1%.

**Discussion:** In view of the past clinical history, secondary deposits of CC-RCC was considered as a differential diagnosis. Histomorphological features, negative mucin stains and immunohistochemical staining profile were consistent with a CC-NET.

**Conclusion:** CC-NET is not prognostically distinctive but mimics many primary and metastatic clear cell tumours of the pancreas.

**Keywords:** clear cell neuroendocrine tumour, pancreas, von Hippel-Lindau syndrome

## CR 41

### Colonic muco-submucosal elongated polyp: a not-so-rare entity

D. Jayasinghe\*, A. Giles

*University Hospital Lewisham, Lewisham and Greenwich NHS Trust, London, United Kingdom*

dineshajayasinghe\_01@yahoo.com

**Introduction:** Colonic muco-submucosal elongated polyps (CMSEPs) are rare, benign polyps first described in Japanese literature in the mid-1990s, and reportedly represent 0.1% of colonic polyps. Herein we report two cases of CMSEPs reported at our department within one month.

#### **Case report:**

Polyp 1: A 55-year-old man presented with abdominal pain and weight loss. Endoscopy revealed a polyp in the ascending colon measuring 15 mm in maximum diameter.

Polyp 2: A 39-year-old woman underwent a site check following a previous tubular adenoma with low and high-grade dysplasia and was found to have a polyp in the sigmoid colon measuring 9 mm in maximum diameter.

Grossly, both were long, pedunculated polyps with a drumstick-like appearance. Microscopy showed elongated polypoid tissue with normal large intestinal mucosa and attenuated muscularis mucosae. Oedematous/hyalinised submucosa contained numerous thick-walled, ectatic blood vessels arranged parallel to the polyp. No dysplasia or malignancy was seen. Both were diagnosed as CMSEPs. The previous polypectomy notes and endoscopic images excluded the possibility of a residual stalk in polyp 2.

**Discussion:** The classic pathological features of CMSEPs are worm-like/drumstick-shaped gross appearance, normal overlying mucosa with absent/diminished muscularis mucosae, and dilated, thick-walled lymphatics and blood vessels in the submucosa with little or no inflammation. The literature describes vascular arrangement parallel to the polyp. Differential diagnoses include prolapse-induced polyps, filiform polyps of inflammatory bowel disease and residual stalk of a previously excised polyp. Misclassification of these polyps as hamartomatous, hyperplastic or juvenile polyps is also described in literature. The distinctive morphology and careful clinical/endoscopic correlation would differentiate these from CMSEPs.

**Conclusion:** Awareness of this little-known entity would help correct identification of CMSEPs and avoid misclassification as polyps with graver clinical implications.

**Keywords:** benign colonic polyps, ectatic vessels, colonic polyps with normal mucosa

## CR 42

### Leiomyomatosis peritonealis disseminata: a rare entity

D. T. T. Jayasinghe\*, S.K. Liyanage

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[tharakaj.med@gmail.com](mailto:tharakaj.med@gmail.com)

**Introduction:** Leiomyomatosis peritonealis disseminata (LPD) is a rare benign disorder characterized by multiple leiomyomas on the pelvic and abdominal peritoneal surfaces. LPD is usually seen in the reproductive age group and is hormone-dependent. This is a case of LPD, which was found incidentally.

**Case report:** A 53-year-old otherwise healthy woman presented with irregular menstrual cycles. No organic cause was found, and she was treated with exogenous hormones. As the symptoms persisted, a total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. Macroscopically, there were five, firm, well-circumscribed intramural, submucosal and subserosal nodules measuring 3-20 mm in diameter, with white and whorled cut surfaces. Eight similar, small nodules were seen in the right parametrium, separated from the uterus, and on the surfaces of both ovaries, measuring up to 4 mm in diameter. The histology of all these nodules showed interlacing fascicles of uniform smooth muscle fibres, compatible with leiomyomas. None of these nodules show cellular atypia, increased mitoses or tumour necrosis. The endometrium showed progesterone-induced changes.

**Discussion and conclusion:** LPD is a rare disorder characterized by the dissemination of multiple leiomyomas on the pelvic and abdominal peritoneal surfaces, usually less than 1 cm in diameter. It is postulated that LPD occurs with metaplasia of submesothelial smooth muscles or mesenchymal cells, probably following the action of initiated or promoted estrogen and progesterone. Coexisting endometriosis and endosalpingiosis have been reported occasionally. LPD can be treated with GnRH agonists or surgery. Incomplete excision of nodules or discontinuation of GnRH agonists leads to recurrences. Malignant transformation occurs rarely.

**Keywords:** leiomyomatosis peritonealis disseminata, leiomyoma



## CR 43

### Extensive granulomas with Schaumann bodies: a rare occurrence in Crohn disease

D. T. T. Jayasinghe\*, T.P.M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[tharakaj.med@gmail.com](mailto:tharakaj.med@gmail.com)

**Introduction:** Schaumann bodies (SB) are calcium and protein inclusions seen in granulomatous diseases. They are known to occur in 10% of Crohn disease (CD). This case is reported to highlight the histological characteristics and distribution of SB in CD, which will help overcome the diagnostic pitfalls in the interpretation of colonic biopsies.

**Case report:** A 56-year-old man diagnosed with CD since 2016, underwent resection of the small bowel due to frequent relapses and features of small intestinal obstruction. The resected small bowel specimen measured 460mm in length and showed areas with a cobblestone appearance, focal ulceration of the mucosa and stricture formation. Histology revealed patchy mucosal inflammation, fissuring ulcers, transmural lymphoid aggregates with Crohn rosary, and numerous granulomas. These granulomas were predominantly seen along the myenteric plexus, focally in the submucosa and rarely in the lamina propria. In all these sites brown/black to purple laminated, concentric round structures were seen in the cytoplasm of Langhans or foreign-body giant cells. These were positive with Perl stain and von Kossa stain. Most of the granulomas were well-formed, especially the granuloma in the myenteric plexus, but ill-formed ones were frequently seen in the submucosa and the lamina propria. In these sites, only giant cells with SB closely mimicking the calcified ova of *Schistosoma* were seen. Considering the presence of characteristic histological features, the previous diagnosis of CD and no history of travel to an endemic area, the possibility of *Schistosoma*-related colitis was excluded, and CD disease was confirmed.

**Discussion and conclusion:** SB bodies are rarely seen in CD and can be a potential pitfall with *Schistosoma*-related colitis, specially in mucosal biopsies as highlighted in this case.

**Keywords:** Crohn disease, Schaumann bodies, *Schistosoma*-related colitis

CR 44

**Uterine tumour resembling ovarian sex cord-stromal tumour: a rare entity with diagnostic challenges**

D. T. T. Jayasinghe\*, S.M. Fernandopulle

Department of Pathology, Colombo North Teaching Hospital, Sri Lanka

[tharakaj.med@gmail.com](mailto:tharakaj.med@gmail.com)

**Introduction:** Uterine tumours resembling ovarian sex cord-stromal tumour (UTROSCT) are rare indolent stromal neoplasms with unclear histogenesis. This is a case of UTROSCT which was found incidentally.

**Case report:** A 54-year-old woman presented with heavy menstrual bleeding of six months duration. Total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. Macroscopically, there was a well-circumscribed subserosal nodule with a homogeneously tan to yellow cut surface, measuring 20mm in maximum diameter. The histology revealed a circumscribed, unencapsulated lesion composed of nests, interconnecting trabeculae and small solid sheets of atypical cells, lying in a fibroblastic stroma. The constituent cells showed uniform ovoid, grooved nuclei and scanty pale cytoplasm. There was no mitotic activity or tumour necrosis. There was no admixed endometrial stromal component. The tumour cells showed strong nuclear positivity with WT1 and cytoplasmic positivity with AE1/AE3. The rest of the specimen showed two intramural leiomyomas and endometriosis of the left fallopian tube.

**Discussion and conclusion:** UTROSCT is a rare uterine mesenchymal neoplasm which resembles an ovarian sex cord tumour and lacks a recognizable endometrial stroma. Endometrial stromal tumour (EST) with sex cord-like elements, leiomyoma and endometrial carcinoma with spindled and chorded patterns are the main differential diagnoses, which can be differentiated by histomorphology and cytogenetics. UTROSCTs do not express *JAZF1-SUZ12* seen in EST, and they may show fusions involving *ESR1* and *GREB1*. UTROSCT has a polyphenotypic immune profile and shows variable positivity for sex cord-stromal markers, epithelial markers, ER, PR, SMA and CD 10. Hysterectomy with bilateral salpingo-oophorectomy or hysteroscopic tumour resection are considered suitable treatment options. Close follow-up is essential to prevent occasional extrauterine and lymph node metastasis or recurrence due to unknown tumour characteristics.

**Keywords:** uterine tumour resembling ovarian sex cord-stromal tumour (UTROSCT), hysterectomy

## CR 45

### A rare case of papillary thyroid carcinoma arising in a struma ovarii

S.S. Jayasundara<sup>1</sup>, D. Walisinghe<sup>1</sup>, S. Rajauru<sup>2</sup>

<sup>1</sup>*Histopathology Unit, National Hospital, Kandy, Sri Lanka*

<sup>2</sup>*Gynecology and Obstetric Unit, National Hospital, Kandy, Sri Lanka*

[sudharikajaya@gmail.com](mailto:sudharikajaya@gmail.com)

**Introduction:** Struma ovarii is a monodermal teratoma comprising exclusively or >50% of benign thyroid tissue. This rare germ cell tumour accounts for 2% of all mature teratomas. Papillary thyroid carcinoma arising in a struma ovarii is even rarer, occurring in less than 5% of struma ovarii cases.

**Case report:** A 53-year-old woman presented with lower abdominal pain for one month. US scan abdomen revealed left side complex ovarian cyst. The CA125 level was normal. A total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Macroscopically, the left ovary showed a 25 mm diameter multiloculated cyst with cystic and solid areas and a focus of warty papillary projections. The initial microscopic examination revealed an ovarian cyst composed of benign thyroid tissue. Further sampling from the papillary areas revealed a microscopic lesion measuring 6 mm in diameter, composed of well-formed papillae lined by follicular epithelial cells showing characteristic nuclear features of papillary carcinoma. There were scattered psammoma bodies. No other teratomatous areas were present. These features were compatible with papillary thyroid carcinoma arising in a monodermal mature teratoma, FIGO stage 1A. Postoperatively, the patient was surveilled for primary thyroid carcinoma and the possibility of metastasis to the ovary was excluded.

**Discussion and conclusion:** Papillary thyroid carcinoma arising from struma ovarii is rare. However, adequate sampling is necessary to arrive at a correct diagnosis. The complete surgical excision of stage IA tumours shows a good prognosis.

**Keywords:** struma ovarii, papillary thyroid carcinoma

## CR 46

### Extra-gastrointestinal stromal tumour of the pancreas: a rare entity

D. R. Karunaratne, M. M. A. Jayawickrama

*Department of Pathology National Hospital of Sri Lanka, Colombo, Sri Lanka*

[rangana\\_k@hotmail.com](mailto:rangana_k@hotmail.com)

**Introduction:** Extra-gastrointestinal stromal tumours (EGISTs) represent 5% -10% of all GISTs. EGISTs arising from the pancreas are extremely rare with around 5% arising in the pancreas.

**Case report:** A 51-year-old woman was found to have an abdominal mass while awaiting a hernia repair. CECT showed a well-defined, low-density lesion within the pancreas. Intraoperatively, the mass was found to be in the distal pancreas and a distal pancreatectomy with splenectomy was performed. The excised specimen showed a circumscribed solid tan-yellow tumour measuring 10 cm in maximum diameter, confined to the body and tail of the pancreas. Microscopically, the tumour was composed of fascicles of spindle cells with elongated, bland nuclei. Some areas showed nuclear palisading and a lymphocytic infiltrate. Mitoses were 1/5 mm<sup>2</sup>. Tumour necrosis, lymphovascular invasion or perineural invasion were not seen. The tumour was confined to the pancreas. The spleen was normal. The differential diagnosis included an EGIST, inflammatory myofibroblastic tumour (IMFT) and a pancreatic schwannoma. The neoplastic cells were positive for CD117 and S100 and negative for ALK. The tumour was diagnosed as an EGIST.

**Discussion:** Histopathological, immunohistochemical and/or molecular assessment is necessary for the confirmation of an EGIST. In this case, positivity for CD117 and negativity for ALK excluded schwannoma and IMFT, respectively. S100 can show variable positivity in about 5% -10% of GISTs.

**Conclusion:** EGISTs, even in their rarity, should be considered in the differential diagnosis of pancreatic neoplasms.

**Keywords:** extra-gastrointestinal stromal tumour, pancreas

## CR 47

### Composite phaeochromocytoma-ganglioneuroma in the adrenal gland: an uncommon diagnosis

D. R. Karunaratne\*,<sup>1</sup> S.R Constantine

*Department of Pathology, National Hospital of Sri Lanka, Sri Lanka*

[rangana\\_k@hotmail.com](mailto:rangana_k@hotmail.com)

**Introduction:** Composite phaeochromocytomas constitute 3% - 9% of all phaeochromocytomas. They are more common in the adrenal medulla but may arise in extra-adrenal sites.

**Case report:** A 43-year-old woman was incidentally found to have a right suprarenal mass on an ultrasound scan. Her urine metanephrine levels were significantly elevated. Contrast-enhanced CT revealed a well-defined, complex right suprarenal tumour, and a right adrenalectomy was performed. The excision specimen showed a yellowish mass of tissue with haemorrhagic, cystic and pale solid areas measuring 11cm in maximum dimension. Microscopically, the tumour was composed of a phaeochromocytoma and a ganglioneuroma. The phaeochromocytoma component showed a Zellballen arrangement of small to large cells with pleomorphic nuclei and amphophilic cytoplasm. Mitoses were 8/2 mm<sup>2</sup>, including atypical forms. The ganglioneuroma component showed ganglion cells in a schwannian stroma. Necrosis, capsular or lymphovascular space invasion were not seen. Normal adrenal cortical tissue was seen at the periphery. The pheochromocytomatous areas were positive for chromogranin and the ganglioneuroma component was positive for S100. The tumour was diagnosed as a composite phaeochromocytoma-ganglioneuroma with a PASS score of 9/20.

**Discussion:** Phaeochromocytomas may rarely combine with other neurogenic elements such as ganglioneuroma (70% - 80%), neuroblastoma or ganglioneuroblastoma. In this case, the histological and immunohistochemical features were compatible with a composite phaeochromocytoma-ganglioneuroma with a PASS score of 9/20, indicating an increased malignant potential.

**Conclusion:** As composite phaeochromocytomas are rare tumours, careful examination of the specimen is essential to prevent underdiagnosis.

