Annual Academic Sessions of the The College of Pathologists of Sri Lanka 2021

Proceedings Booklet

RECENT UPDATES IN PATHOLOGY; TRANSLATION TO PRACTICE

Virtual Conference

13TH - 14TH OCTOBER 2021

Organized by
The College of Pathologists of Sri Lanka

Virtual Conference

On

RECENT UPDATES IN PATHOLOGY;

TRANSLATION TO PRACTICE

Organized by

The College of Pathologists of Sri Lanka

13th - 14th October 2021

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MESSEGE FROM THE PRESIDENT

COLLEGE OF PATHOLOGISTS OF SRI LANKA

As the president of the College of Pathologists of Sri Lanka, 2021, I am honoured and privileged to share this message with all of you on behalf of the members of the council and the organizing committee.

The Annual Academic Sessions 2021 is held as a virtual event, at a time when the dark shadows of the Covid-19 pandemic are looming over Sri Lanka, and it has been an extremely challenging task to organize this conference amidst these uncertainties.

The theme of this year's conference is "*Recent Updates in Pathology; Translation to practice"* and the sessions have been planned to cover a wide range of topics over a period of two days from 13th to 14th of October 2021 with the virtual participation of many eminent resource persons from Sri Lanka as well as from many parts of the world.

My deep appreciation is extended to Senior Professor Sudantha Liyanage, Vice Chancellor of the University of Sri Jayewardenepura and Professor Aloka Pathirana, Dean, Faculty of Medical Sciences, University of Sri Jayewardenepura for accepting my invitation to be the Chief Guest and the Guest of Honour at the Inauguration of the Sessions.

I am extremely grateful to all the resource persons, who readily obliged to be part of this conference at a time when the entire world is experiencing the effects of the pandemic. I am sure your contribution towards professional development of the pathologists in Sri Lanka will be appreciated by all the participants.

I wish to extend my gratitude to our Platinum, Gold, Silver and Bronze sponsors as well as 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 to the two coordinators of this event Dr. Charisma Shahi Fernando and Dr. Jayanjana Asanthi, the joint secretaries, the treasurer and the members of the council, organizing committee, academic committee and the abstract committee for their commitment and to everyone else who contributed in many ways to make this conference a reality amidst many uncertainties.

Prof. Dulani Beneragama

President, College of Pathologists of Sri Lanka, 2021

MESSEGE FROM THE CHIEF GUEST

I wish to thank the President, Council, and the organizing committee of the College of Pathologists of Sri Lanka for inviting me to be the Chief Guest at the Annual Academic Sessions, 2021 and I take this opportunity to send my best wishes to the College of Pathologists on this occasion.

With the increase of cancer patients locally as well as globally, pathologists have been performing a silent service to help in the management of these patients. The College of Pathologists of Sri Lanka, with a history of more than 40 years, has been the academic body and the driving force behind the professional development of pathologists in Sri Lanka.

The Annual Academic Sessions is the primary event responsible for dissemination of knowledge on the academic work carried out by its members during the past years and it is the forum designed to update knowledge on the latest developments in pathology.

The annual academic sessions this year is held with the participation of many eminent local and foreign resource persons with an impressive academic programme and updates. I congratulate the College of Pathologists of Sri Lanka for organizing this conference, at a time when the Covid-19 pandemic has restrained physical meetings to prevent spread of the disease. I am sure your efforts will not be in vain, and this event will help to upgrade the knowledge of the pathologists and thereby help to improve the quality of pathology services in Sri Lanka.

I congratulate the President, Council, and the organizing committee of the College of Pathologists of Sri Lanka for their dedication and commitment in organizing this event and sincerely hope for a successful two days which will be educative and memorable for all of you.

Senior Professor Sudantha Liyanage

BSc (Hons) (USJ), PhD (Cardiff), C Chem, FRSC, FIChem C, FPRISL

Vice-Chancellor – University of Sri Jayawaradenepura

MESSEGE FROM THE CONFERENCE COORDINATORS

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

As we move into the second year of the pandemic, we have tried our utmost not to let the current chaotic situation impede us in our quest for knowledge and further learning. This is our second year hosting a virtual conference, through which we bring to you all, a very esteemed and learned group of resource persons from all around the world namely UK, Australia, and India, including our very own home bred speakers who will be imparting their own take on recent updates in pathology and its translation to practice.

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 virtual event once again reiterates the absolute dedication and commitment of the college.

The objective of this year's program is not only to achieve high standards in the field of pathology through sharing of knowledge and expertise, but also to develop holistic pathologists through topics covering ethics in pathology and special inputs from other clinical fields including oncology and radiology.

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.

Dr. Charisma Shahi Fernando

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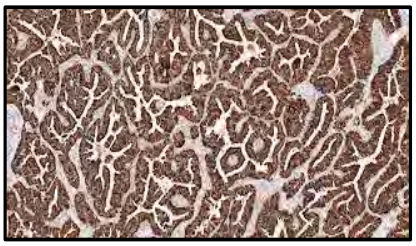

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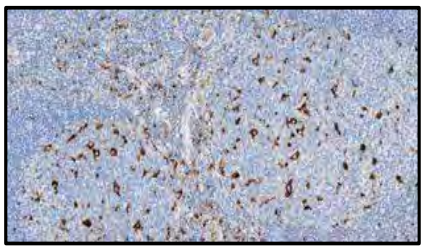

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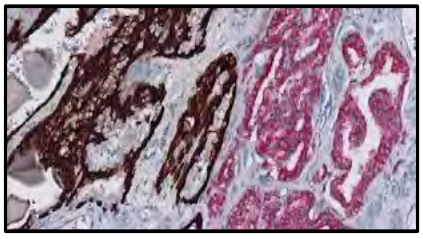

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THE PRESDENTIAL ADDRESS 2021

I wish to extend my heartfelt gratitude to the College of Pathologists of Sri Lanka for appointing me as the President for the year 2021. I am deeply humbled by this great honour bestowed upon me and I assure you that I will do my best to work towards achieving future goals of the college and its members. I solemnly pledge that I will serve the College of Pathologists of Sri Lanka to the best of my ability, worthy of the trust you have placed upon me during these very challenging times to steer the college in the right direction.

I wish to acknowledge the great work done by all the past presidents of the College who have steered this college to where it stands today, and I thank each and every one of them for a job well done. I also wish to extend my deep appreciation to the Immediate Past President, Dr. Shanika Fernandopulle, who has been a pillar of strength to me.

The theme of this year's academic sessions is *Updates in pathology; Translation to Practice* which is aimed at improving the quality of the diagnostic services in our country.

The academic sessions will be held virtually with the participation of many eminent guest speakers from Sri Lanka and across the world and I thank each and every one of them for having accepted our invitation to be part of this virtual conference.

I take this opportunity to congratulate the orator, Senior Professor Lakmini Mudduwa who will be delivering the Prof. Daphne Attygalle Memorial Oration this evening.

As we emerge from yet another wave of the Covid-19 pandemic and a month-long, nationwide lockdown, I thought it was appropriate to address you on the importance of *Resilience*, which seems to be the way forward towards a brighter future.

In December 2019, the beautiful city of Wuhan in the Hubei Province of China was engulfed by a dark cloud, and within a few days this once beautiful city was turned into a dead city by an unknown disease taking a heavy toll on the lives of the people in the city of Wuhan.

Very soon the entire world was to experience the evil effects of this ugly grey cloud causing havoc in countries big and small, and squeezing the very life out of people and the economies in these countries.

The effects were mostly felt in the developing countries like Sri Lanka with struggling economies and limited resources to deliver optimum care required for the patients. At this point, due to the lack of knowledge on this new and deadly disease, Sri Lanka, along with many other countries, had to enforce island wide lockdowns to save the lives of its people.

Despite taking steps to curtail the spread of the disease, the frontline health care workers and members of the forces have been overwhelmed by the emerging waves which have repeatedly engulfed the country from time to time and many health care workers have already succumbed to the disease. I take this opportunity to extend my deep appreciation to all my colleagues working on the front line and wish a speedy recovery to the critically ill patients across the country.

The covid-19 pandemic had major effects on the economy of our country which depends heavily on tourism and exports and with global lockdowns and discontinuation of air travel both these avenues came to a standstill.

During the last two years, our lives have changed dramatically, we have got accustomed to a new normal way of living, and we have forgotten, to certain extents, what normal was before.

Resilience, which by definition, is the capacity to recover quickly from difficulties seems to be the only way forward towards a better and brighter future.

I quote from the book *Fires of Heaven* by Robert Jorden *"The oak fought the wind and was broken; the willow bent when it must and survived."* This can be related to our current lives as well, emphasizing the fact that flexibility and adaptability has greater strength than rigid adherence to the past norms.

A quote by Elizabeth Edwards says *"Resilience is accepting your new reality, even if it's less good than the one you had before. You can fight it, you can do nothing but scream about what you've lost, or you can accept that and try to put together something that's good."*

To survive like the Willow, we must adapt, and adhere to the health care guidelines and social distancing and learn to live with our new reality.

The College of Pathologists has shown extreme resilience and has changed accordingly. The close consultative discussions, teaching and peer discussions around a microscope head have now been largely replaced by remote consultations.

The pen which has long been a close companion of the pathologist is now slowly being replaced by e-signatures and as the world is shifting to a digital platform, the hard copies of reports are now being stored as soft copies in databanks.

With the implementation of social distancing, meetings have become virtual or hybrid with minimum participation. Online education has become the new norm and the bonds between extended family has been via remote platforms.

Resilience is learning to accept these changes as part of the survival process and using these changes for a productive purpose while enjoying the beauty of life.

If you have ever felt darkness around you, be assured that you are not alone and always think of the light that is invariably present at the end of the tunnel.

Your capacity of resilience should not be deterred by the obstacles before you. It should be aimed at fighting the disease keeping aside political and ideological differences which will hamper your chances of survival.

Avoid the IDIOTs obstructing your path of survival. These IDIOTs are not the ones that generally comes to mind, but instead refers to 'Internet Derived Information Obstructing Treatment.' Discard the myths that reach you daily and continuously through your social media platforms sharing unverified and unscientific stories of survival.

Instead, embrace evidence-based medicine and guidelines recommended by the World Health Organization and the Centre for Disease Control.

Maya Angelou once said, "I can be changed by what happened to me, but I refuse to be reduced by it". The college has been resilient in facing this challenge, by continuing its academic activities on virtual platforms. The conventional seminars were converted to webinars, and we have had many such webinars with the participation of resource persons from Sri Lanka and abroad who showed equal resilience by rising to the occasion and mastering the challenges in technology.

The college has not stayed idle during this crisis and used these recurrent lockdowns to improve and uplift the pathology services in the country to improve the quality of cancer diagnosis and reporting. We have completed five guidelines, and these will be printed by the Ministry of Health to be distributed among the pathologists working in the peripheries.

During this year the college was also able to formulate its own Continuous Professional Development Scheme for the benefit of its members.

In addition, as a stepping-stone towards accreditation of the histopathology laboratories in government sector hospitals, the college has conducted a series of quality assurance workshops in collaboration with Sri Lanka Accreditation Board, and I take this opportunity to thank all the resource persons and the coordinators for the extreme resilience shown by conducting these workshops virtually for the very first time.

A captivating quote by Caryn Sullivan says, “In the face of adversity, we have a choice: We can be bitter, or we can be better” and I am happy to say that the College of Pathologists of Sri Lanka has thrived to become better in this face of adversity.

There is a silver lining in every dark cloud, and in this pandemic, there have been many silver linings. One such silver lining is the exponential advances in Information Technology which has paved the way from the toddler to the elderly to adapt to their new normal lives. It is important to reflect upon these silver linings and the lessons learnt from the pandemic and carry these forward to build a better tomorrow.

The social distancing norms have taught us meeting etiquette during our day-to-day work and we have seen unruly crowd behavior being slowly replaced by orderly, disciplined behavior.

Death and disease around us have forced us to live a life of simplicity, to live without any regrets and to be happy with how you look and what you have.

Until we had to be suffocated by wearing masks, the fresh air we breathe seemed so inexpensive. This pandemic has taught us to respect our environment and strive hard to conserve Mother Nature. Little did we realise, that the newspapers that we read and then threw away, the documents that filled our houses and ended up in the bins, and the piles of books that were stacked on our tables never being looked at, were at the cost of thousands of trees cut daily throughout the world. Thanks to the pandemic, the newspapers, documents, and books have now been largely replaced by e-documents, e-papers and e-books, paving the way for a greener world.

COVID has taught us the value of money and shown us that conferences can be held equally well and even better via a virtual platform, saving not thousands but millions of rupees spent on lavish but unhealthy food and unnecessary glamour. It has taught us to appreciate that a good home-cooked meal is better than the unhealthy food that we were accustomed to in the pre-covid era.

Social distancing has taught us the importance of family and to cherish and enjoy your family while you can.

If you feel that you are struggling at any time, laugh at yourself as you muddle through, and believe that the worst is over. We hope and pray that we will be able to move towards a better and a brighter future with children back in schools and in play grounds, the healthcare workers relieved from their suffocating masks and that we would be able to share a meal with our family and friends without any restraints.

Through resilience learn to be a survivor, to tell your story of survival to your children, grandchildren and may be to your great-grandchildren.

Prof. Dulani Beneragama

Presidential Address,

College of Pathologists of Sri Lanka, 2021.

THE COLLEGE OF PATHOLOGISTS OF SRI LANKA

FELLOWSHIP AWARDS 2021

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National Hospital of Sri Lanka

Dr. Palitha Ratnayake

Department of Pathology,
National Hospital Kandy

Professor Lakmini Kumari Boralugoda Mudduwa

Department of Pathology,
Faculty of Medicine,
University of Ruhuna.

PROFESSOR DAPHNE ATTYGALLE MEMORIAL ORATION

ORATOR:

Professor Lakmini Mudduwa

MBBS, Dip Path, MD Path (Histopathology)

Department of Pathology,

Faculty of Medicine,

University of Ruhuna.

Prognosis research in breast cancer; lessons from Southern Sri Lanka

Abstract

Prognosis research are important in identifying and validating prognostic and predictive factors of diseases. They are useful to make health care more effective and to implement new standards of high-quality care and improve patient outcomes. However, there is a gap between the potential and actual impact of prognosis research on health. This gap is attributable to many factors which include the inconsistent use of definitions in clinical outcome measures and the pitfalls in outcome analysis leading to limitations in cross comparison of prognosis research.

Four prognosis research carried out on a large cohort of breast cancer patients are discussed highlighting the value of collecting refined outcome data of breast cancer patients.

GUEST SPEAKERS

Dr. Chinthaka Appuhamy

Consultant Radiologist
Colombo North Teaching Hospital
Sri Lanka

Prof. Wendy Cooper

Clinical Professor
Royal Prince Alfred Hospital
& University of Sydney,
Australia

Dr. Ian Cree

Senior Pathologist,
Section & Group Head
WHO Tumour Classification Group
WHO - IARC, France

Prof. Chandu de Silva

Senior Professor of Pathology
University of Colombo
Sri Lanka

Dr. Anusha Ginige

Consultant Pathologist
District General Hospital Vavuniya
Sri Lanka

Prof. Janaki Hewavisenthi

Senior Professor of Pathology
University of Kelaniya
Sri Lanka

Prof. Thomas Jacques

Prof of Paediatric Neuropathology
UCL GOS Institute of Child Health

Dr. Zane Jaunmuktane

Consultant Neuropathologist
University College London Hospitals

Dr. Amritha Jay

Consultant Histopathologist
University College London Hospitals
United Kingdom

Dr. Nuradh Joseph

Consultant Oncologist Teaching Hospital Batticaloa Sri Lanka

Prof. Priyanthi Kumarasinghe

Professor & Consultant Pathologist University of Western Australia PathWest QEII Medical Centre, Australia

Prof. Dilani Lokuhetty

Senior Professor in Pathology University of ColomboSri Lanka

Prof. Santosh Menon

Consultant Pathologist Tata Memorial Centre India

Dr. Angeles Montero

Consultant Respiratory Pathologist Manchester University Hospitals United Kingdom

Prof. Marco Novelli

Prof of Gastrointestinal Pathology University College London Hospitals United Kingdom

Dr. Lakmali Paranahewa

Interventional RadiologistThe Central Hospital, Colombo Sri Lanka

Dr. Isha Prematilleke

Consultant Pathologist University of Sri Jayawardenepura Sri Lanka

Dr. Niluka Ranathunga

Consultant PathologistDistrict General Hospital Mannar Sri Lanka

Dr. Gayani Ranaweera

Consultant Pathologist University of Colombo Sri Lanka

Prof. Bharat Rekhi

Professor & Consultant Pathologist Tata Memorial Centre India

Dr. Manuel Rodriguez-Justo

Consultant Pathologist University College London Hospitals United Kingdom

Dr. Shirani Samarathunga

Consultant Pathologist District General Hospital Nuwara Eliya, Sri Lanka

Dr. Anna Saparamadu

Consultant Pathologist Northampton General Hospital United Kingdom

Dr. Katherine Vroobel

Consultant Histopathologist The Royal Marsden Hospital United Kingdom

Dr. Harshima Wijesinghe

Consultant Pathologist University of Colombo Sri Lanka

PROGRAMME

Day 1 **13.10.21**	**7.50 - 8.00**	**Welcome address** **Prof. Dulani Beneragama** **President, College of Pathologists of Sri Lanka, 2021**
	Day 1- Session 1 **Chairpersons**	**8:00 - 10:30** **Prof. Gayana Mahendra** **Prof. Dulani Beneragama**
	8:00 - 08:40	**Pitfalls in the diagnosis of bone tumours** ***Prof. Chandu de Silva***
	8:40 - 09:20	**Recent updates in WHO 2020 classification of soft tissue tumours** ***Prof. Bharat Rekhi***
	9:20 - 10:00	**Prostate cancer pathology; from basics to the future** ***Prof. Santosh Menon***
	10:00 - 10:30	**Prostate cancer pathology; from basics to the future** ***Dr. Isha Prematilleke***
	10.30 - 10.45	**Tea break**
	Day 1- Session 2 **Chairpersons**	**10.45 - 11:30** **Dr.Palitha Rathnayake** **Dr. Anusha Ginige**
	10.45 – 11.00	**Radiological correlation of breast lesions** ***Dr. Chinthaka Appuhamy***
	11.00-11.15	**Radiological correlation of thyroid lesions** ***Dr. Chinthaka Appuhamy***
	11.15-11.30	**Radiological correlation of hepatobiliary lesions** ***Dr. Lakmali Paranahewa***
	11:30 - 12:15	**Towards precision medicine in breast and gynaecological cancer; the role of the pathologist** ***Dr. Gayani Ranaweera, Dr. Harshima Wijesinghe, Dr. Nuradh Joseph***
	12.15 -13.00	**Lunch break**
	Day 1- Session 3 **Chairpersons**	**13.00 - 15:30** **Prof. Chandu de Silva** **Prof. Dulani Beneragama**
	13.00 – 13.30	**Approach to classification of tumours** ***Dr. Ian Cree***
	13.30- 14.00	**An update on the 5th series of WHO Classification of CNS tumours** ***Prof. Thomas Jacques***
	14.00- 14.30	**An update on the 5th series of WHO Classification of thoracic tumours** ***Prof. Wendy Cooper***
	14.30 – 15.00	**Pivotal role of the pathologist in driving tumour classification** ***Prof. Dilani Lokuhetty***
	15.00 - 15.30	**Q & A session** ***Dr Ian Cree, Prof. Thomas Jacques, Prof. Wendy Cooper & Prof. Dilani Lokuhetty***
	15.30 - 15.45	**Tea break**
	Day 1- Session 4 **Chairpersons**	**15.45 - 16:45** **Dr. Roshana Constantine** **Dr. Jayanjana Asanthi**
	15.45 – 16.15	**An update in interstitial lung disease** ***Dr. Angeles Montero***
	16.15 - 16.45	**Molecular diagnostics of adult CNS tumours with conventional and advanced technologies** ***Dr. Zane Jaunmuktane***

Day 2 **14.10.21**	**Day 2- Session 1** **Chairpersons**	**8:00 – 9.30** **Oral Presentations** **Prof.. Dulani Beneragama** **Prof. Priyani Amarathunga**
	Day 2 – Session 2 **Chairpersons**	**9.30 – 10.45** **Dr. Shanika Fernandopulle** **Dr. Gayani Ranaweera**
	9:30 – 10.00	**How important is humanities in Pathology?** ***Prof. Janaki Hewavisenthi***
	10.00 – 10.45	**Case discussion** **Dr. Niluka Ranathunga, Dr. Shirani Samarathunga, Dr. Anusha Ginige**
	10.45 - 11.00	**Tea break**
	Day 2- Session 3 **Chairpersons**	**11.00 - 12:30** **Dr. Isha Prematilleke** **Dr. Harshima Wijesinghe**
	11.00 – 11.45	**Tips and tricks in gastrointestinal pathology** ***Prof. Priyanthi Kumarasinghe***
	11.45-12.45	**Cervical and vaginal tumours; an update** ***Dr. Anna Saparamadu***
	12.45 -13.30	**Lunch break**
	Day 2- Session 4	**13.30 - 14:00** **Pathology Quiz** **Conducted by Dr. Dinesha Jayasinghe**
	Day 2- Session 5 **Chairpersons**	**14.00 -15.00** **Prof. Janaki Hewavisenthi** **Dr. Charisma Shahi Fernando**
	14.00 – 14.30	**Role of local resections in early neoplastic disease of the oesophagus and stomach** ***Prof. Marco Novelli***
	14.30- 15.00	**Lower gastrointestinal pathology; an update on molecular targets** ***Dr. Manuel Rodriguez –Justo***
	15.00 - 15.15	**Tea break**
	Day 2- Session 6 **Chairpersons**	**15.15 - 16:15** **Dr. Mihiri Madurawe** **Dr. Charisma Shahi Fernando**
	15.15 – 15.45	**Recent advances in endometrial pathology** ***Dr. Katherine Vroobel***
	15.45 - 16.15	**Salivary gland tumours; an update** ***Dr. Amritha Jay***
	16.15 -17.00	**Closing Ceremony** **Awards** **Vote of thanks**

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Chairpersons: Professor Dulani Beneragama **Professor Priyani Amarathunga**		
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RP 9	Proliferative epithelial changes associated with the novel breast carcinogenesis molecular models: a Sri Lankan study on women with breast cancer	**G.I.M. Jinadas**a, H.D. Wijesinghe, M.M.A. Jayawickrama, M.D.S. Lokuhetty
RP 11	Diagnostic utility of ultrasound scanning, fine needle aspiration cytology and cell block technique in the pre-operative diagnosis of suspicious thyroid nodules	**Kulatunga KMHH**, Pathirana AA, Fernando SSN, Gamage BD, Epa A, Sampath MKA, Sosai C, Seneviratne BS*
RP13	Comparison of oestrogen receptor (ER) and progesterone receptor (PR) status of breast cancer tissue, processed in-house and in external laboratories: an internal audit.	**F.N. Nawas**, N. Meedeniya, A. Ruwan, A.A.H. Priyani*
RP 16	Comparison of expression of ER, PR and p53 immunohistochemical stains between endometrial curetting / biopsy and resected endometrial carcinoma specimens	**K. G. H. Silva*[1]**, B. A. G. G. Mahendra[1], D. Ediriweera[2], J. Hewavisenthi[1]
RP 17	A comparison of cytology and radiological score with histology in patients with breast lumps; what guides the clinicians best	**H. G. D. Thilini***, A. Perera, L. Mudduwa,

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RP 01

The correlation between ß-catenin expression pattern and adverse prognostic factors in a cohort of invasive breast carcinoma patients in Sri Lanka.

L.D.S. De Silva*[1], P.R. Rathnayake[2], A. Vithanage [2,] E.H. Siriweera[1]

[1]*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[2]*Department of Pathology, National Hospital Kandy, Sri Lanka*

shashids84@gmail.com

Introduction and objectives: Breast carcinoma (BCa) is the most common malignancy among females and the second most common cause of cancer related deaths. Due to heterogeneity of BCa, morphological classifications with molecular parameters are being studied widely, the subcellular location of ß-catenin being one of them. This study was carried out to determine the expression pattern of ß catenin and its association with adverse prognostic factors in a cohort of BCa patients in Sri Lanka.

Method: A cross sectional descriptive study was performed on 147 cases of BCa, including mastectomies and wide local excisions received at two teaching hospitals. Details of age, histological grade, lymphovascular invasion, Paget disease, ductal carcinoma in-situ (DCIS) lymph node involvement and hormone receptor status were retrieved from the request forms and reports. The expression of ß-catenin at the membranous, cytoplasmic and nuclear location was assessed by immunohistochemistry on a representative block. The intensity of expression was scored +1 to +3 in comparison to selected controls. The histoscores were calculated by multiplying the intensity score by the percentage of positive cells. The correlation between ß-catenin expression and adverse prognostic factors were calculated using Pearson correlation.

Results: Out of 147 cases, 123 (84.2%), 83 (57.3%) and 18 (12.3%) cases showed membranous, cytoplasmic and nuclear ß-catenin expression, respectively. There was a statistically significant correlation between nuclear ß-catenin histoscores and lymph node positivity and Paget disease.

Conclusion: Lymph node positivity, a known adverse prognostic factor, showed significant correlation with nuclear ß-catenin expression. Nuclear ß-catenin positivity showed a positive correlation with Paget disease, but its prognostic significance is yet to be determined and merits further studies.

RP 02

A descriptive study of histopathological features of breast carcinoma: a single center experience

K. Dharani *[1], D.N.U. Jayatunge[1], B.N.L. Munasinghe[2], B.A.G.G. Mahendra[1]

[1]Department of Pathology, [2]Department of Surgery, Faculty of Medicine, University of Kelaniya, Sri Lanka

dharani_ghs@yahoo.com

Introduction: Breast carcinoma (BC) has increased in incidence over the years, and it is the most common cancer among females in Sri Lanka.

Objective: To describe the histopathological features of BC in a tertiary care setting in Sri Lanka.
Method: Clinicopathological data was retrieved from requests and histopathological reports of BC resection specimens, at the Unit from January 2018 to March 2021.

Results: 100 cases of BC were analysed. The mean age at presentation was 57.5 years (27-86). 86 (86%) invasive BC of no special type (NST), 5 (5%) metaplastic carcinoma and 3 (3%) mixed lobular-ductal type were identified. Mucinous, lobular and micropapillary carcinoma accounted for one case each. 18 (18%), 35 (35%) and 47 (47%) were of histological grade 1, 2 and 3, respectively. 21.43%, 56.12%, 20.41% and 2.04% were pT1, pT2, pT3 and pT4, respectively. 24.1% were N1, 21.9% were N2, while 42.6% were negative for nodal metastasis. An in-situ component was seen in 40.7%. Lymphovascular and perineural invasion were seen in 40.8% and 6.4%, respectively. 12% showed tumour at the resection margin. The average tumour size was 36.6 mm (11-105) while the average Nottingham prognostic index (NPI) was 4.86 (2.2-7.9).

Discussion and conclusion: The mean age at presentation of BC is 57.5 years, which is lower than the mean age in a previous study in Sri Lanka in 2010. The commonest type is invasive BC-NST (86%). Most of the patients presented at the pT2 stage and most were histological grade 3.

RP 03

An audit on muscle biopsies in a tertiary care centre in Sri Lanka

G. S. S. Hegoda*, L.D.S. De Silva, N. V. I. Ratnatunga, R Waduge, T.R.D.S.K. Tennakoon

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

sanwimck@gmail.com

Introduction and objectives: Muscle biopsies are valuable in evaluating muscle disorders. The Department of Pathology, Faculty of Medicine, Peradeniya is the only center in the country which perform enzyme assays and immunohistochemistry in muscle biopsies. This study aims to determine the prevalence of diseases diagnosed in muscle biopsies.

Method: This is a retrospective study carried out using muscle biopsies received from 2009 to 2020. Clinicopathological data and diagnoses were extracted from the archives, recorded and analyzed using MS Excel software. Patients under 18 years were categorized under paediatric group.

Results: 593 muscle biopsies have been reported during the study period. There were 364 paediatric biopsies, out of which 38% (n=137) were muscular dystrophies. Out of muscular dystrophies, 29% (n=40) were diagnosed as Duchenne muscular dystrophy and 11.7% (n=16) as Becker muscular dystrophy. 19% (n=70) of paediatric biopsies showed a congenital myopathy. Out of 229 adult biopsies, 26% (n=59) were diagnosed as muscular dystrophy, 31.4% (n=72) as inflammatory myopathies and 10.5% (n=24) were non-specific myopathies.

Discussion and conclusion: Worldwide prevalence rates of muscle diseases vary with age, location and study population. Duchenne muscular dystrophy is the commonest muscular disease among children which is the same in our study, and adult prevalence rates are variable among different studies. No studies on prevalence rates among Sri Lankans have been carried out to date, highlighting the need for prevalence studies on muscle diseases in Sri Lanka.

