MELANOTIC NEUROECTODERMAL TUMOUR OF INFANCY: UNUSUAL HISTOLOGICAL FEATURES IN A RARE TUMOUR

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Description: Melanotic neuroectodermal tumour of infancy (MNTI) is a rare, locally aggressive but usually benign tumour typically arising in the bones of the head and neck in infants and young children. Histologically, MNTI is a biphasic tumour of neural crest origin, composed of neuroblastic and pigmented epithelial components.

Findings: We present a case arising from the skull of an 18-month-old boy requiring extensive neurosurgical resection. Histological examination confirmed MNTI but with unusual features, viz: (a) the 'small-round-blue-cell' neuroblastic component, usually bland with rare mitoses, showed morphological features suggestive of malignancy, including abundant mitoses (up to 67/10 hpf) and karyorrhexis, foci of necrosis, and a high Ki67 proliferative index (~40%); and (b) the neuroblastic component, in addition to showing typical 'small-round-blue-cell' morphology, included large areas of neuropil containing neuroblastic cells showing varying degrees of ganglionic differentiation.

Discussion: Less than 500 MNTIs are reported in the published literature; only 7% are malignant, the latter based on the presence of metastatic disease.¹ There are currently no defined histological criteria for malignant MNTI,² and there was no evidence of metastatic disease in our patient. Ganglionic differentiation is described in MNTI^{3,4} but, to our knowledge, this usually constitutes a minor component.

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SYNCHRONOUS SESSILE SERRATED ADENOMA AND GOBLET CELL CARCINOID IN THE APPENDIX: A CASE REPORT AND LITERATURE REVIEW

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Introduction: Appendiceal neoplasms are rare, with the majority being adenomas and neuroendocrine tumours. Sessile serrated adenoma (SSA) is associated with the serrated pathway of neoplasia. Less than 50 cases of SSA in the appendix have been reported. Goblet cell carcinoid (GCC) is almost exclusively found in the appendix and has a worse prognosis than other carcinoid variants. We present an interesting case of synchronous SSA and GCC within the appendix.

Case report: A 49-year-old male presented with acute right iliac fossa pain. The appendicectomy specimen measured 55 mm in

length and 12 mm in diameter. Histopathology showed irregular serrated glands, cryptal branching and dilated 'L' shaped crypt bases in the mucosa. There was a second infiltrating tumour within the muscularis propria composed of discrete clusters of goblet like cells. Immunohistochemically, the SSA and GCC showed positive staining for AE1/3, CK20 and polyclonal CEA. The goblet cell tumour was also positive for synaptophysin and CD56.

Discussion: This case of two rare synchronous appendiceal neoplasms highlights the importance of adequate sampling. Although SSAs are rarely described in the appendix, it should be recognised due to a higher risk of malignant transformation. It is further important to differentiate appendiceal goblet cell carcinoma from the more aggressive mucinous adenocarcinoma.

MALIGNANT PROLIFERATING TRICHILEMMAL TUMOUR: A CASE REPORT AND LITERATURE REVIEW

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Introduction: Malignant proliferating trichilemmal tumour (MPTT) is a rare neoplasm with less than 50 cases documented in the literature. It is more common on the scalp and in females. We report a case of MPTT and review the histopathology, differential diagnoses and management.

Case report: An 84-year-old male presented with a tan subcutaneous scalp nodule and underwent an elliptical skin excision (30×22×15 mm). Histopathology showed a well circumscribed dermal mass with a multi-loculated cystic appearance. It was lined by squamous epithelium with an absent granular layer showing evidence of abrupt keratinisation. The keratinocytes showed marked cytologic atypia. Numerous mitoses including atypical mitotic figures were seen. The lesion extended to the margins. The morphological features were consistent with a MPTT.

Discussion: It is important to recognise this rare entity as MPTT tends to recur and metastasise more frequently than the histological mimics of squamous cell carcinoma (SCC) and trichilemmal carcinoma (TLC). Features favouring MPTT over SCC include presence of trichilemmal type keratinisation and the lack of a premalignant epidermal lesion. TLC is distinguished by a lobular proliferation of clear cells and continuity with the epidermis. Wide excision of MPTT with an adequate clearance margin and close follow up are the mainstays of treatment.