**Keywords:** composite phaeochromocytoma, ganglioneuroma, adrenal

CR 48

## Functional ampullary somatostatinoma with somatostatin syndrome: a rare presentation

P. G. S. P Karunarathne\*, T. P. M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[supipinayani@gmail.com](mailto:supipinayani@gmail.com)

**Introduction:** Somatostatinoma is a rare neuroendocrine tumour (NET) involving the pancreas, ampulla and duodenum, which can be functional or non-functional. Functional somatostatinomas produce somatostatin syndrome (SmSn) characterized by diabetes mellitus (DM), cholelithiasis, and steatorrhea. Ampullary somatostatinomas (ASM) are rare and often non-functional.

**Case report:** A 65-year-old man with DM for two years presented with significant loss of weight, loss of appetite, dyspeptic symptoms, constipation, abdominal pain and anaemia for eight months. Endoscopy showed duodenal luminal narrowing. CECT abdomen revealed a periampullary mass for which he underwent Whipple resection. This specimen showed a periampullary exophytic growth measuring 7.5 cm in maximum diameter, involving the pancreatic head and duodenum. The gallbladder showed cholelithiasis. Microscopically, the tumour comprised tubules, rosettes and solid nests of monomorphic tumour cells showing small round nuclei with salt and pepper chromatin and moderate granular eosinophilic cytoplasm. Psammomatous calcifications were seen throughout the tumour. The tumour cells expressed synaptophysin and CD56 and a Ki-67 index of <3%. Posterior pancreaticoduodenal (3/12) and hepatic artery (3/4) lymph nodes showed tumour metastasis with extranodal extension. Diagnosis of well-differentiated NET of ampulla (pT3N1) was made. Following surgery, the patient recovered from DM, and the follow-up CT was uneventful. Preoperative serum somatostatin level was not done as NET was not suspected clinically.

**Discussion and conclusion:** Diagnosis of functional ASM is difficult as nonspecific symptoms can mimic other disease entities. In this case, recent onset DM, cholelithiasis, and gastrointestinal symptoms favour SmSn. Clinicopathological correlation helps arrive at the diagnosis of somatostatinoma, even without the preoperative serum somatostatin level. The prognosis of ASM depends on the tumour size, amount of residual tumour and the extent of metastasis, indicating a high risk of recurrence in this patient due to lymph node metastasis.

**Keywords:** ampullary somatostatinoma, somatostatin syndrome, neuroendocrine tumour

## CR 49

### Appendiceal endosalpingiosis mimicking a neoplasm: a rare case

P. G. S. P. Karunaratne\*, T. P. M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

supipinayani@gmail.com

**Introduction:** Endosalpingiosis is the presence of ectopic fallopian tubal epithelium, commonly found in the serosal surfaces of the pelvis and peritoneum. It is more common than endometriosis in older women. Appendiceal endosalpingiosis (AES) is rare, with only a few reported cases in the literature. This is a case of AES mimicking a neoplasm.

**Case report:** A 54-year-old woman who underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy for pelvic endometriosis four years back presented with chronic lower abdominal pain. CT scan of the abdomen showed an enlarged appendix suspicious of a neoplasm. The appendectomy showed an appendix with a dilated tip measuring 25x20x10mm. The cut surface showed small cysts, up to 3 mm in diameter, in the wall filled with mucinous material. The rest of the appendix measured 35 mm in length and 5mm in diameter, which showed no focal lesions. Microscopy showed multiple foci within the muscularis propria comprising glands and papillae lined by tubal epithelium with ciliated and non-ciliated columnar cells and intercalated cells compatible with endosalpingiosis. Markedly dilated cystic glands show a flattened tubal-type epithelial lining. Intraluminal papillae formation is noted focally. There was no atypia in the lining epithelium. These epithelial cells expressed strong and diffuse nuclear positivity for ER immunostain, confirming the diagnosis of AES. The patient's symptoms were declined following the appendectomy.

**Discussion:** Awareness of endosalpingiosis is mandatory as it is associated with chronic pelvic pain and carries a risk of developing ovarian serous neoplasms. AES is extremely rare and can present with a cystic mass involving the tip of the appendix, raising possibilities of appendiceal mucinous neoplasms and peri-appendiceal implants of mucinous neoplasm of the ovary. Endosalpingiosis can coexist with or may mimic endometriosis.

**Keywords:** appendix, endosalpingiosis



## CR 50

### A case of oesophageal melanocytosis

P. G. S. P. Karunaratne\*, T. P. M. Bopagoda

*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[supipinayani@gmail.com](mailto:supipinayani@gmail.com)

**Introduction:** Oesophageal melanocytosis (EM) is a rare, benign condition with a reported incidence of 0.07-2.1%. EM is usually an incidental endoscopic finding, characterized by non-atypical melanocytic proliferation and increased melanin deposition in the oesophageal mucosa, usually involving the middle and lower third of the oesophagus.

**Case report:** A 75-year-old woman on herbal medication for diabetes mellitus for the last 15 years presented with dyspeptic symptoms. Endoscopy showed unhealthy lower oesophageal mucosa, the biopsy of which showed non-dysplastic oesophageal mucosa with focal parakeratosis, exocytosis of neutrophils, lymphocytes, and occasional eosinophils associated with mild intercellular oedema and congested papillae. The lamina propria showed mild oedema, congestion and scattered mixed inflammatory cells and pigment-laden macrophages. A coarse brown colour crystalline pigment is noted intracellularly within the basal layer and lying freely in the submucosa. The submucosa showed a few scattered inflammatory cells and submucosal glands. PAS and Perl stains are negative for fungal hyphae/lipofuscin and iron, respectively. The intracellular pigment was confirmed as melanin by the melanin bleach procedure. The diagnosis of EM associated with reflux oesophagitis was made.

**Discussion and conclusion:** The aetiology and pathogenesis of EM remain uncertain. The oesophagus lacks any melanocytes. Aberrant migration of melanocytes into oesophageal squamous epithelium is likely to result from reflux esophagitis and chronic stimuli that damage the oesophageal mucosa. In this patient, the long-term use of herbal medications is the most likely chronic causative stimulus. As EM has been suggested as a precursor of primary oesophageal melanoma, recognition of this uncommon entity is vital. Progression from EM to malignant melanoma has not been elucidated due to its rarity.

**Keywords:** oesophageal melanocytosis, reflux oesophagitis, oesophageal melanoma

## CR 51

### **An incidental finding of a primary carcinoid tumour arising in a mature cystic teratoma of the ovary: a rare case**

P. G. S. P Karunarathne\*, S. M. Fernandopulle

*Department of Pathology, Colombo North Teaching Hospital, Ragama, Sri Lanka*

[supipinayani@gmail.com](mailto:supipinayani@gmail.com)

**Introduction:** Primary ovarian carcinoid tumour (POC) is a rare low-grade neuroendocrine tumour (NET) and is considered a monodermal teratoma. A few cases of POC arising in mature cystic teratoma (MCT) were reported in the literature.

**Case report:** A 57-year-old postmenopausal woman was investigated for abdominal pain, dyspeptic symptoms and fascial oedema. The USS of the pelvis showed a suspicious left ovarian cyst, and she underwent a total abdominal hysterectomy. Macroscopy revealed an intact multilocular left ovarian cyst containing sebaceous material, hair, and yellow and white solid areas involving about 40% of the cyst. The cystic areas were lined by stratified squamous keratinized epithelium. The cyst wall showed mature tissue from all three germ layers. Microscopically, there was a circumscribed tumour measuring 8 mm in diameter, composed of nests, cords and trabeculae of uniform cells having small centrally located round nuclei with salt and pepper chromatin, moderate granular eosinophilic cytoplasm and low mitotic activity. Tumour cells showed cytoplasmic positivity with synaptophysin and a Ki-67 index of 5%, confirming the diagnosis of POC arising in an MCT.

**Discussion and conclusion:** Ovarian carcinoid tumour can be primary or metastasis from a NET elsewhere. Tumour bilaterality, multiple ovarian nodules and known NET elsewhere favour a metastatic tumour. The neuroendocrine cells of the gastrointestinal-type epithelium are the most likely cell of origin POC arising in MCT. Morphologically, POC shows trabecular, strumal, mucinous and insular patterns. The insular pattern is usually associated with carcinoid syndrome. As in this patient, POC in MCT limited to the ovary has a good prognosis following total surgical resection. Postoperative follow-up imaging is needed to assess any possible recurrence of the disease.

**Keywords:** carcinoid tumour, mature cystic teratoma, carcinoid syndrome, neuroendocrine tumour

## CR 52

### **Acute oxalate nephropathy caused by consumption of *Averrhoa bilimbi* juice in a healthy adult: a rare case**

P. G. S. P. Karunaratne\*, S. M. Fernandopulle

*Department of Pathology, Colombo North Teaching Hospital, Ragama, Sri Lanka*

[supipinayani@gmail.com](mailto:supipinayani@gmail.com)

**Introduction:** Acute oxalate nephropathy (AON) is a well-known but rarely reported cause of acute kidney injury (AKI) in Sri Lanka (SL). According to the published data, *Averrhoa carambola* (star fruit) is the most common cause. The occurrence of AON due to *Averrhoa bilimbi* (bilimbi), which is consumed as a fruit and is also used as a medicine, is well-recognized, however, only one paediatric case was reported in SL. This is a case of AON in an adult male following ingestion of bilimbi juice.

**Case report:** A 52-year-old man presented with loin pain, nausea and oliguria for three days after drinking a juice made of mashed bilimbi fruits. His serum creatinine was increased (1315  $\mu\text{mol/L}$ ). The urine full report showed pus cells of 15-20/HPF, red blood cells of 80-100/HPF, presence of protein (1+) and few crystals. No casts or dysmorphic red cells were present. The ultrasound scan revealed kidneys of normal size and echogenicity. His renal biopsy revealed dilated tubules with intratubular polarizable fan-shaped crystals, focal tubulitis, granular casts, features of acute tubular injury and moderate mixed inflammatory cell infiltrate and oedema in the interstitium, favouring the diagnosis of AON. This patient fully recovered following four cycles of haemodialysis and supportive care and now has normal renal functions.

**Discussion and conclusion:** Other aetiological causes for hyperoxaluria were excluded clinically in this patient. The urinary oxalate level was not available at the time of admission to support the diagnosis. This case emphasizes the importance of obtaining a detailed dietary history in patients with AKI and educating the public about the renal effects of bilimbi.

**Keywords:** *Averrhoa bilimbi*, oxalate nephropathy, acute kidney injury

## CR 53

### **Bullous variant of mycosis fungoides: a rare case**

K. V. N. D. Kumarasinghe\*, P. N. Amaraweera

*Department of Histopathology, Teaching Hospital, Karapitiya, Sri Lanka*

[88navoda.dk@gmail.com](mailto:88navoda.dk@gmail.com)

**Introduction:** Mycosis fungoides (MF) is the most common cutaneous lymphoma with variable clinical and histological forms. Bullous formation is a rare finding associated with an aggressive clinical course and poor prognosis.

**Case report:** A 48-year-old woman presented with ulcerated skin nodules on the trunk and limbs. Tumour stage of MF was diagnosed following clinical, histological and immunohistochemical assessment. There was no evidence of generalized lymphadenopathy or hepatosplenomegaly. There were no atypical cells in the peripheral blood or the bone marrow biopsy. She was started on chemotherapy, and on post-chemotherapy day ten, she developed fever with generalized vesicles and blisters. Varicella virus PCR was negative. Microscopy of a skin biopsy revealed focal parakeratosis, intracorneal neutrophilic microabscesses, multiple subcorneal and intraepithelial vesicles containing atypical lymphoid cells, acantholytic keratinocytes and neutrophils, and marked spongiosis. Sheets of atypical lymphocytes with convoluted nuclei and frequent mitoses were present in the dermis. There was no dermal infiltration of neutrophils, viral cytopathic effects, vasculitis or leucocytoclasia. Neoplastic lymphoid cells were strongly positive for CD3 and negative for CD20 and CD30.

**Discussion and conclusion:** The initial clinical differential diagnoses, disseminated varicella infection and drug-induced Sweet syndrome, were not supported by the skin biopsy findings. Negativity for CD30 excluded the possibilities of primary cutaneous CD30-positive T-cell lymphoproliferative disorder and primary cutaneous anaplastic large-cell lymphoma. Peripheral T-cell lymphoma was unlikely due to lack of nodal and bone marrow involvement. The diagnosis of a bullous variant of MF was made on clinicopathological grounds and available investigation findings. However, for the completeness of the diagnostic workup, immune-mediated vesicobullous disorders need to be excluded by immunofluorescence, which was not available in the local setting.

**Keywords:** mycosis fungoides, disseminated varicella, bullous variant

## CR 54

### **Florid microglandular adenosis presenting as clinically and radiologically suspicious breast lesion: a case report**

K. V. N. D. Kumarasinghe\*, H. A. S. Perera

*Department of Histopathology, Teaching Hospital, Karapitiya, Sri Lanka*

88navoda.dk@gmail.com

**Introduction:** Microglandular adenosis (MGA) is a rare benign proliferative breast lesion. Atypical forms and carcinomas arising in MGA have also been described. MGA is suggested as a nonobligate precursor of triple-negative breast carcinoma based on molecular and cytogenetic similarities.

**Case report:** A 63-year-old woman presented with a clinically suspicious left breast lump. The mammogram revealed a BIRADS IVc lesion. Tru-cut biopsy of the lesion was reported as a benign breast lesion with adenosis. The wide local excision specimen showed an irregular, firm, white lesion measuring 23x22x11 mm. Histology revealed haphazardly infiltrating, small round to oval open glands in a fibrous stroma. Glandular lumina contained PAS-positive, diastase-resistant, thick eosinophilic secretory material. The glands were devoid of myoepithelial cell layer and lined by a single layer of cuboidal epithelial cells with bland nuclei and vacuolated cytoplasm. Mitoses were scanty. No apocrine snouts or necrotic foci were noted. The lesion involved superior and deep resection margins. The neoplastic epithelial cells showed diffuse strong positivity for S100 and were negative for ER, PR and EMA.

**Discussion and conclusion:** The main differential diagnosis of MGA is with tubular carcinoma of the breast (TCB) due to infiltrative growth pattern and absence of myoepithelial cell layer. It is crucial to differentiate these two entities to avoid unnecessary treatment. The absence of irregular, angulated glands and negativity for ER, PR, and EMA excluded the possibility of TCB. However, considering the role of MGA as a precursor lesion, complete margin clearance was recommended in this case.