RP 04

Evaluating the usefulness of liver biopsies in paediatric liver diseases

A.G.B.A. K Jayarathna*[1], J. Hewavisenthi[1], S.A. Gunaratne[2], P. Jayakody[3]

[1]*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

[2]*Department of Pathology, Lady Ridgeway Hospital for Children, Colombo, Sri Lanka*

[3]*Ministry of Health, Sri Lanka*

achanijayarathne@gmail.com

Introduction: Novel diagnostic methods in the diagnosis of paediatric liver diseases have developed rapidly, including electron microscopy, imaging, serology, and genetic studies. Nevertheless, conventional diagnostic methods like liver biopsy continue to play a pivotal role.

Method: The clinical diagnosis and histological diagnosis of liver biopsies received at Lady Ridgeway Hospital were reviewed during a one-year period beginning from 01st March 2020. These were categorized as: Category 1 - non diagnostic (due to inadequate or unsatisfactory specimens), Category 2 - neither exclude/confirm the clinical diagnosis nor provide prognostic or therapeutic information (e.g. descriptive reports), Category 3 - confirms the clinical diagnosis but does not provide further information such as aetiology, Category 4 - confirms the clinical diagnosis and provides further information important for patient management and Category 5 - changes the clinical diagnosis completely. Thus, categories 1 and 2 were deemed as having no impact on the management of the patient, while categories 3, 4, and 5 were considered as valuable in the management of the patient.

Results: There were 61 liver biopsies. No unsatisfactory/inadequate samples were received (Category 1). Only 8.2% (5/61) of the biopsies generated descriptive reports (Category 2). 29% (18/61) liver biopsies confirmed the clinical diagnosis without providing additional information (Category 3) and 42% (26/61) of the biopsies provided further useful information (Category 4). The histology changed the clinical diagnosis in 20% (12/61) of the biopsies (Category 5).

Conclusion: 92% (56/61) of the biopsies had a significant impact on the diagnosis and management of paediatric liver diseases.

RP 05

What are the best colours and colour combinations useful in inking surgical margins?

D. N. U. Jayathunge*[1], K.G.H Silva[1], S. Thilakaratne[1], D. Ediriweera[2], J. Hewavisenthi[1]

[1]Department of Pathology, [2]Centre for Health Informatics, Biostatistics and Epidemiology, Faculty of Medicine, University of Kelaniya, Sri Lanka

nishaniu@kln.ac.lk

Background: The involvement of surgical margins is a key factor determining the prognosis and management of malignancies. Some specimens require inking up to six margins.

Objectives:

1. To determine the best colours to ink margins using two freely available local paint brands, Fabrica [FB] and Fevicryl [FV].
2. To determine the best colour combinations to assess two adjacent surgical margins.

Method: Initially, the circumference of a total of 140 sections of lipomatous tissue, (1x1cm), were painted with black (BK), blue (BL), brown (BR), green (GR), orange (OR), red (RE) and yellow (YE) using FV and FB. Two investigators separately scored the colour retained [circumferential score 1-4 (1-25%-1, 26-50%-2, 51-75%-3, 76%-100%-4)], the intensity and the original colour score. A composite grade was obtained by the sum of the Z-scores of these three. Secondly, the four best colours, FV-BK, FV-GR, FB-BL and FV-RE, were used to paint the circumference of 20 sections of lipomatous tissue (2x2 cm) to get six combinations (BK-GR, BK-BL, BK-RE, GR-BL, GR-RE and RE-BL). The Z-scores were derived from the differentiation scores given by two independent investigators. The intensity score, original colour score and differentiation scores (1-4) were graded as:1- barely visible X400, 2 - visible X400, 3 - visible X100, 4 - visible X40. Two-way analysis of variance (ANOVA) and a post hoc mean comparison (Tukey HSD) were performed.

Results: Higher standardized composite scores were obtained for FV compared to FB, except for BL and OR. FV-BK had the highest score, while FV-OR scored the lowest. The best colour combinations were BK-GR, GR-RE and BL-RE, while GR-BL was the worst.

Conclusion: Black, green, red, brown (Fevicryl) and black, blue, orange (Fabrica) were the seven best colours. The best combinations were black-green, green-red (Fevicryl), and blue (Fabrica) with red (Fevicryl).

RP 06

Is online teaching of pathology for undergraduates effective?

D. N. U. Jayathunge*, S. Thilakaratne, J. Hewavisenthi

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

nishaniu@ymail.com

Background: The COVID-19 pandemic compelled universities to adopt online teaching (OT). The efficacy of OT needs to be determined and student feedback is one method.

Objective: To compare student perception on OT in pathology with traditional on-site teaching (TOST) and with OT of other disciplines (e.g. parasitology, microbiology, pharmacology, etc.).

Method: Two batches of students who had experienced both OT and TOST in Pathology from the Faculty of Medicine, University of Kelaniya, were selected. A Google form was used to obtain feedback.

Results: Response rate was 58.5% (n=196) after two reminders. 41.3% (n=81) liked only OT, 43.4% (n=85) liked both OT and TOST while, 15.3% (n=30) liked TOST only. The majority (64%; n=103) liked OT due to the availability of recorded study material for future use. 59.8% (n=116) favoured lectures pre-recorded, 8.8% (n=17) on Zoom and 31.4% (n=61) both. Online practicals were considered less effective, similar, and more effective by 42.3% (n=83), 20.4% (n=40) and 37.2% (n=73), respectively, while tutorial classes in OT were regarded more effective, similar and less effective by 57.1% (n=112), 17.3% (n=34), 25.5% (n=50), respectively, than in TOST. The following OT activities in pathology were rated as good, very good or excellent in comparison with other disciplines: lectures 82.7% (n=162), modified practicals 68.4% (n=134), tutorials 80.1% (n=157) and interactive sessions (quizzes, discussion forums, lessons) 77.4% (n=151). When asked as a closed-ended question, if there was a difference in the quality of teaching in pathology between OT and TOST apart from technical errors, 82.4% (n=159) did not perceive a difference.

Conclusion: Most responders liked OT due to the availability of recorded-study material. While a majority considered online practicals less effective, online tutorial classes were rated effective compared to TOST. Students perceived no difference in the quality of teaching in Pathology by the two methods. They rated OT in pathology effective compared to OT of other disciplines.

RP 07

The development of a simple protocol for successful inking of surgical margins with easily accessible colouring agents

D. N. U. Jayathunge*[1], K.G.H Silva[1], S. Thilakaratne[1], D. Ediriweera[2], J. Hewavisenthi[2]

[1]Department of Pathology, [2]Centre for Health Informatics, Biostatistics and Epidemiology, Faculty of Medicine, University of Kelaniya, Sri Lanka

nishaniu@ymail.com

Background: Inking enables precise assessment of the surgical margins, which has important therapeutic implications.

Objectives: To determine the impact of using acetic acid and the method of air drying and fixation on the retention, intensity and the original colour of the agent used.

Method: A total of 120 lipomatous tissue sections measuring 1x1x0.5 cm were used, 60 sections were painted with Fevicryl-black and 60 sections with Fevicryl-orange, the best and worst colours as determined in a previous study, respectively. 40/120 sections were used to assess the effect of acetic acid (20 with and 20 without acetic acid), and 20 sections each to assess four different methods of air drying and fixation. All sections were routinely processed and stained.

Three scores were obtained by two independent investigators; the retention along the circumference from 1-4 (1-25%-1, 26-50%-2, 51-75%-3, 76%-100%-4), the intensity and preservation of the original colour (each graded as 1 - barely visible at x400, 2 - visible at x400, 3 - visible at x100, 4 - visible at x40). A composite grade was obtained by the sum of the Z-scores of these three. Two-way analysis of variance (ANOVA) and a post hoc mean comparison (Tukey HSD) were performed to analyse the data.

Results: There was a significant difference in standardized composite scores with respect to the use of acetic acid in case of the worst colour, orange ($p = 0.0483$) but none when considering the four methods adopted in air drying/ fixation.

Conclusion: It is recommended that acetic acid is used routinely when inking specimens to obtain optimal results.

RP 08

Metaplastic breast carcinoma: analysis of clinical and pathologic characteristics

D.N.U. Jayathunge*, D.T.T. Jayasinghe, N. Wickramarathne, T.P.M. Bopagoda, B.A.G.G. Mahendra

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

nishaniu@ymail.com

Introduction and objectives: Metaplastic breast carcinoma (MBC) is a rare, aggressive entity accounting for less than 1% of breast malignancies. It is characterized by the differentiation of neoplastic cells towards squamous and/or mesenchymal elements and has a poor prognosis due to its triple-negative status. The objective of the study was to analyse the clinicopathological characteristics of MBC in a tertiary care setting.

Method: The records of all cases diagnosed with MBC within one year (02/2020- 02/2021) at the Unit were reviewed.

Results: Out of 88 breast malignancies diagnosed during the period, five were MBC (5.68%). The mean age at presentation was 47 years (34-62). All MBC presented as painless lumps and were BIRADS 5 on radiology. The mean tumour size was 52 mm (25-95). All tumours were of histological grade 3. Four were biphasic tumours, while one was monophasic with pure squamous elements. Squamous (4/5), sarcomatoid (3/5), rhabdoid (1/5) and chondroid (1/5) areas were seen. Two tumours had invasive carcinoma of no special type (NST) elements, 40% and 70%, respectively. None had lymphovascular or perineural invasion. Two (2/5) patients had axillary node involvement at presentation. The mean NPI was 5.6 (4.5-7.2). Four (4/5) tumours were triple-negative, while one showed hormone receptor positivity. One patient developed a local recurrence after 10 months, following eight cycles of chemotherapy.

Discussion and conclusion: MBC in our setting appears to show a higher incidence and a younger age at presentation. As the data available from our study is limited, we need to study a larger series of patients with survival data for better understanding of the behaviour of this aggressive breast cancer.

RP 09

Proliferative epithelial changes associated with the novel breast carcinogenesis molecular models: a Sri Lankan study on women with breast cancer

G.I.M. Jinadasa*[1], H.D. Wijesinghe[2], M.M.A. Jayawickrama[1], M.D.S. Lokuhetty[2]

[1] *Department of Histopathology, National Hospital of Sri Lanka*

[2]*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

maheshi.jinadasa@gmail.com

Background and objective: The novel multistep molecular model of breast carcinogenesis is based on the oestrogen receptor (ER) status of the tumour. Its two main arms comprise ER-positive and ER-negative breast carcinomas (BCa), associated with different pre-neoplastic/high-risk proliferative epithelial changes: columnar cell lesions (CCL), atypical ductal hyperplasia (ADH), lobular carcinoma in-situ (LCIS), low-grade ductal carcinoma in-situ (LG-DCIS) with ER-positive tumours and microglandular adenosis (MGA), pleomorphic LCIS (PLCIS), high-grade DCIS (HG-DCIS) with ER-negative tumours. This study aims to describe the association between proliferative epithelial changes in tissue adjacent to BCa in Sri Lankan women in relation to the ER status of the tumour.

Method: A descriptive cross-sectional study of 420 cases, including wide local excision and mastectomy specimens of BCa handled by the National Hospital of Sri Lanka, Colombo, between 2017–2019. The tissue adjacent to BCa (within 10 mm distance from tumour) was histologically assessed for proliferative epithelial changes. Tumour ER status assessed by immunohistochemistry was reviewed. The associations between proliferative epithelial changes and the ER status were analysed by univariate analysis.

Results: ER-positive BCa (n=322) showed significant associations with columnar cell hyperplasia (27.32% vs 17.34%, p=0.04), flat epithelial atypia (16.77% vs 8.16%, p=0.035) and LG-DCIS (41.30%vs11.22%, p<0.001). PLCIS, though more frequent in ER-positive tumours, did not attain statistical significance. ER-negative BCa (n=98) showed a significant association with HG-DCIS (43.87%vs30.74%, p=0.016). MGA was not detected.

Conclusion: Pre-neoplastic/high-risk epithelial changes evaluated in tissue adjacent to BCa in our local setting support the two recently described molecular models of BCa carcinogenesis. Identification of these proliferative epithelial components in a core biopsy that is negative for BCa should therefore prompt the pathologist to advise the clinician on close clinicoradiological correlation, and if necessary, to perform a repeat biopsy of suspicious lesions.

RP 11

The diagnostic utility of ultrasound scanning, fine needle aspiration cytology and cell block technique in the pre-operative diagnosis of suspicious thyroid nodules

K.M.H.H. Kulatunga[1], A.A. Pathirana[2], S.S.N. Fernando[3], B.D. Gamage[2], A. Epa[4], M.K.A. Sampath[5], C. Sosai[6], B.S. Seneviratne[7]*

[1]*Department of Biomedical Science, Faculty of Health Sciences, KIU*
[2]*Department of Surgery,* [3]*Department of Microbiology,* [7]*Department of Pathology, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*
[4] *Department of Radiology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*
[5]*Department of Basic Sciences, Faculty of Allied Health Sciences, University of Sri Jayewardenepura, Sri Lanka*
[6]*Department of Pathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka*
bimalka03@yahoo.com

Introduction: Ultrasound scanning (USS) and fine needle aspiration cytology (FNAC) are first-line investigations to assess thyroid diseases, which assist in recognizing the suspicious thyroid nodules (*SusTNs)* pre-operatively.

Objectives: To determine the diagnostic utility of USS, FNAC and cell block technique in pre-operative diagnosis of *SusTNs*.

Method: A descriptive cross-sectional study, conducted at Colombo South Teaching Hospital and Department of Pathology, University of Sri Jayewardenepura. Patients with ultrasonically detected *SusTNs* were included. Direct smears and cell blocks were prepared from the USS guided FNAC samples of each patient. The TIRADS classification was used for radiological assessment and the Bethesda system for cytological interpretation. Specimens with insufficient cellularity were excluded.

Results: Seventy-one patients (15-76 years) with a mean age of 49.8 were included, the majority were females (84.5%). When USS findings of the *SusTNs* were compared with that of FNAC, the sensitivity was 50%. 18.9% of TR4a, 66.6% of TR4b and 100% of TR4c specimens were diagnosed as malignant (Bethesda V and VI) in FNAC samples. Cell block findings in 82% of the cases matched with the FNAC results, which was statistically significant ($p<0.001$).

Discussion: TIRADS 4 in USS denotes moderate suspicion for malignancy. The subcategories TR4a, TR4b and TR4c denote undetermined nodules, suspicious nodules and highly suspicious nodules, respectively. There is a statistically significant association between TR4c nodules and thyroid carcinoma (100%).

Conclusion: FNAC and cell block techniques complement each other in the pre-operative assessment of *SusTNs*. Cytology results were useful to separate TIRADS 4 into the subcategories in which TR4c demonstrated significant predictability of malignancy. The pre-operative assessment of *SusTNs* is imperative to minimize unnecessary thyroid surgeries.

RP 12

The histopathology of endometrial biopsies done for abnormal uterine bleeding

M.A.D.N. Munasinghe*[1], S.M. Fernandopulle[1], J. Hewavisenthi[2]

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

[2]*Department of Pathology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

nayanganee@gmail.com

Introduction: The main indication for endometrial curettage and pipelle aspiration is abnormal uterine bleeding (AUB). Histological assessment is important in determining the various structural and non-structural aetiologies for AUB.

Objectives: To describe the histomorphology of the endometrium in different age groups of females presenting with AUB and determine the percentage having organic causes for AUB in each of these age groups.

Method: All pipelle aspiration/curettage specimens received during the year 2020 were included. Evacuated products of conception were excluded. The patient characteristics, clinical information and histopathological findings were obtained. All cases were stratified into age groups 20-39 (A), 40-50 (B) and >50 years (C). The histological findings were classified as normal pattern (NP), ovulatory dysfunction (OD), exogenous hormonal effects (EHE), endometrial polyp (EP), chronic endometritis (CE), atrophic endometrium (AE), disordered proliferative endometrium (DPE), endometrial hyperplasia (EH) and endometrial carcinoma (EC). EP, CE, EH and EC were considered structural causes.

Results: A total of 355 specimens were analysed. The age range of patients was 24-74 years (A-49, B-191, C-115). In the 20-39 age group 13/49 (26.53%) accounted for organic causes (EP-22.4%, CE-2%, EH-2%). In the 40-50 age group 25/191(13%) were due to organic causes (EP-7.32%, CE-1.57%, EH-3.66%, EC- 0.52%). In the >50 age group 29/115 (25.2 %) were due to organic causes (EP-13.04%, EC-10.43%, EH-1.73%). The commonest histological finding for AUB in the > 50 age group was AE-23/115 (23%), in the 20-39 age group EHE-13/49 (26.5%) and in the 40-50 year age group NP-55/191(28.79%).

Conclusion: There is a variation in the histomorphological findings for AUB among different age groups, and non-structural findings are commoner than organic ones.

RP 13

A comparison of oestrogen receptor and progesterone receptor status of breast cancer tissue processed in-house and in external laboratories: an internal audit

F.N. Nawas, N. Meedeniya, A. Ruwan, A.A.H. Priyani*

Lanka Hospital Diagnostics Laboratory, Narahenpita, Sri Lanka.

priyaniaah@gmail.com

Introduction: Optimum fixation/ processing of tissue is required to avoid false negative results (FNR) in breast cancer (BC) biomarkers.

Objectives: This audit compares oestrogen receptor (ER) and progesterone receptor (PR) status of BC tissue processed in-house, under controlled conditions, and in external laboratories (ELs).

Method: BC reported for ER and PR at Lanka Hospital Diagnostics from January to December 2020 were included. The test result, place of processing of tissue, and for cases processed in-house, cold ischemia time, type of fixative and duration of fixation were obtained from the worksheets. Patients of 50-years and above were considered postmenopausal.

Results: Ninety-nine cases were included; 57 were processed in-house, 42 were received as wax tissue blocks from ELs. In all in-house cases, the fixative was 10% NBF, the cold ischemia time was <1 hour, and the duration in formalin ranged from 24-96 hours. Fixation details were not known for those processed in ELs. ER-negative rates (ERNR) were 30% (17/57) and 57% (24/42) for in-house cases and ELs, respectively. For postmenopausal women, those were 23% (10/43) and 54% (13/24), respectively. PR negative rates (PRNR) were 38% (21/55) and 63% (26/41), respectively.

Discussion and conclusion: BC tissue processed in-house showed ERNR and PRNR close to CAP benchmarks (ERNR - overall <30%, ERNR – postmenopausal <20%, PRNR – overall <45%). High ER and PR negative cases received from ELs, exceeding CAP recommendations, could be FNRs as the fixation details are not known. Identification of centres with high negative rates, giving instructions for the pre-analytical phase and monitoring of fixation details when receiving wax tissue blocks from ELs are recommended to avoid FNR in future.

RP 14

A descriptive analysis of Gleason grades of prostate carcinoma in a tertiary care centre in Sri Lanka

D.M.T. Chandramali[1], M.S. Manjika [2], I.V. Prematilleke*[1]

[1]*Department of Pathology,* [2]*Department of Community Medicine, Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

ishaprem@sjp.ac.lk

Introduction: Prostate cancer (PC) is the 5th commonest malignancy in Sri Lankan men. Whilst limited studies exist on PC grades in Sri Lanka (SL), none have used the modified Gleason grading.

Objectives:

1. To describe the Gleason patterns (GP), scores (GS) and grade groups (GG), with a view to identifying the commonest by age group.
2. To compare with data in SL and foreign studies.

Material and method: Eighty-eight PCs reported between 2016-2020 (72-TRUS biopsies, 11-TURP and 5-radical prostatectomies) were retrieved. Data collected included type of sample, age, GP, GS, GG, and were analysed using SPSS 15.0.

Results: Ages ranged from 54-90 years (mean - 70.78), majority (n=67;76%) in the 61-80 age group. The commonest were GS-7 (n=34;38.6%), GP-4 (n=48;54.5%) and GG-IV (n=23;26.1%). GS-6 PCs=11.4%, ≥8=50. GS-7 was the commonest in ages ≤60 (n=7;63.6%), 61-70 (n=14;38.9%) and >80 (n=5;50%). GS-9 was the commonest in ages 71-80. GG-II and GG-IV were the commonest in ages ≤60 (n=4;36.4%) and >80 (n=3;30%). GG-III and GG-IV were the commonest in ages 61-70 (n= 9;25%), and GG-V in ages 71-80 (n=11;35.5%).

Discussion: The mean age was comparable to previous studies in SL and the United Kingdom (UK). Percentages of GS-6, 7 and ≥8 in this study were close to those of a Sri Lankan study, the widest difference being 4.2% more GS ≥8 PCs in our sample. We had fewer GS-6 PCs (11.4%) compared to studies from the UK (31.1%), Finland (>20%), the US (41%) and India (25% - old grading).

Conclusion: This study suggests that our population has a higher proportion of GS >6 PCs. Further studies are warranted for confirmation.

RP 15

The usefulness of Alcian blue stain in confirming goblet cells in oesophageal biopsies with columnar metaplasia

M.M.F. Rimashana[1], L.D.K. Bulathgama[2], Sulochana Wijetunge*[2]

[1]*Medical Laboratory Sciences, Faculty of Allied Health Sciences, University of Peradeniya, Sri Lanka*

[2]*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

sulochana.wijetunge@med.pdn.ac.lk

Introduction: Intestinal metaplasia in the oesophagus is a premalignant complication of gastro-oesophageal reflux. Although using Alcian blue (AB) stain to confirm goblet cells in oesophageal biopsies is a common practice, it is discouraged due to high false positive rates. The objective of this study was to assess the reliability of AB in identifying goblet cells in oesophageal biopsies.

Method: This is a cross-sectional study of 38 samples of gastro-oesophageal junctional biopsies obtained from patients undergoing upper gastrointestinal endoscopy for dyspeptic symptoms. These were assessed initially with haematoxylin and eosin-stained sections, and 30 samples of cardiac mucosa with pooled mucin mimicking goblet cells and 8 with true goblet cells were selected. Ten normal antral biopsies were taken as negative controls. All were stained with AB (pH 2.5). The Fisher's exact test (at $p<0.05$) was used to assess statistical differences.

Results: The AB staining pattern in goblet cell mimics showed negative (10), mild (16) and moderate (4) staining; none had strong staining. All 8 cases with goblet cells were positive with AB, 7 had strong and one had moderate staining. All antral biopsies were negative. When any degree of AB staining was regarded as positive, positive rates in true goblet cells and its mimics were statistically not significant ($p=0.08$). When only strong AB staining was regarded as positive, the difference was significant ($p<0.0001$).

Discussion: The false positivity rate with AB among goblet cell mimics was 66.7% (n=20/30), which is unacceptably high. However, all false positive cases had either mild or moderate staining. Strong positivity was seen only in true goblet cells ($p<0.0001$).

Conclusion: AB can be used to confirm the presence of goblet cells only if strong staining is considered as positive. Interpretation of any degree of staining as positive leads to false positive diagnosis of goblet cells.

RP 16

A comparison of expression of ER, PR and p53 immunohistochemical stains between endometrial curetting/biopsy and resected endometrial carcinoma specimens

K. G. H. Silva[*1], *B. A. G. G. Mahendra*[1], *D. Ediriweera*[2], *J. Hewavisenthi*[1]

[1]*Department of Pathology,* [2]*Centre for Health Informatics, Biostatistics and Epidemiology, Faculty of Medicine, University of Kelaniya, Sri Lanka*

g.hiroshi@yahoo.com

Introduction and objective: The expression of immunohistochemical (IHC) stains is negatively affected by poor fixation. Different markers behave variably under suboptimal pre-analytical conditions with crucial diagnostic and therapeutic implications. The objective was to compare the expression of oestrogen-receptor (ER), progesterone-receptor (PR) and p53 in curettings/ endometrial biopsies (C/EB) with the corresponding hysterectomy in endometrial carcinoma (EC).

Method: Fifty cases of EC diagnosed by C/EB of which the resections received at the same centre, between 2017-2020 were retrieved from four tertiary care hospitals. IHC for ER, PR and p53 were performed on both C/EB and resection. Hormone-receptors (HR) were scored using Allred-score (ARS) (staining of >1% cells, ARS >3: positive). Strong nuclear staining in >80% or complete negative staining was considered mutant p53.

Results: 47/50 (94%) showed concordance of histological typing between C/EB and resections [endometrioid - EC:35/36 (97.2%), serous - EC:11/12 (91.6%), clear cell carcinoma:0/1 (50%), carcinosarcoma:1/1 (100%)]. ER and PR were positive in 33/50 (66%), 32/50 (64%) C/EB and in 28/50 (56%), 33/50 (66%) resections (ER, P=0.303; PR, P=0.209 >0.05), respectively. 46/50 (92%) showed concordance between ER and PR expression. ARS for ER/PR in C/EB was higher than in resections in 20 (40%) and 14 (28%) cases, respectively. The difference in ARS for ER between the two groups was statistically significant (ER, P=0.005; PR, P=0.133). The p53 expression pattern did not show a significant difference between C/EB and resections (P=0.480).

Discussion and conclusion: The underexpression of HR in resections is best explained by poor-fixation, although tumour heterogeneity may contribute. p53 expression appears to be unaffected. Immediate opening of the hysterectomy specimens allowing adequate fixation and preventing over-fixation is crucial in preventing the loss of hormone receptor expression.

RP 17

A comparison of cytology and radiological score with histology in patients with breast lumps: what guides the clinicians best?

H. G. D. Thilini*[1], A. Perera[1], L. Mudduwa[2]

[1]*Department of Pathology, Teaching Hospital, Karapitiya, Sri Lanka*

[2]*Department of Pathology, Faculty of Medicine, University of Ruhuna, Sri Lanka*

t.gamage1983@gmail.com

Introduction: Breast lumps are among the commonest surgical presentation. There are mismatches between Fine Needle Aspiration Cytology (FNAC) and Breast Imaging-Reporting and Data System (BIRADS) categories of triple assessment of breast lumps, leading to unnecessary surgical interventions and management delays.

Objectives: This study was designed to compare FNAC and BIRADS grading using histopathology as the gold standard.

Method: One hundred and six breast lumps were analysed in a descriptive cross-sectional setting, at a single tertiary care centre, from 2018-2019. Their FNAC smears were assessed and compared with BIRADS (B2-B5) categories and with Histopathology.

Results: A comparison of BIRADS (n=75 cases, 70.7%; X^2=33.2, p<0.001) and FNAC (n=80 cases, 75.5%, X^2=95.6, p<0.001) grading with Histopathology revealed majority of cases with true positive diagnosis. The concordance of Histopathology with BIRADS was moderate (Cohen's kappa=0.55, Standard Error (SE)=0.09, p<0.001), while discordance was not significant (McNemer test significance; p=0.80); whereas with cytological grading was stronger (Cohen's kappa=0.94, SE=0.03, p<0.001) and discordance was not significant (McNemer's test significance; p=0.50). Sensitivity, specificity, positive and negative predictive values and accuracy between radiological and cytological grading were 91%,62%,89%,68%,85% and 97%,100%,100%,92%,98%, respectively.

Discussion: The agreement between histopathology and cytological grading revealed stronger concordance compared to the agreement with BIRADS grading. The sensitivity, specificity, positive and negative predictive values and accuracy of cytological grading was more in comparison to the BIRADS grading. Therefore, in situations with discordant results, FNAC grading should be considered a more accurate and valid investigation over BIRADS grading.

Conclusion: FNAC guides the clinician best in managing patients with breast lumps

CR 01

Carcinoma showing thymus-like differentiation of the thyroid (CASTLE): an extremely rare presentation of thyroid malignancy

A.A.S.D Abeygunawardhane*, K. Samalai, M.M.A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

sakutcc@gmail.com

Introduction: Carcinoma showing thymus-like differentiation of the thyroid (CASTLE) is a very rare thyroid carcinoma that affects middle-aged adults. The female to male ratio is nearly 1:1. There are about 82 reported cases in the literature to date.

Case report: A 46-year-old man presented with progressive dysphagia for eight months. Ultrasonography revealed a hypoechoic nodule in the right lobe of the thyroid. Fine needle aspiration cytology (FNAC) of the nodule was reported as papillary thyroid carcinoma (Thy5, Bethesda VI). Macroscopically there was a large, solid, white, non-encapsulated tumour in the right lobe measuring 45x40x25 mm. Microscopically it was composed of solid nests of squamoid cells in a lymphocyte-rich stroma. Mitoses were frequent. Keratin pearl formation, sarcomatoid differentiation and follicular or papillary architecture were not seen. Extrathyroidal extension and margin involvement were present. The background thyroid tissue showed a papillary microcarcinoma and features of chronic autoimmune thyroiditis. The tumour cells were strongly positive for CD5, CD117, CK5/6 and CK19. Synaptophysin was positive focally. TTF1 and calcitonin were negative. The patient is currently undergoing radiotherapy.

Discussion: Clinical features of CASTLE can vary and are non-specific to the disease. FNAc is challenging as it is difficult to differentiate from other thyroid malignancies. CASTLE is characterized by its indolent biological behaviour. The absence of nodal metastasis and invasion of adjacent structures are favourable prognostic factors. Postoperative external radiation is effective.

Conclusion: CASTLE is an extremely rare, distinct malignant entity of the thyroid gland. Preoperative and the histological diagnosis can be difficult, and the definitive diagnosis needs immunohistochemical analysis. However, the definite diagnosis is a must as this entity has a favourable outcome.

