Rosai-Dorfman disease is a rare non-malignant proliferation of histiocytes typically involving the cervical lymph nodes. A small subset of cases present with extranodal manifestations. Case series and individual case reports discuss occurrences of Rosai-Dorfman disease in virtually every anatomical site. We discuss a case of a 63-year-old male presenting with multiple raised nodular lesions on multiple cutaneous sites. Biopsy of these lesions ultimately suggested a diagnosis of cutaneous extranodal Rosai-Dorfman disease. We discuss the histopathological, clinical and immunohistochemical features that allowed for this diagnosis. This is a rare pathological diagnosis with variable manifestations. Clinical suspicion and recognition of its classical histological features are essential for correct diagnosis, and in avoiding unnecessary treatment.

References

1. Komaragiri M, Sparber L, Dardik M. Extranodal Rosai–Dorfman disease: a rare soft tissue neoplasm masquerading as a sarcoma. *World J Surg Oncol* 2013; 11: 63.
2. Mantilla J, Goldberg-Stein S, Wang Y. Extranodal Rosai-Dorfman disease: clinicopathologic series of 10 patients with radiologic correlation and review of the literature. *Am J Clin Pathol* 2016; 145: 211–22.

CLEAR CELL CARCINOMA OF THE HARD PALATE – A CASE REPORT

Sebastian Rosca, Michael Christie, Seve Kranz, Anand Murugasu

Anatomical Pathology Department, Royal Melbourne Hospital, Vic, Australia

Salivary gland neoplasia is a challenging area of diagnostic surgical pathology due to significant histomorphologic overlap between benign and malignant entities with the resultant risk of misdiagnosis and inappropriate treatment.

Clear cell carcinoma is a rare infiltrative, low grade salivary gland neoplasm with an indolent clinical course, little metastatic potential and a propensity for local recurrence.

This case reports a 32-year-old woman who presented with a 1-year history of a mass on her hard palate. The clinical history, histomorphologic, immunophenotypic and molecular features are described and discussed with emphasis on diagnostic pitfalls.

A LIPOMATOUS VARIANT OF SCROTAL ANGIOMYOFIBROBLASTOMA – A CASE REPORT

Priya Susan Roy, Sagarika Tripathy

Anatomical Department, Australian Clinical Labs, Bendigo, Vic, Australia

Angiomyofibroblastoma-like tumour of the male genital tract is a rare benign mesenchymal tumour arising in the scrotum, perineum or inguinal region. It is usually seen in females of reproductive age. Clinical presentation has no pathognomic findings. In the largest series to date, Iwasa and Fletcher¹ reported 25 cases in men with the mean age being 52 years. This case report is of an 18-year-old male with a scrotal angiomyofibroblastoma that was 35 cm in maximum dimension, in the paratesticular tissue separate from testicular parenchyma. Macroscopically, the lesion was a fibrous, fatty and mucoid mass. Histologically, it showed a lobulated pattern of epithelioid and spindle cells arranged around various calibre vessels.

Other differential diagnoses including myxoid liposarcoma and rhabdomyosarcoma were ruled out with the help of FISH studies for FOX 10 and PAX3. DDIT3 and FUS were negative. To date, in all open-access literature, there has only been 34 cases reported of this rare tumour, of which there is only one reported case <20 years of age; this will be the second case in the <20 age group in men. The poster will describe the clinical presentation, imaging, macroscopic findings, histology and immunohistochemistry along with a literature review.

Reference

1. Iwasa Y, Fletcher CD. Cellular angiofibroma: clinicopathologic and immunohistochemical analysis of 51 cases. *Am J Surg Pathol* 2004; 28: 1426–35.

EXTRAMAMMARY PAGET'S DISEASE WITH LYMPH NODE METASTASIS – ROLE OF IMMUNOHISTOCHEMISTRY AND DIAGNOSTIC PITFALLS

Hema Parag Salkade¹, Tan Chien Sheng¹, Ng Kok Kit², Poh Wee Teng¹

¹Department of Laboratory Medicine, Changi General Hospital, Singapore; and ²Department of Urology, Changi General Hospital, Singapore

Extramammary Paget's disease (EMPD) is an uncommon skin disease that is limited to the epidermis but has the potential to invade the dermis and metastasise. We describe a case of primary scrotal EMPD with lymph node metastasis.