**Keywords:** microglandular adenosis, tubular carcinoma, myoepithelial cell layer

CR 55

## Papillary thyroid carcinoma, hobnail variant

P. D. K. Kumari<sup>1,2\*</sup>, K. K. S. Lakshika<sup>1,2</sup>, M. D. S. Lokuhetty<sup>1,2</sup>, M. V. C. de Silva<sup>1,2</sup>

<sup>1</sup>*Department of Histopathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>2</sup>*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*

[wijegunawardanepdkk@gmail.com](mailto:wijegunawardanepdkk@gmail.com)

**Introduction:** Thyroid carcinomas are the most common endocrine malignancy, with papillary thyroid carcinoma (PTC) being the commonest histological type. It generally has a good outcome and prognosis with current treatment. However, some types of PTCs show aggressive behaviour. One such rare variant is the hobnail variant which has higher rates of distant metastasis, recurrences, multifocality and mortality. The hobnail variant (HVPTC) requires the presence of hobnail features in at least 30% of the tumour.

**Case report:** A 48-year-old previously healthy woman presented with a progressively enlarging lump in the anterior neck. Ultrasound scan showed a hypoechoic TIRADS V lesion in the isthmus. FNAC was reported as Bethesda category V, suspicious for papillary thyroid carcinoma. The total thyroidectomy contained an irregular whitish solid lesion measuring 14x11x7 mm, involving the isthmus and the left lobe of the thyroid. Microscopically, the tumour was predominantly composed of papillae and micropapillae lined by poorly cohesive epithelial cells with eosinophilic cytoplasm and apically located nuclei with prominent nucleoli. The cells were elongated, focally columnar and contained overlapping nuclei with occasional nuclear grooves and nuclear inclusions. Some cell clusters infiltrated the surrounding fibrous stroma. The tumour was pT1b (tumour >1 cm but not > 2cm, limited to the thyroid). There was no lymphovascular invasion, multifocality or extrathyroidal extension.

**Discussion and conclusion:** It is important to identify HYPTC due to its rarity, aggressive behaviour, increased radioactive iodine refractoriness and poor prognosis. Its histological hallmark is elongated cells with a high nuclear/cytoplasmic ratio, hobnail appearance, micropapillary pattern and increased nuclear atypia. Most of these tumours harbour BRAF mutation followed by mutations in TP53, TERT promoter, PIK3CA or CTNNB1.

**Keywords:** papillary thyroid carcinoma, hobnail variant

**CR 56**

**Sessile serrated lesion with an unusual stromal proliferation: a rare occurrence**

R. L. Lawrence<sup>1</sup>, T. S. Dissanayake<sup>1</sup>, D. L. Piyarisi<sup>2</sup>, A.S. Rodrigo\*<sup>1</sup>

<sup>1</sup>*Department of Pathology, Sri Jayewardenepura General Hospital, Sri Lanka*

<sup>2</sup>*Surgical Unit, Sri Jayewardenepura General Hospital, Sri Lanka*

[sonalirodrigo@yahoo.com](mailto:sonalirodrigo@yahoo.com)

**Introduction:** Sessile serrated lesions (SSL) are epithelial proliferations in 30 % of colonic polyps and are diagnosed on morphology using strict diagnostic criteria.

**Case report:** A 78-year-old woman was found to have an ascending colon polyp on a colonoscopy. A snare polypectomy specimen showed a polyp measuring 15x12x8 mm. Microscopy revealed a sessile serrated lesion (SSL) fulfilling the strict diagnostic criteria of serrated columnar epithelium with goblet cells and unequivocal architectural distortion with asymmetrical glands, dilatation of crypt base and horizontal growth along the muscularis mucosa. There was no evidence of dysplasia. Stromal proliferation was noted in the lamina propria and the cellular features were compatible with Schwann cells with scattered ganglion cells. There was no nuclear atypia, mitoses, necrosis or stromal neuroblasts. The conclusion was SSL with no dysplasia and ganglioneuroma-like stromal proliferation in the lamina propria.

**Discussion and conclusion:** SSLs are known to be associated with unusual stromal proliferations, commonly perineural-like stromal changes. However, ganglioneuroma-like stromal changes, as seen in this case, were not documented. Differential diagnoses include mucosal perineuroma and ganglioneuroma which show similar stromal proliferations around the glands that do not show serrated morphology. Although positivity for EMA and CD34 in the stroma can differentiate perineural-like stromal changes from ganglioneuroma-like stromal changes, where ganglion cells and stroma are positive for NSE and S100, morphological features alone are sufficient for a definite diagnosis. This proliferation is likely a reaction to serrated epithelium rather than a true neoplastic proliferation of stromal cells. No clinical significance is attributed to this unusual association of SSL.

**Keywords:** sessile serrated lesions, ganglioneuroma, perineuroma



CR 57

**Limitations of tru-cut biopsy in the evaluation of low-grade breast lesions: a case report**

R. L. Lawrence<sup>1</sup>, P. K. Arambepola<sup>2</sup>, D. Y. Abeywardhana<sup>2</sup>, A. S. Rodrigo<sup>1\*</sup>

<sup>1</sup>Department of Pathology, <sup>2</sup>Department of Radiology, Sri Jayewardenepura General Hospital, Sri Lanka  
[sonalirodrigo@yahoo.com](mailto:sonalirodrigo@yahoo.com)

**Introduction:** Tru-cut biopsy (TCB) is widely popular when diagnosing breast lesions. Differentiating benign and malignant lesions and subtyping malignant tumours are not difficult on most occasions. Occasionally, the limitation of tissue may give rise to diagnostic dilemmas.

**Case report:** A 68-year-old woman presented with breast pain, and mammography revealed neoplasms of BIRADS category 4b in both breasts. A TCB of the right breast lesion showed a tumour with solid clusters and nests of round to ovoid, monomorphic cells with eccentric bland nuclei and moderate cytoplasm. Signet ring cell morphology with intracytoplasmic mucin was seen. Mitoses were sparse. A cluster of tumour cells was seen floating in extracellular mucin. A biopsy from the left breast lesion showed similar morphology. A diagnosis of a low-grade breast carcinoma was made, favouring bilateral solid papillary carcinoma (SPC). The patient underwent wide local excision of both lesions, and histology revealed a mucinous carcinoma of both breasts with low-grade DCIS of solid and cribriform morphology.

**Discussion and conclusion:** Although TCB has revolutionized the management of breast lesions, limitations are observed when evaluating heterogeneous lesions that require accurate quantification of tumour components for a definitive diagnosis. In this case, SPC was favoured as the biopsy had solid clusters of cells with low-grade nuclear morphology with focal signet ring-like cells and focal extracellular mucin. Mucinous carcinoma needs a pure mucinous component of more than 90% for the diagnosis. Low-grade DCIS can mimic a low-grade carcinoma. Partially biopsied papillary lesions need a complete evaluation. Awareness of the limitations of this test and judicious reporting can minimize the diagnostic pitfalls.

**Keywords:** solid papillary carcinoma, mucinous carcinoma, tru-cut biopsy

## CR 58

### Duodenal gangliocytic paraganglioma: a report of two rare cases

H. Maddumage\*, P. Pingle

*Department of Cellular Pathology, Royal Surrey Hospital, NHS-Foundation Trust, United Kingdom*

[hansikamaddumage@gmail.com](mailto:hansikamaddumage@gmail.com)

**Introduction:** Duodenal gangliocytic paragangliomas (DGP) are rare neuroendocrine neoplasms in the peri-ampullary area. Most tumours are asymptomatic and rarely cause symptoms. Excision of DGP is curative; however, there is a rare possibility of regional nodal metastasis.

**Case report:** Here, we report two cases of DGPs presenting as polyps in a 46-year-old woman (case 1) and a 62-year-old man (case 2). Grossly, case 1 and case 2 were polypectomy and Whipple specimens, respectively. Both specimens contained 65 mm-sized stalked polyps with a fleshy tan cut surface. Both polyps were circumscribed mucosal tumours, composed of schwannian-type spindle-shaped cells and ganglion-like cells that were positive for S100 and SOX10 and nests and trabeculae of epithelioid neuroendocrine-type cells positive for cytokeratin, CAM 5.2 and neuroendocrine markers. The mitotic activity was low. In case 2, one regional lymph node was positive for the neuroendocrine component of the gangliocytic paraganglioma.

**Discussion:** Gangliocytic paragangliomas are rare solitary neuroendocrine tumours that arise in the peri-ampullary region in the third to ninth decade of life. They are mostly sporadic, and rarely associated with neurofibromatosis type 1. These tumours exhibit triphasic cellular differentiation: epithelioid neuroendocrine, ganglion-like and schwannian-type spindle cells. Negativity of CD117/DOG1 and positivity of neuroendocrine markers excluded the possibility of gastrointestinal stromal tumour and schwannoma respectively.

**Conclusion:** Here we report two DGPs; one of which had regional nodal metastasis. There is no consensus regarding the prognosis or optimal follow-up of these cases since the number of reported cases remains low. Further reports and data relevant to gangliocytic paragangliomas are necessary for the determination of appropriate management and surveillance following surgery.

**Keywords:** duodenum, gangliocytic paraganglioma, regional metastasis

**CR 59**

**Unilateral deep infiltrative ureteric endometriosis: a silent cause of hydronephrosis**

M. D. J. C. Malagala\*, P.D. K. Thiranagama, C. S. P. Sosai, A. M. Abeygunasekara

*Department of Histopathology, Colombo South Teaching Hospital, Sri Lanka*

malagalajayani@gmail.com

**Introduction:** Endometriosis of the urinary tract is a rare entity, affecting the bladder, ureter and kidneys in a relative frequency of 40:5:1, respectively. Intrinsic ureteric endometriosis is considered a deep invasive form that may be undiagnosed and results in chronic renal damage.

**Case report:** A 42-year-old woman presented with recurrent right loin pain for nine months. Her menstrual cycles were regular, and there was no dysmenorrhea. Examination and family history were insignificant. USS showed hydronephrosis of the right kidney and a periureteric soft tissue mass. Bilateral adnexa were normal. CT scan showed dilation of the ureter up to S1 level and a soft tissue lesion measuring 19x15 mm at the mid to proximal part of the right ureter. The biopsy of this soft tissue lesion showed features of endometriosis. A nephroureterectomy was performed due to the non-functioning kidney. Macroscopically, the right kidney was 120x60x35 mm. The pelvicalyceal system was markedly dilated. The ureter was 200 mm in length, with a dilated proximal part and distal narrowing with periureteric soft tissue thickening by an irregular, firm, white lesion. Microscopy revealed ureteric endometriosis involving the submucosa, muscular wall and adventitia.

**Discussion and conclusion:** Although histopathological features are typical, clinical diagnosis of ureteric endometriosis is challenging due to its unusual location and non-specific symptoms. This patient had no history of endometriosis or cyclical pain, which caused a delay in the diagnosis and led to chronic renal damage. Endometriosis should be considered a possible cause of hydronephrosis in women. Imaging techniques are not always effective in establishing a correct preoperative diagnosis.

**Keywords:** endometriosis, ureter

**CR 60**

**Angiomyomatous hamartoma arising in an inguinal lymph node: a report of a rare entity**

I. W. G. A. L. Malhasi\*, I. V. Prematilleke

*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

malhasig@gmail.com

**Introduction:** Angiomyomatous hamartoma (AMH) is a benign vascular lesion of the lymph node that primarily involves the inguinal nodes. Involvement of other lymph node groups is extremely uncommon. It is characterized by the replacement of the parenchyma with blood vessels, smooth muscle and fibrous tissue. We report an AMH arising in the left inguinal region of a 69-year-old man.

**Case report:** A 69-year-old man presented with a left groin mass. Ultrasound scan showed a subcutaneous, solid, tubular mass in the left inguinal region. The radiological impression was of a matted lymph node mass involved by granulomatous lymphadenitis. The excision biopsy comprised a 2cm firm mass. Microscopy revealed a lymph node mass, the parenchyma of which was replaced by haphazard bundles of smooth muscle, confirmed by Masson trichrome stain, together with fibrous tissue and fat. Thick-walled blood vessels were present in the hilum and proliferating thin-walled vessels were seen within fibromuscular tissue. Small nodules of residual lymphoid tissue consisting of a polymorphous population of lymphoid cells and tingible-body and haemosiderin-laden macrophages were identified at the periphery. Cellular pleomorphism, granulomata, necrosis, mitoses, atypical lymphoid cells or epithelial cells were not seen.

**Discussion and conclusion:** It is suggested that AMH represents a disordered angiogenic process which starts in the hilum and extends towards the cortex, causing normal lymphatic tissue to become displaced and atrophic, leading it to be overdiagnosed as Castleman disease. Although AMH of lymph nodes is very rare, its recognition is important for differentiating it from other angiomatous lesions such as angiomyolipoma, vascular transformation of lymph node sinuses and angiolipomatous hamartoma which may mimic this entity.

**Keywords:** hamartoma, lymph node, inguinal

## CR 61

### **Uterine leiomyoma with chondroid metaplasia: a rare phenomenon in a common neoplasm**

I.W.G.A.L. Malhasi<sup>1\*</sup>, B. Seneviratne<sup>1</sup>, R.C. Fernandopulle<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

<sup>2</sup>*Department of Obstetrics and Gynaecology, Colombo South Teaching Hospital, Sri Lanka*

[malhasig@gmail.com](mailto:malhasig@gmail.com)

**Introduction:** Leiomyoma is the commonest uterine tumour with a wide spectrum of variants. Adipose metaplasia is a relatively common finding in leiomyomas. However, chondroid metaplasia, as in this case, is a rare phenomenon.

**Case report:** A 49-year-old woman presented with menorrhagia for four months. The ultrasound scan revealed multiple uterine leiomyomas. There were no differences in the radiological appearances of these nodules. She underwent a hysterectomy and bilateral salpingectomy. Macroscopically, the uterus revealed multiple (15), well-circumscribed, subserosal and intramural nodules. The largest nodule was 65 mm in diameter and showed solid, glistening, myxoid-like areas. The other nodules showed greyish-white, whorled cut surfaces. No necrosis or haemorrhages were seen. The endometrium was 2 mm in thickness and was unremarkable. Histologically, the largest nodule predominantly showed chondroid areas comprising chondrocytes in a myxohyaline matrix merging with the interlacing fascicles of regular smooth muscle cells at the periphery. Extensive sampling did not reveal nuclear atypia, necrosis or mitoses in the chondroid areas. No sarcomatous differentiation or malignant epithelial elements were seen. The other nodules comprised interlacing fascicles of smooth muscle fibres. None of these nodules showed nuclear atypia, necrosis or increased mitoses. The endometrium showed progesterone-related effects.