CR 02

Malignant paraganglioma: a rare case in lumbar spine

L.A.H.A.Albert*, H.N.Thewarapperuma, J Wickramasooriya

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

harshanianakshi@gmail.com

Introduction: Paraganglioma is a rare benign neuroectodermal tumour, usually arise in the adrenal medulla or extra-adrenal paraganglia. In the spinal cord, it frequently involves cauda equina/filum terminale. The malignant nature of paragangliomas depends on its ability of metastasis.

Case report: A 29-year-old woman presented with numbness and weakness of the lower limbs, which gradually worsened over five months. Imaging studies showed an L1-L3 intradural extramedullary lesion with bone involvement. The laminectomy specimen contained multiple pieces, measuring 5.4x5.1x 4.5 cm. The histology revealed a tumour infiltrating bone and soft tissue, which comprised small nests and cohesive clusters (zellballen) of medium-sized round to oval cells having mild nuclear atypia and scanty clear to pale eosinophilic cytoplasm. Occasional mitoses were noted. There was no necrosis. The tumour cells were positive for synaptophysin, CD56 and vimentin, but negative for AE1/AE3, S-100, CD99, GFAP, Melan-A and EMA. These features concluded the lesion as a paraganglioma. However, four months later, the patient presented with new lesions in the thoracic spine with similar image resonance, highly suggestive of a metastatic deposit.

Discussion and conclusion: Although generally considered benign, 10-20% of paragangliomas show metastatic potential. However, there are no definite histological features to predict the behaviour. Infiltrative and malignant lesions may lack sustentacular cells. Paragangliomas rarely express cytokeratin, in that case, need to exclude metastatic deposits of small cell carcinoma, carcinoid tumours and neuroendocrine carcinoma. Spinal paragangliomas are extremely rare but need to be considered in the differential diagnosis of spinal tumours with a nested pattern.

CR 03

Adult granulosa cell tumour of the ovary associated with endometrial hyperplasia with atypia in an elderly patient

N. T. Amarasinghe*[1], S. Wijetunge[1], M. N. Jiffry[2]

[1] *Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[2] *Department of Pathology, Teaching Hospital Peradeniya, Sri Lanka*

nuwaniamarasinghe@gmail.com

Introduction: Granulosa cell tumour (GCT) is an indolent low-grade malignant sex cord-stromal tumour. They occur at any age but are most common in perimenopausal women. Endometrial hyperplasia is a known association due to the oestrogenic effects of hormones secreted by this tumour.

Case report: A 70-year-old woman presented with postmenopausal bleeding. The ultrasound scan revealed a right ovarian cyst. She underwent a hysterectomy, right ovarian cystectomy, left oophorectomy and bilateral salpingectomy. The predominantly cystic right ovarian cyst had a smooth wall with a mural nodule measuring 7 mm. The endometrial thickness was 15 mm. Microscopy of the ovarian cyst showed a neoplasm composed of ovoid cells with minimal atypia, occasional nuclear grooves and scanty cytoplasm. The cells were arranged in sheets with scattered Call-Exner bodies. Mitotic activity was sparse. The cells were positive for calretinin and inhibin. The Ki-67 index was 5%. A diagnosis of adult GCT was made. The endometrium showed multiple foci of complex and irregular glands arranged in a back-to-back manner, lined by columnar cells containing moderately pleomorphic nuclei. The remaining endometrium showed endometrial hyperplasia without atypia (EHWOA). Sampling of the entire endometrium showed no myometrial invasion, and a diagnosis of endometrial hyperplasia with atypia (EHWA) was made.

Discussion and conclusion: Adult GCT and hyperoestrogenaemia associated pre-malignant and malignant conditions are rare in elderly. In this case, EHWOA may have occurred as a result of prolonged oestrogen secretion. Genetic mutations result in EHWA, and this is exacerbated by hyperoestrogenaemia. This highlights the importance of extensive endometrial sampling in patients with granulosa cell tumours in order to exclude EHWOA, EHWA or endometrial carcinoma.

CR 04

A concurrent occurrence of adrenocortical adenoma and a neuroendocrine tumour in the pancreas

N. T. Amarasinghe*, R. Waduge

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

nuwaniamarasinghe@gmail.com

Introduction: Adrenocortical adenomas (ACA) and pancreatic neuroendocrine tumours (PanNET) are uncommon tumours with incidences of 0.35% and 0.21%, respectively. Concurrent occurrence of ADA and PanNET is rare.

Case Report: A 54-year-old woman diagnosed with non-cirrhotic portal hypertension, hypersplenism and adrenal mass was found to have an inoperable lesion in the peripancreatic region. The adrenal mass was circumscribed and 1.5 cm in diameter. Peripancreatic mass and liver biopsies, adrenalectomy and splenectomy were performed. The adrenal lesion showed an encapsulated tumour comprising cells with empty spaces resembling zona fasciculata and compact cells resembling zona reticularis arranged as cords and nests. Mitoses, necrosis and vascular invasion were absent. These cells were positive for synaptophysin and Melan-A. Chromogranin was negative. The Ki-67 proliferative index was 1%. Reticulin stain revealed intact pattern resembling normal adrenal tissue. A diagnosis of ACA was made. Pancreatic biopsy revealed an infiltrating neoplasm comprising nests and organoid structures of atypical cells that were positive for pancytokeratin, synaptophysin, chromogranin and CD56. CK7, CK20, CK19 and CEA were negative. The Ki-67 proliferative index was 3%. A diagnosis of PanNET was made. Liver biopsies and spleen were negative for tumours.

Discussion: Though the concurrent occurrence of ADA and PanNET is rare, this can occur in multiple endocrine neoplasia type-1 (MEN1). This patient's investigations were negative for parathyroid, pituitary and gastrointestinal tract tumours. However, knowing the possibility of later presentation of other tumours related to MEN1, further follow up was advised.

Conclusion: Though this is likely to be sporadic, it could still be familial. There is no specific molecular test to confirm MEN1, as MEN 1 gene could be positive in both familial and sporadic cases.

CR 05

Adenoid cystic carcinoma of breast: a rare entity

T. Arudselvan*, P.N. Yogananda, M. M. A Jayawickrama, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

thadsha89@gmail.com

Introduction: Adenoid cystic carcinoma of the breast is a rare invasive carcinoma of the breast composed of neoplastic epithelial and myoepithelial cells. Immunomorphologically it is similar to salivary adenoid cystic carcinoma and is composed of tubular, cribriform and solid patterns of basaloid cells.

Case report: A 71-year-old woman presented with a painful left breast lump of 2 months duration. Mammography revealed a BIRADS III lesion. The tru-cut biopsy of the lesion revealed a tumour composed of epithelial and myoepithelial cells arranged in tubular and cribriform structures containing eosinophilic material within the lumina. The constituent cells had angulated hyperchromatic nuclei. The tumour cells showed membrane positivity for CD 117 and the Ki-67 proliferation index was 28%. The tumour was negative for ER, PR and HER2 (triple-negative). These features were in keeping with an adenoid cystic carcinoma. The patient underwent a mastectomy and was referred for adjuvant radiotherapy.

Discussion and conclusion: Adenoid cystic carcinoma is a rare, slow growing highly malignant tumour accounting for 0.1-3.5% of all breast cancers. It mostly affects elderly women and is usually a solitary lesion. It is differentiated from other invasive breast carcinomas morphologically by the presence of myoepithelial cells and immunohistochemically by positive luminal component for CD117 and triple negativity. These tumours show favourable prognosis with a rare possibility of local and distant metastases.

CR 06

Peritoneal deciduoid mesothelioma: a rare variant of epithelioid mesothelioma

T. Arudselvan*, P. G. N. L. Chandrasiri, H.D. Wijesinghe, M.V.C. De Silva

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

thadsha89@gmail.com

Introduction: Deciduoid mesothelioma is a rare variant of malignant epithelioid mesothelioma, an entity first described in 1985. It was initially reported exclusively in the peritoneum of young women without a history of asbestos exposure. However, later, cases were reported involving the pleura and peritoneum of men and women with and without a history of asbestos exposure.

Case report: A previously healthy 35-year-old man presented with low-grade fever, associated with dry cough, night sweats and weight loss for one month. CT abdomen and pelvis revealed enlarged upper mesenteric lymph nodes. Laparoscopy revealed multiple peritoneal and omental nodules. Biopsies of these nodules showed infiltrating sheets and nodules of large polygonal tumour cells and a lymphoplasmacytic infiltrate in the fatty tissue. The tumour cells showed eosinophilic glassy cytoplasm with distinct cell borders and enlarged vesicular nuclei with prominent nucleoli. Mitoses were not prominent. There was no necrosis. The tumour cells showed diffuse positivity for immunohistochemical stains calretinin, CK5/6, AE1/AE3 and EMA, focal positivity for CK7, and negative for desmin, CK20, Melan-A and HepPar-1. A diagnosis of malignant deciduoid mesothelioma was made.

Discussion and conclusion: Histologically, deciduoid mesothelioma comprises round to polygonal cells resembling decidual cells. Clinicomorphologically, it varies significantly from classic epithelioid mesothelioma which occurs mostly in elderly men with a history of asbestos exposure. The distinct cytomorphological features, presence of fat infiltration and the immunohistochemical profile help to differentiate epithelioid mesothelioma from metastatic carcinoma and reactive mesothelial hyperplasia.

CR 07

Papillary meningioma: a rare variant posing a diagnostic challenge on intraoperative crush smear

C. M. Baranasuriya*, S. Vinothika, R. Constantine

Department of Histopathology, National Hospital of Sri Lanka

chamalib@gmail.com

Introduction: Papillary meningioma is a rare, aggressive variant of meningioma accounting for less than 1% of all meningiomas.

Case report: A 45-year-old man presented with recent onset seizures. Magnetic resonance imaging (MRI) scan showed an extra-axial, dural based, heterogeneously enhancing mass with perilesional oedema, raising a differential diagnosis of atypical meningioma and a metastatic deposit. The intraoperative crushed smears showed many papillary structures with radially arranged tumour cells around blood vessels. Cells contained oval nuclei with fine chromatin and some cells show intranuclear inclusions. Cells with rhabdoid cytology were also present. Whorls or psammomatous calcifications were not evident. A diagnosis of papillary meningioma was made on the cytological features of the crush smears. The tumour was excised, and the histology showed similar features as above. Tumour necrosis and brain invasion were evident. The tumour cells showed positivity for EMA and vimentin. A diagnosis of papillary meningioma (WHO grade III) was made.

Discussion: Papillary meningioma occurs in a younger population than conventional meningiomas. Imaging shows an irregular tumour brain interface, heterogeneous enhancement and marked peritumoural oedema. The prognosis is poor with a high incidence of brain invasion, recurrence, and metastasis. The main differential diagnosis includes ependymoma, choroid plexus tumours, astroblastoma and metastatic carcinoma. The extra-axial location on MRI is a helpful feature. In most instances, classic patterns of meningioma are demonstrable. However, as in this case, if the papillary pattern predominates, a diligent search for meningothelial cytological features is crucial.

Conclusion: Although rare, the awareness of this entity is important, as it is often misdiagnosed on crush smears.

CR 08

Osteosarcoma with chondroblastoma-like areas: a potential pitfall for diagnosis

C. M. Baranasuriya[1]*, M. M. A. Jayawickrama[1], S. Thoufeek[2], M. V. C. de Silva[2]

[1] *Department of Histopathology, National Hospital of Sri Lanka*

[2] *Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

chamalib@gmail.com

Introduction: Chondroblastoma-like osteosarcoma (OS) is an exceedingly rare variant of osteosarcoma, which mimics a benign tumour.

Case report: A 37-year-old man presented with painful swelling of the right lower end of femur for one year. MRI scan showed a heterogeneously enhancing 6 cm lesion with cortical breach in the distal femoral epiphysis, extending into the meta-diaphyseal region. Biopsies revealed sheets of plump cells admixed with scattered giant cells, permeating between pre-existing bony trabeculae. Cells contained moderately pleomorphic nuclei and eosinophilic cytoplasm. Mitoses were frequent with atypical forms. Chondroblastoma-like areas with chicken-wire calcifications were present. On careful examination, neoplastic osteoid deposition was observed. The Ki-67 proliferative index was 50%. A diagnosis of conventional OS was made. The patient was referred for chemotherapy.

Discussion: Chondroblastoma-like osteosarcomas are reported commonly in the third decade. Common sites include metatarsals, femur and tibia. Radiologically, a lytic expansile lesion with cortical destruction is seen. The differential diagnoses include, giant cell tumour, chondroblastoma and aneurysmal bone cyst. The main histological differential diagnosis is chondroblastoma. Features favouring OS include older age, extension of the lesion to the metadiaphyseal region, permeative growth pattern, presence of atypical mitoses and malignant osteoid. In addition, H3F3B K36M point mutation, which can be detected by immunohistochemistry, is identified in 95% of chondroblastomas. Although scattered giant cells were seen, the presence of malignant osteoid helped to distinguish this lesion from giant cell tumour.

Conclusion: Awareness of this rare entity is important as both radiological and histological features may appear deceptively benign.

CR 09

Wilms tumour metastasizing to spine: an uncommon occurrence of a common tumour

C. M. Baranasuriya*, D. T. T. Jayasinghe, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

chamalib@gmail.com

Introduction: Nephroblastoma is the commonest paediatric renal tumour. The mean age at diagnosis is 3-4 years. At diagnosis, metastases are present in 10% of nephroblastoma cases. The commonest sites are lung, liver, and lymph nodes. Metastases to the bones and spinal cord is extremely rare and is seen only in 1% of cases.

Case report: A 20-month-old baby presented with a left lumbar mass and difficulty in walking. CECT scan showed a mass lesion in the left kidney and an extradural soft tissue mass causing spinal cord compression at D8. A laminectomy and excision of the spinal mass were performed. Intraoperative crush smears showed small round blue cells with scattered tubules. Histology showed a triphasic tumour composed of undifferentiated blastema, epithelial tissue with tubules and mesenchymal elements with fascicles of spindled cells. Anaplastic features were not seen. WT-1 was strongly positive in the blastemal and spindle cell nuclei. Tubules were highlighted by CD56. vimentin was positive in the mesenchyme. This tumour was concluded as a metastatic deposit of a triphasic nephroblastoma.

Discussion: The commonest paediatric malignancies that metastasize to the spinal canal include neuroblastoma, Ewing sarcoma, osteosarcoma, rhabdomyosarcoma and lymphoma. Among the paediatric renal tumours, clear cell sarcoma has a higher propensity for bone metastases. The characteristic triphasic morphology and nuclear positivity for WT-1 helps in differentiating Wilms tumour from other differential diagnoses.

Conclusion: Although rare, Wilms tumour needs to be considered in the differential diagnosis of paediatric spinal tumours as prompt multimodal therapy can improve patient outcome even in the setting of advanced disease.

CR 10

Microcystic serous cystadenoma coexisting with pancreatic intraepithelial neoplasia

D.M.T Chandramali*, Bimalka Seneviratne

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

tharanga.dissanayake88@gmail.com

Introduction: Microcystic serous cystadenoma is a rare epithelial neoplasm of the pancreas commonly arising in the pancreatic body or tail. It has no communication with the pancreatic duct. Pancreatic intraepithelial neoplasia is a microscopic non-invasive lesion confined to the pancreatic ducts, which acts as a precursor lesion of ductal adenocarcinoma.

Case report: A 50-year-old woman who presented with upper abdominal pain was found to have a lesion in the pancreatic tail in close proximity to the spleen. Radiological findings were suggestive of a neuroendocrine tumour. A distal pancreatectomy with splenectomy was performed. Macroscopically there was a well-defined pancreatic lesion measuring 38x37x30 mm with a microcystic cut surface. Histologically the lesion was composed of multiple small cysts with irregular contours lined by non-dysplastic cuboidal epithelium comprising cells with small round regular nuclei. There is no cellular atypia. The adjacent non-neoplastic pancreatic tissue showed a dilated duct with low-grade pancreatic intraepithelial neoplasia.

Discussion: Serous cystadenoma accounts for 1-2% of all pancreatic neoplasms, and the microcystic variant is the commonest variant. Pancreatic intraepithelial neoplasia is a microscopic lesion, usually <5 mm, which is graded as low-grade and high-grade according to the degree of cytoarchitectural atypia. Rarely serous cystadenoma can coexist with other pancreatic pathologies such as neuroendocrine tumours, adenocarcinoma and intraductal papillary mucinous neoplasm. Synchronous serous cystadenoma with pancreatic intraepithelial neoplasia is even more rare with very few cases reported in the literature.

Conclusion: Microcystic serous cystadenoma can rarely coexist with other pancreatic tumours, including preinvasive lesions. Therefore, careful assessment of the surrounding pancreatic tissue is also equally important when handling pancreatic resection specimens.

CR 11

Renal findings of partial lecithin-cholesterol-acyltransferase deficiency: an extremely rare cause of nephrotic syndrome

L. J. De Silva*, A. V. T. Damayanthi, M. V. C. De Silva

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

lalanidesilva7@gmail.com

Introduction: Lecithin-cholesterol-acyltransferase (LCAT) is one major enzyme in the cholesterol metabolism pathway. Partial LCAT deficiency is a rare autosomal recessive disorder characterized by nephrotic-range proteinuria (NRP), anaemia and corneal opacity (CO). Due to the characteristic manifestations in the cornea, it is also known as 'fish-eye disease'. This case highlights the renal findings of two siblings with congenital LCAT-deficiency.

Case report: The two patients are the second and third siblings of a family with second-degree consanguinity. The elder, six-year-old boy presented with steroid-resistant NRP and hypercholesterolaemia at two years of age, and the genetic testing confirmed the partial-LCAT-deficiency. He has developed end-stage renal failure and is currently on dialysis. His two-year-old sister presented with similar symptoms. Renal biopsies of both showed mostly viable glomeruli and a few partially sclerosed glomeruli. The mesangial matrix was expanded with cells containing pale eosinophilic bubbly cytoplasm. Capillary lumina were obliterated by enlarged mesangial cells in worst-affected glomeruli. The basement membranes were irregular and wrinkled, and the silver stain highlighted their bubbly appearance. Many tubules and some vascular endothelial cells showed cytoplasmic vacuolations. Immunofluorescence was negative. Both patients are being followed up for visual impairment, although the characteristic CO is not yet established.

Discussion and conclusion: LCAT-deficiency is either congenital due to genetic mutation in Ch-16q22 or acquired due to inhibitory autoantibodies to LCAT. Testing of LCAT-antibodies is required, particularly in older patients. Renal manifestation is a major cause of morbidity. The renal findings of LCAT-deficiency are similar to the late-stage of membranous glomerulopathy (MG). However, with the presence of bubbly cytoplasm and negative immunofluorescence, MG is unlikely in this patient.

CR 12

A diagnostic dilemma of breast carcinoma in a core biopsy with dimorphic cells

L. J. De Silva *, K. V. C. Wijegunaratne, I. W. G. A. L. Malhasi, M. D. S. Lokuhetty

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

lalanidesilva7@gmail.com

Introduction: Breast carcinoma (BCa) with dimorphic cells is a rare but distinct entity. These tumours comprise malignant epithelial cells with a subpopulation having clear cytoplasm resembling myoepithelial or glycogen/lipid-rich cells. Herein we report three BCa cases with dimorphic pattern on core biopsies, causing a diagnostic dilemma.

Case report:

Case 1: A 32-year-old woman, recent painful breast lump

Case 2: A 64-year-old woman, mammographically malignant breast lump for two months

Case 3: A 46-year-old woman, mammographically malignant breast lump for six months

Core biopsies of all three tumours showed nests of cells comprising two distinct populations: clusters of malignant epithelial cells with irregular hyperchromatic nuclei, high nuclear/cytoplasmic ratio and moderate eosinophilic cytoplasm and cell clusters with abundant clear cytoplasm resembling myoepithelial or glycogen/lipid-rich cells. Careful examination showed similar nuclei in both cell populations. Clear cells were negative for glycogen by PAS stain. Both cell types showed similar immunoprofiles with hormone receptor, ER, PR and AR positivity and negativity for HER2 and basal/myoepithelial markers, p63 and CK5/6, confirming the diagnosis as BCa with a dimorphic pattern.

Discussion: The dimorphic pattern in BCa could create a diagnostic dilemma. The differential diagnoses include duct carcinoma in-situ and malignant adenomyoepithelioma, excluded by negative basal/myoepithelial stains. The lipid-rich pattern is unlikely with ER/PR positivity and glycogen-rich pattern was excluded by negative PAS stain. The dimorphic pattern was first described in papillary lesions in 1994 and later in DCIS and invasive carcinoma as well.

Conclusion: Awareness of this rare pattern is important, especially in core biopsies of breast. Careful attention to atypical nuclear features in clear cells and supportive ancillary stains facilitate correct diagnosis.

CR 13

Primary small intestinal lymphoma presenting as acute intestinal perforation: a rare case

W.M.A.S. De Silva*, A. Vithanage, T. Jayamaha, R.M.P. Ratnayake

Department of Pathology, National Hospital, Kandy, Sri Lanka

achaladesilva83@gmail.com

Introduction: Primary small intestinal lymphomas (SmIL) are rare, accounting for about 15%-20% of all small intestinal malignancies. Diffuse large B-cell lymphoma (DLBCL) is the commonest subtype (55-69%) commonly affecting middle age to elderly men. The symptoms and signs are mainly non-specific and comprise abdominal pain, ileus, diarrhoea, weight loss, gastrointestinal bleeding and palpable masses.

Case report: A 69-year-old man presented to the emergency treatment unit with features of acute abdomen, and the exploratory laparotomy revealed evidence of ileal perforation. A short segment of the ileum, 23 cm in length, was received, which macroscopically revealed an intramural tumour at the perforation site obstructing the entire lumen, measuring 5.5 cm in maximum diameter. Multiple mesenteric nodules up to 2.5 cm in diameter were present. Microscopically, the tumour revealed a transmural infiltrate of loosely cohesive sheets of round to oval medium-sized cells with vesicular nuclei, occasional conspicuous nucleoli and scanty cytoplasm. Mitoses were frequent, including atypical forms. Two of the eleven paraileal mesenteric lymph nodes were involved by the tumour. The tumour cells were positive for immunohistochemical stains LCA, CD20, BCL6 and MUM1, negative for CD117 and CD10 and the Ki-67 index was 67%. These features are compatible with diffuse large B-cell lymphoma of activated B-cell type according to the Hans algorithm classification.

Discussion and conclusion: Acute intestinal perforation is a rare clinical presentation of SmIL. When gene expression technologies are not available, immunohistochemical findings are considered an acceptable alternative to determine the cell of origin in DLBCL, which is a requirement as the cell of origin subtyping affects the prognosis and treatment modality.

CR 14

Malakoplakia presenting as haematuria: a rare case

A. W. K. Dilrukshi*, T. G. Liyanage

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

kaushikadilrukshiaw@gmail.com

Introduction: Malakoplakia is a rare histiocytic disorder most commonly found in the genitourinary tract. It is believed to occur due to defective bacterial digestion by macrophages leading to calcium and iron accumulation on residual bacterial glycolipids. Histologically this lesion is characterized by the formation of Michaelis-Gutmann bodies. Malakoplakia of the bladder usually presents with recurrent urinary tract infection (UTI) and haematuria and is often not diagnosed until symptoms have persisted for some time.

Case report: A 42-year-old woman presented with recurrent UTI for two years. Repeated urine cultures were performed, and she was treated accordingly with antibiotics. CT abdomen/pelvis was normal with no upper urinary tract abnormalities. Later she developed haematuria. She was investigated with cystoscopic examination, which showed multiple soft, raised mucosal plaques. No solid lesions were identified. Bladder biopsy showed collections of histiocytes with abundant granular acidophilic cytoplasm in the lamina propria. Some histiocytes showed rounded basophilic intracytoplasmic Michaelis-Gutmann bodies, which were highlighted by positive Perls stain. No granulomas or fungal hyphae were noted. Positive immunostaining with CD68 confirmed the histiocytic nature of these cells.

Discussion: The distinction between malakoplakia and other causes of histiocytic collections in the bladder mucosa such as xanthogranulomatous cystitis, tuberculosis and fungal infections is important due to different treatment modalities. Xanthograulomatous cystitis is morphologically similar to malakoplakia but lacks Michaelis-Gutmann bodies. The absence of fungal hyphae and granulomas excluded the possibility of tuberculosis and fungal infections.

Conclusion: Malakoplakia of the genitourinary tract is a rare condition which requires careful histological assessment for the correct diagnosis.

CR 15

Extranodal marginal zone B-cell lymphoma of mucosa associated lymphoid tissue (MALT lymphoma) presenting as a rectal polyp

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

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

dilushi.da@gmail.com

Introduction: MALT lymphoma is the second commonest lymphoma involving the gastrointestinal (GI) tract, majority involving the stomach. Their incidence is 60-70% in the stomach compared to 2.5% in the rectum. We report a case of rectal MALT lymphoma mimicking adenocarcinoma clinically, endoscopically and on imaging.

Case report: A 69-year-old man presented with bleeding per rectum and altered bowel habits for six months. Colonoscopy showed a sessile polypoidal mass (type IIa, nongranular lesion) suspicious of invasive carcinoma. Contrast-enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) were suggestive of lower rectal carcinoma extending to the anal canal with regional lymph node metastasis. As two prior biopsies were negative for carcinoma, polypectomy was performed. Macroscopically it was a mucosa covered, smooth brown polyp measuring 60 mm in maximum dimension with a tan cut surface. Histology revealed sheets and occasional nodular aggregates of monotonous small to medium sized lymphoid cells markedly expanding the mucosa and infiltrating the submucosa. The tumour cells were CD20 and BCL2 positive and Cyclin D1, CD3, CD10 and CD5 negative. The Ki-67 proliferation index was around 10%. The diagnosis of a low-grade B-cell lymphoma favouring a marginal zone lymphoma was made.

Discussion: MALT lymphoma cannot be distinguished from carcinoma clinically or on imaging. Endoscopically, it may mimic invasive carcinoma appearing as a sessile polyp or an ulcer. This case emphasizes that although a definitive diagnosis of lymphoma of the GI tract requires histology, superficial biopsies are often inadequate.

CR 16

Nephrotic syndrome in systemic lupus erythematosus with minimal histological changes: an atypical clinical presentation

S.B.K. Gamage*, S.A. *Rodrigo*

Department of Histopathology, Sri Jayewardenepura General Hospital, Sri Lanka

shathieka.gamage@gmail.com

Introduction: Lupus nephritis (LN) is a common manifestation of SLE, classified as classes I-VI. Classes I-II show minimal clinical and histological changes. Classes III-IV present with nephrotic range proteinuria, haematuria, and active sediments. Immune complexes on immunofluorescence are seen in all classes.

Case report: A 25-year-old woman treated for rheumatoid arthritis for two years presented with persistent joint pain and alopecia. Investigations showed a persistently high ESR (>100 mm/1st hour), low haemoglobin (6.6 g/dL), positive serum ANA and dsDNA, low serum C3 and C4 levels and normal serum creatinine. Her urine full report showed microscopic haematuria, urine protein to creatinine ratio (UPCR) was 6907 mg/g and no active sediments in urine. Her renal biopsy showed six viable normocellular glomeruli. There was no basement membrane thickening, no crescents, no interstitial inflammation or significant IFTA. Immunofluorescence staining showed focal non-specific trace positivity of IgM and C3 in glomerular basement membranes, while IgG, IgA, and C1q were negative.

Discussion: Nephrotic syndrome in LN is usually associated with proliferative glomerulonephritis. This patient's renal biopsy showed normal histology and immunocomplex negativity, which is not diagnostic of LN- Class I. Therefore, diagnoses such as NSAID use, under sampled FSGS, coincidental minimal change disease (MCD) and lupus podocytopathy (LP) should be considered. NSAID use was excluded in this patient. She responded well to steroids. LP, under sampled FSGS and MCD show diffuse foot process effacement in electron microscopy with an absence of immune complex deposition. Differentiating these is not essential as all are steroid-sensitive. Vigilant follow-up is mandatory due to the natural history of this disease's flare and dormancy.

CR 17

Congenital cystic adenomatoid malformation/congenital pulmonary airway malformation of the lung: perinatal autopsy finding

S.B.K. Gamage*, A.S. Rodrigo

Department of Histopathology, Sri Jayewardenepura General Hospital, Sri Lanka

shathieka.gamage@gmail.com

Introduction: Congenital cystic adenomatoid malformation (CCAM)/ congenital pulmonary airway malformation (CPAM) is a rare hamartomatous lung disorder reported in 1 per 25 000 births, which is associated with stillbirth, neonatal distress and bronchial atresia.

Case report: A 29-year-old primi mother with a 28-week POA presented with intrauterine death. A silent heart and air-filled cystic spaces were detected in the right lung on the US scan. The autopsy showed a dead male fetus with an enlarged right lung weighing 20 g, causing a mediastinal shift and compressing the left lung, which is 5 g in weight. The cut surface of the right lung showed multiple cysts measuring 5-10 mm in diameters. The left lung was macroscopically normal. Microscopy of the right lung depicted haphazardly arranged micro and macro cysts lined by ciliated columnar cells, and surrounded by a fibrous wall and separated by alveolar spaces. Microscopy of the left lung showed prominent bronchi with cartilaginous walls. Histology confirmed a CCAM/CPAM-type 2 of the right lung with absent cartilage, which is characteristic of this entity. No other external or internal congenital malformations were observed.