A 78-year-old male presented with an irregular thickening of the right scrotal skin with lymphadenopathy. Fine needle aspiration (FNA) of the inguinal lymph node revealed a poorly differentiated metastatic malignancy. Metastatic melanoma and poorly differentiated adenocarcinoma were considered in the differential diagnosis.

Subsequently, an incisional biopsy of the scrotal lesion showed epidermal acanthosis, acantholysis and elongation of the rete ridges with atypical cells. The tumour cells were diffusely positive for CK 5/6 and CK7, and p63 highlighted the basal cells. Acantholytic squamous cell carcinoma was our initial diagnostic consideration. However, in view of coexpression of CK5/6 and CK7, and a metastatic lymph node, the case was reviewed and additional immunohistochemical stains were performed. The lesional cells were immunoreactive for mCEA, BerEP4 and GATA3, and negative for CK20 and MelanA stains.

Immunohistochemistry was helpful in arriving at the final diagnosis of EMPD. EMPD although rare, should also be considered in the differential diagnosis of metastatic poorly differentiated adenocarcinoma in appropriate clinical settings.

PARATHYROID ADENOMA WITH PROMINENT LYMPHOCYTIC INFILTRATION: A POTENTIAL PITFALL IN FROZEN SECTIONS AND CYTOLOGY REPORTING

Nassim Saremi^{1,2}, Admire Matsika^{1,3}

¹Anatomical Pathology, Mater Pathology, South Brisbane, Qld, Australia; ²Pathology Queensland, Gold Coast university Hospital, Southport, Qld, Australia; and ³Biomedical Sciences, Faculty of Medicine, University of Queensland, Herston, Qld, Australia

Introduction: Parathyroid adenoma with prominent lymphocytic infiltration is rare, with only 12 cases reported in the English scientific literature. All these cases were not related to synchronous autoimmune or IgG4-related disease and presented clinically with primary hyperparathyroidism.

Case presentation: A 65-year-old woman presented with hypercalcemia and a neck mass, suggestive of parathyroid adenoma. She underwent parathyroidectomy and histopathological examination of the specimen showed a hypercellular parathyroid nodule comprising mature lymphoid cells admixed with parathyroid chief cells. A comprehensive immunohistochemistry panel confirmed the benign nature of the lesion. The patient remains asymptomatic a year after the surgery.

Discussion: Parathyroid adenoma with prominent lymphocytic may be misdiagnosed as a lymph node at frozen section or cytology smears by an unsuspecting pathologist or cytologist, if the lymphoid component is predominant on the slide examined. Additionally, the morphology could be misinterpreted as a metastasis of parathyroid carcinoma into a peri-thyroidal lymph node. For this distinction, correlation with clinical and radiological findings is crucial and clues to adenoma include presence of a rim of uninvolved parathyroid parenchyma, low mitotic activity (<5%) and positive staining for parafibrin, bcl-2 and MDM2 on immunohistochemistry.

EOSINOPHILIC SOLID AND CYSTIC RENAL CELL CARCINOMA WITH MONOSOMY OF CHROMOSOMES X AND 3

Nassim Saremi^{1,2}, Admire Matsika^{1,3}, Bhuvana Srinivasan^{1,3}

¹Anatomical Pathology Department, Mater Health, South Brisbane, Qld, Australia; ²Pathology Queensland, Central Laboratory, Queensland Health, Herston, Qld, Australia; and ³School of Biomedical Sciences, University of Queensland, Qld, Australia

Introduction: Eosinophilic solid and cystic renal cell carcinoma (ESC-RCC) is a recently described sporadic counterpart to tuberous sclerosis complex (TSC)-associated RCC that is not yet included in the 2016 World Health Organization (WHO) classification of renal neoplasms. The first series was reported in 2016 and, to date, just over 60 cases have been reported in the literature. The tumour is often asymptomatic and almost exclusive to female patients with a median age of 57 years. ESC-RCC constitutes approximately 0.2% of all renal cell carcinomas and most cases reported have shown a better prognosis than clear cell renal cell carcinoma. About 10% of reported cases have metastasised with four of them dying of the malignancy.