**Discussion and conclusion:** The exact nature of these chondroid foci is not fully understood, however, it may involve the reprogramming of stem cells or undifferentiated mesenchymal cells. Such metaplastic changes have no malignant implications, however, may lead to the consideration of differential diagnosis with other malignant mesenchymal tumours such as chondrosarcoma and carcinosarcoma of the uterus. Thorough sampling and clinico-radiological correlation are recommended not to overlook a malignant mesenchymal tumour with heterologous differentiation.

**Keywords:** leiomyoma, cartilaginous metaplasia, uterus

**CR 62**

**Rosai-Dorfman disease of the breast: a rare case leading to a diagnostic dilemma**

J. D. S. Manoharee, M.G.A.I. Maveekumbura, S.M. Fernandopulle\*

*Department Of Pathology, North Colombo Teaching Hospital, Ragama, Sri Lanka*

[shanikamadonna@gmail.com](mailto:shanikamadonna@gmail.com)

**Introduction:** Rosai-Dorfman disease (RDD) is a histiocytosis that usually involves cervical lymph nodes in children and young adults. Extranodal RDD is encountered in over 40% of patients. Localization to the breast is rare and often causes difficulties in the differential diagnosis of breast malignancies.

**Case report:** A 23-year-old woman presented with a non-tender, 2x2 cm lump in the upper outer quadrant of her right breast for three weeks. Ultrasound scan revealed a hypoechoic lesion with irregular margins. Magnetic resonance imaging showed a suspicious lesion suggestive of an inflammatory malignancy with possible axillary lymph node involvement. A tru cut biopsy from the mass was reported as lymphocytic mastitis. Since the lesion was clinically and radiologically suspicious the patient underwent a wide local excision of the lesion. The cut surface of the specimen showed a firm tan lesion measuring 30 mm in maximum diameter. Histology showed an inflammatory lesion in the breast composed of lymphocytes, plasma cells and histiocytes. The cytoplasm of some of the histiocytes contained engulfed intact plasma cells and lymphocytes, denoting emperipolesis. No granulomatous inflammation, epithelial or lymphoproliferative malignancy were seen. On immunohistochemistry, the histiocytes with emperipolesis showed diffuse cytoplasmic positivity for S100. The immunohistochemical features confirmed the diagnosis of RDD of the breast.

**Discussion and conclusion:** RDD is caused by benign proliferation of histiocytes, and involvement of the breast is a rare occurrence that may mimic malignancy on ultrasound scan and mammogram. The histological differential diagnoses of RDD include granulomatous mastitis, IgG4-sclerosing mastitis and breast lymphoma with plasmacytic differentiation. In this case, the final diagnosis was made by histological assessment with immunohistochemical staining.

**Keywords:** Rosai-Dorfman disease

## CR 64

### **Dedifferentiated ovarian carcinoma with a differentiated component of endometrioid carcinoma: a case report**

G. W. C. M. Silva, P. N. Amarabandu\*

*Department of Histopathology, Apeksha Hospital, Maharagama, Sri Lanka*

[chanikamadhushani1988@gmail.com](mailto:chanikamadhushani1988@gmail.com)

**Introduction:** Dedifferentiated carcinoma of the ovary is an uncommon tumour, comprising less than 0.5% of ovarian carcinomas. It is a biphasic tumour with a differentiated endometrioid carcinoma and an undifferentiated component.

**Case report:** A 48-year-old woman with a history of hysterectomy for fibroid uterus presented with progressive abdominal distension. The CECT showed a well-defined mass in the pelvis. The resected specimen comprised multiple pieces of tan-coloured tissue, collectively measuring 140x130x45 mm. Microscopy revealed ovarian tissue with a tumour showing an abrupt transition between the two morphologically different malignant components. One component showed sheets of large atypical polygonal cells with moderately pleomorphic vesicular nuclei, prominent nucleoli, moderate eosinophilic cytoplasm and frequent mitoses. Focal tumour necrosis was evident. The other component was composed of glandular and papillary structures lined by atypical columnar epithelial cells containing moderately pleomorphic vesicular nuclei and moderate eosinophilic cytoplasm. The glandular component showed diffuse nuclear positivity for immunohistochemical stain ER and was negative for vimentin, WT1, CD117 and PLAP. The tumour cells in the solid area showed focal positivity for PLAP and were negative for ER, WT1, CD117 and vimentin. Tumour cells in both components were positive for EMA. Currently, the patient is being treated with chemotherapy.

**Discussion and conclusion:** The initial differential diagnoses on the features of the H&E-stained sections included high-grade endometrioid adenocarcinoma with solid areas, dedifferentiated carcinoma and mixed adenocarcinoma. The abrupt transition between the two components and EMA and CD117 negativity in the solid component favoured dedifferentiated carcinoma. This is an aggressive tumour with a poor prognosis.

**Keywords:** dedifferentiated carcinoma, endometrioid-type adenocarcinoma, vimentin, WT1, CD 117 and PLAP



## CR 65

### Primary angiosarcoma of the urinary bladder

H.A.S Perera, V.A.D.N. Samarawickrama\*

*Department of Pathology, Teaching Hospital, Karapitiya, Sri Lanka*

svadnilanthi@gmail.com

**Introduction:** Primary angiosarcoma of the urinary bladder is an uncommon aggressive vascular neoplasm.

**Case report:** A 55-year-old farmer who was a heavy smoker and had a history of using agrochemicals for more than 30 years presented with lower abdominal pain and gross painless haematuria. Cystoscopic examination identified a mass lesion with large areas of necrosis arising in the postero-lateral wall of the bladder and occupying the bladder cavity. Ultrasound scan of the abdomen and pelvis revealed a bladder mass without extraperitoneal spread. Contrast-enhanced CT showed a locally advanced lesion with no evidence of metastasis. Salvage cystoprostatectomy and ileal conduit urinary diversion was done. The specimen was composed of fragments of tissue which revealed a tumour with sheets of epithelioid, rhabdoid and spindle-shaped cells with areas of haemorrhage and necrosis. The cells showed highly pleomorphic nuclei, multiple prominent nucleoli, abundant eosinophilic cytoplasm and occasional intracytoplasmic red blood cells. Benign urothelium was identified focally. The main differential diagnoses were sarcomatoid urothelial carcinoma, rhabdomyosarcoma, leiomyosarcoma and melanoma. The tumour was sampled extensively. On immunohistochemical evaluation, CK7 and AE1/AE3 were positive in the normal urothelium. The tumour cells showed diffuse, strong immunostaining with vimentin and complete membranous staining with CD31. CD99 was focally and weakly positive in some tumour cells. MyoD1, SMA, desmin, BCL2, melan A, EMA and CK20 were negative.

**Discussion and conclusion:** Considering the morphology and immunohistochemical markers, the final diagnosis was given as primary angiosarcoma of the urinary bladder which is an exceedingly rare tumour. Sarcomas constitute less than 1% of all malignant neoplasms of the urinary bladder and angiosarcomas comprise 2% of all sarcomas. The reported five-year survival rate ranges from 10-35%.

**Keywords:** primary angiosarcoma, intracytoplasmic vacuoles.

CR 66

**Infantile galactosialidosis complicated by nephrotic syndrome: a rare case**

M. K. Piyasinghe<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, R. S. Thalgahagoda<sup>2</sup>, S. Y. Dissanayake<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Department of Pediatrics, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[mekupi@gmail.com](mailto:mekupi@gmail.com)

**Introduction:** Galactosialidosis is a rare autosomal recessive lysosomal storage disease with renal manifestations. This is a case of galactosialidosis with nephrotic syndrome (NS), demonstrating foamy cells in renal biopsy.

**Case report:** An eight-month-old boy, the firstborn of a non-consanguineous marriage, was investigated for generalized body swelling and treated for steroid-resistant NS from the age of two months. This baby had dysmorphic features, including coarse facies and flat nasal bridge, hepatosplenomegaly, and evidence of developmental delay. Investigations revealed enlarged kidneys, abnormal spinal vertebrae, hypertrophic cardiomyopathy, and a metabolic disease was suspected. The biochemical tests revealed low albumin, high ALP, increased glutamine levels and altered renal function tests. The renal biopsy showed foamy epithelial cells filling the Bowman space and replacing the tubular epithelium, suggestive of a storage disorder. However, interstitial foamy cells were not seen. Later, genetic testing identified a CTSA gene abnormality, associated with galactosialidosis. He was treated with steroids and given supportive care. Parents were offered genetic counselling.

**Discussion and conclusion:** NS is the most common renal manifestation of galactosialidosis and can be associated with many histological patterns, including minimal change disease, FSGS, and proliferative glomerulonephritis, with/without features of excess metabolic product deposition. Foamy cell infiltration in the kidney could be due to different conditions, but in galactosialidosis, it is due to sialyloligosaccharides deposition, leading to nephromegaly and steroid-resistant NS. This usually results in end-stage renal failure, and supportive care is the only available management. CTSA gene is involved in producing lysosomal catalytic enzymes, and its defect is associated with lysosomal storage disorders. Enzyme replacement therapy and gene therapy are still under investigation.

**Keywords:** galactosialidosis, foamy cells, renal biopsy, nephrotic syndrome, CTSA-gene

## CR 67

### **Dedifferentiated endometrioid carcinoma with sarcomatous differentiation arising in a background of endometriosis and presenting as a posterior vaginal mass**

M. K. Piyasinghe<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, C. Rathnayake<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*Department of Obstetrics and Gynaecology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

mekupi@gmail.com

**Introduction:** Endometriosis is a risk factor for carcinomas of the gynaecological tract. Dedifferentiated endometrioid carcinomas are extremely rare. This is a case of a dedifferentiated endometrioid carcinoma with a predominant sarcomatous component and background endometriosis.

**Case report:** A 72-year-old postmenopausal woman, who is obese, presented with lower abdominal pain. She had a hysterectomy and salpingo-oophorectomy (HSO) 15 years ago for dysfunctional uterine bleeding, the histology of which had not shown any significant pathological findings. CT revealed a vaginal mass attached to the sigmoid colon, but her colonoscopy was normal. A sigmoid colectomy specimen was received with a 7 cm necrotic mass. Microscopy showed a biphasic tumour composed of well-formed endometrioid-type glands (20%) and a high-grade tumour showing sheets of undifferentiated cells (80%), in a myxoid and necrotic stroma, with background endometriosis. There were areas that showed rhabdoid morphology. On immunohistochemistry analysis, EMA, pancytokeratin, vimentin and CD10 were positive in both components. The Ki-67 index was 100% in undifferentiated cells. immunohistochemistry for mismatch repair (MMR) genes, MLH1, MSH2, MSH6, and PMS2, did not show any microsatellite instability.

**Discussion and conclusion:** Dedifferentiated endometrioid carcinomas are rare and aggressive cancers with low-grade adenocarcinoma and high-grade undifferentiated tumour components. Some are associated with mutations of MMR genes in both components, which helps in differentiating it from other high-grade carcinomas. Although these tumours usually arise from the native/metaplastic endometrial epithelium of the uterus/ovaries, this tumour originated from active peritoneal endometriosis which was present in the patient even after HSO, due to obesity-associated high oestrogenic status. Extensive sampling and a broad panel of immunohistochemical stains are essential to identify the line of differentiation in such undifferentiated tumours.

**Keywords:** dedifferentiated endometrioid carcinoma, endometriosis, microsatellite instability

## CR 68

### Recurrence of oxalate nephropathy, mimicking an acute transplant rejection

M.K. Piyasinghe<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, A.W.M. Wazil<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*National Hospital Kandy, Sri Lanka*

[mekupi@gmail.com](mailto:mekupi@gmail.com)

**Introduction:** Oxalate nephropathy is a rare metabolic disease, forming calcium oxalate crystals in renal tubules, leading to chronic kidney disease (CKD). We report a case of recurrent primary hyperoxaluria noted in a transplant renal biopsy, mimicking graft rejection clinically and histologically.

**Case report:** A 32-year-old man presented with features of severe CKD. Increased urinary oxalate crystals were identified during investigation. His sister had similar findings in her urine with preserved renal functions. Following a non-cadaveric allograft renal transplant, he presented with rising serum creatinine, clinically suggestive of acute rejection. Renal biopsy on Day 8 revealed tubulitis, suggestive of acute cell-mediated rejection. Occasional tubules showed oxalate crystals. As there was no response despite increasing steroids, a repeat renal biopsy was done on Day 45. The second biopsy showed numerous oxalate crystals, which were fluorescent under polarized light, in tubules with mild tubular and interstitial inflammation.

**Discussion:** Occasional oxalate crystals are common in transplant renal biopsies. However, if the history is supportive, it can be subtle evidence of a recurrence of hyperoxaluria which is difficult to separate from graft failure clinically. Due to sampling errors, the renal core biopsy may only show the cellular reaction against it, manifesting as tubular and interstitial inflammation, which is histologically identical to acute cell-mediated rejection.

**Conclusion:** Although the occurrence of occasional oxalate crystals in renal biopsy is mostly insignificant, its presence can be an indicator of a recurrence in primary hyperoxaluria. Therefore, diagnosis of tubular reaction as acute rejection should be done with great caution in such cases, as it is a common mimicker.

**Keywords:** oxalate nephropathy, allograft transplant, renal biopsy, acute rejection

## CR 69

### A rare case of primary malignant melanoma of the bladder

H. K. U. Piyamalika\*, B. Senevirathna

*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

[aruni1987125@gmail.com](mailto:aruni1987125@gmail.com)

**Introduction:** The urethra and penis are the most common sites of primary malignant melanoma (PMM) in the genitourinary tract, where metastatic melanomas are more common than PMM. This is a case of PMM of the bladder, which is exceedingly rare and has a poor prognosis.

**Case report:** A 76-year-old woman presented with haematuria. Cystoscopy showed multiple black lesions in the bladder mucosa. The patient underwent transurethral resection of the bladder tumour. Microscopy showed a poorly differentiated malignancy composed of large polygonal-shaped cells arranged in nests and sheets. The constituent cells displayed round to oval nuclei with prominent nucleoli and eosinophilic cytoplasm. Some areas showed intracytoplasmic brown pigments. The histological differential diagnosis included poorly differentiated urothelial carcinoma and malignant melanoma. On immunohistochemistry, the cells were negative for pan cytokeratin and showed positive staining for melanotic markers, HMB45 and S100. As no primary focus of skin melanoma was identified on clinical examination and no other lesions were identified on CT scan and magnetic resonance imaging, the diagnosis was confirmed as PMM of the bladder.