Discussion: CCAM/CPAM is of unknown aetiology, commonly seen in males and is detected on the antenatal US scan. CCAM/CPAM is classified into five groups based on clinicopathological features. CCAM/CPAM-Type 2 is associated with other anomalies such as renal agenesis, pulmonary hypoplasia and diaphragmatic hernia, although they were not evident in our case. Awareness of this entity is vital as surgical resection in new-borns and in utero drainage of macrocystic lesions in the fetus is lifesaving if promptly detected.

CR 18

Diffuse lipomatosis of thyroid gland: a rare entity

A. P. Ginige[1*], V. Varanitharan[2]

[1]*Department of Histopathology,* [2]*Surgical Unit, District General Hospital, Vavuniya, Sri Lanka*

prabhamalie@yahoo.com

Introduction: Diffuse lipomatosis of thyroid gland is a rare entity. It is characterized by diffuse fatty tissue infiltration in thyroid stroma.

Case report: A 56-year-old man was investigated for thyroid enlargement. He was clinically euthyroid. Ultrasound scan revealed diffused thyroid enlargement, and fine needle aspiration cytology of the thyroid glands revealed mature adipose tissue and stromal tissue. Macroscopically, the total thyroidectomy specimen showed a yellowish-brown colour enlarged lobular gland weighing 300 g. The right lobe measured 85x40x35 mm, the left lobe measured 80x40x25 mm and the isthmus measured 35x25x25 mm. The cut surfaces revealed a yellowish-brown colour appearance. Microscopic examination revealed extensive replacement of thyroid tissue by mature adipose tissue. Scattered thyroid follicles with colloid were seen within the adipose tissue. There was no evidence of a malignancy. The diagnosis of diffuse lipomatosis of the thyroid was made. The postoperative course was uneventful.

Discussion and Conclusion: Except for few adipocytes that may be found near the capsule and in the perivascular location, diffuse lipomatosis of thyroid is rare. This condition is usually seen in middle-aged group with no sex predilection. Natural history of diffuse lipomatosis of thyroid is unknown. Rarely, it has been reported in association with malignant thyroid neoplasms. Therefore, thorough sampling and further follow-up required.

CR 19

A rare case of cutaneous polyarteritis nodosa presenting as digital gangrene in a young female

N. W. Gunatilake*, J. Liyanage, H. Perera, K. Selvaratnam, T. Jeevasanthi

District General Hospital, Trincomalee, Sri Lanka

nisayuri@gmail.com

Introduction: Cutaneous polyarteritis nodosa (CPAN) is a rare form of necrotizing vasculitis of unknown aetiology relating to small-to-medium-sized arteries. It is limited to the skin and lacks progression to visceral involvement. Clinical manifestations include tender subcutaneous nodules, cutaneous ulcers and necrosis.

Case report: A 28-year-old woman presented with bluish discolouration of toes and fingers with poor response to treatment for Raynaud's disease, and later progressed to left big toe gangrene. She had no diabetes mellitus, no systemic symptoms of vasculitis, and distal pulses were normal. Her basic investigations including lower limb arterial duplex scan, and CT angiogram of abdomen and lower limbs were normal. ANA, C-ANCA, P-ANCA and hepatitis serology were negative. Biopsy from the edge of the lesion revealed leukocytoclastic vasculitis of thickened, medium-sized vessels at mid-deep dermis with neutrophil infiltration. Subcutis was normal. Eosinophils, thrombi, fibrosis or granulomata were not seen.

Discussion A deep biopsy is needed as CPAN involves medium-sized vessels in the deep dermis and subcutis. Histological findings can be discontinuous, therefore, examination of deeper levels is important. Lesions at four histological stages have been described: early lesions show fibrinoid necrosis, leukocytoclasia, and thickened vessel walls infiltrated by neutrophils, eosinophils and lymphocytes, progressing to thrombi and aneurysm formation with necrosis. Mature lesions show vessel wall fibrosis and occlusion. Small-medium vessel neutrophilic vasculitis with negative ANCA serology without systemic involvement differentiate CPAN from other differential diagnoses.

Conclusions: This is an unusual presentation of CPAN. However, after excluding common causes of digital gangrene clinically and with investigations, pathologists should consider CPAN disease spectrum and specific histologic features so that this rare entity is not missed. Further, clinical and laboratory findings must be correlated to make a definitive diagnosis.

CR 20

A case highlighting the possibility of diagnosing subcutaneous panniculitis like T-cell lymphoma on cytology

N. W. Gunatilake*, K. Selvaratnam, T.Jeevasanthi, A. Jayathilake

Department of Pathology, District General Hospital, Trincomalee, Sri Lanka

nisayuri@gmail.com

Introduction: Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare cytotoxic T-cell lymphoma accounting for < 1% of all non-Hodgkin lymphomas and characterized by multiple subcutaneous nodules occurring in the trunk and extremities. This is a case of SPTCL suspected on fine needle aspiration cytology (FNAC) and confirmed by histomorphology with immunohistochemistry.

Case report: A 33-year-old man presented with fever of five weeks and subcutaneous nodules in the epigastric region of three days duration. Full blood count was normal, erythrocyte sedimentation rate (ESR) was 24 mm/1hr, antinuclear antibodies (ANA) were negative and the ultrasound scan abdomen was normal. FNAC of the subcutaneous nodules revealed cellular smears comprising moderately pleomorphic medium-sized lymphoid cells with hyperchromatic irregular nuclei having clumped chromatin and mitoses with abnormal forms. These cells were closely admixed with fat lobules and inflammatory cells, including histiocytes and macrophages, in a necrotic background. Plasma cells or epithelioid histiocytes were not seen. These features were suspicious for SPTCL. Tru-cut biopsy of subcutaneous nodules revealed adipose tissue infiltrated by medium-sized atypical lymphocytes in a predominantly lobular pattern. These cells showed hyperchromatic irregular nuclei with abnormal mitoses. There was rimming of adipocytes by atypical cells. The background showed evidence of fat necrosis. The atypical cells showed positivity for CD3 and CD8 and were negative for CD20, CD4, CD30 and CD56. The Ki-67 proliferative index was 70-80%. These features were consistent with SPTCL. The patient's bone marrow was negative. He was given chemotherapy and is now in remission.

Discussion and conclusion: The diagnosis of SPTCL on cytology is rare. It needs to be differentiated from panniculitis, lupus erythematosus panniculitis and other lymphomas. The definitive diagnosis is assisted by the characteristic histomorphology and immunohistochemistry.

CR 21

A rare case of pancreatic carcinosarcoma

N. W. Gunatilake*, K. Selvaratnam, U. Samarajeewa, S. Subathra

Department of Pathology, District General Hospital, Trincomalee, Sri Lanka

nisayuri@gmail.com

Introduction: Pancreatic carcinosarcoma is a rare tumour with a dismal prognosis.

Case report: A 61-year-old woman presented with obstructive jaundice. CT scan revealed a mass in the pancreatic uncinate process. Pancreaticoduodenectomy was performed, which showed an irregular whitish firm lesion measuring 5.5 cm involving the pancreatic head and uncinate process. After thorough sampling, sections revealed a high-grade biphasic tumour with areas of necrosis. There was an admixture of predominant malignant spindle cell component (SpC) and focal areas of malignant epithelial components (EpC). SpC showed high-grade sarcoma with multinucleated tumour giant cells but no heterologous elements, and was positive for vimentin and negative for desmin, S100, CD117, BCL2 and CD34**.** EpC showed a predominantly high-grade adenocarcinoma and focal squamous cell carcinoma. EpC was positive for pancytokeratin, adenocarcinoma for CK7, and squamous cell carcinoma for CK5/6. Tumour was locally advanced**,** involving duodenum and pancreatic resection margins with positive lymph nodes. The postoperative period was uneventful, chemotherapy (Gemcitabine) was started and is currently on the second cycle.

Discussion: In the presence of sarcomatoid areas, and abundant cellular desmoplastic stroma in a pancreatic carcinoma, the possibility of carcinosarcoma needs to be considered. Primary pancreatic undifferentiated pleomorphic sarcoma and sarcomatoid carcinoma must be considered in the differential diagnosis of carcinosarcoma.

Conclusions: Adequate sampling is necessary to identify different tumour elements as in this case, where the epithelial elements were focal. Immunohistochemistry is necessary to confirm biphasic nature and to identify different sarcomatous elements. Accurate diagnosis is important as it influences further patient management. However, most patients have <12 months survival as the majority are locally advanced tumours.

CR 22

Metastatic carcinoma obscured by fibroblastic proliferation in the bone marrow

G. S. S. Hegoda*, D.M. Dissanayake, S. Wijetunge.

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

sanwimck@gmail.com

Introduction: Bone marrow (BM) deposits can be associated with a fibrotic stromal response that may results in a dry tap in BM aspiration or masking of the malignant cells in trephine biopsy.

Case report: A 42-year-old woman presented with dyspeptic symptoms and vomiting for one month. On examination, she had a mild pallor. On her initial diagnostic workup, she had bicytopenia. Her upper gastrointestinal endoscopy showed a gastric-outlet obstruction, and the CECT abdomen showed irregular thickening of the gastric pylorus extending into the 1st part of the duodenum. The initial biopsy of the gastric pylorus was inconclusive. A BM biopsy was performed due to bicytopenia, which showed BM tissue almost replaced by fibrous tissue, simulating myelofibrosis. On close examination, a few singly scattered suspicious cells were noted within this fibrous tissue, which were almost bland and could have been easily missed. However, some cells showed intracytoplasmic positivity with Alcian blue stain and the immunohistochemical stains showed strong cytoplasmic positivity with Pan CK. A diagnosis of a metastatic deposit of a poorly differentiated adenocarcinoma was made. The repeat biopsy of the pyloric lesion showed a poorly differentiated adenocarcinoma arising from the gastric pylorus.

Discussion and conclusion: Metastatic deposits of BM elicit a stromal response that may obscure the malignant cell population. A high degree of suspicion and clinical correlation is required to order special stains and immunohistochemical stains, which are invaluable to highlight the atypical cells in these instances.

CR 23

A case report of mucinous cystic neoplasm of the liver: a rare case associated with bile duct dilatation

H.H.W.S.B. Herath*, M.M.A. Jayawickrame

Department of Histopathology, National Hospital of Sri Lanka

wsbherath@gmail.com

Introduction: Mucinous cystic neoplasm of the liver (MCN-L) was defined by the WHO tumour classification system (2019) as a cyst-forming epithelial neoplasm, typically showing no communication with the bile ducts, lined by cuboidal to columnar, variably mucin-producing epithelium, associated with ovarian-type subepithelial stroma. MCN-L is rare, and the presence of bile duct dilatation is an even rare finding.

Case report: A 52-year-old woman presented with epigastric pain for eighteen months. Contrast enhanced computed tomography revealed a multiloculated, low-dense cystic lesion in the liver segment IV measuring 8.4x6.6x5.4 cm, causing compression of the common hepatic duct. A left hemi-hepatectomy with cholecystectomy was performed. The cyst was multiloculated and contained mucoid material, and there was no communication with the bile ducts. Microscopically, the cyst was lined by a single layer of columnar mucinous epithelium, which showed PAS positive, PAS-D resistant and Alcian blue positive cytoplasmic contents. Epithelial cells were positive for immunohistochemical stains CK7 and CK19. The underlying ovarian-like hypercellular stroma showed positivity for PR, focal positivity for ER and negativity for CD10. The postoperative course was uncomplicated.

Discussion: A biliary immunophenotype with the expression of CK7 and CK19 is characteristic of benign MCN-L. The presence of ER, PR positive ovarian type stroma excluded an intraductal papillary neoplasm and a simple hepatic liver cyst. CD10 negativity in stromal cells excluded endometriosis with mucinous metaplasia. Intrahepatic biliary dilatation, which was present in this patient, has only been seen in few reported cases of MCN-L.

Conclusion: Definite diagnosis of MCN-L relies on histological features. The prognosis of non-invasive biliary MCN is excellent.

CR 24

Sporadic variant of Burkitt lymphoma presenting as acute appendicitis

G.S.Hettiarachchi*, H.N.Thewarapperuma, D.J.Wickramasooriya, H.A.S.Perera

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

gayanisiluni@gmail.com

Introduction: Burkitt lymphoma (BL) is a highly aggressive lymphoma of follicular B-cell origin. Three epidemiological variants are recognized as endemic, sporadic and immunodeficiency associated BLs. Sporadic BL accounts for 30-50% of childhood lymphomas in western Europe and the USA. Most extra nodal cases present as abdominal masses, the ileocecal region being the commonest site. Lymph node involvement is exceedingly rare. This is a rare case of sporadic BL clinically presented as acute appendicitis.

Case report: An 11-year-old previously healthy boy had presented with acute onset abdominal pain and tender abdomen at the right iliac fossa, and an appendicectomy had been performed, which had shown a grossly inflamed appendix. Microscopy showed an intact surface epithelium of the mucosa, and there is diffuse infiltration of medium-sized monomorphic lymphoid cell population with brisk mitoses. Tingible body macrophages imparted a starry-sky pattern. The monomorphic lymphoid cells showed round nuclei with nucleoli and basophilic cytoplasm. These cells were immunoreactive for CD20, CD10, BCL6 and were negative for BCL2 and TdT. The Ki-67 proliferative index was >95%, confirming a BL. Immunostain with EBER expressed positive nuclear staining. Imaging studies (CT) showed diffuse thickening of the peritoneum, necrosis of hepatic parenchyma and bilateral mild pleural effusion. No mass lesions were noted. The child is awaiting chemotherapy.

Discussion: Pathologists should be aware of the rare pathologies present as acute appendicitis, and careful sampling is required. The Epstein-Barr virus is detected in 20-30% of sporadic BLs. The molecular hallmark is MYC translocation. BL is highly aggressive, but intensive chemotherapy leads to long-term survival in 70-90% of cases. The prognosis is better in children than in adults.

CR 25

A rare case of a dedifferentiated chordoma

P.M.S. Hiroshini*, T. Jayamaha, R.M.P. Ratnayake

Department of Pathology, National Hospital Kandy, Sri Lanka
surangi.hiroshini@yahoo.com

Introduction: Chordoma is a malignant tumour arising from embryonic notochordal remnants. Histological subtypes described are chordoma NOS, chondroid chordoma and dedifferentiated chordoma. A dedifferentiated chordoma is a biphasic tumour with a chordoma NOS component and a high-grade sarcomatous component.

Case report: A 71-year- old woman presented with pain in the lower back for two weeks, and recent weight loss. Per-rectal examination revealed a soft tissue mass in the presacral region. Computed tomography scan of the abdomen showed a solid and cystic lesion in the presacral region. The resected specimen contained a mass of gelatinous tissue measuring 50x70x80 mm with areas of haemorrhage and necrosis. The histology revealed two distinct morphological patterns. One area revealed a tumour composed of epithelioid cells with abundant bubbly cytoplasm in the background of myxoid material. The other area was a high-grade spindle cell sarcoma component, accounting for 10% of the tumour. Immunohistochemically the cells strongly expressed pancytokeratin, S100 and EMA with reduced expression of S100 and EMA in the high-grade sarcomatous area. Proliferative activity was elevated up to 40% in the high-grade areas. The tumour cells were negative for CK 7, CK20, GFAP, CDX2 and desmin.

Discussion: The differential diagnoses of the epithelial component were chordoma, chordoid meningioma, myxopapillary ependymoma, chondrosarcoma and metastatic deposits from a mucinous adenocarcinoma. However, in our patient, morphology and immunohistochemistry were suggestive of a chordoma with a focal high-grade spindle cell sarcoma component (dedifferentiated chordoma). Brachyury is the best marker to diagnose chordoma, in which the expression is weaker in sarcomatous area.

Conclusion: Diagnosis of dedifferentiated chordoma is established using a combination of clinical, radiological, histopathological and immunohistochemical studies. Precise diagnosis is of paramount importance for prognostic and treatment implications.

CR 26

A rare case of a pancreatic lipoma

P. M. S. Hiroshini*, A. Vithanage, R.M.P. Ratnayake

Department of Pathology, National Hospital Kandy, Sri Lanka

surangi.hiroshini@yahoo.com

Introduction: Pancreatic neoplasms are of epithelial, mesenchymal or non-ductal origin. The majority are epithelial, with adenocarcinoma accounting for about 85%. Mesenchymal tumours of the pancreas are rare and account for about 1%. The lipomatous tumours are the rarest of the mesenchymal tumours.

Case report: A 65-year-old woman presented with abdominal discomfort and loss of appetite for four months. Examination revealed tenderness in the epigastrium and the right hypochondrium. CT scan showed a well-defined mass with fat density within the abdomen extending from the sub hepatic region to S1 level measuring 192x184x126 mm. The excised lesion showed a well-demarcated lobulated mass of fatty tissue. Microscopy revealed lobulated mature adipose tissue with widely interspersed small islands of normal pancreatic tissue. There was no cellular pleomorphism, mitoses or necrosis. Lipoblasts were not seen. Other mesenchymal elements were not identified.

Discussion: Pancreatic lipoma must be distinguished from lipomatous pseudohypertrophy, teratoma and liposarcoma. Histologically lipoma is an encapsulated tumour composed of mature adipocytes. Lipomatous pseudohypertrophy of the pancreas is an unencapsulated lesion characterized by replacement of pancreatic tissue with adipose tissue, leaving only scattered clusters of pancreatic elements. Liposarcoma predominantly consists of mature looking fat tissue, which shows size and shape variation of adipocytic element with the presence of numerous multivacuolated lipoblasts. Teratoma often shows elements derived from ectodermal, endodermal origins and mesodermal components. Considering radiological findings and morphological features, the lesion was diagnosed as a benign lipomatous lesion consistent with a lipoma.

Conclusion: Since a benign lipomatous lesion is an important mimicker of malignancy, exact diagnosis is crucial for patient management.

CR 27

Follicular thyroid carcinoma metastasizing to frontotemporal bone and sphenoidal wing: a case report

P.M.S. Hiroshini*, A. Vithanage, R.M.P. Ratnayake

Department of Pathology, National Hospital Kandy, Sri Lanka

surangi.hiroshini@yahoo.com

Introduction: Thyroid carcinomas (TC) account for approximately 1.5% of all carcinomas in women and 0.5% in men. Follicular thyroid carcinoma (FTC) is a well-differentiated TC accounting for 6-10% of all TCs.

Case report: A 68-year-old woman presented with a well-circumscribed nodule in the right frontotemporal region of the skull for three weeks, which measured 3.5x3x1.5 cm. CT head revealed an infiltrating tumour involving the sphenoidal wing, extending to the right orbit, right ethmoid sinus and frontotemporal region. FNAC of the nodule revealed sheets and clusters of epithelial cells, with moderate anisonucleosis, forming numerous micro follicles. The histology of the excised nodule revealed follicular/acinar structures lined by cuboidal cells with rounded nuclei showing moderate nuclear pleomorphism. Some cells showed clear cell change. Intracytoplasmic stainable mucin was not identified. Immunohistochemically the tumour cells were strongly positive for CK7, TTF1 and thyroglobulin, confirming this as a metastatic deposit from an FTC. Subsequent USS thyroid showed a solitary nodule in the right lobe of the thyroid measuring 3x3x2.5 cm. The histology of the total thyroidectomy specimen confirmed this lesion as a widely invasive FTC.

Discussion and conclusion: The histological differential diagnoses of the initial biopsy were sinonasal adenocarcinoma, metastatic deposits from FTC, renal cell carcinoma and parathyroid carcinoma. Metastases may be the presenting lesion in some FTCs as in this case. Clinicopathological correlation supported by imaging studies and immunohistochemical markers helped to arrive at the correct diagnosis. The prognosis of FTC presents with metastases is generally poor.

CR 28

A malignant granular cell tumour of the thigh with distant metastases: a rare tumour

P.M.S. Hiroshini*, A. Vithanage, R.M.P. Ratnayake

Department of Pathology, National Hospital Kandy, Sri Lanka

surangi.hiroshini@yahoo.com

Introduction: Granular cell tumour (GCT) is a rare soft tissue neoplasm of neural origin, accounts for about 0.5% of all soft tissue neoplasms. Malignant GCT (MGCT) is an exceedingly rare and aggressive tumour, representing less than 1 - 2% of all GCT.

Case report: A 33-year-old woman presented with a painless left thigh mass of six months duration, which was rapidly enlarging for the last two weeks. CECT revealed a soft tissue sarcoma arising from the adductor compartment, with evidence of lymph node and bone metastases. Chest X-ray revealed multiple lung metastases. Macroscopically, the tan-coloured firm mass measured 80x60x60 mm. Microscopy revealed a tumour composed of nests and sheets of polygonal cells with increased nuclear-cytoplasmic ratio, vesicular and pleomorphic nuclei with prominent nucleoli, a mitotic count of 4/10 high power fields (HPFs) and abundant granular eosinophilic cytoplasm. The tumour was 1 mm from the closest resection margin. The Periodic acid-Schiff stain was positive, giving a strong magenta colour to the cytoplasm. The tumour cells strongly expressed immunohistochemical stain S-100 and were negative for desmin and MyoD1. The features were compatible with an MGCT.

Discussion: The differential diagnoses included MGCT, rhabdomyosarcoma, leiomyosarcoma with granular cell change and alveolar soft part sarcoma. Histological features met the Fanburg-Smith criteria to diagnose this tumour as MGCT.

Conclusion: Malignancy in GCT is debatable. The presence of metastases is currently considered the only unequivocal sign of true malignancy. In this patient, the diagnosis was arrived at after evaluation of clinical and radiological findings and the histological appearance and immunohistochemical pattern of the tumour.

CR 29

Metastatic high-grade goblet cell carcinoma of the appendix mimicking primary ovarian tumour: a case report

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

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

tharakaj.med@gmail.com

Introduction: Appendiceal goblet cell adenocarcinoma (A-GCC) is a rare subtype of appendiceal mucinous neoplasms (AMNs) included in new WHO tumour classification and no longer considered a neuroendocrine tumour. It neither forms a discrete tumour nor an enlarged appendix. Its ovarian metastases are usually bilateral. We report a metastatic A-GCC presenting as a unilateral ovarian mass mimicking primary ovarian tumour.

Case report: A 60-year-old woman presenting with abdominal pain was found to have a right adnexal mass on USS. Serum CA125 was 14 IU/mL. The total abdominal hysterectomy and bilateral salpingooophorectomy showed a right ovarian mass measuring 150 mm in maximum dimension, with a nodular, intact surface. The cut surface was white and solid with a few small cystic spaces. Tumour was confined to the ovary and comprised of infiltrating cords, nests, sheets, clusters, glandular structures and signet ring cells in extracellular mucin pools. Tumour cells were CK20 and CEA positive and CK7, ER and WT1 negative. Metastatic mucinous carcinoma was favoured and excluding primary appendiceal/gastrointestinal carcinoma was recommended. Upper and lower gastrointestinal endoscopy were normal. Subsequent laparoscopic appendicectomy showed firm thickening of body and tip with no discrete tumour. Histology revealed a circumferentially infiltrating tumour morphologically similar to the ovarian tumour with minimal stromal response in the wall and mostly intact mucosa. Synaptophysin and CK19 were focally positive in the goblet cells in the tumour.

Discussion: Ovarian metastases in AMNs are usually bilateral, with grossly enlarged appendix and/or pseudomyxoma peritonei. This case highlights that A-GCC can present with unilateral ovarian involvement and grossly normal appendix, mimicking primary ovarian malignancy. Thus, excluding primary appendiceal tumour intraoperatively is paramount in preventing delays in diagnosis and treatment.

CR 30

An abdominal mass mimicking an aortic aneurysm: a histological challenge

S.S.Jayasundara*, R.M.P.Ratnayake, C.Weerasinghe

Department of Histopathology, National Hospital Kandy, Sri Lanka

sudharikajaya@gmail.com

Introduction: Solid sclerosing retroperitoneal masses are a diagnostic challenge for pathologists since different histological entities can have a similar presentation. We report a case of a 61-year-old man who presented with a paraaortic mass, which was radiologically suspected to be an aortic aneurysm.

Case report: A 61-year-old man presented with chronic back pain for seven months. Radiological evaluation suggested an infrarenal aortic aneurysm measuring 4.2x3.9 cm with an associated thrombus. Consequently, he underwent a mini laparotomy and a sclerosed retroperitoneal mass encircling the aorta was detected and excised. Macroscopy revealed firm white tissue measuring 15x10x10 mm. Histology showed storiform fibrosis and a dense infiltrate of plasma cells, lymphocytes and eosinophils. The presence of storiform fibrosis and immunohistochemical confirmation with CD138 and IgG, favoured the diagnosis of IgG-4 related disease. Elastic van Gieson stain highlighted the vessels with obliterative phlebitis. Other connective tissue disorders were not suspected, and the patient is currently being investigated.

Discussion: The differential diagnoses include inflammatory myofibroblastic tumour, low-grade lymphoma, Castleman disease (plasma cell variant) and IgG-4 related disease. A mixed population of T and B-lymphocytes highlighted by CD3 and CD20 and the absence of Reed Sternberg cells excluded the possibility of lymphoma. Negativity of ALK immunomarker made inflammatory myofibroblastic tumour unlikely. Involvement of the aortic wall is one of the commonest presentations of IgG4 related disease. Serum IgG4 levels were requested to confirm the diagnosis.

Conclusion: Sclerosed paraaortic masses pose a diagnostic challenge. IgG-4 related disease is a possible differential diagnosis.

CR 31

Pure invasive micropapillary carcinoma: a case report on a rare entity in the male breast

G Jayathilake[1], R. D.K. Medonsa*[2]

[1]*Department of Pathology, Base Hospital Matale, Sri Lanka*

[2] *Department of Pathology, Faculty of Medicine and Allied Sciences, University of Rajarata*

dharanikdw@gmail.com

Introduction: Pure invasive micropapillary carcinoma of male breast is exceedingly rare. This morphologically distinctive, aggressive subtype often accompanies an in-situ or conventional duct carcinoma (CDC) component.

Case report: A 53-year-old man presented with a painful mass in the left breast for one week. Ultrasound scan revealed a hypoechoic lobulated mass, and FNAC was reported as malignant (C5). The subsequent radical mastectomy revealed a well-defined subareolar tumour measuring 35 mm in maximum diameter. Epithelial structures were composed of micropapillae, hollow tubules and morula in empty spaces with cells arranged in reversed polarity. Despite extensive sampling, no in-situ or CDC component was found. Lymphovascular emboli were seen but all four axillary lymph nodes were tumour free. Tumour cells were strongly and diffusely positive for ER and faintly positive for PR. HER2 showed complete 3+ and incomplete moderate basolateral staining in around 90% and 10% of cells, respectively. EMA showed an inside-out membranous staining. Patient has not yet complied with treatment but five months after surgery he is free of recurrence.

Discussion: The definitive diagnosis of IMPC is imperative due to its poor prognosis with low recurrence-free survival compared to stage-matched CDC of breast. Characteristic immunohistochemistry helps in differentiating IMPC from metastatic micropapillary carcinoma, especially from the lung and bladder. This is a unique case of pure IPMC in an adult man. Due to the scarcity of entities such as IPMC among men, available information on the behaviour of these malignancies is minimal. However, precise diagnosis of this rare entity is important in deciding the follow-up protocol.

CR 32

Clear cell variant of follicular thyroid carcinoma: a rare entity

P.G.A.N. Jayathilaka*, M.A.D.N. Munasinghe, S.M. Fernandopulle

Department of Histopathology, Colombo North Teaching Hospital, Ragama, Sri Lanka

amalinijaya@gmail.com

Introduction: Clear cell variant of follicular thyroid carcinoma is a rare subtype of thyroid cancer.

Case report: A 43-year-old woman presented with a multinodular goitre. USS revealed two nodules in each lobe and FNAC was compatible with Thy2 (colloid goitre). Thyroidectomy specimen showed colloid nodules and an encapsulated 35 mm nodule in the left lobe. Microscopy of this nodule revealed solid nests and microfollicles with a prominent vascular network, completely surrounded by a fibrous capsule. Vascular invasion was identified but capsular invasion was absent. Almost all the tumour cells (>50%) had central small nuclei and clear cytoplasm. Nuclear clearing, overlapping, grooves or pseudo-inclusions were not present. These cells were positive for TTF1 and thyroglobulin, and negative for CD10, vimentin, synaptophysin and chromogranin A. Hence, this was diagnosed as clear cell variant of angio-invasive follicular thyroid carcinoma

Discussion: Morphologically clear cell change in a thyroid tumour may be either of primary thyroid or extra-thyroid origin. Thyroid neoplasms include follicular adenoma and carcinoma, papillary carcinoma, medullary carcinoma and oncocytic neoplasms. Metastatic tumours include clear cell renal cell carcinoma and parathyroid neoplasms. In this case, thyroid origin was confirmed by positivity for TTF1 and thyroglobulin. Clear cell variant of follicular thyroid carcinoma, although rare (0.14 -3%), was considered in the absence of overt nuclear features of papillary thyroid carcinoma. Negativity for CD10 and vimentin with radiologically normal kidneys excluded a metastatic renal cell carcinoma. Negative neuroendocrine markers excluded a parathyroid neoplasm and medullary thyroid carcinoma.