Case report: A 63-year-old woman presented with an incidental 40 mm, ill-defined nodule in the lower pole of the right kidney. Partial nephrectomy was performed and gross examination of the tumour showed a well-circumscribed neoplasm with solid-cystic cut surface. Microscopy revealed acinar and nested architecture with the neoplastic cells showing intracytoplasmic stippling ('Leishmania bodies-like') within the deeply eosinophilic cytoplasm. Macro- and micro-cysts in other fields were lined by hobnailed, eosinophilic tumour cells. Immunohistochemically, the neoplasm showed strong and diffuse staining for CK20 and AE1/AE3. CK7 was negative. Karyotyping of tumour cells showed previously unreported monosomy of chromosomes X and 3. The patient remains alive and well, without evidence of disease progression a year after surgery.

CLINICAL UTILITY OF THE TFE3 BREAK-APART FLUORESCENCE IN SITU HYBRIDISATION (FISH) ASSAY IN MALIGNANT TUMOURS

T. H. Lim¹, S. T. A. Lim¹, Y. S. Y. Yeap¹, Y. J. Ng¹, C. H. What¹, K. K. J. Ho¹, S. L. Tien¹, S. Selvarajan²

¹Department of Molecular Pathology, Singapore General Hospital, Singapore; and ²Department of Anatomical Pathology, Singapore General Hospital, Singapore

TFE3 break-apart FISH probe, located on Xp11.23 is designed to detect TFE3 gene rearrangements in many tumours including renal cell carcinomas (RCC) and alveolar soft part sarcoma (ASPS), and some variants of perivascular epithelioid cell neoplasm (PEComa). FISH test may be deployed as an adjunct tool for diagnosis. Generally, the FISH probe will give a typical FISH signal pattern but unusual patterns can also be encountered. We aimed to evaluate the break-apart FISH patterns and incidence of TFE3 gene rearrangements in soft tissue tumours such as RCC and ASPS.

As the morphological assessment may have overlapping features, TFE3 FISH test is a useful ancillary tool in the confirmation or exclusion of differential diagnosis.

MALIGNANT SOLITARY FIBROUS TUMOUR OF UTERINE CERVIX, MIMICKING HIGH GRADE ENDOMETRIAL STROMAL SARCOMA

Pooja Singhal, Raghwa Sharma, Spinderjeet Samra

Tissue Pathology and Diagnostic Oncology, ICPMR, Westmead Hospital, NSW, Australia

Solitary fibrous tumour (SFT) is an uncommon mesenchymal neoplasm of fibroblastic origin which can be found in any organ, however, its occurrence in the female genital tract (FGT) is extremely rare. To date, around 50 cases of SFTs originating in FGT have been reported (as case report or small case series) of which 12 are in the uterus and 5 in the cervix. These are rare tumours and should be considered in the differential diagnosis of spindle cell lesion in the female genital tract when the immunohistochemistry is not supportive of common lesions. We are reporting a case of a malignant SFT involving primarily the cervix and extending into the isthmus uterus, involving the bladder, presenting with recurrence and distant metastasis to the lung and mesentery.

OLFACTORY NEUROBLASTOMA WITH DIVERGENT DIFFERENTIATION

Pooja Singhal, Narinder Singh, Spinderjeet Samra, Hedley Coleman

Tissue Pathology and Diagnostic Oncology, ICPMR, Westmead Hospital, NSW, Australia

Olfactory neuroblastoma (ONB) is an uncommon malignant neoplasm of the nasal cavity that arises from olfactory epithelium. Rare cases of focal histologic changes of divergent cell populations have been reported in ONB. However, these changes are seen mainly in tumours treated with chemo radiation or with distant metastases.