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A RARE CASE OF AN EPITHELIOID HAEMANGIOENDOTHELIOMA PRESENTING IN THE SKULL OF AN 11 YEAR OLD GIRL

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Epithelioid haemangioendothelioma is a rare low grade malignant vascular tumour with an estimated prevalence of less than one in one million. It may occur at any age with a mean age of presentation of 36 years and most commonly affects the liver and lung. Approximately 14% of epithelioid haemangioendotheliomas occur as primary lesions of bone of which the long bones of the extremities are the most common sites of involvement. We present a rare case of an epithelioid haemangioendothelioma involving the skull of an 11 year old girl. The patient initially presented with a protuberant mass in the parietal-occipital region on a background of recurrent headaches. MRI and CT scanning revealed a large partially calcified temporal-parietal-occipital mass in continuity with the calvarium. Biopsy of the lesion was performed which revealed a vasoformative tumour composed of polygonal and spindled cells in a focally chondromyxoid stroma. No multilayering of cells was seen and mitotic activity was inconspicuous. Immunohistochemistry showed co-expression of CD31 and cytokeratin. The patient underwent tumour embolisation and subsequent craniotomy with subtotal tumour resection. The clinical and pathological features of this case will be discussed with a review of the literature.

A RARE CASE OF RENAL LEIOMYOMA – CASE REPORT AND LITERATURE REVIEW

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Introduction: Renal Leiomyomas are rare tumours of the kidney with a prevalence of 4.2–5.2% based on incidental findings at autopsy. They represent 1.5% of benign and 0.3% of overall renal tumours.

Case history: A 47-year-old female presented with 4 weeks history of ongoing right iliac fossa pain, weight loss and nocturnal diarrhoea. An MRI of the abdomen showed a 158×110×110 mm mass arising from the upper pole of the kidney and compressing the adrenal gland. There was no evidence of renal vein invasion or lymph node metastasis. The preoperative diagnosis was primary renal cell carcinoma and the patient proceeded to right radical nephrectomy and adrenalectomy.

Macroscopically, the upper pole of the right kidney showed a well circumscribed mass, 145×95×90 mm, with a tan cream, homogenous, whorled cut surface, arising from the renal capsule. The histology showed a leiomyoma of the kidney, comprising of low grade spindled cells with eosinophilic cytoplasm and rare mitoses. On immunohistochemistry, the spindled cells were

positive for h-caldesmon, colponin and negative for HMB45 and Melan A, thus a diagnosis of leiomyoma was made.

Discussion: Leiomyomas of the kidney mostly arise from the smooth muscle of renal capsule, vessels or renal pelvis. Kidneys are the most common site in the genitourinary tract. The main histological differential is angiolipoma.

PLEOMORPHIC HYALINISING ANGIECTATIC TUMOUR: A CASE REPORT

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Pleomorphic hyalinising angiectatic tumour (PHAT) is a rare non-metastasising soft tissue neoplasm of uncertain lineage usually occurring in subcutaneous tissue of the lower extremities. There is 30–50% rate of recurrence and no metastasis has been reported. Histologically it is characterised by clusters of ectatic thin-walled vessels throughout a sheet-like proliferation of spindle cells. Organising thrombus is frequently present within the vessels and fibroind and hyaline material in the vessel wall is a common finding. We report a case of PHAT in a 79-year-old male who presented with three months history of slow growing painless inguinal lump. He had surgical excision and microscopy showed a circumscribed and unencapsulated tumour composed of scattered thin-walled ectatic vessels. The vessels contained intra and extravascular fibrin associated with perivascular hyalinisation. The immunohistochemistry showed diffuse positivity for CD34 and vimentin. S100 was negative. The six month follow up showed no evidence of local recurrence.

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SINONASAL EPITHELIAL-MYOEPITHELIAL CARCINOMA: A CASE REPORT

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Epithelial-myoepithelial carcinoma is a rare tumour representing around 1% of the salivary gland tumours. We present a rare case of epithelial-myoepithelial carcinoma of the sinonasal cavity in an 82-year-old female. She presented with intermittent nasal obstruction and nasal bleeding. The CT scan of the sinuses revealed a polypoid lesion in the posterior nasal space 16 mm in diameter. Microscopy

showed a polyp composed of numerous ductal structures lined by a double layer of cells comprising an inner layer of bland ductal epithelium and an outer layer of myoepithelial cells.