Conclusion: Careful morphological examination with immunohistochemistry, clinical and radiological correlation is mandatory for diagnosis of this rare variant.

CR 33

Metastasis of hepatocellular carcinoma presenting as a soft tissue swelling, diagnosed on fine needle aspiration cytology

D. N. U. Jayathunge*[1], P. Vishwajith[2], T.P.M. Bopagoda[1].

[1]Department of Pathology, [2]Department of Surgery, Faculty of Medicine, University of Kelaniya, Sri Lanka

nishaniu@ymail.com

Introduction: Rapid diagnosis of extrahepatic metastasis of hepatocellular carcinoma (HCC) without delay is crucial for planning potential therapy, as the prognosis is poor. We report a rare case of HCC presenting as a progressively enlarging lump over the left sternoclavicular joint, which was diagnosed by fine needle aspiration cytology (FNAC).

Case report: A 65-year-old man presented with a progressively enlarging lump on the anterior chest wall for three months. On examination, it was a firm, pulsatile lump located over the left sternoclavicular joint. Radiological investigations revealed a 3.5×2.9 cm, heterogeneously echogenic, sternal soft tissue mass with internal vascularity and destruction of the underlying bone. FNAC of the lump showed cellular smears comprising sheets, clusters, and trabeculae of round to polygonal cells. These cells showed abundant, eosinophilic, granular cytoplasm and centrally located large nuclei with vesicular chromatin and prominent nucleoli. Endothelial cells were seen traversing some of the cell clusters and some cells showed cytoplasmic bile pigments. The cytological features were suggestive of an HCC. Imaging of the liver revealed multifocal lesions in both lobes with typical arterial phase enhancement with portal venous wash-out on computerized tomography (CT), confirming the radiological diagnosis of an HCC. No ancillary investigations, including alpha-fetoprotein levels or histological confirmation was carried out as the radiology and the cytology was consistent with HCC.

Discussion: Hepatocellular carcinoma can present clinically as a palpable soft tissue swelling. This case highlights the use of FNAC in the diagnosis of such lesions.

CR 34

Portal vein tumour thrombosis in pancreatic neuroendocrine tumour: a rare finding

D.N.U. Jayathunga*[1], R.M.P. Vishwajith[2], T.P.M. Bopagoda[1]

[1]Department of Pathology, [2]Department of Surgery, Faculty of Medicine, University of Kelaniya, Sri Lanka

nishaniu@kln.ac.lk

Introduction: Tumour thrombosis is an uncommon finding seen in about 5% of pancreatic neuroendocrine tumours (PanNETs). It is frequently underreported on pre-operative imaging and is associated with increased post-surgical morbidity and mortality as thrombosis of peri-pancreatic vessels may cause spleno-porto-mesenteric hypertension, leading to life-threatening upper gastrointestinal bleeding. This is a case of a PanNET with tumour thrombosis of the portal vein**.**

Case report: A 58-year-old woman presented with obstructive jaundice for one month. The CECT of the abdomen revealed a pancreatic neoplasm arising from the uncinate process encasing the superior mesenteric vessels and compressing the common bile duct. The Whipple resection specimen revealed a circumscribed tumour in the head of the pancreas measuring 80x60x55 mm. The tumour involved the superior mesenteric vein with tumour thrombosis in the portal vein, 3.6 mm away from the vessel resection margin. The microscopic examination revealed a grade 3 PanNET encasing and involving the wall of the superior mesenteric vein. There was perineural invasion, vascular emboli and tumour thrombosis of the portal vein. The tumour involved the anterior surface, inferior resection margin and the transected margin of the pancreas. All thirteen regional lymph nodes were free of tumour metastasis. The patient was free of complications related to tumour thrombosis.

Discussion: Peripancreatic vessel thrombosis caused by direct infiltration of tumour determines surgical unresectability, even in neuroendocrine tumours. Therefore, it is vital to detect this either pre-operatively or within the resected specimen by the pathologist to alert the clinicians for close post-operative monitoring and reassessment of the patient to minimize complications.

CR 35

A case of xanthogranulomatous endometritis: a rare entity

M. P. G. N. S. Jayawardhana*, B. H. Perera, L. C. W. Samaranayake.

Base Hospital, Thambuttegama, Sri Lanka

sanjeewanijayawardhana77@gmail.com

Introduction: Xanthogranulomatous endometritis is a rare form of endometritis clinically mimicking endometrial carcinoma which mainly affects elderly women. Even though several cases have been reported globally, case reports from Sri Lanka are not encountered in the literature.

Case report: A 70-year-old woman presented with a 2-week history of yellow vaginal discharge. Ultrasonically a pyometron was diagnosed. Fragments of white-brown endometrial curettings revealed a few resting endometrial glands in a stroma containing sheets of foamy histiocytes with plasma cells, lymphocytes, focal neutrophils, foreign body-type multinucleated giant cells and siderophages. A pathological diagnosis of xanthogranulomatous endometritis was made. Macroscopically the uterus was not enlarged and cut sections revealed yellow brown material in the endometrial cavity with irregular yellow coloured endometrium of up to 5 mm in thickness. Microscopy of the contents of the endometrial cavity showed sheets of foamy histiocytes admixed with inflammatory cells and necrotic debris. The endometrium had similar histological features of endometrial curettings, which confirmed the diagnosis.

Discussion: Xanthogranulomatous inflammation is characterized by the presence of sheets of foamy macrophages with other inflammatory cells. It is mainly associated with haematometra, pyometra or after radiation therapy due to cervical obstruction.

Conclusion: The rare entity of xanthogranulomatous endometritis is a strong mimicker of endometrial carcinoma and requires histological assessment for a definitive diagnosis.

CR 36

Blastoid mantle cell lymphoma with isolated extranodal disease presenting as a rapidly enlarging tonsillar mass

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

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

supipinayani@gmail.com

Introduction: Mantle cell lymphoma (MCL) is an uncommon, mature B-cell non-Hodgkin lymphoma, predominantly affecting elderly men. It commonly involves lymph nodes. Extranodal disease occurs less frequently in gastrointestinal tract and Waldeyer's ring. We present a rare case of blastoid MCL with isolated extranodal disease (IED) involving tonsils at the time of diagnosis.

Case report: A 73-year-old man investigated for generalized ill-health was found to have lymphocytosis, high ESR and monoclonal B-cell lymphocytosis in peripheral blood flow cytometry. Bone marrow showed no evidence of a lymphoma. Serum LDH was 318 U/L. The CT and MRI showed mild splenomegaly with low attenuated lesions and the apical fibrosis of the left lung suggestive of old tuberculosis changes. There was no evidence of lymphadenopathy. Screening for tuberculosis was negative. He defaulted follow up and presented four months later with odynophagia and an enlarging left tonsillar mass. The tonsillar biopsy showed diffuse infiltration of monotonous, medium-sized, lymphoid cells and scattered large cells with dispersed chromatin. Abundant mitoses and focal spotty necrosis were seen. The tumour cells were strongly positive for CD20, CD5, Cyclin D1 and BCL2 and were negative for CD3, BCL6, CD10, TdT and CD23. The Ki-67 proliferative index was 90%. A diagnosis of blastoid mantle cell lymphoma was made. He has clinically improved with four cycles of R-CHOP chemotherapy.

Discussion and conclusion: Blastoid MCL with IED at the time of diagnosis is a rare event and the outcome of this disease is not well documented. Few studies were carried out on MCL with IED, which showed that this variant has an indolent course and good prognosis compared to classical nodal type.

CR 37

Bilateral ovarian serous cystadenofibromas presenting as a malignancy

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

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

supipinayani@gmail.com

Introduction: Ovarian cystadenofibroma is a rare benign tumour composed of both epithelial and fibrous stromal components, accounting for 1.7% of all benign ovarian tumours. We present a case of bilateral ovarian cystadenofibromas in a postmenopausal woman, which were radiologically and intraoperatively suspicious of ovarian malignancy.

Case report: A 65-year-old woman presented with lower abdominal pain for six months. The CT scan showed bilateral complex cystic and solid ovarian masses suspicious of malignancy. She underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy. On gross examination, both ovarian masses were multicystic with firm, yellowish-white solid areas amounting to more than 75% of the tumour. The histology of both masses showed tumours predominantly composed of stromal elements with scattered glands lined by tall columnar ciliated cells resembling tubal epithelium. The stroma was composed of short fascicles of spindle cells with bland elongated nuclei and scanty eosinophilic cytoplasm, arranged in a storiform pattern in a collagenous background. There was no evidence of cellular atypia, mitoses, or necrosis. Reticulin stain highlighted the individual spindle cells of the stroma. Omental tissue was normal.

Discussion and conclusion: The appearance of cystadenofibroma on routine imaging like USS and CT often resembles a malignant tumour which can lead to unnecessary, extensive surgery, especially in young patients, as this tumour tends to occur in a wide age range of 15–65 years. An awareness of this rare entity and its characteristic MRI findings of dense fibrous stromal proliferation with scattered small cystic glandular structures on the T2-weighted images and the use of intra-operative frozen sections can prevent unnecessary surgery in young patients.

CR 38

Bowen disease arising in seborrheic keratosis

N. P. Karunathilake[1*], S. Wijetunge[1], A. Dharmapala[2]

[1]Department of Pathology, [2] Department of Surgery, Faculty of Medicine, University of Peradeniya, Sri Lanka

Prasadini87@yahoo.com

Introduction: Seborrheic keratosis (SK) is a benign skin lesion, with no gender predilection and rarely (7%) shows malignant transformation into non-melanotic-carcinomas (5.4%) like basal cell carcinomas and squamous cell carcinomas (SCC), melanomas, keratoacanthomas and eccrine carcinoma. Frequency of Bowen disease (BD) in SK is rare (1.4%). SK is usually a coin-like, exophytic/flat, sharply demarcated lesion in head and neck region (78.3%).

Case report: A-72-year-old man presented with a painless nodular skin lesion over the right shoulder for two years, with recent enlargement and appearance of satellite nodules over three months. Examination revealed three nodules with irregular margins with white and dark discolouration. Excision biopsy was performed to rule out a malignancy. Macroscopy revealed a skin ellipse with underling fatty tissue measuring 20x25x10 mm. There were three nodules: the larger lesion is 6 mm, and two satellite nodules were 3 mm each. Microscopy of the larger nodule revealed epidermis with marked basaloid cell proliferation and numerous pseudohorn cysts. High power examination revealed areas of cellular pleomorphism of varying degrees with increased mitoses involving all layers. The satellite nodules revealed a similar degree of dysplasia. All lesions were entirely processed, and invasion was absent. Hence the diagnosis of BD arising in a SK was made.

Discussion: BD is a form of SCC in-situ with the potential for significant lateral spread. Though SK is not premalignant, malignancy can arise from basal layer cells, spinous layer cells and melanocytes. A long-standing lesion with recent changes indicates that the dysplastic changes are a recent development.

Conclusion: Careful histological evaluation of SK is important to avoid missing rare complications such as dysplasia developing in SK, especially with atypical clinical features.

CR 39

A case report of dyshormonogenetic goitre

V. Kemanilamiya*, M.M.A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

hemanilamiya456@gmail.com

Introduction: Dyshormonogenetic goitre (DG) is a rare cause of congenital hypothyroidism occurring due to lack of enzymes necessary for the synthesis of thyroid hormones.

Case report: A 17-year-old girl presented with a diffuse enlargement of the thyroid gland and the ultrasound scan showed a coarse echogenic gland with increased vascularity. Fine needle aspiration was suspicious of a papillary carcinoma (PTC). Investigations revealed a markedly increased thyroid stimulating hormone (TSH) level of 128.828 μU/mL and a very low free T4 level of less than 0.01 ng/dL. A thyroidectomy was performed, and macroscopic examination revealed a diffusely enlarged firm gland with a white cut surface. Histology revealed multiple nodules separated by fibrous septae. The nodules and inter nodular areas were composed of follicles, trabeculae and papillary infoldings. Focal areas showed nuclear features of PTC with nuclear clearing, overlapping and grooves. True papillae and psammoma bodies were not seen.

Discussion and conclusion: DG is a rare benign condition that may lead to overdiagnosis of PTC in cytology and histology. Associated follicular carcinoma and PTC have been reported in patients with DG. DG needs to be considered in young patients presenting with goitre and it is important to be aware of the overlapping cytological and histological features with PTC. Confirmation with molecular diagnosis allows genetic counselling and the identification of asymptomatic carriers.

CR 40

Cutaneous ALK-1 negative anaplastic large cell lymphoma with lymph node metastasis

V. Kemanilamiya*, S. R. Constantine

Department of Histopathology, National Hospital of Sri Lanka

hemanilamiya456@gmail.com

Introduction: Cutaneous ALK-1 negative anaplastic large cell lymphoma (ALCL) is a CD30-positive T-cell lymphoma. It is the second commonest cutaneous lymphoma.

Case report: A 56-year-old woman with diabetes mellitus had itchy skin lesions for 10 years. Initially these lesions started on the forearm then gradually spread to other areas. She had no systemic symptoms. Previous skin biopsies had varying benign diagnoses and there were no large cells or features of mycosis fungoides. There was no response to treatment. The most recent presentation was with unilateral cervical lymphadenopathy. We received biopsies from a skin nodule and cervical lymph node. Microscopically of both showed similar morphology with diffuse sheets of large, atypical cells in a background of inflammatory cells predominantly comprising eosinophils. The cells contained kidney-shaped vesicular nuclei with prominent nucleoli. Multinucleated cells and atypical mitoses were seen. The large cells were positive for CD3 and CD30 and negative for CD20, CD15, PAX5 and ALK-1. The Ki-67 proliferative index was 90%.

Discussion and conclusion: The presence of large atypical lymphoid cells and strong positivity for CD3 and CD30 with negative CD15, CD20, PAX5 and ALK differentiate the cutaneous ALK-1 negative ALCL from classic Hodgkin lymphoma. The presence of skin lesions and absence of systemic symptoms favour cutaneous ALCL rather than nodal ALCL with skin involvement. Patients with skin lesions and involvement of regional lymph nodes have a prognosis similar to that of patients with only skin lesions, and a 10-year disease specific survival rate of 90%.

CR 41

Mucinous cystadenocarcinoma of breast: a case report of a newly recognized rare malignant breast tumour

K. Kopinath*[1], C. Senarath[2], K. Dissanayake[1]

[1]*Department of Histopathology, National Hospital of Sri Lanka*

[2]*Department of Pathology, Teaching Hospital, Ratnapura, Sri Lanka*

kathirkopi@gmail.com

Introduction: Mucinous cystadenocarcinoma of the breast (MCCB) is an extremely rare subtype of invasive breast carcinoma, with limited reported cases in the literature. MCCB is composed of multiple cystic spaces lined by columnar cells with variable atypia, containing abundant intracytoplasmic and extracellular mucin.

Case report: A 45-year-old woman presented with a left side breast lump for six months. On examination, there was a hard lump in the subareolar region measuring 10 cm in diameter. Ultrasound scan revealed a heterogeneously enhancing solid lesion with cystic components. Fine needle aspiration cytology showed malignant cells. The mastectomy specimen showed a solid tumour with a gelatinous cut surface. Microscopy revealed multiple irregular cystic spaces filled with mucin and contained papillary structures with fibrovascular cores lined by simple and stratified columnar cells. The tumour cells contained abundant intracellular apical mucin and moderately pleomorphic basally located vesicular nuclei and scattered mitoses. There was cancerization of lobules and an adjacent in-situ carcinoma component with mucinous differentiation. The tumour was positive for immunohistochemical stains mammaglobin and CK7 and negative for CK20, ER, PR and HER2. The Ki-67 proliferation index was 26%.

Discussion: MCCB shows similar morphology of ovarian and pancreatic cystadenocarcinoma. Most of the MCCBs are negative for ER, PR and HER2. They are positive for CK7, mammaglobin and GCDFP-15. The presence of in-situ carcinoma component and negativity for CK20 help to exclude the possibility of metastatic deposit of pancreatic cystadenocarcinoma.

Conclusion: MCCB is included as a new entity in the latest WHO classification of tumours of the breast as it has a unique morphology and favourable prognosis.

CR 42

Mixed epithelial and stromal tumour of kidney: a case report of a rare benign tumour of the kidney

K. Kopinath*, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

kathirkopi@gmail.com

Introduction: Mixed epithelial and stromal tumour (MEST) of the kidney is a rare benign tumour. It is a biphasic tumour composed of epithelial and stromal components. MEST predominantly occurs in perimenopausal women as a solid and cystic tumour.

Case report: A 44-year-old woman presented with a history of left loin pain of six months duration. Computerized tomography intravenous urogram (CT-IVU) showed a 5 cm complex cyst with a solid component (BOSNIAK category III cyst) in the lower pole of the left kidney. A partial nephrectomy was performed, which showed a well-circumscribed unencapsulated solid and cystic tumour with a rim of normal renal parenchyma. Microscopy revealed varying sized cysts and glandular structures lined by cuboidal to columnar epithelium and focal stratification. Haemorrhage, mitoses and necrosis were not seen. The stroma was composed of plump spindle cells with focal areas resembling ovarian stroma and bundles of smooth muscle. Condensation of stromal cells around the cysts was also noted. The stroma showed strong nuclear positivity for ER and PR. The smooth muscle bundles and stromal cells showed strong cytoplasmic positivity for SMA. CD10 showed positivity in the epithelium and stromal cells surrounding the epithelium.

Discussion and conclusion: Even though MEST is considered a benign neoplasm with a good prognosis, it has a potential for malignant transformation. Careful microscopic examination is essential as malignant transformation may occur in both stromal and epithelial components. Nephron sparing surgery is the main stay of treatment. Cystic nephroma, cystic partially differentiated nephroblastoma, angiomyolipoma and biphasic synovial sarcoma need to be considered in the differential diagnosis.

CR 43

Granulomatosis with polyangiitis presenting with involvement of the nasal septum and skin: a case report

P. D. K. Kumari*, K. Samalai, K. Kopinath, C. M. Baranasuriya, M. M. A. Jayawickrama,

K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

wijegunawardanepdkk@gmail.com

Introduction: Granulomatosis with polyangiitis (GPA), previously known as Wegener granulomatosis, is a rare, potentially lethal vasculitis affecting small to medium-sized blood vessels. Hallmark histological features are necrotizing granulomata and pauci-immune vasculitis, most commonly affecting the upper respiratory tract, lungs and kidneys. Typically, GPA is found in middle age but may occur in people of all ages.

Case report: A 63-year-old man, who was previously well, presented with headache for three months and bilateral leg ulcers. Rigid nasal endoscopic examination detected a necrotic nasal septum with surrounding ulceration and sinus X-ray and CECT showed the left maxillary and frontal sinus opacity. Biopsies were taken from the nasal septum and the leg ulcers. Microscopically, the fragmented nasal mucosal tissue showed geographic necrosis, thrombosed and necrotic blood vessels containing vasocentric granulomas with giant cells and dense mixed inflammation. The skin biopsy showed leukocytoclastic vasculitis of small to medium-sized vessels, vasocentric granulomas and prominent areas of geographical necrosis. The background showed mixed inflammation with scattered multinucleated giant cells. Grocott and PAS stains for fungi and Ziehl-Neelsen stain for mycobacteria were negative. During the follow-up, the patient was detected to have a left lung lesion and positivity for c-ANCA. He was started on immunosuppressive treatments, which improved the symptoms.

Discussion and conclusion: GPA principally presents with lung involvement at the time of diagnosis. The main morphologic changes are vasocentric granulomas and geographical necrosis. Rarely, sinonasal involvement may be the first manifestation, as in this case. Therefore, GPA needs to be considered in the presence of extensive necrosis and vasocentric granulomata in biopsies from the upper respiratory tract.

CR 44

Multiple uterine lipoleiomyomas: a case report

S. K. Liyanage*, P. N. Managoda

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

saumyak40@yahoo.com

Introduction: Uterine lipoleiomyoma is a rare variant of leiomyoma, composed of smooth muscle cells admixed with variable proportions of mature adipose tissue. The incidence varies from 0.03% to 0.2%. This is an unusual case of two uterine lipoleiomyomas in a postmenopausal woman, predominantly containing mature adipose tissue.

Case report: A 63-year-old postmenopausal woman diagnosed with diabetes mellitus and hypertension presented with progressively worsening pelvic pain for two years. The CT scan revealed a mass in the pouch of Douglas with two well-defined fat containing areas and was radiologically suspicious for malignancy. A total abdominal hysterectomy and bilateral salpingo-oophorectomy was performed, which revealed two separate firm, circumscribed nodules in the uterus, one subserosal and the other intramural, measuring 50mm and 60mm in maximum diameters, respectively. The cut surfaces were homogenously fatty. Microscopically, both nodules were composed predominantly of mature adipose tissue intermingled with occasional bundles of regular spindle cells. There was no significant size variation in lipocytes, no atypical cells, mitoses, necrosis, epithelial or heterologous elements. The spindle cell component was positive for immunohistochemical stains smooth muscle actin and desmin. These features were compatible with lipoleiomyomas with mature adipose tissue as the main component. There was no evidence of endometrial hyperplasia or any other additional pathology.

Discussion: Lipoleiomyomas, although benign, cause diagnostic difficulties due to rapid growth and the appearance on imaging studies, particularly in postmenopausal women. Therefore, a definite diagnosis can only be achieved by histological assessment. Lipoleiomyomas are known to occur frequently in hyperoestrogenic states, including diabetes mellitus.

CR 45

Invasive micropapillary urothelial carcinoma: a rare aggressive subtype presenting with multifocal disease

K. S. Maheshika*, M.A.D.N. Munasinghe, N.G.M.M Amarasinghe, S.M. Fernandopulle

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

saramaheshika@gmail.com

Introduction: Invasive micropapillary urothelial carcinoma is an aggressive subtype, presents at higher stage and carries a poor survival. This is a well-defined entity which needs to be diagnosed distinctly from ordinary urothelial carcinoma. Multifocality is frequent.

Case report: A 73-year-old woman was investigated for one episode of gross haematuria. Imaging showed upper tract urothelial carcinoma in the left kidney, a synchronous lesion in the bladder and regional lymph node metastases. Other organs were normal. Transurethral resection of the bladder tumour showed a carcinoma predominantly composed of (70%) small, tight nests within retracted connective tissue spaces forming lacuna. Reverse nuclear polarity marked nuclear pleomorphism and prominent nucleoli were seen. Tumour cells revealed strong, diffuse positivity for CK7 and patchy positivity for CK 20. The left nephroureterectomy, specimen revealed two synchronous tumours in the renal medulla and mid ureter showing similar histology to that of bladder carcinoma. Lymphovascular tumour emboli and hilar and paraaortic lymph node metastases were present. The patient has improved symptomatically in the six months of follow up and is waiting adjuvant therapy.

Discussion and conclusion: Invasive micropapillary urothelial carcinoma should be diagnosed accurately as these patients require vigorous treatment. The carcinogenesis pathway is different to that of conventional urothelial carcinoma, and they show a luminal type molecular profile. They are positive for CK 7, CK 20, EMA, uroplakin II, GATA 3 and PPARγ. They commonly express strong basal MUC 1 positivity and show more HER2 over-expression than conventional urothelial carcinoma with retraction artifact.

CR 46

Ovarian stromal hyperthecosis mimicking ovarian neoplasms: report of two cases

M.D.J.C Malagala*, D.H. Beneragama

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

malagalajayani@gmail.com

Introduction: Ovarian stromal hyperthecosis (OSH) is defined by the presence of luteinized cells in the ovarian stroma. It occurs in elderly, obese females and is associated with virilization and insulin resistance.

Case report: Case 1- A 68-year-old woman presented with postmenopausal bleeding. Ultrasound scan showed increased endometrial thickness. Curettings were suspicious of endometrial carcinoma. Total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH and BSO) was performed. The uterus showed irregularly thickened endometrium. Multiple 2-3 mm white nodules were seen in both ovaries mimicking tumour deposits. Histology showed an endometrial carcinoma (FIGO I) in the uterus, and nodules of luteinized stromal cells in both ovaries.

Case 2- A 79-year-old woman diagnosed with breast carcinoma on tamoxifen, was found to have a 3x4 cm left ovarian cyst on imaging, suspicious of a metastatic tumour. CA125 was 5 U/mL. TAH and BSO was performed. The left ovary showed a 3 cm, haemorrhagic tumour with focal tan-coloured areas. Histology of the uterus showed endometrial hyperplasia. There was a left ovarian haemangioma with sheets of luteinized stromal cells positive for inhibin.

Discussion and Conclusion: OSH is typically bilateral and defined as differentiation of ovarian stromal cells into luteinized, steroidogenically active, androgen producing cells. Peripheral aromatization of androgens results in oestrogenic effects with increased risk of endometrial carcinoma (Case 1), especially in postmenopausal women. Tamoxifen induced oestrogenic effect (Case 2) can result in endometrial hyperplasia, but an ovarian haemangioma is a rare occurrence. This may possibly be due to stimulation of oestrogen-sensitive endothelial cells. Mechanical compression by the expanding haemangioma might have resulted in stromal luteinization. As OSH mimics metastatic deposits clinically and macroscopically, histology is required to confirm the diagnosis.

CR 47

An atypical teratoid/rhabdoid tumour: a case report

I.W.G.A.L. Malhasi *, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

malhasig@gmail.com

Introduction: Atypical teratoid/rhabdoid tumour (AT/RT) is a malignant central nervous system (CNS) embryonal tumour occurring in the paediatric population. It comprises poorly differentiated elements including rhabdoid cells. We report an AT/RT occurring in the right cerebello-pontine angle (CPA) of a boy presenting with ipsilateral facial nerve paralysis.

Case report: A 4-year-old boy presented with deviation of the mouth to the left side and inability to close the right eye for two weeks. MRI revealed a cystic tumour in the right CPA causing a mass effect. He underwent surgery. Histology revealed an infiltrative, necrotic tumour composed of cells arranged in nests and sheets. Constituent cells had abundant eosinophilic cytoplasm and eccentric, vesicular nuclei with prominent nucleoli. Mitotic activity was brisk with atypical forms. Rosette formation and microvascular proliferation were not seen. Immunohistochemistry showed positivity for vimentin, EMA and SMA. The Ki-67 proliferation index was 56%. The tumour cells were negative for GFAP, PLAP, synaptophysin and chromogranin. A diagnosis of CNS embryonal tumour with rhabdoid features was made based on morphology and immunohistochemistry.

Discussion and conclusion: Although rare, AT/TR is one of the differential diagnoses for primary CNS tumours in the paediatric population when presenting with specific clinical features and symptoms of a short duration. Loss of nuclear expression of SMARCB1 protein (INI) demonstrated by molecular genetic studies or by immunohistochemistry is essential for diagnosis of AT/RT. Combined assessment of cytology, histology and immunohistochemistry in the appropriate clinical and radiological context is the key to accurate diagnosis of CNS tumours, especially when specific mutational analysis and molecular studies are not available for confirmation.

CR 48

Histiocytic sarcoma of brain: a case report

I. W. G. A. L. Malhasi *, M. M. A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

malhasig@gmail.com

Introduction: Histiocytic sarcoma is an aggressive tumour composed of cells resembling mature histiocytes, and the involvement of the central nervous system (CNS) is rare. We report a histiocytic sarcoma occurring in the left frontal lobe of an adult male.

Case report: A 58-year-old man presented with headache, episodes of altered level of consciousness and seizures for three months. The MRI of the brain revealed a high-grade glioma in the left frontal lobe. During surgery, the tumour was located in the frontal lobe with extension into the left ventricle. Macroscopy revealed a white, fleshy tumour. Histology revealed a cellular tumour composed of sheets of large, non-cohesive, moderately pleomorphic cells with abundant foamy cytoplasm, resembling histiocytes and bizarre, multinucleated cells with necrosis and haemorrhage. There was no microvascular proliferation. Immunohistochemically, the tumour cells were positive for CD68 and S-100. Vimentin and CD34 were positive in some cells. Tumour cells were negative for GFAP, EMA, AE1/AE3, synaptophysin, chromogranin and Melan-A. The Ki-67 proliferation index was 22%. Immunohistochemistry and morphology confirmed a histiocytic sarcoma. Patient died on the tenth postoperative day due to sepsis caused by pneumonia.

Discussion and conclusion: Histiocytic sarcoma can occur at any site within the CNS, including brain parenchyma, meninges and spinal cord, which mimic a high-grade glioma radiologically and on imprint smears. The histiocytic nature and appropriate immunophenotype assessment confirm the diagnosis. Histiocytic sarcoma can rarely metastasize and has a poor prognosis.

CR 49

An osteoblastoma occurring in the acetabulum: an uncommon tumour in an uncommon site

I.W.G.A.L. Malhasi*, M.M.A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

malhasig@gmail.com

Introduction: Osteoblastoma is an uncommon bone-forming tumour that usually arises in the spine, sacrum, femur and tibia. This is a case report of an osteoblastoma in the left acetabulum, a rare site with only a few reported cases in the literature.