**Discussion and conclusion:** It is essential to rule out metastasis from a distant primary before confirming the diagnosis of PMM of the bladder. PMM has microscopic variability; it can be epithelioid, spindle-shaped, or extremely bizarre. Microscopically, these lesions, particularly the hypomelanotic to amelanotic melanomas, may mimic urothelial carcinoma. Careful evaluation for the presence of melanin pigment and nucleolar prominence and performance of melanotic markers assist in achieving the correct diagnosis.

**Keywords:** primary malignant melanoma, bladder

## CR 70

### Peritoneal deciduosis: a rare but alarming mimicker of peritoneal carcinomatosis

A. Rajapakse<sup>1\*</sup>, R. Punchihewa<sup>2</sup>, P. A. Denegama<sup>3</sup>, P. Samarasinghe<sup>1</sup>, M. L. Amarasena<sup>3</sup>

<sup>1</sup>Department of Pathology, Base Hospital, Mawanella, Sri Lanka

<sup>2</sup>Department of Pathology, National Hospital for Respiratory Disease, Sri Lanka

<sup>3</sup>Department of Obstetrics and Gynaecology, Base Hospital, Mawanella, Sri Lanka

[a.d.rajapakse5@gmail.com](mailto:a.d.rajapakse5@gmail.com)

**Introduction:** The presence of ectopic decidual tissue is defined as deciduosis. Gross deciduosis peritonei is a rare, benign, self-limited, metaplastic condition of the peritoneum, usually seen in pregnancy, involving the ovaries, uterus and cervix. This is a rare presentation with peritoneal involvement, incidentally discovered during a caesarean section.

**Case report:** A previously healthy 26-year-old woman was admitted for the delivery of her first baby. An emergency caesarian section was performed due to foetal distress. During surgery, numerous white nodules were noticed on the omentum, large bowel and bilateral ovaries. Multiple biopsies were taken from these nodules, and the specimen consisted of fatty tissue containing pale to white firm nodules of varying sizes. Microscopic examination of haematoxylin and eosin-stained sections showed numerous nodular collections of large, oval to spindle-shaped cells with enlarged nuclei and abundant cytoplasm, resembling decidual cells. No atypical features were seen. The origin of these cells was confirmed by positivity for ER and CD10 and negativity for pancytokeratin. A healthy baby was delivered, and the postoperative period of the mother was uneventful.

**Discussion and conclusion:** Deciduosis occurs due to a progesterone-mediated stimulus, causing metaplasia of subserosal stromal cells. Gross peritoneal deciduosis is usually asymptomatic and resolves spontaneously 4-6 weeks after delivery. Rarely, features of appendicitis, intraperitoneal haemorrhage, tubo-ovarian abscess and bowel obstruction may be present. The differential diagnoses include tuberculosis, mesothelioma and carcinomatosis. It can raise suspicion of tubercles and carcinomatosis. Histological assessment is needed for the diagnosis.

**Keywords:** deciduosis, peritoneal carcinomatosis

## CR 71

### An uncommon presentation of solitary anorectal metastasis of breast carcinoma

M. M. Rajapakse\*, A. Perera, P. Kariyawasam, A.W.K. Dilrukshi

*Department of Histopathology, Teaching Hospital, Karapitiya, Sri Lanka*

[madhara77@gmail.com](mailto:madhara77@gmail.com)

**Introduction:** Metastasis of breast carcinoma (BC) to the gastrointestinal (GI) tract is infrequent (6%-18%), with invasive lobular BC (4.2%) being the commonest subtype to metastasize. Anorectal metastasis is extremely rare and associated with advanced staging and unfavourable outcome. Diagnosis may be challenging due to nonspecific GI symptoms and a prolonged asymptomatic period.

**Case report:** A 56-year-old woman with a history of invasive multifocal lobular BC treated with mastectomy and chemoradiotherapy presented with difficulty in defecation for 10 months. She had been asymptomatic with negative screening since 2016. Her CT scan showed a tumour in the anorectal region, infiltrating the bladder and cervix. Colonoscopy revealed a tight anal tumour, 6 cm above the anal verge. Mucosal biopsies revealed disclosive, singly dispersed atypical cells infiltrating the rectal mucosa. The individual cells had a plasmacytoid appearance with enlarged, hyperchromatic, eccentrically placed nuclei. A few signet ring-like cells were present. On immunohistochemistry, these cells were AE1/AE3 positive, LCA negative and showed strong and diffuse positivity for ER, PR and CK7 with negative HER-2. The immunohistochemical staining pattern was the same as the primary BC.

**Discussion and conclusion:** This case was diagnosed as an anorectal deposit from lobular BC. Invasive BC may give rise to unusual metastasis mimicking an anorectal growth. Therefore, the patient's history of BC played a pivotal role in the correct diagnosis. Immunohistochemistry is beneficial in diagnosing and confirming the primary in suspicious cases.

**Keywords:** breast carcinoma, metastasis, ano-rectal, immunohistochemistry



CR 72

**Langerhans cell histiocytosis of thyroid gland: a rare case**

P.P. Rajawasam\*, P.N. Amaraweera<sup>2</sup> A. Perera

*Department of Pathology, Teaching Hospital, Karapitiya, Sri Lanka*

pprajawasam@gmail.com

**Introduction:** Langerhans cell histiocytosis (LCH) is a rare malignancy characterized by the monoclonal proliferation of Langerhans cells. LCH manifests as a unifocal disease or a multifocal disease with multisystem involvement. Involvement of the thyroid gland is extremely rare.

**Case report:** A 30-year-old woman with hypothyroidism presented with a goitre for six months. Ultrasonography revealed diffuse enlargement of the thyroid gland, with evidence of thyroiditis. FNAC smears were cellular and composed of singly dispersed cells and loosely cohesive clusters of atypical histiocyte-like cells. The cells had round to oval vesicular nuclei with intranuclear grooves and nuclear indentations. Some binucleated and multinucleated cells were seen. Scattered eosinophils and lymphocytes were present in the background. Papillaroid clusters were not seen. FNAC was reported as a haematolymphoid tumour infiltrating both lobes of the thyroid gland, favouring LCH. On gross examination, the thyroid gland was diffusely enlarged with a rubbery, white cut surface. Histology demonstrated an infiltrating lesion composed of diffuse sheets of cells with a histiocytoid morphology. These cells had vesicular nuclei with prominent grooves and indentations and abundant eosinophilic cytoplasm. Scattered eosinophils were seen. The tumour cells were positive for S100 and CD1a and negative for CD3 and CD20. The patient responded well to treatment with combined chemotherapy and radiotherapy.

**Discussion and conclusion:** The rarity of LCH of the thyroid poses diagnostic challenges. It can be easily mistaken for lymphoma, undifferentiated carcinoma, lymphocytic thyroiditis or granulomatous thyroiditis. The positivity of tumour cells for CD1a and S100 confirmed LCH in this case. Early recognition, accurate diagnosis, appropriate and timely treatment, and follow-up are vital for a better prognosis. Unifocal disease is associated with a good prognosis. Involvement of other organs must be assessed since multifocal multisystem disease requires aggressive therapy.

**Keywords:** Langerhans cell histiocytosis, thyroid gland, rare case

## CR 73

### **“Sea-anemone-like” ciliated tumour cells in a serous ovarian carcinoma: a rare cytological finding**

J. N. Reddy, M. Schubel, A. Darlington, [D. Samarasinghe\\*](#)

*Department of Histology and Cytology, Te Whatu Ora Waikato, Waikato Hospital, New Zealand*

[dushy.samarasinghe@waikatodhb.health.nz](mailto:dushy.samarasinghe@waikatodhb.health.nz)

**Introduction:** Ciliated cells are seen in various cytological specimens, including those from the respiratory tract, reproductive tract and digestive tract. The presence of cilia is usually indicative of a benign process. Very rarely is the presence of ciliated cells encountered in malignancies.

**Case report:** A 78-year-old woman presented with ascites. No other pertinent clinical history was provided with this specimen. The ascitic fluid sample contained 10 ml of orange-colour fluid. Four smears were prepared from the deposit of the centrifuged fluid and stained with Papanicolaou and Diff-Quik methods. These smears showed papillary aggregates of polyhedral cells with dense, hyperchromatic, pleomorphic nuclei and abundant cytoplasm. Isolated cells within these aggregates and some single-lying cells displayed unipolar cilia/“sea-anemone-like” cells. Immunohistochemical stains were performed on sections of the cell block, and these cells were positive for PAX8, CK7 and WT1.

**Discussion and conclusions:** Cytological features, in conjunction with the immunohistochemical results, led to the diagnosis of metastatic serous carcinoma of müllarian origin. CT scan showed widespread peritoneal disease, ascites and a mixed-density pelvic lesion. The histological features of the omental core biopsy were compatible with a serous carcinoma, consistent with the initial cytological findings.

**Keywords:** ciliated cells, effusion cytology, ovary, serous carcinoma

## CR 74

### **Dedifferentiated chondrosarcoma arising from preexisting exostosis in a middle-aged man: a rare case**

K. B. Rojika<sup>1</sup>, N. Samarawickrama<sup>2\*</sup>

<sup>1</sup>*Faculty of Medicine, University of Ruhuna, Galle, Sri Lanka,*

<sup>2</sup>*Department of Pathology, District General Hospital, Matale, Sri Lanka*

[rojikariyawasam@gmail.com](mailto:rojikariyawasam@gmail.com)

**Introduction:** Dedifferentiated chondrosarcoma is a high-grade malignancy with a poor prognosis. It usually occurs in the fifth to seventh decade, with male preponderance. Dedifferentiated chondrosarcoma is most often located in the pelvis and long bones, such as the femur or tibia. Dedifferentiation may originate from enchondroma, low-grade chondrosarcoma or chondrosarcoma arising from a preexisting osteochondroma.

**Case report:** A 61-year-old man with multiple exostoses presented with a rapidly enlarging mass over his right proximal tibia. Radiological studies revealed a destructive bone lesion in the proximal tibia, extending into the surrounding soft tissue. The initial biopsy was reported as a high-grade sarcoma. The tumour was resected with the proximal part of the tibia. Macroscopy revealed a destructive tumour composed of solid areas with necrotic and haemorrhagic foci, involving the tibia and the surrounding soft tissue. Definite cartilaginous areas were not identified. Microscopy revealed a predominantly high-grade sarcomatoid spindle cell component with cells showing rhabdoid morphology. Focal lobular architecture was present. Subsequent sampling revealed a minor grade II chondrosarcoma component with an abrupt transition to the predominant high-grade sarcomatoid component. A diagnosis of dedifferentiated chondrosarcoma was made.

**Discussion:** Preexisting exostosis is known to cause chondrosarcoma and subsequent dedifferentiation. Therefore, adequate sampling of such lesions is paramount to avoid diagnostic difficulty and the usage of immunohistochemical studies.

**Conclusion:** If high-grade sarcomatous areas are seen in an otherwise nonspecific bone-involving tumour, it is prudent to sample the tumour adequately to identify any chondrosarcomatous component.

**Keywords:** dedifferentiated chondrosarcoma, chondrosarcoma, exostoses

## CR 75

### Well-differentiated liposarcoma of the tongue: a case report

K. Samalai<sup>1,2\*</sup>, H.D. Wijesinghe<sup>1,2</sup>, S. Wadusinghearachchi<sup>3</sup>, M.V.C De Silva<sup>1,2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>2</sup>*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*

<sup>3</sup>*National Dental Hospital, Colombo, Sri Lanka*

[bparththipan@gmail.com](mailto:bparththipan@gmail.com)

**Introduction:** Well-differentiated liposarcoma (WDLS) is the commonest soft tissue sarcoma, frequently involving the retroperitoneum and extremities. Involvement of the head and neck region is rare.

**Case report:** A 43-year-old man presented with a gradually enlarging lesion on the tongue for two years. Examination revealed a firm nodule in the left lateral part of the tongue measuring 2x2 cm. There was no mucosal ulceration. A wide local excision was performed, and the cut sections of this specimen showed a circumscribed, yellowish-white, lobulated lesion extending to a depth of 2 cm. Microscopy revealed a circumscribed lesion composed of variable-sized adipocytes admixed with lipoblasts with enlarged, indented, hyperchromatic nuclei, spindled cells and occasional multinucleated tumour giant cells. Mitoses were infrequent. The surrounding skeletal muscle bundles were compressed. The resection margins were clear. The differential diagnoses included WDLS, intramuscular lipoma with regressive changes and clear cell carcinoma of the minor salivary glands. Special stains PAS and PAS-D were negative. The tumour cells showed strong and diffuse nuclear and cytoplasmic staining for S100 and scattered strong nuclear positivity for MDM2 and CDK4. A diagnosis of lipoma-like WDLS was made. There was no recurrence following nine months of surgery.

**Discussion and conclusion:** WDLS is a locally aggressive tumour with no metastatic potential unless there are dedifferentiated areas. WDLS bears supernumerary ring and giant marker chromosomal anomalies leading to amplification of MDM2 and CDK4 genes. Overexpression of these proteins can be recognised via immunohistochemistry. In contrast to WDLS elsewhere, lesions involving the oral cavity are smaller, well-circumscribed and have indolent behaviour. Inadequate excision might lead to recurrence and the potential to dedifferentiate and metastasize. Wide local excision with adequate free margins is the treatment of choice. In view of possible recurrence, close follow-up was suggested.

**Keywords:** well-differentiated liposarcoma, MDM2, oral-cavity, indolent

## CR 76

### Myopericytoma/myofibroma of the gingiva: a case report

K. Samalai<sup>1,2\*</sup>, A.A.H. Priyani<sup>1,2</sup>, A. Silva<sup>3</sup>, P.R. Jayasooriya<sup>4</sup>, M.V.C. De Silva<sup>1,2</sup>

<sup>1</sup>Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

<sup>2</sup>Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka

<sup>3</sup>Teaching Hospital, Kulliyapitiya, Sri Lanka

<sup>4</sup>Department of Oral Pathology, Faculty of Dental Sciences, University of Peradeniya, Sri Lanka

[bparththipan@gmail.com](mailto:bparththipan@gmail.com)

**Introduction:** Myopericytomas/myofibromas are benign pericytic tumours. Any age group can be affected, and there is a male predominance. Distal extremities are the commonest location followed by proximal extremities, neck and trunk. Involvement of the oral cavity is a rare presentation.

**Case report:** A 32-year-old man presented with an enlarging, 3x3 cm, firm submucosal nodule in the third retromolar region of the mandible associated with an impacted tooth. Computed tomography revealed a mildly enhancing soft-tissue lesion. On histology, the subepithelial connective tissue contained a circumscribed spindle cell lesion composed of short fascicles of spindle cells with intervening haemangiopericytomatous vessels. The cells contained ovoid to elongated nuclei with finely dispersed chromatin and moderate eosinophilic cytoplasmic processes. Mitoses were rare. Most vascular spaces were surrounded by myxofibrous and hyalinized stroma. The underlying fatty tissue was uninvolved. The margins were clear. The tumour cells showed strong and diffuse cytoplasmic staining for SMA and were negative for CD34, desmin, h-caldesmon, and S100. An immunomorphological diagnosis of benign myopericytoma/ myofibroma was made.