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ATYPICAL MICROGLANDULAR ADENOSIS POST-NEOADJUVANT CHEMOTHERAPY

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Aim/Introduction: The progression from microglandular adenosis (MGA) to atypical microglandular adenosis (AMGA) then to ductal carcinoma has been documented in case reports and series.^{1–6} We report here the first case of AMGA and residual invasive ductal carcinoma in the context of post-neoadjuvant chemotherapy.

Case/Methods/Results: A 57-year-old female presented with a 70×55 mm left breast lump. Her medical history included hysterectomy and bilateral oophorectomy for dysfunctional uterine bleeding and fibroid uterus. A breast needle core biopsy showed invasive carcinoma of no special type with areas of high grade ductal carcinoma *in situ*. Focal areas of MGA were noted. The tumour was ER–, HER2– and PR+(30%). A left axillary lymph node fine needle aspirate showed cohesive fragments of malignant epithelial cells consistent with metastatic carcinoma. The patient was initially treated with neoadjuvant chemotherapy (adriamycin, cyclophosphamide and docetaxel). The breast lump reduced in size to 10 mm in maximum dimension. Following review with a breast surgeon she underwent a left total mastectomy and axillary clearance.

Macroscopic examination of the breast specimen revealed an ill-defined nodular area measuring 80 mm in maximum dimension. Microscopically there was an atypical glandular proliferation with glands arranged haphazardly in fibrous stroma and fat. The glands showed a range of proliferative changes with areas of solid and cribriform architecture. These changes were consistent with a spectrum ranging from MGA to AMGA and ductal carcinoma *in situ*. The glands were devoid of a myoepithelial layer on immunoperoxidase stains. A 12 mm foci of invasive carcinoma in the form of nests and cords without tubule formation was also present which was similar to the initial core biopsy. All seventeen lymph nodes were negative for malignancy.

Discussion: Microglandular adenosis is an uncommon benign proliferative glandular lesion. Case studies have shown that carcinoma arises in cases of MGA^{1,3–6} and there is molecular evidence of its progression to malignancy.⁷ What is unclear are the effects of chemotherapy on MGA. We report here the first case of atypical MGA in the context of post-neoadjuvant chemotherapy.

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CUTANEOUS ROSAI DORFMAN DISEASE: A CASE REPORT AND LITERATURE REVIEW

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Purpose: Rosai-Dorfman disease (RDD) is a benign, multi-organ, non-Langerhans cell histiocytosis that was described as a distinct entity by Rosai and Dorfman¹ in 1969. Purely cutaneous RDD (C-RDD) has been described, but is considered a rare occurrence.² The aim of this study was to present a case of C-RDD and perform a literature review on the subject.

Material: A 62-year-old female presented with a single 7 mm nodule on her mid back. The lesion was completely excised.

Result and conclusion: The sections show a dermal based tumour composed of histiocytes admixed with a prominent mixed inflammatory infiltrate. Some of the histiocytes engulf inflammatory cells (emperipolesis). The lesional cells express S100 and CD68 and are negative for CD1a, consistent with C-RDD. Only a single cutaneous lesion has been detected and there is no systemic involvement.

In the literature, C-RDD is a histiocytosis of uncertain histogenesis, a distinct entity with no systemic involvement.³ Usually cutaneous lesions do not require any treatment besides excision as they show a self-limiting course and tend to regress spontaneously.

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SOLITARY FIBROUS TUMOUR OF THE PROSTATE: ASSESSMENT OF MALIGNANCY POST TREATMENT

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Aim: We present a case of solitary fibrous tumour (SFT) in an unusual site (prostate) in a patient who has undergone neoadjuvant radiotherapy.

Case summary: A 54-year-old man presented with renal colic symptoms and was incidentally found to have a 6 cm mass posterior to his prostate. Core biopsy revealed a mixed spindle cell lesion raising the possibility of a sarcomatoid tumour. The patient was treated with neo-adjuvant radiotherapy and subsequently a pelvic exenteration was performed. The histopathology of the resection specimen confirmed SFT.

Discussion: Solitary fibrous tumour is an uncommon fibroblastic mesenchymal tumour that was initially described in pleura, but can occur in any site. Due to the relative rarity of SFT in the prostate, there is still a limited understanding of the clinical course of this tumour. Several general criteria have been