Case report: A 25-year-old man presented with left hip pain for three months. CT scan revealed a lytic lesion in the left acetabulum suggestive of a giant cell tumour (GCT). The lesion was resected, and the intra-operative frozen section assessment revealed a giant cell-rich lesion, cytologically compatible with GCT. The lesion was received as multiple bony and soft tissue pieces, 75x50x20 mm in aggregate. Microscopy revealed woven bony trabeculae rimmed by osteoblasts in a vascularized stroma. Numerous osteoclast-like multinucleated giant cells (MNGCs), haemosiderin laden histiocytes and extravasated red blood cells were noted. There was no nuclear pleomorphism, permeative growth pattern or mitoses. The overall appearance was of an osteoblastoma. Re-evaluation of the imaging studies concluded that the features were more in favour of osteoblastoma than GCT.

Discussion and conclusion: The rarity of the site and overlapping radiological features led to a radiological diagnosis of GCT, which was favoured in the cytology due to the presence of MNGCs. Radiological correlation is essential in diagnosing bone lesions, however, histopathologists should not be carried away by the radiological impression alone. Many non-neoplastic and neoplastic bone lesions harbour MNGCs and their presence should be interpreted in an appropriate context. Although the prognosis is good, osteoblastoma has a propensity for local recurrence.

CR 50

Haemangioma of thyroid gland: a rare entity

I.W.A.G.L Malhasi *, M.M.A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

malhasig@gmail.com

Introduction: Haemangioma is a benign vascular neoplasm that rarely occurs in the thyroid gland. We present a case of haemangioma in a multinodular goitre.

Case report: A 61-year-old woman presented with a long-standing neck lump. Ultrasound scan revealed a multinodular goitre. Fine needle aspiration (FNA) was not performed as there was no evidence of suspicious nodules. The patient underwent thyroidectomy. Macroscopic examination showed an enlarged nodular thyroid. The cut surface revealed a well-defined, haemorrhagic, cystic lesion in upper pole of the right lobe measuring 16 mm. Microscopy revealed multiple ectatic vascular spaces lined by a single layer of endothelial cells. No nuclear atypia, mitoses or necrosis was seen. The adjacent tissue revealed haemorrhage. The lesion was well demarcated from the background thyroid which showed features of a colloid goitre. The histological diagnosis of a colloid goitre with a haemangioma in the right lobe was made. Re-evaluation of the imaging was not performed.

Discussion and conclusion: Haemangioma of the thyroid gland is mostly considered a reactive vascular proliferation, which may follow needle aspiration, trauma or changes during development of a nodular goitre. This can be diagnosed or suspected on ultrasonography and hence it is important to perform FNAC of thyroid lesions after imaging studies. If suspected on imaging, FNA is better avoided due to the risk of haemorrhage and since the yield is only blood rather than cellular material. Haemangioma should be included in the differential diagnosis when a lesion appears haemorrhagic. The treatment of choice is lobectomy.

CR 51

Invasive micropapillary carcinoma of breast: an aggressive carcinoma with high locoregional recurrence

I. W. G. A. L. Malhasi *, A. V. T. Damayanthi, P. G. N. L. Chandrasiri, N. R. Perera

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

malhasig@gmail.com

Introduction: Invasive micropapillary carcinoma is a rare, aggressive type of breast carcinoma co-exist with axillary lymph node metastases at presentation. Although areas of micropapillary morphology may occur in invasive breast carcinoma of no special type (IBC-NST), pure invasive micropapillary carcinomas are rare.

Case report: A 70-year-old woman presented with a palpable left breast lump. Mammography revealed a BIRADS-5 lesion with axillary lymph node involvement. Fine needle aspiration or tru-cut biopsy were not performed. She underwent a left-sided mastectomy with axillary clearance, which macroscopically revealed a hard, white tumour in the lower outer quadrant, measuring 35 mm in maximum dimension. Microscopy showed an invasive carcinoma composed of well-formed tumour morules of cuboidal cells lying in empty spaces within a spongy stroma. These morules had no fibrovascular cores. The epithelial clusters displayed a unique reverse polarity. The nuclear grade was intermediate. There were no IBC-NST areas or in-situ carcinoma components. Peritumoral lymphovascular space invasion was noted within the tumour. Four out of thirteen left axillary lymph nodes showed metastatic tumour deposits (4/13). The tumour cells were positive for ER and PR and negative for HER2. EMA was positive in the peripheral cell membrane. The Ki-67 index was 24%.

Discussion and conclusion: Patients with invasive micropapillary carcinomas have a higher rate of locoregional recurrence compared to IBC-NST. This is thought to be due to its aggressive nature and the high frequency of lymphovascular space invasion, which are characteristic of this tumour. Awareness of this entity and its behaviour are highlighted.

CR 52

Myeloid sarcoma presenting as a gastric mass: a case report

I.W.G.A.L. Malhasi*, D.R. Jayasinghe

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

malhasig@gmail.com

Introduction: Myeloid sarcoma (MS) is a tumour mass composed of myeloid blasts occurring at sites other than bone marrow with effacement of tissue architecture. We report a rare case of MS presenting as a gastric mass in a young man with acute myeloid leukaemia (AML).

Case Report: This is a 22-year-old man was on consolidation treatment when he presented with abdominal pain. Computed tomography revealed a mass in the lesser sac. Upper gastrointestinal endoscopy showed a large, infiltrating mass in the lesser curvature of the stomach extending from the gastro-oesophageal junction to the antrum. The mucosal biopsies revealed non-specialized gastric mucosa with intact foveolar architecture. The lamina propria was expanded with a diffuse infiltrate of monomorphic cells with a blastoid appearance, containing medium, round, hyperchromatic nuclei, and scanty cytoplasm. The glandular epithelium was histologically unremarkable. A differential diagnosis of MS and lymphoma was made. Immunohistochemically, the neoplastic cells showed strong and diffuse positivity for MPO, CD34 and CD117 confirming the diagnosis of MS. LCA, CD3 and CD20 were negative.

Discussion and conclusion: The possibility of MS should always be considered when a patient with AML presents with a tumour mass, although its incidence is only 2-9% of all AML cases. Approximately half of the cases are initially misdiagnosed, most often, as a lymphoma. Careful histomorphological evaluation combined with immunohistochemistry and clinicopathological correlation aid in reaching an early diagnosis. Initiating treatment is important as MS may be the initial manifestation of a relapse of AML.

CR 53

Mucinous adenocarcinoma arising from urachal remnants: a case report of a rare carcinoma

I. W. G. A. L. Malhasi*-[1], G. G Ranaweera[1], W. A. S. de Silva [2]

[1]*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[2] *Department of Urological Surgery, National Hospital of Sri Lanka*

malhasig@gmail.com

Introduction: Urachus is a fibrous remnant of the allantois, a canal draining the urinary bladder of the fetus during embryonic life. It undergoes involution after third trimester. We report a mucinous adenocarcinoma arising in urachal remnants, an extremely rare tumour accounting for <1% of all bladder malignancies.

Case report: A 59-year-old woman presented with a suprapubic mass and urinary symptoms for six months. Imaging revealed a large, solid necrotic growth involving the dome of bladder extending up to the umbilicus. The epicentre of the tumour was in the bladder. The uterus and ovaries were normal. Cystoscopy revealed a bladder mass. Colonoscopy was normal. A superficial biopsy contained multiple tan-brown fragments of tissue admixed with gelatinous material. Histology revealed fragmented bladder mucosa infiltrated by small nests, papillary structures and glands floating in pools of mucin. Constituent cells contained mildly pleomorphic, hyperchromatic nuclei and moderate, pale mucinous cytoplasm. Mitoses were present. Lymphovascular or perineural invasion was absent. The uninvolved urothelium was histologically unremarkable. No other primary malignancy was found elsewhere. With the given clinical, radiological and histomorphological features and according to the criteria proposed by Gopalan et al., the diagnosis of an invasive mucinous adenocarcinoma arising from urachal remnants was made.

Discussion and conclusion: Urachal adenocarcinoma has to be differentiated from adenocarcinoma rising in the adjacent organs such as the colon and ovary and distant sites such as the stomach and breast. Immunohistochemistry may not be helpful due to lack of specificity and overlapping positivity among the entities. Clinical, radiological and histological correlation plays a crucial role in the diagnosis.

CR 54

Fungal endocarditis and fungal pneumonia causing an infant death: a perinatal post-mortem evaluation

I. W. G. A. L. Malhasi*, A. A. H. Priyani

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

malhasig@gmail.com

Introduction: Disseminated fungal infections (DFIs) are associated with high mortality among patients admitted to intensive care units (ICU). Neonates are highly vulnerable due to extreme prematurity and immunosuppression. Hence awareness and a high index of suspicion are required for early detection. This premature infant who died of respiratory distress while under intensive medical care was found to have DFI at the post-mortem.

Case report: A mother with dichorionic-diamniotic twin pregnancy underwent an emergency Caesarean section due to absent diastolic flow at 28 weeks of gestation. Both babies were admitted to the neonatal ICU due to prematurity. Twin 2 developed grunting and was treated with intravenous antibiotics and oxygen and managed as acute respiratory distress. Despite treatment, the baby expired in 35 days. A small, yellow-brown, friable mass attached to the luminal side of the left atrial wall was noted at the post-mortem. Microscopically, it was an infective vegetation comprising fibrinous material encasing inflammatory cells. Non-septate fungal hyphae, positive for Periodic Acid Schiff and Grocott methenamine silver stains were present within giant cells. Sections from the right lung showed granulomas composed of epithelioid histiocytes, lymphocytes and Langhan type giant cells. The giant cells contained fungal hyphae similar to those in the heart, and a diagnosis of DFI with endocarditis and pneumonia was made.

Discussion and Conclusion: Clinical diagnosis of DFIs is difficult. Patients are very ill and require aggressive treatment with intravenous antifungals. If detected early, adequate treatment could prevent death. Surgical management with the removal of infected nidus has been undertaken to supplement antifungal therapy in almost all survivors reported in the literature.

CR 55

Osteosarcoma of infratemporal fossa with chondromyxoid fibroma-like areas: a potential pitfall

I.W.G.A.L. Malhasi*[1], R.G.S. Rassagala [2], M.M.A Jayawickrama[2], O.R. Gunasinghe[3], M.V.C. de Silva[1].

[1]*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[,2] *Department of Histopathology, National Hospital of Sri Lanka*

[3]*Department of Histopathology, Apeksha Hospital, Maharagama.*

malhasig@gmail.com

Introduction: De novo-primary osteosarcoma of the jaw and cranial vault is rare. We report an osteosarcoma arising in base of the skull of a young man showing unusual chondromyxoid fibroma (CMF)-like areas.

Case Report: A 27-year-old man presented with left side facial pain and numbness. Imaging revealed a tumour in the left infratemporal fossa eroding pterygoid plates and the left maxillary sinus with intratumoral bone formation. Histology revealed a tumour with an abundant myxoid stroma rich in osteoid and woven bone showing a permeative growth pattern. No osteoblast rimming was seen. Stromal cellularity was variable, ranging from areas with low cellularity to areas with cellular crowding. Neoplastic cells comprised spindle-shaped cells, stellate cells, small round cells and scattered larger, pleomorphic cells arranged haphazardly. Mitoses, including atypical forms, were present. No chondroid areas or necrosis were seen. Tumour cells were positive for CD99 and S100 focally. The Ki-67 proliferation index was 50%. Tumour cells stained negatively with EMA, BCL2, AE1/AE3, GFAP, CD34, desmin and MyoD10. Based on above findings, a diagnosis of a conventional osteosarcoma with CMF-like myxoid areas was made.

Discussion and Conclusion: Osteosarcomas may show benign-looking areas, which can potentially lead to diagnostic pitfalls. Features of CMF including a lobular pattern with hypocellular and hypercellular areas comprising stellate and spindle cells in a myxoid background were noted in this tumour. However, the presence of osteoid with a permeative growth pattern, atypical mitoses, infiltrative nature on imaging and the high Ki-67 index favoured a malignant tumour over a benign entity. The overall impression considering clinico-radiological and immuno-histomorphological features led to the correct diagnosis of osteosarcoma over CMF.

CR 56

Hobnail variant of papillary thyroid carcinoma: a rare aggressive variant

I.W.G.A.L. Malhasi*, D.R. Jayasinghe, M.V.C. de Silva

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

malhasig@gmail.com

Introduction: Papillary thyroid carcinoma (PTC) is the commonest thyroid malignancy which has many histological variants. The hobnail variant of papillary thyroid carcinoma (HVPTC) is a rare aggressive variant, which is diagnosed when >30% of the tumour cells show hobnail features.

Case Report: A 16-year-old girl presented with a goitre for five months with recent rapid enlargement. Ultrasound scan showed a TIRADS IV nodule in the right lobe with features suspicious of cervical lymph node involvement. Fine needle aspiration revealed cells from a papillary thyroid carcinoma with eosinophilic cytoplasm. The total thyroidectomy with cervical lymph node dissection showed an encapsulated, tan-white papillary lesion with focal solid areas in the right lobe of the thyroid measuring 28 mm in maximum dimension. Histology confirmed the lesion as a HVPTC with >80% of the tumour having papillary and micropapillary structures lined by cells showing loss of polarity and cohesion. Tumour cells had large, irregular apical nuclei with thick membranes and prominent nucleoli, and eosinophilic cytoplasm. Typical PTC features, nuclear crowding, overlapping, grooves and intranuclear pseudo-inclusions, were not prominent. The solid areas showed Hurthle cell change with more typical PTC nuclear features. Capsular and lymphovascular invasion and microscopic extrathyroidal extension were present. One lymph node contained deposits from the HVPTC.

Discussion and conclusion: HVPTC frequently exhibits aggressive features including micropapillae, extrathyroidal extension, nodal/distant metastases and a likelihood of poor response to radioiodine treatment. These factors contribute to poor survival. Identification of this rare variant of PTC is important for therapeutic decision-making, close clinical follow-up, and monitoring of patients to detect recurrence.

CR 57

A primitive neuroectodermal tumour of the abdominal wall: a rare presentation with a different clinical impression

J. D. S. Manoharee*, T. P. M. Bopagoda

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

jdsmanoharee@gmail.com

Introduction: Primitive neuroectodermal tumours (PNETs) are malignant small round cell tumours of neuroectodermal origin. They belong to the Ewing sarcoma family and occur in children and young adults. Peripheral PNET is rare, and only a few cases involving the soft tissues of the abdominal wall have been reported. We present a case of peripheral PNET involving the anterior abdominal wall of a middle-aged woman

Case report: A 45-year-old woman presented with a solitary non-tender lump in the right anterior abdominal wall for eight months. This lesion was 2 mm in diameter and was clinically suspected as a lipoma, therefore radiological assessment was not carried out. The excised specimen was composed of multiple pale tan pieces of tissue measuring 30x25x10 mm in aggregate. The histology revealed an infiltrative tumour composed of small, monotonous cells with round, hyperchromatic nuclei and vacuolated scant cytoplasm. Scattered mitoses were noted. No tumour necrosis was seen. Tumour cells were diffusely PAS-positive. Immunohistochemistry showed strong, diffuse CD99 positivity, focal BCL2 positivity and negativity for LCA, EMA, AE1/AE3 and CD56. The presence of EWSR1 gene rearrangement was confirmed by dual-probe break-apart fluorescent in-situ hybridization (FISH), and the diagnosis of Ewing/ PNET sarcoma was confirmed. Postsurgical CT scan of neck, chest and abdomen showed no residual tumour or evidence of metastasis. The patient was referred to the oncologist for further management.

Discussion: This case demonstrates an unusual and rare presentation of a PNET involving the anterior abdominal wall of an adult leading to a diagnostic dilemma.

CR 58

Autoimmune encephalitis associated with an ovarian teratoma: a rare clinical presentation

J. D. S. Manoharee*, T. P. M. Bopagoda

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

jdsmanoharee@gmail.com

Introduction: Anti-*N*-methyl-D-aspartate (NMDA) receptor encephalitis is a form of autoimmune encephalitis, in which the NMDA receptors located in the forebrain and hippocampus region are targeted by antibodies resulting in a progressive decline of NMDA receptor-associated synaptic functions and neurological and psychiatric manifestations.

Case report: A 21-year-old previously healthy young woman presented with acute onset generalized tonic-clonic seizures and deteriorated level of consciousness. Cranial MRI was suspicious of encephalitis with no evidence of infarction or haemorrhage. CSF cytology revealed lymphocytosis with elevated protein levels. The patient was managed initially as post-infectious encephalitis. As her condition deteriorated despite treatment and viral studies were negative, she was investigated with anti-NMDA-receptor antibodies, which were positive. CECT pelvis detected a left-sided ovarian cyst. Laparoscopic cystectomy was performed nearly one month after the onset of symptoms. Macroscopically the cyst measured 25 mm in maximum dimension. It was unilocular and filled with hair and sebaceous material. Histology confirmed the diagnosis of a mature cystic teratoma with a prominent glial tissue component. The patient showed no improvement following the cystectomy, immunoparesis and immunomodulating drugs. Repeat MRI brain showed bilateral hippocampal atrophy and damaged lentiform and caudate nuclei, suggesting sequelae of autoimmune encephalitis.

Discussion: Anti-NMDA-receptor encephalitis should be considered in young women presenting with primary psychiatric disorders and infective encephalitis. Early detection by performing anti-NMDA-receptor antibodies, abdominal imaging and treatment with pulse immunoglobulins, immune modulators and tumour resection will give a better chance of recovery. Due to the rarity and lack of awareness of this disease like in our case, some patients have poor-outcome with permanent morbidity.

CR 59

Laryngeal adenoid cystic carcinoma presenting as a solitary thyroid nodule

R.D.K. Medonsa*[1], M.M.A. Jayawickrama [2]

[1]*Department of Pathology, Faculty of Medicine and Allied Sciences, University of Rajarata, Sri Lanka*

[2]*Department of Histoathology, National Hospital of Sri Lanka*

dharanikdw@gmail.com

Introduction: Laryngeal adenoid cystic carcinoma (ACC) is very rare and accounts for <1% of all laryngeal tumours, with only 120 reported cases. Laryngeal ACC invading the thyroid gland and presenting as a solitary nodule has not been previously reported in Sri Lanka.

Case report: A 41-year-old man presented with dyspnoea, hoarseness of voice and enlargement of thyroid for five months. Computerized tomography revealed two separate tumour nodules in the trachea and the right lobe of the thyroid. Thyroid FNAC was concluded as atypia of unknown significance (Bethesda III/Thy 3). Subsequent total thyroidectomy and laryngeal mass excision revealed a solid laryngeal tumour measuring 3.5 cm in maximum diameter and a separate right thyroid nodule measuring 2.5 cm in diameter. Both tumours were diagnosed as grade 2 ACC on haematoxylin and eosin-stained sections. The tumours showed typical microscopic features of an ACC; coexisting bi-layered ducts, cribriform structures and pseudocysts with abundant basophilic mucin and inner low-grade basaloid epithelial cells with low proliferation activity resting on myoepithelial cells. Perineural invasion was evident.

Discussion and conclusion: As the histological features were characteristic of ACC, no additional investigations were performed. Laryngeal ACC metastasis into the thyroid gland is very rare and easily overlooked in the FNAC especially if typical cytological features are absent. Primary ACC in the thyroid gland is exceptional. However, in unusually aggressive tumours with an accompanying history of tumour elsewhere or atypical cytopathological features, possibility of nonthyroidal neoplasms involving the thyroid should be considered due to different prognostic and therapeutic implications.

CR 60

A case report of urachal adenocarcinoma: a rare entity

M.A.D.N. Munasinghe*, P.G.A.N. Jayathilake, S.M. Fernadopulle

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

nayanganee@gmail.com

Introduction: The urachus is the embryonic remnant originating from the involution of the allantois and cloaca and extends from the umbilicus to the bladder dome. Primary urachal adenocarcinoma (UC) is a rare entity arising from urachal remnants.

Case report: A 67-year-old man presented with haematuria for six months. CT scan showed a contrast- enhancing nodular area in the soft tissue of the anterior bladder wall and an adjacent urachal cyst. The surgically removed specimen was comprised of the urachus, umbilicus and the anterior bladder wall with an irregular whitish tumour of 32 mm in maximum dimension. The cyst adjacent to the tumour was 5 mm in diameter. Histology of the tumour revealed an adenocarcinoma-NOS infiltrating the bladder wall. No evidence of cystitis cystica/cystitis glandularis or urothelial dysplasia was noted. The separate cyst was lined by dysplastic columnar epithelium. The tumour cells were positive for immunohistochemical stains CK20 and CK7. A primary tumour elsewhere was excluded clinically and radiologically.

Discussion and conclusion: Primary UC has to be differentiated from adenocarcinoma arising from other organs such as colon, urinary bladder and urothelial carcinoma with glandular differentiation. According to WHO tumour classification (2016), the criteria for pathological diagnosis of UC includes tumour localized in the dome/anterior wall of the bladder, the epicentre of the tumour in the bladder wall, absence of florid cystitis cystica/cystitis glandularis or absence of urothelial dysplasia, and absence of known primary elsewhere. The presence of the urachal remnant in association with the tumour is supportive of the diagnosis. Immunohistochemical stains aid in differentiating UC from adenocarcinoma extending from organs in the vicinity.

CR 61

High-grade myxofibrosarcoma presenting as a slow growing small subcutaneous lump

A.R. Pathiravasam*, T.G. Liyanage.

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

arundikapathiravasam@gmail.com

Introduction: Myxofibrosarcoma is a relatively rare subtype of soft tissue sarcoma that is commonly seen in elderly patients. High-grade myxofibrosarcoma is rare and usually presents as a large tumour.

Case report: A 52-year-old man presented with a slow-growing subcutaneous lump in the right arm over a one-year duration. Macroscopically it was a circumscribed nodule measuring 25x24 x10 mm with a whitish and yellowish cut surface. Histopathology showed a multinodular lesion with variable cellularity. Hypocellular nodules showed scattered spindle and stellate cells with mild atypia within a myxoid stroma. Some nodules were cellular with solid and less myxoid areas. These nodules showed cells with hyperchromatic pleomorphic nuclei, scattered large cells with bizarre nuclei and multinucleated giant cells. Scattered mitoses with many atypical forms were seen. Thin-walled curvilinear vessels with perivascular condensation of tumour cells were prominent. The tumour infiltrated the adjacent adipose tissue. The tumour cells were strongly positive for vimentin and negative for S-100. Therefore, a neural tumour was excluded, and the diagnosis of high-grade myxofibrosarcoma was made.

Discussion: Multinodular lesion with infiltrative margins, myxoid stroma, curvilinear vessels together with high-grade features; cellular solid areas, pronounced cellular pleomorphism with bizarre and multinucleated giant cells and numerous atypical mitoses justified the diagnosis of high-grade myxofibrosarcoma. The specificity of immunohistochemistry in myxofibrosarcoma is low and usually demonstrates vimentin positivity.

Conclusion: High-grade myxofibrosarcoma is a rare entity with poor outcomes. Inadequate sampling may miss high-grade areas. Recognizing the histopathological features and adequate sampling are crucial in establishing the correct diagnosis.

CR 62

Secondary haemophagocytic lymphohistiocytosis: a preventable fatality

H. K.U. Piyamalika, P. Thiranagama, H.P. Maddumage, C.S.P Sosai*

Department of Pathology, Colombo South Teaching Hospital, Kalubowila, Sri Lanka

cherinesosai@yhoo.com

Introduction: Haemophagocytic lymphohistiocytosis (HLH) is a rare, multifaceted syndrome induced by the aberrant activation of macrophages and cytotoxic T cells. Secondary HLH due to infections, autoimmune disease (AD) or malignancy results in hypercytokinaemia, causing irreversible organ damage that is rapidly progressive and frequently fatal if delayed or left undiagnosed.

Case report: A 21-year-old woman presented with a three-week history of intermittent fever, fatigue, arthralgia and myalgia. Examination revealed a body temperature of 104^0 F and bilateral cervical lymphadenopathy. Intra parotid lymph nodes, bilateral pleural effusion and mild hepatomegaly were present on USS. FNAC and biopsy of the cervical lymph node were performed. Further investigations revealed CRP-99 mg/dL, peripheral bicytopenia, serum ferritin 10,000 ug/L, bone marrow aspirate with HLH, negative dengue and retroviral screen, Hepatitis B Antigen, CMV and EBV IgG positivity, negative serum fibrinogen and absent organisms in cultures of urine and blood. Patient deteriorated rapidly and expired while in intensive care pending reports of FNAC, which showed HLH and lymph node biopsy which was involved by ALK-positive anaplastic large cell lymphoma. The post-mortem revealed generalised lymphadenopathy, cerebral oedema, bilateral pneumonia, pulmonary oedema and a nutmeg liver. Microscopy showed alveolar lumina, liver sinusoids, splenic sinuses and bone marrow with extensive HLH and atypical lymphoid cells in the interstitium. All lymph nodes showed HLH and atypical lymphoid cells.

Discussion: In view of lymph node HLH, Rosai-Dorfman disease and Langerhan cell histiocytosis were excluded by the lack of large histiocytes and lack of grooved nuclei and eosinophils in the latter. Causes of secondary HLH was elucidated by exclusion of infections including HIV, AD and other neoplasms by clinicopathological correlation and the presence of ALK-positive neoplastic lymphoid cells.

CR 63

Melioidosis complicating an ovarian teratoma: a case report

U. V. V. Ranathunga*[1], U. N. Wijenayake[2], L. J. De Silva[1], P. G. N. L. Chandrasiri[1], A. Kaluarachchi[2,] M. V. C. De Silva[1]

[1]*Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka*

[2]*University Gynaecology and Obstetrics Unit, National Hospital of Sri Lanka, Sri Lanka*

uvvranathunga@gmail.com

Introduction: Melioidosis is an endemic infection in South-East Asia, caused by a soil associated saprophyte bacterium *Burkholderia pseudomallei.*

Case report: A 38-year-old woman who had a recent travel history to India and North Central province presented with low-grade fever, abdominal pain, and weight loss for one month. She had outdoor activities and bathing in tanks during her travels. Initial investigations revealed high CRP levels and neutrophil leukocytosis, and the blood and urine cultures were negative. Melioidosis antibodies were positive at a titre of 1:1280. The extensive radiological evaluation did not reveal any infective focus, except for a solid and cystic lesion in the left adnexa, suggestive of a teratoma. She was treated with intravenous meropenem for four weeks. In the absence of any improvement, the teratoma was suspected harboring melioidosis infection. A left side oophorectomy was performed, which macroscopically showed solid, firm, gritty cut surface with areas of cystic degeneration. Microscopy revealed an immature teratoma (grade-II) with evidence of gliomatosis peritonei. There were areas of suppurative inflammation within the teratoma and within the glial elements in the peritoneum. Gram negative bacilli were not identified. Stains for fungi and mycobacteria were negative. The patient improved following oophorectomy, and the melioidosis serology became negative.

Discussion and conclusion: The probability of melioidosis infection within teratoma was confirmed by histological findings of suppurative inflammation and marked postoperative clinical improvement. Melioidosis is known to cause suppurative inflammation in deep organs, but gynaecological manifestations are limited to a few reported cases, presenting as tubo-ovarian abscess, pelvic inflammatory disease and cervicitis.

CR 64

A mixed germ cell tumour with non-gestational choriocarcinoma, mature teratoma and an adenocarcinoma arising from the teratomatous component

G.G. Ranaweera*[1], S Thanansayan [1], M.V.C de Silva[1], K Karunanayake [2]

[1]*Department of Pathology,* [2]*Department of Obstetrics and Gynaecology, Faculty of Medicine, University of Colombo, Sri Lanka*

gayani@path.cmb.ac.lk

Introduction: Non-gestational choriocarcinoma (NGC) accounts for <1% of ovarian tumours. There are only eleven reported cases of somatic adenocarcinoma arising from a teratoma. Simultaneous occurrence of both is extremely rare.

Case report: A 12-year-old girl presented with abnormal vaginal bleeding and abdominal distension for 3 months, and the ultrasound scan showed a left ovarian mass. Serum AFP, beta-hCG, and LDH levels were raised, being 35.44 ng/mL, 42,446.47 mIU/ml and 537 IU/l, respectively. The resected ovarian tumour measured 170x150x110 mm and had yellowish friable areas, spongy areas and haemorrhages. Approximately 30% of the tumour comprised sheets of malignant cytotrophoblastic and syncitiotrophoblastic cells arranged in a plexiform pattern with brisk mitotic activity and large areas of haemorrhage and necrosis. The syncitiotrophoblastic component showed positivity for beta-hCG. Other areas comprised a mature teratoma (MT) showing a benign vascular proliferation (50%), mature skin and intestinal glandular tissue (10%). Glandular epithelium showed focal high-grade dysplasia. One focus showed infiltrative angulated glands surrounded by a desmoplastic stroma. The invasive focus occupied <one low power field. Immature elements were absent. The diagnosis was a mixed germ cell tumour comprising NGC, MT showing a benign vascular proliferation and malignant transformation of the intestinal glandular component.

Discussion and conclusion: Both NGC and adenocarcinoma components pose a risk for recurrence and metastatic disease. NGC responds less well to chemotherapy compared to gestational choriocarcinoma. In the present case, the adenocarcinomatous component was negligible and thus the focus of chemotherapy was targeted towards NGC. Meticulous sampling of germ cell tumours is necessary to detect small areas of divergent differentiation.