**Discussion:** Myopericytoma is an uncommon benign perivascular myoid neoplasm that forms a morphological spectrum with myofibroma. The presence of gaping vessels and surrounding hyalinized stroma suggested a differential diagnosis of solitary fibrous tumour and schwannoma but they were excluded by negativity for CD34 and S100, respectively. Myopericytoma rarely exhibits malignant changes. Recurrence is unusual, thus local excision is sufficient.

**Conclusion:** Awareness that this entity may occur within the oral cavity will help to avoid misdiagnosis.

**Keywords:** myopericytoma, myofibroma, pericytic tumours, gingiva, haemangiopericytomatous vessels

**CR 77**

**Dyshormonogenetic features in a hyperplastic nodule: a caveat in thyroid pathology**

P. C. U. Samaraweera\*<sup>1</sup>, I. W. G. A. L. Malhasi<sup>1</sup>, D. H. Beneragama<sup>1</sup>, M. V. C. de Silva<sup>2</sup>

<sup>1</sup>*Department of Pathology and Cancer Research Centre, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

<sup>2</sup>*Centre for diagnosis and research in cancer, Faculty of Medicine, University of Colombo, Sri Lanka*

[udeshika\\_samaraweera@yahoo.com](mailto:udeshika_samaraweera@yahoo.com)

**Introduction:** Dyshormonogenetic goitre is genetically determined thyroid hyperplasia due to enzyme defects in thyroid hormone synthesis.

**Case report:** A 43-year-old woman was investigated for left midline neck swelling. The ultrasound scan showed a prominent nodule in the left lobe of the thyroid compatible with TIRADS 4. Cytology revealed features suspicious of papillary carcinoma, corresponding to Bethesda category V. A total thyroidectomy was performed. The macroscopy showed a tan-brown, well-circumscribed nodule in the left lobe measuring 22 mm in maximum diameter. Microscopically, this was an unencapsulated nodule composed of colloid-filled microfollicles lined by plump thyroid epithelial cells displaying varying degrees of cellular pleomorphism, prominent nucleoli and pale eosinophilic cytoplasm. The interfollicular areas contained focal acellular eosinophilic material. The background thyroid tissue showed features of a colloid nodular goitre. The possibilities of a hyperplastic nodule, medullary thyroid carcinoma and a noninvasive follicular thyroid neoplasm with papillary nuclear features (NIFTP) were considered. Congo red stain for amyloid was negative. Immunohistochemistry showed negativity for HMBE1 and CK19. This was concluded as a hyperplastic nodule with dyshormonogenetic features.

**Discussion and conclusion:** Dyshormonogenetic goitre is a rare benign condition in a patient with thyroid enlargement and hypothyroidism which may lead to overdiagnosis of neoplastic lesions due to overlapping cytoarchitectural features. The presence of acellular material in the stroma raised the suspicion of medullary carcinoma, however, was excluded by negative congo red stain. CD56 and calcitonin were requested but were not performed due to limited resources. The negativity of these two stains would further confirm the absence of a NIFTP and a medullary carcinoma, respectively. As this was a solitary nodule, dyshormonogenetic features in a hyperplastic nodule was concluded as the final diagnosis.

**Keywords:** dyshormonogenetic, goitre, hyperplastic nodule

## CR 78

### Mesonephric hyperplasia of uterine cervix: a diagnostic challenge

P.C.U. Samaraweera<sup>1\*</sup>, M. Gunawardana<sup>1</sup>, D.H. Beneragama<sup>1</sup>, M.V.C. de Silva<sup>2</sup>

<sup>1</sup>*Department of Pathology and Cancer Research Centre, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

<sup>2</sup>*Centre for Diagnosis and Research in Cancer (CeDARC), University of Colombo, Sri Lanka*

[udeshika\\_samaraweera@yahoo.com](mailto:udeshika_samaraweera@yahoo.com)

**Introduction:** Mesonephric hyperplasia (MH) of the uterine cervix is a well-known but uncommon entity that arises from mesonephric remnants. Differentiation of MH from lobular endocervical glandular hyperplasia (LEGH) and endocervical adenocarcinoma is diagnostically challenging. We report a case of MH mimicking endocervical adenocarcinoma.

**Case report:** A 59-year-old woman was investigated for perimenopausal bleeding. Abdominal examination was normal. Hysteroscopy revealed an irregular endometrium with a normal cervix. A total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed. The cervix was macroscopically unremarkable. However, microscopy showed groups of small, discrete glands infiltrating the deep cervical stroma in a diffuse pattern with a minimal desmoplastic reaction. The entire cervix was sampled and there was no back to back arrangement of glands. The glands were lined by cuboidal cells with bland nuclei. There was no karyorrhexis, mitotic activity or nuclear atypia. Some of the glandular lumina were filled with a deeply eosinophilic material. Angio or perineural invasion was not found. The lesion did not involve the lower uterine segment. The rest of the uterus showed non-reactive endometrial glands, decidualized stroma and adenomyosis. Both fallopian tubes and ovaries were histologically unremarkable. The differential diagnosis included MH and endocervical adenocarcinoma. The immunohistochemical profile showed CD10 and GATA3 positivity. WT1 was negative and the Ki-67 index was very low. A diagnosis of MH was made based on the immunomorphological features.

**Discussion:** MH can be misdiagnosed as LEGH or malignancy, and a panel of immunohistochemistry (IHC) markers are helpful as supplementary tools. A minimal panel of IHC, including CD 10, WT1, GATA 3 and Ki-67, was performed in this case. Negativity of CEA, p53, ER and PR would also support the diagnosis of MH. The sampling of the entire cervix is mandatory to exclude any co-existing neoplastic conditions.

**Keywords:** mesonephric hyperplasia



**CR 79**

**Cytological diagnosis of intraosseous odontogenic keratocyst in the mandible: a case report**

P. C. U. Samaraweera<sup>1\*</sup>, H. K. U. Piyamalika<sup>1</sup>, C. S. P. Sosai<sup>1</sup>, P. Kirupakaran<sup>2</sup>

<sup>1</sup>Department of Histopathology, <sup>2</sup>Oromaxillaryfacial Unit, Colombo South Teaching Hospital, Sri Lanka

[udeshika\\_samaraweera@yahoo.com](mailto:udeshika_samaraweera@yahoo.com)

**Introduction:** Cytological diagnosis of intraosseous cysts of the jaw is challenging, as causative pathology is diverse and rare, making management modes variable. Intraosseous odontogenic keratocyst (OKC) should be distinguished from ameloblastoma and other tumours in order to aid in appropriate management.

**Case report:** A 37-year-old man presented with swelling of the mandible and loose anterior teeth for one month. On examination, a diffuse bony hard swelling was present in the submental region. The orthopantomogram revealed a well-defined, unilocular, radiolucent cyst within the anterolateral part of the mandible in relation to the roots of bilateral lower anterior teeth. The clinical diagnosis was ameloblastoma. Fine needle aspiration cytology (FNAC) revealed clusters of benign columnar cells arranged in a picket fence appearance, clusters of anucleate squames, nucleated squamous cells and multinucleated giant cells, compatible with OKC. The histology of the incisional biopsy measuring 5x3x1 mm showed a cyst lined by keratinized squamous epithelium and basal columnar cells in a picket fence arrangement containing stratified, hyperchromatic, elongated nuclei, confirming the diagnosis of OKC.

**Discussion and conclusion:** The differential diagnoses of cystic jaw lesions are vast, including ameloblastoma, myeloma, Langerhans cell histiocytosis, cemento-ossifying fibroma, aneurysmal bone cyst, giant cell and fibro-osseous lesions and malignant tumours. FNAC is a valuable preoperative diagnostic tool to determine the nature of intraosseous cysts as benign or malignant, as in this case. The presence of anucleate squames, columnar cells and lack of basaloid cells favoured OKC over ameloblastoma on cytology. The sparsity of giant cells and lack of eosinophils and plasma cells helped to exclude the other lytic lesions.

**Keywords:** OKC, cysts, intraosseous, cytomorphology

**CR 80**

**Exuberant granulomatous reaction masking the diagnosis of Hodgkin lymphoma: a diagnostic pitfall**

A. Senevirathne\*, R. M. N. D. Ratnayake

*Department of Pathology, National Hospital Kandy, Sri Lanka*

[aanusenevirathne@gmail.com](mailto:aanusenevirathne@gmail.com)

**Introduction:** Hodgkin lymphoma is a malignant lymphoid proliferation of crippled neoplastic B cells in a mixed inflammatory background. Hodgkin lymphoma can cause a granulomatous reaction, which can either mimic or coexist with tuberculosis occurring due to immunosuppression.

**Case report:** A 42-year-old man presented with pyrexia of unknown origin, cervical lymphadenopathy and hepatosplenomegaly. Initial investigations revealed an elevated erythrocyte sedimentation rate and a positive Mantoux test. Bone marrow biopsy showed granulomatous inflammation. On these grounds, antituberculosis therapy was started. The patient showed a poor response to treatment, and his serum transaminase levels started to rise. Antituberculosis therapy was halted at this point and a cervical lymph node biopsy was performed. Histology revealed multiple granulomas composed of epithelioid histiocytes and multinucleated giant cells with central eosinophilic, granular necrosis resembling tuberculous granulomas. As the patient clinically deteriorated, a review of the histology was done, during which a few scattered large cells with lobulated nuclei and prominent nucleoli were noted. These cells showed membrane and Golgi positivity to CD30 and bluish positivity to PAX5. These cells were negative for CD20 and CD3. Accordingly, a diagnosis of Hodgkin lymphoma was made, and the patient was referred for oncological management.

**Discussion and conclusion:** A granulomatous reaction in Hodgkin lymphoma could be either due to the lymphoma itself or due to concurrent tuberculosis infection. Sometimes, the prominent granulomatous reaction can mask the presence of lymphoma. Careful histological evaluation combined with immunohistochemical assessment and clinical and radiological correlation avoid misdiagnosis.

**Keywords:** Hodgkin lymphoma, granulomatous reaction, tuberculosis

## CR 81

### **Oncocytic papillary cystadenoma of the parotid gland: a rare salivary gland neoplasm**

K. A. H. Seneviratne\*, T. G. Liyanage

*Department of Pathology, Faculty of Medicine, University of Ruhuna, Galle, Sri Lanka*

[hiruniseneviratne@gmail.com](mailto:hiruniseneviratne@gmail.com)

**Introduction:** Oncocytic papillary cystadenoma (OPC) is a rare benign salivary gland neoplasm, that accounts for 4% of all salivary gland neoplasms.

**Case report:** A 44-year-old man presented with a lump in the left preauricular region for one month. There was no increase in size or associated pain. Ultrasonography showed a well-defined, hypoechoic lesion in the left parotid gland. Cytology revealed collections of cyst macrophages, scattered haemosiderin-laden macrophages and lymphocytes. A few benign salivary duct epithelial cell clusters were identified. No squamous elements, mucinous cells or magenta colour stromal fragments were identified. The superficial conservative parotidectomy specimen showed a cystic lesion measuring 6x5x4 mm. Microscopy revealed the fibrous wall of a collapsed, unilocular cyst with intraluminal papillary projections. These papillae were lined by a cuboidal epithelium with bland nuclei and oncocytic cytoplasm. No complex papillary architecture, mucinous cells, lymphoid stroma, cytological atypia or mitoses were identified. The lesion was completely excised.

**Discussion and conclusion:** OPC should be distinguished from other benign and malignant tumours, including intraductal papilloma, low-grade mucoepidermoid carcinoma, Warthin tumour and cystadenocarcinoma. Intraductal papilloma is a well-defined unilocular cystic lesion with the duct lumen completely filled with broad papillae. Papillae in papillary cystadenoma occupy the lumen to a lesser degree. Low-grade mucoepidermoid carcinoma has a cystic and solid growth pattern with squamoid, mucinous and intermediate cells and an infiltrative growth pattern in contrast to OPC. Warthin tumour has a prominent lymphoid stroma. Cystadenocarcinoma shows mild nuclear atypia and has an infiltrative growth pattern. OPC has indolent behaviour and a good prognosis after complete excision.

**Keywords:** oncocytic papillary cystadenoma, salivary gland, benign neoplasm

**CR 82**

**Leiomyomatosis peritonealis disseminata in a patient with a high-grade endometrioid adenocarcinoma and extensive endometriosis**

G. W. C. M. Silva\*, P. H. Abeygunasekara

*Department of Histopathology, Apeksha Hospital, Maharagama, Sri Lanka*

[chanikamadhushani1988@gmail.com](mailto:chanikamadhushani1988@gmail.com)

**Introduction:** Leiomyomatosis peritonealis disseminata is a condition characterized by the presence of multiple benign smooth muscle nodules on the peritoneal surface of the pelvic and abdominal cavities.

**Case report:** A 47-year-old woman was investigated for abdominal distension and heavy menstrual bleeding for three months, and the USS revealed bilateral complex ovarian masses with cystic and solid components and a bulky uterus. During surgery, tumour deposits were suspected at multiple sites. The laboratory received multiple specimens: The uterus, bilateral ovarian masses and fallopian tubes, and specimens labelled as rectal deposits, small bowel deposits and omentum. Microscopy of the right ovarian mass showed a high-grade endometrioid adenocarcinoma with extensive endometriosis. The tumour showed focal cytoplasmic positivity for vimentin and was negative for ER and Napsin A. p53 showed a wild-type positivity. WT-1 was not available. The left ovarian mass was sampled extensively and showed foci of endometriosis. Microscopic examination of an area suggestive of capsular breach of the right ovarian mass, omentum and rectal and small bowel deposits showed multiple, small, well-defined nodules composed of fascicles of spindle cells with elongated monomorphic nuclei, moderate eosinophilic cytoplasm and indistinct cell borders. Nuclear atypia, mitoses or necrosis were not seen. The spindle cells showed diffuse cytoplasmic positivity for SMA and desmin, confirming the presence of leiomyomatosis peritonealis disseminata.

**Discussion and conclusion:** Although leiomyomatosis peritonealis disseminata is a benign condition, it can macroscopically mimic metastatic tumour deposits. In 10% of cases, it can co-exist with endometriosis, as in our case, and with endosalpingiosis. This condition is usually self-limiting but may recur if incompletely excised. Malignant transformation can occur rarely.

