TEST YOURSELF: ANSWER

Diagnosis: subcutaneous myxopapillary ependymoma

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Discussion

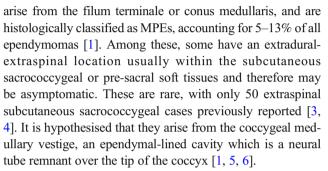
MRI demonstrated a $6.7 \times 6 \times 6$ cm lobular fluid signal intensity mass arising from the midline natal cleft abutting the coccyx, with multiple hypointense internal septa (Fig. 1). The mass was well-circumscribed and confined to the intergluteal fold subcutaneous tissues. The distal thecal sac, cauda equina and conus were normal. An ultrasound-guided biopsy was performed to exclude a soft tissue sarcoma.

Tumour resection histology showed a pseudopapillary architecture with a central core composed of PAS-positive myxohyaline matrix, and small vessels around which relatively uniform cuboidal to columnar cells were arranged. Mild cytological atypia was noted with low mitotic count. Characteristic immunoprofile included S100 and GFAP cytoplasmic positivity, while cytokeratin was negative (Fig. 2). A diagnosis of subcutaneous myxopapillary ependymoma (MPE) was made.

Ependymomas are common spinal tumours, with an average age of 35 years and a male predilection [1, 2]. A minority

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Imaging features can be non-specific but a pure midline location suggests a tumour of congenital origin [7]. Subcutaneous MPEs usually present as large well-defined and encapsulated masses on imaging [1, 4]. Radiographs demonstrate a well-defined, ovoid lesion which may contain punctate calcific foci [4]. CT shows a well-circumscribed soft tissue attenuation mass in the retro-sacral region with possible bony remodelling owing to their chronic slow growth [8]. MRI demonstrates a lobular mass hypo/isointense to the muscle on T1W, hyperintense on T2W, with scattered hypointense fibrovascular septa and a surrounding hypointense fibrous apsule [8]. Although not used in our case, MPEs usually isplay heterogeneous contrast enhancement due to their erivascular mucinous stroma mixed with fibrous tissue. Areas of necrosis and haemorrhage tends to favour malignany [8]. Imaging homogeneity as in our case implies a benign esion.

Most MPEs are classified as WHO grade I and are gener-Ily non-aggressive with a favourable prognosis [1]. However, 5-35% can metastasize to regional subcutaneous soft tissues, one, lungs and pleura via haematogenous spread [5]. Given he relatively non-specific imaging features, subcutaneous APE should be included in the differential diagnosis of perioccygeal masses along with sacrococcygeal teratoma, pilonial cyst and neurogenic tumours [4, 8]. Patients presenting with such masses should be investigated with MRI followed y image-guided biopsy [8]. Once the diagnosis has been established, the remainder of the CNS should be imaged to exclude other ependymomas. Surgical excision is the



preferred treatment with a 98% 5-year survival rate following complete resection. However, there is a 41% chance of local recurrence especially with incomplete excision [5]. Radiotherapy is reserved for cases with incomplete margins, and chemotherapy may be used in the setting of metastatic disease [5].

Compliance with ethical standards

Conflict of interest The authors declare they have no conflict of interest.

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