CR 65

Pleomorphic rhabdomyosarcoma presenting as a psoas abscess

R.G.S. Rassagala*, S.R.Constantine

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

gayathrirassagalas@gmail.com

Introduction: Ninety per cent of rhabdomyosarcomas occur in children and young adults. Pleomorphic rhabdomyosarcoma is a rare high-grade variant, which is most common in the sixth to seventh decade of life, occurring more frequently in men than women. It typically arises in the deep soft tissue of the extremities. This is a rare case of pleomorphic rhabdomyosarcoma in the psoas muscle which radiologically appeared as an abscess.

Case report: A 45-year-old man presented with a lump in the left psoas region for two years and left leg pain for one month. The MRI revealed an "intramuscular collection" in the left psoas muscle suspicious of an abscess. Subsequently, tissue drainage and a biopsy were performed. Histologically the biopsy revealed sheets and clusters of highly atypical cells, exhibiting markedly pleomorphic, hyperchromatic nuclei, scanty eosinophilic cytoplasm and frequent mitoses, compatible with a poorly differentiated malignancy. The initial panel of immunohistochemical stains showed negative staining with pancytokeratin, HMB45, CD3, CD20, S-100, CD31, and positive staining with vimentin, which excluded the possibility of carcinoma, melanoma and lymphoma. In the second panel, the tumour cells showed strong cytoplasmic positivity for desmin and strong nuclear positivity for MyoD1, which lead to the diagnosis of a pleomorphic rhabdomyosarcoma.

Discussion and conclusion: The age and clinical presentation of the patient were unusual, as pleomorphic rhabdomyosarcoma is usually a rapidly growing tumour in elderly patients. The use of a panel of immunohistochemical stains is mandatory to exclude the other possibilities. This is a highly aggressive sarcoma with a high mortality rate.

CR 66

Primary high-grade B-cell lymphoma in the urinary bladder: an uncommon bladder tumour

R.G.S. Rassagala*, M. M.A. Jayawickrama

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

gayathrirassagalas@gmail.com

Introduction: Lymphoma of the urinary bladder is a rare tumour accounting for 0.2% of primary and 1.8% of secondary tumours of the bladder. Most primary bladder lymphomas are of low-grade mucos-associated lymphoid tissue type lymphoma. High-grade B-cell lymphoma is uncommon, and the clinical, radiological and cystoscopic features are non-specific. The diagnosis exclusively depends on histology.

Case report: A 79-year-old man presented with lower abdominal pain and frequency of urination. On examination, there was a palpable pelvic mass. The computerized tomography revealed a large growth infiltrating into the bladder wall, metastatic deposits in the liver and lymph node enlargement. Transurethral resection of the tumour revealed sheets of monomorphic, medium-sized atypical cells containing hyperchromatic nuclei and pale cytoplasm. Mitoses were frequent. Conventional urothelial differentiation was not seen. Tumour cells were positive for LCA, CD20 was strongly positive and CD3 showed a few positive cells. A very high Ki-67 proliferation index of 90% confirmed a high-grade B-cell lymphoma. Pancytokeratin, synaptophysin and chromogranin were negative.

Discussion and Conclusion: The differential diagnoses considered were lymphoma and small cell neuroendocrine carcinoma. Patients with a high-grade B-cell lymphoma of the bladder should be considered to have systemic disease, which requires chemotherapy. Surgery can be beneficial for the alleviation of urinary symptoms. The prognosis is usually favourable and depends on the tumour stage and complications.

CR 67

Maduramycosis: a neglected chronic infection of foot

K. Samalai*, V. Kemanilamiya, M. M. A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Maduramycosis is an uncommon chronic and debilitating infection of tropical and subtropical countries, affecting deep dermis, subcutis and soft tissue. Early detection and aggressive treatment are the mainstay for successful eradication.

Case report: A 60-year-old farmer from Anuradhapura presented with painless, gradual swelling of foot for 20 years. He was medically attended from time-to-time but was non-compliant. Examination revealed a non-tender swelling of the foot with multiple discharging sinuses. CRP was raised to 168 mg/L. A nodular soft tissue mass affecting the underlying bone was identified on imaging. Repeated punch biopsies were negative, and a deeper soft tissue biopsy was performed. Fragments of fibrous tissue containing multiple granulomata with palisading histiocytes surrounding compact colonies of brown-pigmented fungal hyphae were identified microscopically and confirmed with Grocott and PAS stains. Suppuration and necrosis were present. A diagnosis of Maduramycosis with Eumycetoma was made. Debulking surgery was carried out.

Discussion: Maduramycosis is an infrequent deep-seated infection involving the foot and lower legs. It often presents as a chronic disease due to slow progression, misdiagnosis, and non-compliance. Actinomycetes species (filamentous bacteria) and Eumycetoma (fungi) are the causative organisms, with Actinomycetes being two-fold commoner than Eumycetoma. Eumycetoma are commonly associated with soil and therefore agricultural occupation is a risk factor. Histologically the presence of suppurative granulomata with colonies of typical infective organisms is the hallmark. Distinction between the two aetiologies is important as the treatment differs. Extensive eumycetic lesions necessitates surgical intervention followed by triple therapy with itraconazole, terbinafine and potassium iodide. Prolonged follow-up is needed as recurrence is common even after adequate medical and surgical treatment.

Conclusion: Prompt and proper pathological diagnosis of Maduramycosis is essential for early detection, aggressive control and active surveillance.

CR 68

Epithelioid sarcoma mimicking a granulomatous lesion: a case report

K. Samalai*, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Epithelioid sarcoma (ES) is rare, representing <1% of soft-tissue sarcomas. ES has two clinicopathological subtypes, the classic/distal type (ES-DT) most commonly involves the distal upper extremity, and the proximal/large cell type affects the deep soft tissue. The ES-DT is the commoner, and frequently occurs in young men.

Case report: A 19-year-old man presented with multiple pigmented and ulcerated cutaneous nodules in the distal forearm for two years. These lesions showed sporothrichoid spread, and there was ipsilateral axillary lymphadenopathy. The clinical differential diagnosis was cutaneous granulomas. The punch biopsy revealed a nodular infiltrative tumour in the mid-dermis composed of epithelioid cells with central geographic areas of degeneration. The epithelioid cells contained mildly pleomorphic, ovoid vesicular nuclei having prominent nucleoli, and deeply eosinophilic cytoplasm. The periphery of the lesion showed spindling of cells. Mitoses were infrequent. The lesion is surrounded by a lymphocytic infiltrate. Special stains, PAS, Grocott, and Ziehl-Neelsen stain, were negative for organisms. Immunohistochemical stains AE1/AE3 and CK19 showed strong diffuse cytoplasmic positivity and CD34 showed membranous staining in the majority of the cells. A diagnosis of ES-DT was made.

Discussion and conclusion: ES are slowly progressive tumours, which involve both cutaneous/deep tissue. Histologically, DT-ES exhibits a pseudo-granulomatous growth pattern. The mildly atypical epithelioid cells may mimic activated histiocytes. Central degeneration and peripherally infiltrating lymphocytes may mimic necrobiotic granuloma. Therefore, careful histological assessment is vital for accurate diagnosis. ES shows immunoreactivity for epithelial markers, and >50% of cases show CD34 staining. The majority are associated with loss of nuclear expression of SMARCB1(INI1). Though sarcomas usually show a haematogenous spread, ES regularly metastasize to lymph nodes.

CR 69

Asymptomatic silicosis in a road repair worker

K. Samalai*, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Silicosis is an occupational pneumoconiosis caused by inhalation of the crystalline form of silicon dioxide. Histopathological evaluation is essential to rule out progressive fibrosis and tuberculosis.

Case report: A 62-year-old man, who was a road repair worker, was incidentally found to have lung opacities, which were of 10 mm, in the right upper lobe with hilar lymphadenopathy while undergoing pre-operative imaging for coronary artery bypass graft. A lung biopsy and a lymph node were obtained during the thoracotomy procedure. The lymph node showed nodules of varying stages of maturity, comprising entrapped histiocytes and fibroblasts within concentrically arranged hyaline deposits with foci of fibrinoid necrosis. Dust-like vaguely polarizable particles were present within the nodules. Granulomata and caseation were not seen. The lung biopsy showed similar but less well-formed nodules and collections of histiocytes expanding the interstitium. A histopathological diagnosis of silicosis was made.

Discussion: High-risk occupations for silicosis include coal mining, sandblasting and glass/concrete manufacturing. Inhaled dust deposits in terminal bronchioles and alveoli and activates pulmonary macrophages which release cytokines, resulting in tissue damage and recruitment of fibroblasts which ultimately cause interstitial fibrosis. They also interfere with the ability of macrophages to inhibit the growth of mycobacteria, explaining the common association of silicosis and tuberculosis. Symptoms are vague and radiology is indefinite. Accumulation of macrophages and fibroblasts forming concentric sclero-hyaline nodules and fibrinoid necrosis are the typical microscopic findings. Weakly polarizable tiny particles are expected under darkfield microscopy. Removing the source of exposure is the mainstay of treatment.

Conclusion: As the clinical and radiological findings are non-specific, histological assessment becomes the cornerstone for the diagnosis of silicosis and ruling out complications.

CR 70

Sinonasal glomangiopericytoma: a rare tumour of the nasal cavity

K. Samalai*, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Sinonasal glomangiopericytoma (SNG) is a rare low-grade malignant tumour with a perivascular myoid phenotype. It differs from the solitary fibrous tumour (SFT) and other pericytic tumours (PTs) in its location, biologic behaviour and immunomorphological features.

Case report: A 57-year-old man presented with epistaxis and was found to have a unilateral painless lesion in the middle meatus. CT scan showed a 2 cm lesion involving the maxillary sinus and extending into the nasal cavity. Polypoid reddish-grey tissue was received, and histology revealed an unencapsulated tumour beneath the respiratory epithelium composed of cells in sheets, storiform arrangement and short palisades with intervening gaping vessels. The cells contained regular ovoid nuclei with eosinophilic cytoplasm and indistinct cell borders. Mitoses were rare. Thick, acellular hyalinised material was noted around vasculature. The cells were immunoreactive to SMA but lacked CD34 staining.

Discussion: SNG accounts for <0.5% of all sinonasal neoplasms and presents in the seventh decade. They involve paranasal sinuses and nasal cavity, present with epistaxis or nasal obstruction, and <5% are bilateral. They lie beneath the mucosa and show varying architecture and comprise uniform cells with a syncytial arrangement. Haemangiopericytomatous vessels with peritheliomatous hyalinization is characteristic. The cells are positive for SMA, n-beta-catenin and cyclin-D1 and lack the expression of CD34, STAT6 and BCL2. Heterozygous mutations of CTNNB1 have been identified. SNG differs from other PTs as they involve distal extremities, have a benign course, and show different fusion genes. SFT can occur at any site and has a spectrum from benign to malignant. NAB2-STAT6 fusion is pathognomonic for SFT.

CR 71

Thrombotic microangiopathy following kidney transplantation: A case report

K. Samalai*, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Thrombotic microangiopathy (TMA) is a debilitating complication in the post-kidney transplant (post-KT) stage, associated with poor patient and graft outcomes.

Case report: A 29-year-old woman with diabetes mellitus presented with poorly resolving renal function since day six of post-KT. Following renal biopsy, the platelet count dropped drastically. The serum level of tacrolimus was within the normal range. The renal biopsy revealed unequally affected glomeruli with mesangiolysis, double contouring of glomerular basement membranes and fibrin thrombi in glomerular capillaries. Two glomeruli showed segmental capillary collapse, and one contained a cellular crescent. Mild peritubular capillaritis was noted. Endothelial swelling was evident in small arteries. A clinicopathological diagnosis of TMA with a possible antibody-mediated rejection (AMR) was made.

Discussion: Post-KT TMA shows an incidence of 5.6 per 1000 transplant recipients and arises de-novo or due to recurrence, which frequently occurs within the initial 3-6 months. De-novo cases are the commonest, mainly due to AMR and immunosuppressive drugs, which are believed to cause alteration in the complement pathway. Other causes include viral infections, other medications, genetic abnormalities in the complement cascade, etc. The management is targeted at the underlying cause.

TMA causes glomerular and vascular changes. The glomeruli show basement membrane thickening, fibrinoid necrosis and collapse of capillaries with mesangiolysis. Vessels show endothelial swelling, luminal thrombi and fibrinoid necrosis of media. Sclerosis of mesangiolysed foci with myo-intimal proliferation and duplication of the internal elastic lamina are the chronic changes of TMA.

Conclusion: Pathological identification of the route cause for TMA in post-KT renal biopsy is crucial as the treatment is directed towards the aetiology rather than TMA itself.

CR 72

Brown tumour of maxilla mimicking a giant cell tumour of bone

K. Samalai*, M. M. A. Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

bparththipan@gmail.com

Introduction: Brown tumour (BT) is rare but a known sequela of hyperparathyroidism (HPT). The maxilla is an uncommon location for BT, and radiologically and histologically masquerades as a giant cell tumour of bone (GCT).

Case report: A previously healthy 43-year-old woman presented with an intraoral lesion. The X-ray showed a lytic lesion affecting the maxilla suggestive of a GCT. Initial histological assessment was compatible with a GCT. A month later, she presented with acute pancreatitis and was found to have high serum calcium and PTH levels. Four-dimensional computed tomography of the parathyroid revealed a left side parathyroid adenoma, multifocal lytic and expansile bone lesions and bilateral medullary nephrocalcinosis. Upon request, the previous histological slides were reviewed. Fragments of tissue revealed numerous osteoclast-like multinucleated giant cells in a background of an intensely vascular fibroblastic stroma. Haemosiderin deposits were present. The lesion was surrounded by a rim of reactive bone resembling fibrous dysplasia (FD). It was compatible with a BT in the appropriate clinical setting.

Discussion and conclusion: BTs occur in about 25% of patients with HPT. BT commonly affects the ribs, clavicle and pelvis. Although multifocal bone lesions were identified through subsequent investigations, the patient initially presented with a maxillary lesion, which is an uncommon site for both BT and GCT. Unevenly dispersed osteoclasts, areas of fibroblastic proliferation devoid of osteoclasts, stromal haemorrhage and peripheral rimming of FD-like reactive bone are subtle features that differentiate BT from GCT. In the absence of a clinical diagnosis of HPT, a pathological diagnosis of BT is rarely made as it mimics GCT both histologically and radiologically.

CR 73

A case report of carcinoma of unknown primary: Limitations of immunohistochemistry in subtyping of carcinomas

C.L. Samaranayake*, P. Kariyawasam

Department of Histopathology, Teaching Hospital Karapitiya, Sri Lanka

charika.lakmalie@gmail.com

Introduction: Carcinoma of unknown primary (CUP) accounts for 3% of malignancies. Identifying the primary site and subtyping of carcinoma is challenging and is essential for treatment.

Case report: A 63-year-old woman presented with back pain for six months. CT scan revealed bilateral ovarian masses, omental deposits, sigmoid colon involvement, abdominal lymphadenopathy and multiple lung and bone lesions, including the spine. The radiological diagnosis was a malignant ovarian tumour with metastasis. Serum CA 125 level was elevated (13705 U/ml). The patient underwent a total abdominal hysterectomy and right salpingo-oophorectomy with omentectomy. Macroscopy revealed a left ovarian mass measuring 5x4x3.5 cm, a right ovarian mass measuring 3.5x2x2 cm and omental deposits. The uterus and the right fallopian tube were normal. The left fallopian tube was not present. Microscopy revealed an adenocarcinoma involving both ovaries, with involvement of the capsule, and the omentum, composed of infiltrative glands lined by atypical cells having enlarged pleomorphic vesicular nuclei and eosinophilic cytoplasm, some having cytoplasmic vacuoles. Mitoses were 4/10 high power fields. Tumour necrosis was not present. The immunohistochemical stains revealed diffuse strong positivity with CK7, CK19, TTF1 and Napsin A, and negative staining with CK20, ER and WT1. Radiological findings, histomorphology and immunohistochemical findings favoured clear cell ovarian carcinoma-eosinophilic variant.

Discussion: TTF1, although generally considered relatively specific for lung and thyroid carcinomas, is expressed in 33% ovarian clear cell adenocarcinomas. PAX8 and HNF-1b, though essential for further evaluation, was not available in the local setting.

Conclusion: Immunohistochemistry is essential for tumour subtyping. However, low specificity and lack of availability are the main limitations encountered.

CR 74

Langerhans cell histiocytosis presenting as an ulcerative scalp lesion: A diagnostic challenge on cytology

P. G. G. Sanjeewani*, L. A. H. A. Albert, S. Wickramasuriya, P. Kariyawasam

Department of Histopathology, Teaching Hospital, Karapitiya, Sri Lanka

pggsanjeewani@yahoo.com

Introduction: Langerhans cell histiocytosis (LCH) is a disorder of proliferating histiocytes with variable clinical presentations. The solitary form of LCH usually involves bone in children and, the skull is a common site. LCH shows characteristic microscopic and immunohistochemical features.

Case report: A four-year-old boy presented with a progressively enlarging ulcerative scalp lump for two weeks, and the imaging revealed an underlying lytic lesion of the skull. Intraoperative crush smears were cellular and showed cells with vesicular nuclei and moderate eosinophilic cytoplasm. The background shows a mixed inflammatory cell infiltrate admixed with eosinophils and scattered multinucleated giant cells. The cytological features were in favour of an inflammatory lesion, and the differential diagnoses included LCH. The specimen was composed of a flap of skull bone with overlying soft tissue containing an ulcerated lesion, 15x15x3 mm. Histology of the lesion showed sheets of polygonal cells with folded and grooved nuclei having vesicular chromatin, small eosinophilic nucleoli and abundant eosinophilic cytoplasm. Mixed inflammation with eosinophils and multinucleated giant cells were noted. The lesion extended into the scalp muscle and bone. Immunohistochemically, the neoplastic cells were positive for S-100, CD68 and CD1a, confirming LCH. At the time of diagnosis, there was no other system involvement.

Discussion: Crush smear or imprint is used for cytological assessment and is a commonly used rapid diagnostic test in resource-poor settings due to its feasibility, cost-effectiveness and reliable diagnostic accuracy. LCH is a rare disorder and a difficult diagnosis by cytology alone.

Comment: LCH has long been considered a reactive process. But recent insight into molecular pathology favours a neoplastic origin. Its prognosis depends on the clinical type and staging.

CR 75

Sessile serrated lesion with dysplasia in a colectomy with polyposis, predominantly having adenomas

S.M.I. Seneviratne*, M.A.G.N. Gunathilake, A. Vithanage

Department of Histopathology, National Hospital Kandy, Sri Lanka

indu_anuruddha@yahoo@co.uk

Introduction: Polyposis syndromes predominantly having adenomas can be autosomal dominant; Familial adenomatous polyposis-FAP, attenuated familial adenomatous polyposis, polymerase proofreading- associated polyposis and AXIN2-associated polyposis, or autosomal recessive; MUTYH-associated polyposis, NTHL1-associated polyposis and constitutional mismatch repair deficiency.

Case report: A 40-year-old woman underwent a pan colectomy for multiple colonic polyps. The patient did not have any extracolonic manifestations. The colectomy specimen was 660 mm in length and contained more than 100 polyps, ranging 5 mm to 15 mm, and one sessile polyp of 30 mm diameter located in the proximal transverse colon. Microscopically, except for the sessile lesion, the sampled polyps revealed tubular adenomas with low-grade intestinal-type dysplasia. Microscopy of the sessile polyp revealed serrations extending to the crypt base, crypt architectural changes as horizontal growth along muscularis mucosa and dilation of crypt base, in keeping with a sessile serrated lesion (SSL) with dysplasia. The lining epithelium of some crypts showed low-grade dysplasia.

Discussion and conclusion: SSLs are documented rarely in FAP and well-described in MUTYH-associated polyposis and NTHL1-associated polyposis. Data regarding other polyposis syndromes are sparse. Distinct extra-colonic manifestations are identified in all syndromes. Diagnosis of the exact syndrome is important as the mode of inheritance and cancer risk are variable. Genetic analysis is necessary, however, not carried out in this patient, owing to economic constraints. WHO Classification of digestive tract tumours (5^{th} edition) has suggested a diagnostic algorithm for patients with adenomatous polyposis.

CR 76

Malignant myopericytoma: A case report of an exceedingly rare tumour

S.M.I. Seneviratne*, M.A.G.N. Gunathilake, R.M.P. Ratnayake

Department of Histopathology, National Hospital Kandy, Sri Lanka

indu_anuruddha@yahoo.co.uk

Introduction: WHO classification of Soft Tissue and Bone Tumours describes myopericytoma and myofibroma as neoplasms of the same morphological continuum. Myopericytomas are common in adults, while myofibromas are predominant in the first two years of life. Malignant myopericytoma is an extremely rare tumour. A malignant counterpart of myofibroma has not been described.

Case report: A 46-year-old woman presented with a supraclavicular mass of one-year duration. The resected tumour was a lobulated mass measured 90x70x50 mm. The cut surface was whitish and solid. Microscopy revealed an unencapsulated lesion containing nodules of spindle cells concentrically arranged around blood vessels which show a haemangiopericytoma-like pattern. Some areas showed sheets of cells with moderate to markedly pleomorphic nuclei and focal necrosis. The resection margins contained infiltrating tumour nodules. The tumour cells showed diffuse strong positivity for SMA and focal positivity for BCL2. Occasional cells showed weak positivity for desmin. The Ki-67 index was 60%. H-caldesmon was inconclusive. Pancytokeratin, CK7, CK20, EMA, HMB45, CD31, GFAP, LCA and CD34 were negative. A diagnosis of a malignant myopericytoma was made.

Discussion and conclusion: The initial differential diagnoses were carcinoma, melanoma, perivascular tumour, synovial sarcoma, MPNST and low-grade myofibroblastic sarcoma. Moderate to marked nuclear pleomorphism, focal loss of perivascular arrangement, high proliferative index, tumour necrosis and the presence of tumour infiltration were suggestive of a malignant myopericytoma. Atypical myofibroma/ myopericytoma was excluded owing to the markedly increased proliferative activity and the lack of diffuse desmin positivity. Malignant myopericytoma shows a poor clinical outcome with local recurrence and distant metastases.

CR 77

Juvenile xanthogranuloma involving maxillary sinus: A common lesion in an uncommon site

H. R. S. D. Sumansekara[1]*, K. V. C. K. Dharmadasa[2], S. Wijetunge[1], B. Bandaranayake[2]

[1]*Department of Pathology, Faculty of Medicine, University of Peradeniya, Sri Lanka*

[2]*Sirimavo Bandaranayke Specialized Children's Hospital, Peradeniya, Sri Lanka*

sachinthanasumanasekara@gmail.com

Introduction: Juvenile xanthogranuloma (JXG) is an uncommon benign childhood disorder that belongs to a group of non-Langerhans cell histiocytosis. JXG typically involves the skin of the head and neck region. However, there are well-documented cases of extracutaneous locations. This is a rare case of a maxillary sinus JXG in a child.

Case report: A three-year-old boy presented with epistaxis and nasal blockage for five months. Examination under anaesthesia revealed a mass arising from the left side middle meatus. CECT showed a soft tissue lesion in the left side maxillary sinus extending to the nasal cavity. During surgery, no adhesions were noted. Macroscopy showed a soft, yellowish lesion measuring 3x2x1 cm. Microscopy revealed an unencapsulated lesion composed of sheets of foamy histiocytes and Touton type multinucleated giant cells admixed with mononuclear inflammatory cells. There was no cellular pleomorphism or prominent mitotic activity. Necrosis was not present. Grocott-Gomori stain for fungi and Ziehl-Neelsen stain for acid-fast bacilli were negative. The lesional cells were positive for CD68 and were negative for vimentin, SMA and ALK1, excluding the remote possibility of an inflammatory myofibroblastic tumour. These features were consistent with a JXG.

Discussion and conclusion: JXG results from a reactive process following a defective macrophage response to a non-specific injury. Although the current lesion had the typical histology, the unusual location in the maxillary sinus posed a diagnostic challenge. Close clinical correlation, appreciation of benign features, both clinically and histologically, and exclusion of other possibilities help arrive at the correct diagnosis.

CR 78

Extra-skeletal osteosarcoma of the penis: a rare case in world literature

J. M. M. Theepan.*, S. R. Constantine.

Department of Histopathology, National Hospital of Sri Lanka.

jathavani@gmail.com

Introduction: Extra-skeletal osteosarcoma (ESOS) is a rare soft tissue malignancy of elderly, and usual locations include retroperitoneum and extremities. ESOS of the penis is rare and as per our knowledge, this is the eighth case reported in world literature.

Case report: A 92-year-old man presented with a non-healing ulcer at the tip of the penis for 6 months. The partial penectomy showed a growth at the tip of the penis with a gritty cut surface, involving the urethra, corpus cavernosum and corpus spongiosum. Histology revealed a poorly differentiated malignancy with sheets of pleomorphic spindle cells in a myxoid stroma. There were multiple foci of lace-like osteoid surrounded by malignant spindle cells occupying ~40% of the tumour. Foci of mineralized osteoid were noted with malignant spindle cells. Mitoses were frequent with atypical forms. The overlying skin was non-dysplastic; features of squamous differentiation were not identified. The tumour cells in osteosarcomatous and pleomorphic areas showed strong positivity for SMA and CD99. The negativity of tumour cells for AE1/AE3, CK5/6, Melan-A, HMB45, S100, Desmin, CD 31, CD 34 and BCl2 excluded the possibility of a spindle cell squamous cell carcinoma, malignant melanoma, histiocytic neoplasms, pleomorphic leiomyosarcoma, cutaneous angiosarcoma and poorly differentiated synovial sarcoma. A diagnosis of ESOS of the penis was made in view of the presence of malignant osteoid, positivity for CD99 and SMA as reported in osteosarcomas and negativity for epithelial and melanocytic markers.

Discussion and conclusion: ESOS accounts for <1% of soft tissue sarcomas. The major differential diagnosis is undifferentiated pleomorphic sarcoma, which is excluded by the presence of malignant osteoid. They are high-grade and have a poor prognosis with local and distant metastasis. While radical excision seems the best option for local control, the major cause of disease-related death appears to be distant metastasis.

CR 79

Small cell neuroendocrine carcinoma of urinary bladder: a rare tumour

J. M. M. Theepan*, K. Samalai, M. M. A. Jayawickrama

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

jathavani@gmail.com

Introduction: Small cell neuroendocrine carcinoma (SCNEC) of the bladder is a rare and aggressive tumour associated with a poor prognosis. It often presents at a late stage and accounts for less than 1% of bladder malignancies.

Case report: A 53-year-old man presented with haematuria and was found to have a polypoid growth in the lateral wall of the bladder on cystoscopy. Imaging revealed a polypoid growth with early peri-vesical extension. Histology of the bladder chippings showed an infiltrating tumour with solid sheets and nests of undifferentiated cells with hyperchromatic nuclei and scant cytoplasm. Frequent mitoses, crush artefact and apoptotic debris were evident. The tumour invaded the lamina propria and muscularis propria (at least T2). The tumour cells showed immunoreactivity for chromogranin A and synaptophysin, and the Ki-67 proliferative index was 98%, confirming a diagnosis of SCNEC . The patient underwent cystoprostatectomy and pelvic lymph node dissection, which showed a pT3a pN0 tumour.

Discussion and conclusion: The differential diagnosis for an undifferentiated small cell tumour of the bladder includes SCNEC, lymphoma and high-grade urothelial carcinoma. Morphology and the immunohistochemical staining pattern help to differentiate between these small cell tumours. SCNEC of the urinary bladder confers a worse prognosis when present in its pure form in comparison to a mixed tumour with coexistent urothelial carcinoma. The prognosis depends on the performance status and extent of the disease at the time of diagnosis. Overexpression of p53, age of the patient, gender and presenting features do not appear to correlate with prognosis. Neoadjuvant or adjuvant cisplatinum-based chemotherapy has been used with radical cystectomy to potentially improve long-term survival.

CR 80

Lipofibromatosis in extradural space of lumbar vertebrae presenting as a mass lesion with bone involvement

J. M. M. Theepan*[1], S. Vinothika[1], N. C. Marasinghe[2], S. A. Gunaratne[2], K. Dissanayake[1].

[1]*Department of Histopathology, National Hospital of Sri Lanka*

[2]*Lady Ridgeway Hospital for Children, Colombo, Sri Lanka*

jathavani@gmail.com

Introduction: Lipofibromatosis (LF) is a rare paediatric soft tissue tumour with an admixture of mature adipose tissue and fibroblastic elements. Although benign, LF can be locally destructive. The common sites of involvement include extremities and the trunk.

Case report: A 9-year-old boy presented with progressively worsening lower back pain of 4 months duration. Imaging revealed a soft tissue mass in the back involving the extradural spaces of L3, L4, L5 vertebrae and extending through neural foramina of L4 and L5. Repeated guided core biopsies revealed a benign fibroblastic lesion composed of mature adipose tissue traversed by cellular fascicles of spindle cells with bland elongated nuclei. Mitoses or nuclear atypia were not evident. The lesion infiltrated the adjacent skeletal muscles featuring atrophy of the muscle fibres with tumour permeating into bone trabeculae of the vertebral pedicles. There were foci of ossification and calcification with active cartilage fragments. Immunohistochemistry with S100 was negative and excluded a LF-like neural tumour (LF-NT).