**Keywords:** leiomyomatosis peritonealis disseminata, SMA, desmin

**CR 84**

**Cellular variant of focal segmental glomerulosclerosis with foam cells: the rarest variant**

H.R.S.D. Sumanasekara<sup>1\*</sup>, S. Wijetunge<sup>1</sup>, K.H.M. Fernando<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

<sup>2</sup>*District General Hospital, Polonnaruwa, Sri Lanka*

[sachinthanasumanasekara@gmail.com](mailto:sachinthanasumanasekara@gmail.com)

**Introduction:** Focal segmental glomerulosclerosis (FSGS) is a common cause of glomerulopathy that exhibits histological heterogeneity in the quality and location of the glomerular lesions, which can be correlated with response to treatment. The cellular variant is the least common histological variant and can show foam cell proliferation without segmental scars. We report a case of a young boy found to have cellular FSGS with foam cells.

**Case report:** A 14-year-old previously well boy presented with generalized body swelling, frothy urine and increased blood pressure of one week. He showed proteinuria (2+) and microscopic hematuria without dysmorphic red cells or casts. The serum albumin level was low. Serum creatinine and ASOT levels were not elevated. The renal biopsy revealed 15 glomeruli, five showing segmental foamy cells associated with cellular proliferation. Extracellular matrix deposition, collapse of glomerular tufts and tip lesions were absent. Immunofluorescence showed granular capillary positivity (1+) for IgM and negativity for IgG, IgA and C3. Foamy cells were positive for the histiocytic marker CD68. A diagnosis of cellular FSGS with foamy cell proliferation was made. He failed to respond to the initial treatment with oral prednisolone for one month. Therefore, oral tacrolimus was added and continued with oral prednisolone, which was tapered over six months. The proteinuria improved, and the patient is currently in remission.

**Discussion and conclusion:** The presence of foam cells in glomeruli is seen in both tip and cellular variants of FSGS. Diagnosis of the cellular variant is vital as its prognosis is intermediate between other variants and can help to plan more personalized treatment protocols.

**Keywords:** focal segmental glomerulosclerosis, foam cells, tacrolimus

## CR 85

### Secondary cutaneous amyloidosis associated with a syringoma

H.R.S.D. Sumanasekara\*, D. Walisinghe, K. Masinghe

*National Hospital Kandy, Sri Lanka*

[sachinthanasumanasekara@gmail.com](mailto:sachinthanasumanasekara@gmail.com)

**Introduction:** Syringoma is a benign eccrine sweat gland tumour which commonly affects elderly women. It usually presents as a skin-coloured nodule in the lower eye lid or periorbital area. Syringoma is known to be associated with diabetes and Down syndrome, but cases with co-existing amyloidosis are extremely rare. This is a rare case of secondary cutaneous amyloidosis associated with an eyelid syringoma.

**Case report:** A-72-year-old woman with a history of diabetes mellitus and dyslipidaemia presented with a non-painful tender lump in her lower eye lid for four years, which had gradually increased in size. No other skin lesions were present. The lump was excised, and the biopsy measured 7x7x6 mm. Microscopic examination of the skin tissue showed dermal deposition of an amorphous eosinophilic material surrounded by a giant cell reaction. There were a few scattered duct-like and comma shaped structures. Congo red stain highlighted the eosinophilic material that was salmon-pink in colour and showed apple-green birefringence under polarised light. The immunohistochemical stain CK5/6 showed positivity in these areas, indicating keratinocytic origin. The diagnosis of a syringoma with stromal deposition of amyloid was made. Systemic amyloidosis was excluded by the medical team.

**Discussion and conclusion:** A commonly accepted theory of both primary and secondary cutaneous amyloidosis is that the filamentous degeneration of cytokeratins released from apoptotic keratinocytes forms amyloid K protein, and therefore amyloid deposition is seen in tumours of epidermal origin. This theory is demonstrated in this case as well.

**Keywords:** cutaneous amyloidosis, syringoma

**CR 86**

**Fibrosarcomatous dermatofibrosarcoma protuberans masquerading as a vascular malformation**

S. Thilakarathne<sup>1\*</sup>, M.V.C. De Silva<sup>2</sup>, M. Bopagoda<sup>1</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

<sup>2</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[ajithsj7@gmail.com](mailto:ajithsj7@gmail.com)

**Introduction:** Fibrosarcomatous dermatofibrosarcoma protuberans (FS-DFSP) is a form of tumour progression that occurs in 5-15% of DFSP with a high rate of metastasis and recurrence. We report a rare case of FS-DFSP, which mimicked a vascular malformation clinically and on imaging.

**Case report:** A 34-year-old woman presented with a mass in the right flank since childhood, which enlarged rapidly and turned red during her second pregnancy. The ultrasound scan suggested a vascular malformation. Although magnetic resonance imaging (MRI) was recommended, it was not done, instead, an excision was performed. The specimen comprised skin-covered tissue containing a white, nodular lesion with a vague whorled cut surface showing focal brown cystic areas. Microscopy revealed a circumscribed, focally infiltrative, unencapsulated tumour in the dermis, composed entirely of spindle cells predominantly arranged in long fascicles with a storiform pattern of arrangement in some areas. The tumour cells had scanty cytoplasm and uniform spindle-shaped nuclei with vesicular chromatin and indistinct nucleoli. Mitoses were 15/10 HPF. The stroma was minimal, collagenous and focally showed a haemangio-pericytomatous vascular pattern. The lesion infiltrated subcutaneous fatty tissue with a dense fascicular arrangement and focally infiltrated the overlying epidermis. Deep and radial resection margins were involved. Most tumour cells expressed strong diffuse positivity for CD34. However, the infiltrative areas with dense fascicles were negative for CD34. The cells were negative for EMA, S100 and BCL2. Histomorphology and immunophenotyping were suggestive of a FS-DFSP.

**Discussion and conclusion:** Though DFSP has nonspecific imaging characteristics on ultrasonography, a well-defined, hypoechoic, subcutis-based lesion with peripheral vascularity favours DFSP. As no reliable ultrasound features indicate sarcomatous changes, MRI and CT assist in establishing the diagnosis to ensure adequate resection. Proper correlation with clinical, radiological and histological findings is mandatory to minimize errors in the diagnosis of FS-DFSP.

**Keywords:** dermatofibrosarcoma protuberans, fibrosarcomatous change, vascular malformations, soft tissue sarcoma

## CR 87

### A case of hepatic hydatid cyst

H. G. D. Thilini\*, P. Amaraweera

Teaching Hospital, Karapitiya, Sri Lanka

[t.gamage1983@gmail.com](mailto:t.gamage1983@gmail.com)

**Introduction:** Hepatic echinococcosis is a rare disease entity. Human echinococcosis occurs following infestation with larval tapeworms of the genus *Echinococcus*. This disease is endemic in certain countries, including the Mediterranean region, and reported in immigrants from these countries.

**Case report:** A 65-year-old woman, who worked in Jordan presented with abdominal pain elevated liver enzymes and a cystic liver lesion on ultrasound examination. On macroscopic examination, the lesion comprised fragments of a cyst with multiple thin-walled saccules filled with serous to mucoid fluid. Microscopy showed a cyst wall lined by a thick eosinophilic acellular lamellated membrane, surrounded by granulation tissue with formation of fibrous tissue. The saccules contained keratinous material suggestive of parasitic cuticles. A few protoscolices without hooklets were also identified.

**Discussion:** Human infection with *Echinococcus granulosus* leads to development of hydatid cysts, most often in the liver and lungs and less frequently in the bones, kidneys, spleen, muscle and central nervous system. The asymptomatic incubation period can last for many years until hydatid cysts grow to an extent that triggers symptoms. Abdominal pain, nausea and vomiting are common presenting complaints in hepatic echinococcosis. Ultrasonographic imaging is the technique of choice for the diagnosis. Serologic tests for specific antibodies can support the diagnosis. Treatment involves percutaneous aspiration, surgical removal and anti-infective drugs.

**Conclusion:** Correct early diagnosis is vital for early intervention before rupture of cysts. Accurate preoperative diagnosis of hydatid disease can be made from personal history and typical imaging findings in non-endemic areas.

**Keywords:** hepatic hydatid cyst, echinococcosis, *Echinococcus granulosus*



## CR 88

### A rare case of ceruminous adenoma of the ear extending into the mastoid region

H. G. D. Thilini<sup>1\*</sup>, L.A.H.A. Albert<sup>2</sup>

<sup>1</sup>*Department of Pathology, Teaching Hospital, Karapitiya, Galle, Sri Lanka*

<sup>2</sup>*Department of Pathology, Teaching Hospital, Jaffna, Sri Lanka*

[t.gamage1983@gmail.com](mailto:t.gamage1983@gmail.com)

**Introduction:** Ceruminous adenoma is a benign tumour arising from cerumen-secreting modified apocrine glands of the external auditory canal.

**Case report:** A 74-year-old woman presented with right ear discharge and pain for one month. On examination, she was found to have a polypoidal lesion in the right ear canal extending into the mastoid region, which was excised. Macroscopy showed an 11x8x7 mm polypoidal mass. Histomorphology revealed an unencapsulated, well-demarcated lesion with solid and cystic areas. There were glandular structures composed of inner cuboidal cells with apical snouts and an outer spindled myoepithelial cell layer. The inner cells contained a yellow-brown granular cerumen pigment stained positively with PAS stain. There were solid nests of rounded cells with scanty cytoplasm bearing round to oval nuclei and stippled chromatin. Mitoses were sparse. Necrosis or nuclear pleomorphism was not present. Immunohistochemical stains CK7 and CD117 highlighted the luminal cells, while basal cells showed diffuse nuclear staining with P63. Dual cell population and immunohistochemical findings distinguished this tumour from other neoplasms that occur in this region. Excision of the lesion was complete.

**Discussion and conclusion:** Although uncommon, ceruminous adenoma is the most common external auditory canal tumour in the outer portion where the ceruminous glands are present. This must be distinguished from ceruminous adenocarcinoma, which is infiltrative with perineural invasion, irregular glands, nuclear pleomorphism, increased mitoses, including atypical forms and necrosis. Middle ear adenoma and paraganglioma needed exclusion as imaging showed extension into the mastoid region. Complete surgical excision must be achieved as recurrences are common.

**Keywords:** ceruminous adenoma, ear canal lesion

**CR 89**

**Idiopathic pseudotumour of pterygopalatine fossa: a diagnostic challenge**

P. D. K. Thiranagama<sup>1</sup>, M.D.J.C. Malagala<sup>1</sup>, S. Nanayakkara<sup>2</sup>, M.V.C. de Silva<sup>3</sup>, V. Gurusamy<sup>4</sup>,  
C. S. P. Sosai<sup>1</sup>

<sup>1</sup>*Department of Histopathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

<sup>2</sup>*Department of Histopathology, National Eye Hospital, Sri Lanka*

<sup>3</sup>*Department of Histopathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>4</sup>*Department of Ophthalmology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*

[prasangasiriwardena@gmail.com](mailto:prasangasiriwardena@gmail.com)

**Introduction:** Idiopathic pseudotumour is an umbrella term for nonspecific, non-neoplastic, chronic expansile lesions accounting for less than 5 % in the head and neck region. Aggressive behaviour and difficult histological typing make it a critical diagnostic challenge.

**Case report:** A 59-year-old man presented with rapid, progressive impairment of vision and intermittent proptosis of the left eye for three months, and examination revealed proptosis, afferent pupillary defect and mild limitation of extraocular movement. Contrast CT and MRI showed a mass in the pterygopalatine fossa extending to the temporal fossa. The radiological differential diagnoses were en plaque meningioma or lymphoma. ESR and CRP were initially raised but declined, and blood picture and ANCA levels were normal. A biopsy of the lesion revealed lobules of mature adipose tissue, variable stages of fibrosis with short spindle cells, and infiltrates of lymphocytes and plasma cells. Tumour necrosis, nuclear atypia or mitotic activity were absent. Histological differential diagnoses included granulomatous lesions and lymphoid and sclerosing tumours/lesions. Ziehl Neelsen stain was negative. Immunohistochemistry for SMA was positive in the spindle cells and, AE1/AE3, ALK-1, S100 and IgG4 were negative. The patient responded to steroid treatment with a reduction in tumour size.

**Discussion and conclusion:** Investigation findings excluded tuberculosis, IgG4-related disease, plasma cell granuloma, mast cell granuloma and diffuse neurofibroma. SMA positivity was in favour of sclerosing tumours/lesions, including inflammatory myofibroblastic tumours. ALK-1 negativity was reported in orbital inflammatory myofibroblastic tumours. Detection of clonal cytogenetic abnormalities is required for confirmation, which is not available in the local setting.

**Keywords:** inflammatory myofibroblastic tumour, pterygopalatine fossa, idiopathic pseudotumour

**Renin-secreting tumour of the kidney: a rare entity**

J. A. K. Udeshika<sup>1\*</sup>, A. V. T. D. Damayanthi<sup>2</sup>, M. M. A. Jayawickrama<sup>1</sup>

<sup>1</sup>*Department of Pathology, National Hospital of Sri Lanka*

<sup>2</sup>*Department of Histopathology, District General Hospital, Trincomalee, Sri Lanka*

[kaumadim@gmail.com](mailto:kaumadim@gmail.com)

**Introduction:** Renin-secreting renal tumours are rare mesenchymal tumours with a few reported cases in the literature. These tumours are derived from the modified smooth muscle cells in the walls of the afferent arteriole of the juxtaglomerular apparatus.

**Case report:** A 20-year-old woman was investigated for young hypertension and hypokalaemia. Contrast-enhanced computed tomography revealed a tumour in the lower pole of the left kidney, and she underwent a partial nephrectomy. Macroscopically, this specimen revealed a well-circumscribed tumour, measuring 17x12x10 mm, confined to the renal cortex. Microscopy revealed a well-circumscribed, unencapsulated cortical tumour composed of sheets of round to polygonal cells having central round vesicular nuclei and eosinophilic cytoplasm with well-defined cell borders. Atypia, increased mitoses or tumour necrosis were not seen. The stroma contained numerous dilated thin-walled vessels. Hemangiopericytoma-like vasculature was not evident. There was no evidence of capsular or vascular invasion. Immunohistochemistry revealed membranous positivity for CD34 and CD117 and cytoplasmic positivity for vimentin. The patient became normotensive shortly after surgery.

**Discussion and conclusion:** Morphological features and immunohistochemistry findings confirmed the diagnosis of a juxtaglomerular cell tumour. CD34, CD117 positivity and hypertension exclude the possibility of glomus tumour. Although juxtaglomerular cell tumours are rare, they should be considered in the differential diagnosis of renal neoplasms in young hypertensive patients as surgical excision is curative.