To the best of our knowledge twelve cases of epithelial myoepithelial carcinoma of the sinonasal region have been reported in the literature, including the present case. In review of the literature, most cases, including the case presented, were disease free at the 10–40 month post-operative follow up. This neoplasm is a typically a low grade malignant neoplasm, but in rare cases the tumour can metastasise to distant sites.

In conclusion, we illustrate a rare case of epithelial-myoepithelial carcinoma of the sinonasal cavity, a morphologically characteristic tumour in an unusual site.

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A RARE VASCULAR TUMOUR OF THE OVARY: A CASE REPORT

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Case history: A 72-year-old female presented with a 2 year history of a right adnexal mass. Right salpingo-oophorectomy revealed a 40 mm diameter, circumscribed mass with a spongy, haemorrhagic cut surface. Microscopically, the mass was composed of small to medium sized blood vessels within an oedematous and fibrotic stroma, with islands of anastomosing vascular channels lined by atypical endothelial cells showing moderate nuclear pleomorphism and occasional mitotic figures. Focally within some of the anastomosing vascular channels a few haematopoietic cells were seen. The endothelial cells were positive for vascular markers CD31 and CD34. Small nests of luteinised stromal cells, showing inhibin positivity were noted at the periphery of the tumour, consistent with residual ovarian parenchyma.

Discussion: We present this as a case of primary ovarian angiosarcoma. Primary ovarian angiosarcoma is a rare entity. Angiosarcomas may display a wide range of morphological features ranging from well (low grade) to poorly (high grade) differentiated. Poorly differentiated lesions show areas of sarcomatous differentiation with significant nuclear atypia, brisk mitotic activity, necrosis and destructive growth. Differentiating between benign vascular tumours and well-differentiated angiosarcomas may be challenging.

GENOMICS VS PLASIAS: GENETIC EXPLANATIONS FOR THE HISTOPATHOLOGIC FEATURES OF TUMOURS

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Tumours result from genomic disturbances in somatic cells. In genetics, genotypic changes are called ‘mutations’ and phenotypic changes are called abnormal ‘traits’. However, genetic terms are not used for the histopathology of tumours. Instead, 19th century words such as ‘(de)differentiation’, ‘neoplasia’, ‘dysplasia’ and ‘anaplasia’¹ are employed.

Nevertheless, histopathologic phenomena of tumours can be analysed in genetic terms.² First, tumours generally exhibit excessive growth together with other traits, particularly altered specialisation, abnormal cellular spatial arrangements, and invasion and metastasis. The phenomenon of association of traits is known as ‘linkage’. Second, linkage of traits is usually a manifestation of proximity of relevant genes in complex loci. Hence third, if in a somatic cell mutation causes linked traits, the relevant genomic lesion probably involves a complex locus. That is: a larger lesion than a single gene, or in a particular nucleotide in an exon of a particular gene. Several mechanisms for mutations of entire complex loci are known.

Lastly, additional phenomena of tumour types, for example their specificity to kinds of parent cell, and their variabilities, can be explained by genomic mechanisms.

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METASTATIC THYMOMA – A RARE PRESENTATION AS A DURAL-BASED SPINDLE CELL LESION IN THE CRANIAL CAVITY

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Intracranial dural-based spindle cell lesions are most commonly meningiomas, solitary fibrous tumours and haemangiopericytomas. Other primary and metastatic spindle lesions are rare. Metastatic thymoma presenting as an intracranial lesion is exceptional and poses a diagnostic challenge.

We report the case of a 75-year-old Indonesian female who presented with a solitary 2.8 cm dural-based mass in the left parieto-occipital region. Excision showed a cellular spindle cell tumour with a prominent haemangiopericytoma-like vascular pattern. Frequent rosettes were seen. The tumour cells showed bland bipolar nuclei, fine chromatin, inconspicuous nucleoli and rare mitoses. Dispersed small lymphocytes were present. The spindled tumour cells were positive for cytokeratins AE1/3 and Cam5.2, PAX8 and CD20. Terminal deoxynucleotidyl transferase (TdT) and CD5 showed immature T-lymphocytes within the tumour. The spindle cells were negative for synaptophysin, chromogranin, epithelial membrane antigen, CD34, and TTF1. The features were highly suggestive of metastatic thymoma with Type A or Type AB features. Computed tomography showed a large anterior mediastinal mass, consistent with a primary mediastinal thymoma.

Thymic epithelial tumours with intracranial metastases are unusual and most are thymic carcinomas. Thymomas with intracranial metastases are rare. The characteristic morphology and immunoperoxidase profile of thymomas are of value in the differential diagnostic work-up.