Discussion and conclusions: Although having similar morphology to adult lesions, calcification and/or ossification is peculiar in paediatric cases of LF. Paravertebral localization of the tumour is an unusual feature reported in the literature. It may infiltrate the adjacent muscles and grow around nerves and vessels, which may result in pain, tenderness and functional deficits. The major differentials include LF-NT, calcifying aponeurotic fibroma and infantile fibrosarcoma. LF-NT, though morphologically similar to LF, can be distinguished by S100 immunoreactivity and LMNA-NTRK1 gene fusion. The local recurrence rates are around 70%, with an increased tendency of recurrence in the presence of congenital onset of disease, male gender, location in the hand/feet and mitotically active fibroblastic component.

CR 81

Merkel cell carcinoma associated with invasive squamous cell carcinoma

J. M. M. Theepan*, S. Vinothika, S. Thanansayan, K. Dissanayake

Department of Histopathology, National Hospital of Sri Lanka.

jathavani@gmail.com

Introduction: Merkel cell carcinoma (MCC) is an uncommon aggressive cutaneous malignancy with neuroendocrine differentiation, affecting sun-exposed areas of the elderly. MCC associated with another tumour is rarer, and squamous cell carcinoma (SQCC), in-situ or invasive, is the commonest associated malignancy with MCC.

Case report: A 91-year-old woman presented with a pigmented patch in the right lateral thigh with inguinal lymphadenopathy. Cytology of the inguinal lymph node revealed a poorly differentiated malignancy. Wide local excision of the skin lesion showed an ulcerated hyperpigmented, crusted lesion. Histology revealed a poorly differentiated invasive tumour with intermingled small and epithelioid cell components. The epithelioid cells were arranged in sheets and infiltrative cords and contained pleomorphic nuclei with deeply eosinophilic cytoplasm. They were positive for CK5/6. The small cell areas showed nuclei with stippled chromatin and scant cytoplasm and were highlighted by synaptophysin, chromogranin and CK20. Both cell populations were negative for S100. The overlying epithelium showed carcinoma in-situ. Inguinal lymph nodes showed extensive deposits of both tumour components, confirmed by immunohistochemistry. A diagnosis of MCC associated with invasive SQCC, with regional lymph node metastasis of both tumour components was made. The tumour was considered bi-phenotypic, due to the intermingling of the MCC and SQCC areas.

Discussion and conclusions: A small number of primary cutaneous neuroendocrine carcinomas are associated with SQCC or (more rarely) basal cell carcinoma or adnexal tumour. Although collision tumours may occur, there is also evidence that bi-phenotypic tumours exist, as in this case. MCC shows a high rate of recurrence and metastasis with an overall 5-year survival rate of 35% for patients with regional metastases. The overall survival data is limited for such combined tumours.

CR 82

Primary synovial sarcoma of the kidney: An incidental finding in a young woman

J. M. M. Theepan[1*], P. N. Yogananda[1], L. J. De Silva[1], W. R. D. Fonseka[2], N. D. Perera[3],

M. M. A. Jayawickrama[1]

[1]Department of Histopathology, [2]Vascular Surgery Unit, [3]Genitourinary Surgery Unit, National Hospital of Sri Lanka

jathavani@gmail.com

Introduction: Synovial sarcoma (SS) is a rare soft tissue malignancy, primarily involving extremities. Primary renal SSs are even rarer, accounting for <2% of malignant renal tumours with less than 150 reported cases globally.

Case report: A 36-year-old woman was incidentally found to have a left renal mass while being investigated for primary subfertility. Imaging revealed a left renal mass suggestive of a renal cell carcinoma (RCC). The left partial nephrectomy specimen revealed a solid and cystic lesion with necrotic and haemorrhagic areas. Histology revealed a tumour with fascicles of mildly pleomorphic spindle cells admixed with acinar structures lined by columnar epithelial cells with vesicular nuclei and pale eosinophilic cytoplasm. The tumour cells were immunoreactive for CD99 and BCL2, while the epithelial component was positive for AE1/AE3. CD10 was negative. The tumour was confirmed as biphasic SS sarcoma with cytogenetic translocation of SYT-SSX2 on chromosome 18q11.2. Subsequent completion nephrectomy did not reveal residual tumour tissue. The patient was referred for post-operative chemotherapy.

Discussion and conclusions: The pre-operative differentiation of SS from RCC is difficult due to similarities in clinical presentation and radiological features. The differential diagnoses include sarcomatoid RCC, primary Ewing tumour, adult Wilm tumour and undifferentiated carcinoma. Primary SS should be included in the differential diagnosis in spindle cell tumours of the kidney, especially in young adults. The prognosis of the tumour is generally poor. Multimodal treatment with surgery and chemotherapy may achieve curative effects.

CR 83

Primary cutaneous anaplastic large cell lymphoma mimicking cutaneous leishmaniasis

J. M. M. Theepan *, S.R. Constantine

Department of Histopathology, National Hospital of Sri Lanka

jathavani@gmail.com

Introduction: Primary cutaneous anaplastic large cell lymphoma (PC-ALCL) is the second most common cutaneous T-cell lymphoma, accounting for 9% of cases.

Case report: A 24-year-old young man presented with hypopigmented and erythematous nodules over the face and extremities of two years duration, with differential diagnoses of diffuse cutaneous leishmaniasis (CL), sarcoidosis, leprosy and mycosis fungoides (MF) transforming to tumour stage. Skin punch biopsies were performed, which showed a superficial mixed inflammatory infiltrate rich in plasma cells suggestive of CL. A consensus diagnosis of CL was made at the clinicopathological meeting. However, as other ancillary investigations for CL were negative, an incisional biopsy of the elbow lesion was performed. Histology revealed an ALCL with numerous large, atypical cells in a background comprising a nodular infiltrate of lymphocytes and plasma cells. Hallmark cells of ALCL were also present. The neoplastic cells showed strong immunoreactivity with CD30 (in >75% of cells), CD3, CD4, CD5 and MUM1 and variable reactivity with EMA and CD15. They were negative for CD20, CD8 and ALK. The Ki-67 proliferative index was high in neoplastic cells. Considering the history of negative lymph node biopsy and bone marrow examination, a diagnosis of primary cutaneous anaplastic large cell lymphoma was made following a multidisciplinary team meeting. The patient was referred to the National Cancer Institute Maharagama for further management with chemotherapy.

Discussion and conclusions: PC-ALCL is a cutaneous neoplasm composed of large CD30 positive cells (by definition >75% of cells) in a background of mixed inflammatory cells. It is a mimicker of other inflammatory conditions of the skin in the early stages. PC-ALCL needs to be considered a differential diagnosis in nodular ulcerative lesions of the skin, clinically and histologically, for its diagnosis at early stages.

CR 84

Primary central nervous system melanoma

H.N. Thewarapperuma*, S. Wickramasuriya, G.S. Hettiarachchi, H.A.S. Perera

Department of Histopathology, Teaching Hospital Karapitiya, Sri Lanka

thewarapperumaharshi@gmail.com

Introduction: Melanoma is a malignant lesion arising from melanocytes. Primary intracranial melanomas are uncommon and constitute 1% of all melanomas and 0.07% of all brain tumours.

Case report: A 55-year-old previously healthy woman presented with headache and intermittent vomiting for three months duration. Magnetic resonance imaging demonstrated a space-occupying lesion in the right parietal region and was reported as high-grade glioma. Intraoperative cytology revealed dispersed plump spindle cells with intracytoplasmic brownish pigment. The resected specimen showed an infiltrative tumour composed of loosely cohesive epithelioid to spindle cells with granular intracytoplasmic melanin pigment. Moderately pleomorphic nuclei showed prominent nucleoli. Immunohistochemically the cells showed cytoplasmic granular positivity for HMB45 and Melan-A confirming the diagnosis of melanoma.

Discussion: Melanocytic tumours of the central nervous system (CNS) originate from leptomeningeal melanocytes, and include meningeal melanocytosis, melanomatosis, melanocytoma and melanoma. In our case, the diagnosis of melanoma was made on the grounds of nuclear pleomorphism, mitotic activity and necrosis. Initially, cerebral metastasis of melanoma was assumed as it is the 3rd commonest cause of cerebral metastasis. In the absence of an occult primary lesion in the skin, eye, mucus membranes, and lower gastrointestinal tract after thorough evaluation, a diagnosis of primary CNS melanoma was made. Complete surgical resection followed by focal radiotherapy gives better prognosis in primary CNS melanoma compared to metastases, which needs systemic therapy as well.

Conclusion: Definitive diagnosis of primary CNS melanoma is a diagnosis of exclusion, especially in the absence of cutaneous melanosis. A high index of clinical suspicion and good pathology reporting is important in diagnosing these tumours.

CR 85

Rare occurrence of parathyroid carcinoma in a patient with MEN 1 syndrome

S. Thoufeek, H.D.Wijesinghe*

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

harshima@path.cmb.ac.lk

Introduction: Multiple endocrine neoplasia type 1 (MEN1) is an autosomal dominant syndrome caused by germline mutation of the MEN1 gene. Patients with MEN1 present with parathyroid adenomatosis, adenomas of the endocrine pancreas, gastrinomas of the duodenum, pituitary adenomas and neuroendocrine tumours of thymus, stomach, lung and intestine. Only 15 cases of parathyroid carcinoma (PC) have been reported in this setting.

Case report: A 52-year-old man presented with features of hypercalcaemia with a serum calcium level >1000mmol/L, and radiological investigations showed a parathyroid neoplasm, pituitary adenoma, gastrinoma and pancreatic adenoma. He was suspected to have MEN1 and underwent parathyroidectomy, thyroidectomy and thymectomy. A 40x20x20mm firm tan-coloured mass with solid and cystic areas was identified in the left inferior parathyroid. Microscopically a parathyroid tumour was seen comprising lobules of epithelial cells separated by thick fibrous bands. The constituent cells had mildly pleomorphic nuclei with dense chromatin, inconspicuous nucleoli and eosinophilic cytoplasm. Mitoses were not evident. There was no necrosis. There were foci of vascular invasion confirmed by CD34. Perineural or capsular invasion, invasion of the surrounding soft tissue or adjacent thyroid gland were not identified. The other three parathyroid glands showed parathyroid adenomatosis. Multifocal papillary microcarcinoma was present in the thyroid. The thymus was normal. He died of an unrelated cause five months after the diagnosis.

Discussion and conclusion: PC in the setting of MEN1 is rare but should be considered in patients presenting with a parathyroid neoplasm and severe hypercalcaemia. Ideally, the diagnosis of MEN1 should be confirmed with genetic testing. However, the presence of pancreatic, duodenal, pituitary and parathyroid tumours in this patient is highly suggestive.

CR 86

A case of primary ALK-negative anaplastic large cell lymphoma of the bone mimicking tuberculosis

S. Vinothika*, S.R. Constantine

Department of Histopathology, National Hospital of Sri Lanka

smayurran@yahoo.com

Introduction: ALK-negative anaplastic large cell lymphoma (ALK(-)ALCL) was first introduced under the WHO classification in 2008. As opposed to ALK-positive ALCL, which usually affects children and young adults, ALK(-)ALCL affects the middle-aged. Primary bone lymphomas are usually clinically diagnosed as infections or inflammatory diseases due to their rarity.

Case report: A 48-year-old woman presented with back pain and intermittent fever for three months. Computed tomography showed extensive destruction of L2/L3 vertebrae suggestive of tuberculous spondylodiscitis, and antituberculous drugs were started empirically. Biopsy from the L2/L3 vertebrae showed a lesion composed of large pleomorphic cells containing enlarged nuclei and some having eccentric horseshoe-shaped nuclei (hallmark cells). Multinucleated giant cells with wreath-like arrangements were noted. Mitoses were present. The tumour infiltrated the bone. The background showed necrosis and contained numerous neutrophils, lymphocytes, collections of foamy histiocytes. The differential diagnoses were ALCL and classic Hodgkin lymphoma (CHL). The large, atypical cells were positive for immunohistochemical stains CD30 and CD4 showed focal positivity for EMA and were negative for CD3, CD20, PAX5 and ALK. Strong CD30 positivity with equal intensity excluded peripheral T-cell lymphoma-NOS, CD4 positivity excluded a metastatic carcinoma. A primary ALK (-) ALCL of bone was diagnosed and the patient was referred for chemoradiotherapy.

Discussion and conclusions: Histopathological evaluation of bone lesions is imperative to diagnose rare cases of lymphomas. Multiple immunohistochemical markers are needed for confirmation of the T cell origin, as CD3 is frequently negative. When all T cell markers are negative, PAX5 is helpful to rule out CHL as it shows weak expression in CHL and is negative in ALCL.

CR 87

A rare case of sarcomatoid urothelial carcinoma with small cell neuroendocrine carcinoma component

S.Vinothika*, J.M.M Theepan, M.A Jayawickrama

Department of Histopathology, National Hospital of Sri Lanka

smayurran@yahoo.com

Introduction: Sarcomatoid urothelial carcinoma (SUC) is a rare variant of invasive urothelial carcinoma accounting for 0.6% of all bladder tumours with heterologous elements such as chondrosarcoma or osteosarcoma. SUC can be associated with areas of small cell neuroendocrine carcinoma which is even rarer.

Case report: A 59-year-old man presented with gross haematuria, and computerized tomography revealed a bladder tumour extending to the prostate and seminal vesicles. Biopsies were taken from bladder tumour and prostate. Biopsy of the bladder tumour showed nests and sheets of pleomorphic tumour cells and sheets of spindle cells. A chondrosarcomatous component containing malignant cartilage was seen. There were large areas of necrosis. PanCK was strongly positive in urothelial carcinoma and focally positive in sarcomatoid areas. A component of small cell neuroendocrine carcinoma (SmCC) was noted in the bladder tumour and predominantly in the prostatic cores. SmCC showed round to oval cells containing nuclei with salt pepper chromatin and scanty cytoplasm. Crush artefact was seen in those areas. Mitoses were frequent. SmCC was positive for synaptophysin and chromogranin with a high Ki-67 index. A diagnosis of SUC with chondrosarcomatous component admixed with small cell neuroendocrine carcinoma was made. As the prostate was involved, it was staged as pT4a.

Discussion and conclusion: SUC and small cell neuroendocrine carcinoma are aggressive tumours with extensive necrosis and are at an advanced stage. Both have poor prognoses. Histological features with appropriate immunohistochemistry confirm the diagnosis.

CR 88

Bilateral acoustic neuroma: a diagnostic hallmark of neurofibromatosis type 2

W.M.M.A. Wanasinghe*[1], K. Kopinath[2], K. Dissanayake[2]

[1] *Department of Pathology, Apeksha Hospital, Maharagama.*

[2] *Department of Histopathology, National Hospital of Sri Lanka*

drmadumaliwanasinghe@gmail.com

Introduction: Acoustic neuroma or vestibular schwannoma is the most frequent tumour of the cerebellopontine (CP) angle. It represents 8% of all intracranial tumours and 85% of CP angle masses. Around 4% of cases arise in the background of neurofibromatosis type 2 (NF2). Schwannoma is a benign tumour that arises from Schwann cells.

Case report: A 24-year-old woman presented with sudden onset bilateral hearing loss, unsteadiness of gait and slurring of speech. Magnetic resonance imaging (MRI) revealed two extra axial lesions in bilateral CP angle cisterns causing compression of the brain stem, cerebellum and the fourth ventricle. They extended into each internal auditory canal, causing widening of the meatuses. The right-side tumour was excised and received as multiple fragments. Microscopy showed a spindle cell tumour with hypercellular and hypocellular areas. The tumour cells formed fascicles and had spindle-shaped nuclei with dense chromatin, eosinophilic cytoplasm and indistinct cell borders. Nuclear palisades, accompanied by Verocay bodies were evident in hypercellular areas. Schwannoma was diagnosed on morphology.

Discussion and conclusion: Bilateral vestibular schwannoma is one of the first presentations of NF2, which is an autosomal dominant disorder. This patient had no family history or cutaneous manifestations of NF. Other than schwannoma, NF2 patients may develop juvenile cortical cataract, multiple meningiomas, gliomas, spinal cord ependymomas and schwannosis before the third decade of life. It is important to do genetic studies to exclude mutated NF 2 genes in first degree relatives of the affected individuals. Affected patients need long-term follow-up in view of other tumours that can arise in the background of NF2.

CR 89

A rare case of squamous metaplasia in papillary carcinoma of thyroid

I. L.Wickramanayake*, T. G. Liyanage

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

indikalakmali507@gmail.com

Introduction: Squamous metaplasia (SM) in the thyroid gland is rare and seen with neoplastic lesions such as mucoepidermoid carcinoma, squamous cell carcinoma, papillary thyroid carcinoma (PTC) and non-neoplastic lesions like Hashimoto's thyroiditis.

Case report: A 29-year-old woman presented with a thyroid swelling of 6 months associated with cervical lymphadenopathy. An ultrasound scan of the neck revealed a suspicious nodule in the right midzone. Cytology revealed a papillary carcinoma of the thyroid. She underwent total thyroidectomy and cervical lymphadenectomy. There was a solid nodule in the right mid-zone of the thyroid measuring 20x12x8 mm, abutting the deep resection margin. Microscopy showed a classical variant PTC with abrupt transition to a squamous component. The squamous component accounted for 20% of the tumour and was composed of variable-sized nests of squamous cells with no atypia. No diffuse sclerosis or psammoma bodies were noted. Focal lymphocytic thyroiditis was present in the background. Cervical lymph nodes had metastatic deposits of papillary carcinoma, with no squamous metaplasia.

Discussion: Focal or extensive SM in PTC is rarely encountered and represents an important diagnostic pitfall and challenge for the histopathologist. It can be misinterpreted as squamous cell carcinoma (SCC) or anaplastic carcinoma with squamous differentiation. SM is mostly seen in diffuse sclerosing and cribriform morular variants of PTC and is rare in the classical variant. The absence of cytological atypia in the squamous component is essential for the diagnosis of SM.

Conclusion: In view of the therapeutic implications of the various differential diagnoses, awareness of this lesion will help pathologists to make an accurate diagnosis.

CR 90

Xanthogranulomatous hypophysitis: a rare inflammatory condition of pituitary gland

L D Wickramasinghe*, K. Samalai, K. Dissanayake

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

thinulaayan@gmail.com

Introduction: Xanthogranulomatous hypophysitis (XGH) is a rare inflammatory condition that mimics a pituitary neoplasm clinically and radiologically.

Case Report: A 27-year-old woman presented with secondary amenorrhoea for 4 years. Physical examination was unremarkable and found to have panhypopituitarism biochemically. MRI scan of the brain revealed a mass lesion compressing the optic chiasm and was reported as pituitary macroadenoma. She underwent transnasal endoscopic transphenoidal excision.

The specimen was received as multiple tan-coloured pieces of tissue. The entire specimen was processed and examined at multiple levels. Microscopy revealed collections of foamy histiocytes, cholesterol crystals and foreign-body giant cells. There was no caseation or a prominent inflammatory cell infiltrate. Pituitary tissue, viable or necrotic tumour tissue were not evident. Zeihl-Neelsen stain for acid-fast bacilli and Grocott stain for fungi were negative. A histological diagnosis of XH was made.

Discussion: There are five types of pituitary hypophisitis; namely, lymphocytic, granulomatous, xanthomatous, necrotizing and xanthogranulomatous. XGH is the least common type. It can be primary autoimmune or secondary to a craniopharyngioma, Rathke-cleft cyst or due to systemic conditions like tuberculosis. The pituitary adenoma is the major differential diagnosis, which mimics XGH radiologically. Collections of foamy histiocytes, cholesterol clefts and multinucleated giant cells are the important microscopic features for the diagnosis. Exclusion of secondary causes of XGH is also important.

Conclusion: XGH is a rare condition, which mimics a neoplastic pituitary lesion clinically and radiologically. Histology is the mainstay of diagnosis.

CR 91

Multifocal adenomatous oncocytic hyperplasia of the parotid gland: a rare entity

L.D. Wickramasinghe*, S. Rodrigo, D. G. S. Udayanga

Department of Pathology, Sri Jaywwardenepura General Hospital, Sri Lanka

thinulaayan@gmail.com

Introduction: Multifocal adenomatous oncocytic hyperplasia (MAOH)/multifocal nodular oncocytic hyperplasia of the parotid gland is a rare non-neoplastic entity that should be differentiated from other benign and malignant oncocytic lesions of the salivary tissue.

Case report: A 63-year-old man was investigated for swelling of the right parotid gland. MRI scan revealed a lesion within the right parotid gland. FNAC revealed mature adipose tissue only. The excised parotid gland measured 6x4x3 cm, and the cut surface revealed a tan colour nodule measuring 3.5x2.8x2.5 cm surrounded by salivary tissue. Microscopy revealed an unencapsulated lesion with lobules containing acini and central ducts. The constituent cells showed mildly pleomorphic centrally located nuclei with prominent nucleoli and abundant eosinophilic, granular cytoplasm. Mitoses were inconspicuous. No fibrosis or necrosis was noted. The background contained unremarkable salivary tissue. MAOH of the parotid gland was the diagnosis.

Discussion: MAOH accounts for 0.1% of diseases of the parotid gland. Oncocytoma, oncocytic carcinoma, Warthin tumour, diffuse oncocytosis are the differential diagnosis for oncocytic lesions of the salivary tissue. MAOH can be differentiated from other oncocytic lesions by the absence/presence of an incomplete capsule, absence of mitoses, the presence of normal parenchymal structures, absence of necrosis and little or no internal fibrosis of the stroma.

Conclusion: Diagnosis of MAOH is important since it is a benign condition cured by complete surgical excision.

CR 92

Adenocarcinoma arising from a hamartomatous polyp in a patient with Peutz-Jeghers syndrome

D. J. Wickramasooriya*, G. S. Hettiarachchi, P. G. G. Sanjeewani, P. Kariyawasam

Teaching Hospital, Karapitiya, Galle, Sri Lanka, Sri Lanka

wickramasooriyaj@gmail.com

Introduction: Peutz-Jeghers syndrome (PJS) is characterised by mucocutaneous pigmentation and polyps in the gastrointestinal tract. It is associated with a germline mutation in the serine threonine kinase 11 gene (STK11). These patients are at an increased risk of developing both gastrointestinal and extraintestinal malignancies.

Case report: A 45-year-old previously healthy woman presented with intermittent abdominal pain and vomiting for one week. Lower gastrointestinal endoscopy showed multiple pedunculated polyps extending from caecum to rectum with endoscopic features of malignancy in one polyp. Subtotal colectomy was performed, and the resected bowel showed seven pedunculated polyps ranging from 0.5 to 5cm in maximum dimensions. Histology revealed six benign hamartomatous polyps (HMP). The largest polypoidal mass showed areas of hamartoma, dysplasia and invasive carcinoma. Histology of the left ovary, which had been removed for surgical reasons, revealed a sex cord stromal tumour with annular tubules in a small focus. Retrospective physical examination showed mucocutaneous pigmentation. Clinical and histological features directed towards the diagnosis of invasive adenocarcinoma of the colon in a patient with PJS and led to subsequent family screening.

Discussion: HMPs are considered benign polyps. However, HMPs associated with syndromes may have malignant potential. Colorectal adenocarcinomas in these patients are thought to be arising from HMPs, but their carcinogenic pathway is under debate. Sex-cord stromal tumour with annular tubules is characteristically associated with PJS. This case presents evidence for an adenocarcinoma arising from a HMP in PJS hence the importance of frequent surveillance of these patients with HMPs.

CR 93

T-cell lymphoblastic lymphoma: a rare case of adult lymphoma

S. Wickramasuriya*, L.A.H.A. Albert, P.G.G. Sanjeewani, P. Kariyawasam, Sri Lanka

Department of Pathology, Teaching Hospital, Karapitiya, Sri Lanka

wshiranthika@yahoo.com

Introduction: T-cell lymphoblastic lymphoma (TC-LBL) is a neoplasm of immature T cells. It is a rare entity in adults and represents 2% of all non-Hodgkin lymphomas (NHL). It usually presents as a mediastinal mass with pleural effusion.

Case report: A 58-year-old man presented with low-grade fever, cervical lymphadenopathy and a right-sided pleural effusion. Contrast enhanced computed tomography (CECT) revealed an anterior mediastinal mass with a right-sided pleural effusion and enlarged para-aortic lymph nodes. Cervical lymph node biopsy showed totally effaced nodal architecture with extra capsular invasion. The node was diffusely infiltrated by medium sized monomorphic atypical lymphoid cells, with indented nuclei, high nuclear to cytoplasmic ratio, finely dispersed chromatin, inconspicuous nucleoli, and increased mitoses. Cytoplasm was scanty. A diagnosis of high-grade NHL was made. Immunohistochemical studies revealed strong membrane positivity with CD3 and strong nuclear positivity with TdT. CD 20 was positive only in scattered B-cells. MUM1, BCL6 and CD10 were negative. The Ki-67 index was more than 90%. The diagnosis was confirmed as TC-LBL.

Discussion: Even though diffuse large B-cell lymphoma (DLBCL) is the commonest adult NHL, TC- LBL should be suspected in a patient with typical clinical features or classic histopathological findings since TC-LBL is a highly aggressive form of NHL. In such cases immunohistochemical evaluation with TdT in the first panel is appropriate to confirm the diagnosis.

Conclusion: Due to the aggressiveness of TC-LBL, meticulous work-up and timely diagnosis is pivotal for better patient outcomes.

CR 94

Tophaceous pseudogout presenting as a solitary nodule in the middle finger: a rare presentation

T.W.Wijesiri*, T.G. Liyanage

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

thiliniwwijesiri@gmail.com

Introduction: Pseudogout/calcium pyrophosphate dihydrate deposition disease (CPPD) is characterized by the accumulation of CPPD crystals in the intra-articular and periarticular tissue. Deposition of crystals as a nodule/tophaceous pseudogout (TP) is a rare presentation of CPPD. It is common in the temporal mandibular joint, however, rare in extremities.

Case report: A 70-year-old woman presented with a lump over the left middle finger for three years. There was a recent rapid enlargement associated with pain and whitish discolouration over the skin. She had chronic pain in both the large and small joints for ten years, however, was not on regular treatment. On examination, the lump was hard and measured 1.7x1.5x1.5 cm. Radiography of the left middle finger revealed a cloud-like lesion. The surgically excised lesion showed chalky-white material. Histopathology showed a well-circumscribed nodular lesion composed of brown nodules of varying shapes with foreign body type giant cell reaction. There were rhomboid-shaped crystals within these nodules, which were demonstrated by polarized light. Diagnosis of TP was made, and she is under further investigation.

Discussion: TP frequently occurs in middle-aged and elderly patients with a female predominance, and lesions range from 2 to 4 cm. Clinicoradiological features may mimic tophaceous gout, tumoral calcinosis, synovial chondromatosis, soft tissue chondromas and chondrosarcoma. Polarized light demonstrates the characteristic rhomboid crystals.

Comment: Tophus formation in pseudogout is a rare finding. In the presence of mimickers, histopathological evaluation is the key to the correct diagnosis and appropriate management.

CR 95

Diffuse gastric hyperplastic polyposis (DGHP): a rare entity

P. N. Yogananda*[1], S.R. Constantine[2]

Department of Histopathology, National Hospital of Sri Lanka

priy629@gmail.com

Introduction: Gastric polyps are common and detected incidentally during 1-6% of upper gastrointestinal endoscopies (UGIE). 75% of gastric polyps are hyperplastic polyps. However, DGHP or the presence of 50 or more gastric polyps in a person is very rare, with only a few reported cases.

Case report: A 45-year-old man gave a history of recurrent episodes of vomiting and abdominal fullness for 10 years. During previous hospital admission, nine years earlier, his haemoglobin level had been 2.0 g/L necessitating blood transfusion. The UGIE revealed multiple gastric polyps. Colonoscopy was normal. The biopsy showed hyperplastic polyps. Over the next nine years, he was symptomatically treated with proton pump inhibitors and sulindac on and off. He underwent a total gastrectomy in August 2020 as his symptoms gradually worsened. Macroscopic examination of the resected specimen revealed gastric mucosa carpeted with multiple mucoid sessile polyps of varying sizes. The largest pedunculated polyp measured 50 mm and was at the gastric inlet. Microscopic examination of the polyps revealed extensive foveolar hyperplasia with architectural distortion, irregular crypts, crypt dilatation and focal epithelial serration. The epithelium showed abundant cytoplasmic mucin. The lamina propria was oedematous and contained thin wispy bands of smooth muscle extending upward from the muscularis mucosae. Very focally, there was mild cellular dysplasia. However, there was no malignancy. The clinicopathological features did not favour Menetrier disease, juvenile polyposis, Cronkhite Canada Syndrome or fundic gland polyposis. He underwent endoscopic dilatation for a bowel wall stricture at the anastomotic site a few months later. Currently, the patient is asymptomatic and is on nutritional support with oral iron and vitamins.

Discussion and conclusion: DGHP is rare but should be considered in a person presenting with multiple gastric polyps.

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