**Keywords:** juxtaglomerular cell tumour, renin, CD34, CD117, vimentin

## CR 91

### Silent intricacies: when a fatal snake bite unveils cardiac cysticercosis

V. Vaibhav<sup>1\*</sup>, S. Yashpal<sup>1</sup>, R. Meshram<sup>1\*</sup>, A. Singh<sup>2</sup>, G. Khorwal<sup>3</sup>

<sup>1</sup>Department of Forensic Medicine and Toxicology, All India Institute of Medical Sciences, Rishikesh, India

<sup>2</sup>Department of Pathology & Laboratory Medicine, All India Institute of Medical Sciences, Rishikesh, India

<sup>3</sup>Department of Anatomy, All India Institute of Medical Sciences, Rishikesh, India

[vikasvaibhav007@gmail.com](mailto:vikasvaibhav007@gmail.com)

**Introduction:** Snake bites remain a significant health concern in certain geographical regions, especially in the tropics. Prompt and appropriate medical attention is crucial to prevent fatal outcomes.

**Case report:** A 23-year-old man who suffered a snakebite was found dead on admission to hospital. Autopsy revealed significant cardiac abnormalities. The root of the aorta and aortic valve exhibited atheromatous changes with calcification. Notably, multiple collateral vessels were observed, originating from the left main coronary artery and extending along its course. The left main coronary artery showed atheromatous changes with a 40% lumen blockage, situated 0.5 cm distal to its origin. Numerous cystic nodules with calcified margins, containing muddy fluid, were discovered on the surface of the left ventricle, base of the aorta, left atrium and interventricular septum. Histology showed unremarkable cardiac myocytes with focal areas of fibrosis and a dense chronic inflammatory cell infiltrate. A cyst was identified within the myocardium, which was confirmed to contain a cysticercus larva. These features were consistent with cardiac cysticercosis.

**Discussion and conclusion:** Awareness of atypical presentations and simultaneous pathologies is essential in providing a comprehensive understanding of the cause of death and informing future medical practices and interventions. The findings underscore the importance of conducting detailed autopsy examinations to comprehensively evaluate complex clinical cases.

**Keywords:** cysticercosis, cardiac cysticercosis, parasitic infestation, intracardiac cysts

## CR 92

### Phosphaturic mesenchymal tumour: a rare entity causing tumour induced osteomalacia

I. L. Wickramanayake\*, C.M. Baranasuriya, S. Abeygunawardhane, H.D. Wijesinghe, M.V.C.de Silva  
*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*  
*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*  
[Indikalakmali507@gmail.com](mailto:Indikalakmali507@gmail.com)

**Introduction:** Phosphaturic mesenchymal tumours (PMT) are extremely rare benign neoplasms that present with tumour-induced osteomalacia due to the release of phosphatonins, a fibroblastic growth factor 23 (FGF23), which decrease renal reabsorption of phosphate and inhibit 1-alpha hydroxylase.

**Case report:** A 32-year-old man presented with a history of muscle pain, generalized ill health and left hip joint pain for one year. Biochemical studies revealed hypophosphatemia with normocalcaemia. MRI revealed a cortical-based, non-aggressive lesion in the left greater trochanter, and the PET scan concluded this as a benign osseous neoplasm. Additionally, there was a generalized reduction of bone density. Histologically, scooping of the bony lesion revealed tumour fragments composed of bland, spindle-shaped cells which showed haphazard and vague storiform arrangement in a background of a rich capillary-sized vascular network. The cells had small, bland oval nuclei and scanty cytoplasm. Mitosis and necrosis were absent. The tumour cells showed patchy, weak cytoplasmic staining with vimentin and SMA and negative staining with ERG, CD56, and CD34. Immunomorphological (IHC) features and clinicopathological correlation were consistent with PMT. The patient's clinical condition improved with surgical resection and treatments with phosphate buffers.

**Discussion:** The pathologic diagnosis was delayed due to the rarity of PMTs, vague symptoms of their presentation, and histologic overlap with other low-grade mesenchymal tumours such as non-ossifying fibroma, chondromyxoid fibroma, chondroblastoma and aneurysmal bone cyst. Awareness of PMT will help pathologists to make an accurate diagnosis, which has therapeutic and prognostic implications.

**Conclusion:** Clinical correlation with intensity of symptoms, imaging studies and strong positivity for FGFR1 by IHC and serological assessment of FGF23 are useful in the diagnosis.

**Keywords:** neoplasm, osteomalacia, fibroblast growth factors, hypophosphatemia

CR 93

**A rare case of systemic Epstein-Barr virus-positive T- cell lymphoma of childhood**

I. L. Wickramanayake<sup>1\*</sup>, H.D. Wijesinghe<sup>1</sup>, G. Ranaweera<sup>1</sup>, M.V.C. de Silva<sup>1</sup>, S. Gunaratne<sup>2</sup>

<sup>1</sup>*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

<sup>1</sup>*Centre for Diagnosis and Research in Cancer, University of Colombo, Sri Lanka*

<sup>2</sup>*Lady Ridgeway Hospital for Children, Colombo, Sri Lanka*

[Indikalakmali507@gmail.com](mailto:Indikalakmali507@gmail.com)

**Introduction:** Systemic Epstein-Barr virus (EBV)-positive T-cell lymphoma of childhood (SETLC) is a rare, rapidly progressive disease characterized by monoclonal expansion of EBV-positive T cells in tissues or peripheral blood.

**Case report:** A previously healthy four-year-old girl presented with acute onset high fever for four weeks associated with generalized lymphadenopathy and hepatosplenomegaly. Investigations revealed pancytopenia (white blood cell count -  $4 \times 10^9/L$ , platelet count -  $80 \times 10^9/L$ , haemoglobin - 8.4 g/dL) and high serum concentration of EBV/DNA. Bone marrow biopsy revealed haemophagocytosis. The submental lymph node biopsy showed paracortical expansion by histiocytes with haemophagocytosis and a diffuse infiltrate of medium-sized moderately atypical lymphoid cells. These cells were positive for CD3, CD8 and granzyme B and negative for CD4 and CD56.

**Discussion:** SETLC needs to be distinguished from systemic chronic active EBV infection (CAEBVI) with which it shows overlapping clinical and morphological features, including paracortical expansion of lymph nodes, infiltration of liver, spleen and bone marrow by EBV-positive T cells and haemophagocytosis. The morphological changes are less extensive in systemic CAEBVI. The T cells in SETLC are of CD8+ immunophenotype, while in systemic CAEBVI they are usually CD4+. Even in the absence of EBERISH, the acute onset of the disease, histomorphological evidence of haemophagocytosis, moderately atypical lymphocytes with positive staining with CD3, CD8 and granzyme B in the presence of high serum titre of EBV-DNA favoured a diagnosis of SETLC in this case.

**Conclusion:** Awareness of SETLC and systemic CAEBVI helps pathologists make an accurate diagnosis that has therapeutic and prognostic implications. Clinical correlation with the intensity of symptoms, immunohistochemistry and T-cell gene rearrangement studies are useful in the diagnosis.

**Keywords:** Epstein-Barr virus, lymphoma, haemophagocytosis

**CR 94**

**Malignant adenomyoepithelioma of the breast: a rare tumour**

K. V. C. Wijegunaratna\*, H. D. Wijesinghe

*Department of Pathology, CeDARC, Faculty of Medicine, University of Colombo, Sri Lanka*

[wijegunaratnac@yahoo.com](mailto:wijegunaratnac@yahoo.com)

**Introduction:** Malignant adenomyoepithelioma (MAME) is a rare tumour affecting salivary glands, lungs and breasts in a wide age range, with an increasing incidence with age. This is a case of malignant adenomyoepithelioma of the breast.

**Case report:** A 69-year-old woman presented with a left breast lump. Imaging showed a hyperdense lesion with well-circumscribed margins, compatible with a BIRADS 4b lesion. The core needle biopsy showed glandular lumina lined by a dual layer of cells and the stroma containing myoepithelial cells. An infiltrating malignant component was not seen in this specimen, and the differential diagnosis of adenoid cystic carcinoma and adenomyoepithelioma were considered. The myoepithelial cells showed p63 positivity, were negative for CD117, and showed low ER positivity and HER2 negativity. The wide local excision specimen showed a tumour with glandular spaces lined by epithelial and myoepithelial cells, and the stroma contained sheets of myoepithelial cells. The periphery of the lesion showed a Nottingham grade III invasive breast carcinoma of no special type. Focal squamous differentiation was seen.

**Discussion and conclusion:** MAME is a rare malignancy that arises from the malignant transformation of adenomyoepithelioma and shows atypia, infiltrative growth, high mitoses and necrosis compared to adenomyoepithelioma. The diagnosis can be challenging in core needle biopsies, and due to the presence of dual epithelium, the diagnosis can even be missed if the malignant component is not sampled. The differential diagnosis includes papilloma, epithelial myoepithelial carcinoma and adenoid cystic carcinoma. The prognosis depends on the subtype of the invasive tumour. Due to the low incidence, information on prognostic features and treatment is limited.

**Keywords:** adenomyoepithelioma, breast, malignant transformation

**CR 95**

**Neonatal death due to a multiseptated bladder causing complete urinary tract obstruction and pulmonary hypoplasia**

K.V.C Wijegunaratna\*, H. D. Wijesinghe

*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

wijegunaratnac@yahoo.com

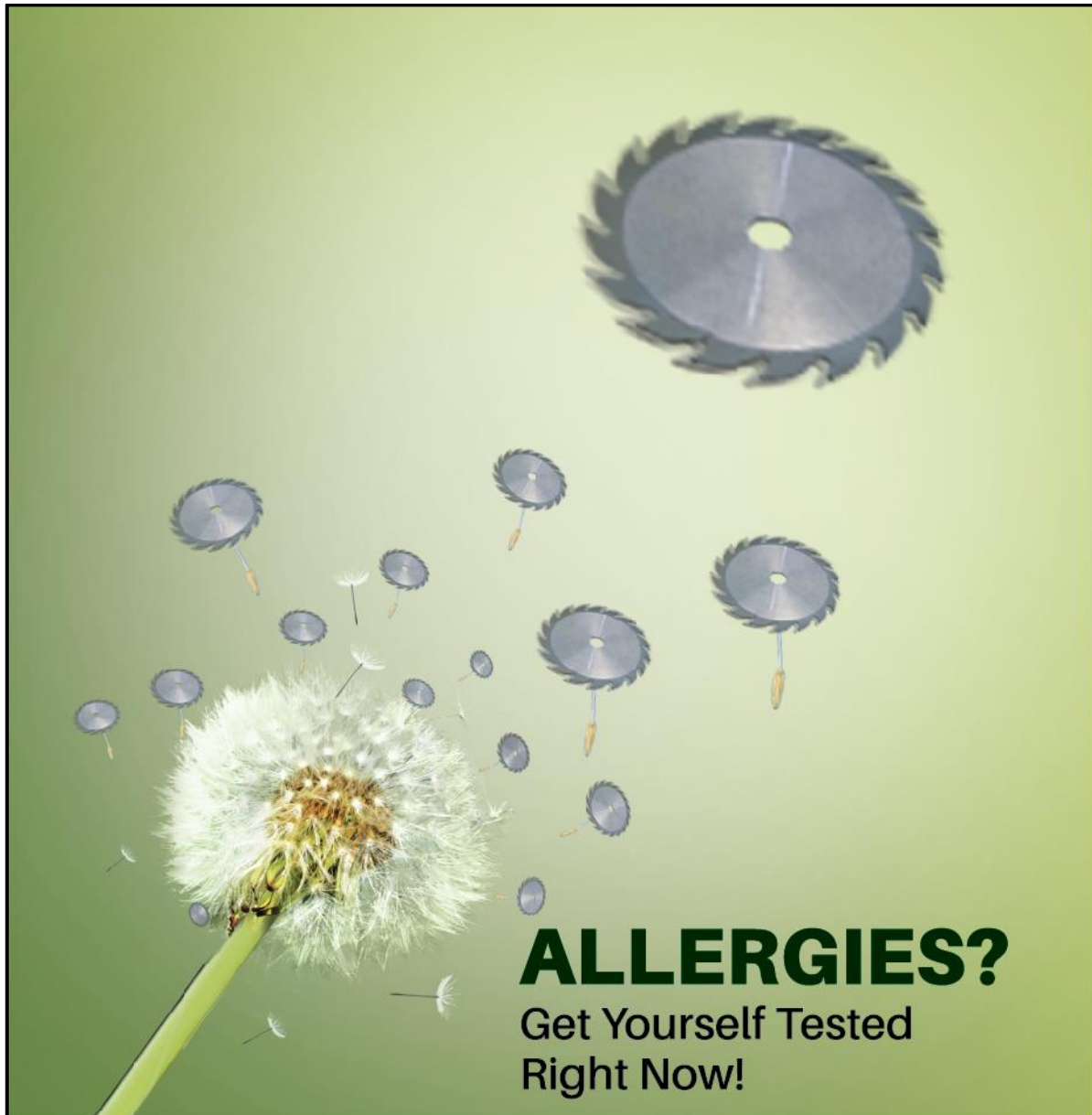
**Introduction:** Multiseptated bladder is a very rare congenital bladder anomaly results in intravesical obstruction and ultimate kidney failure if the septation is complete. Here we report a case of multiseptated bladder causing complete urinary flow obstruction and pulmonary hypoplasia resulting in neonatal death.

**Case report:** A 38-year-old pregnant mother was suspected to have anhydramnios at 30 weeks of period of amenorrhoea (POA). An ultrasound scan done at 31 weeks of POA showed gross dilatation of the urinary tract of the foetus with bilateral vesicoureteric obstruction and anhydramnios. During delivery, no liquor was detected, and the baby died due to respiratory failure. On postmortem examination, both lungs were hypoplastic. The kidneys, both ureters and bladder were grossly dilated. The bladder showed two complete septa separating the bladder into three chambers resulting in complete urinary obstruction. On microscopy, both kidneys showed severe cortical fibrosis. Both lungs showed a reduction of alveoli with fibrosis. No other congenital anomalies were detected.

**Discussion and conclusion:** Congenital anomalies of the bladder include agenesis, hypoplasia, congenital division and megacystis. Bladder septation is a rare congenital anomaly, and depending on whether the septation is complete or incomplete, obstruction can occur. A multiseptated bladder is a very rare form of bladder septation with only a few published case reports. If the septation causes complete obstruction of the urine outflow it results in anhydramnios, bilateral hydronephrosis/hydroureter, lung hypoplasia and uraemia. There is a possibility of reversing it by surgery in utero.

**Keywords:** multiseptated bladder, uraemia, complete bladder obstruction





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