Skeletal Radiology (2020) 49:1175–1176
https://doi.org/10.1007/s00256-020-03408-w

TEST YOURSELF: ANSWER

Diagnosis: subcutaneous myxopapillary ependymoma

Bashiar Thejeel[1] · Ramanan Rajakulasingam[2] · Petra Balogh[3] · Asif Saifuddin[2]

Published online: 14 March 2020

Discussion

MRI demonstrated a 6.7 × 6 × 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 arise from the filum terminale or conus medullaris, and are histologically classified as MPEs, accounting for 5–13% of all ependymomas [1]. Among these, some have an extradural-extraspinal location usually within the subcutaneous sacrococcygeal or pre-sacral soft tissues and therefore may be asymptomatic. These are rare, with only 50 extraspinal subcutaneous sacrococcygeal cases previously reported [3, 4]. It is hypothesised that they arise from the coccygeal medullary vestige, an ependymal-lined cavity which is a neural tube remnant over the tip of the coccyx [1, 5, 6].

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 capsule [8]. Although not used in our case, MPEs usually display heterogeneous contrast enhancement due to their perivascular mucinous stroma mixed with fibrous tissue. Areas of necrosis and haemorrhage tends to favour malignancy [8]. Imaging homogeneity as in our case implies a benign lesion.

Most MPEs are classified as WHO grade I and are generally non-aggressive with a favourable prognosis [1]. However, 15–35% can metastasize to regional subcutaneous soft tissues, bone, lungs and pleura via haematogenous spread [5]. Given the relatively non-specific imaging features, subcutaneous MPE should be included in the differential diagnosis of pericoccygeal masses along with sacrococcygeal teratoma, pilonidal cyst and neurogenic tumours [4, 8]. Patients presenting with such masses should be investigated with MRI followed by 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

The case presentation can be found at doi:10.1007/s00256-020-03407-x

✉ Bashiar Thejeel
thejeel@ualberta.ca

Ramanan Rajakulasingam
ramanan.rajakulasingam1@nhs.net

Petra Balogh
petra.balogh@nhs.net

Asif Saifuddin
asif.saifuddin@nhs.net

[1] Department of Radiology and Diagnostic Imaging, University of Alberta, 16 & 85 Ave, Edmonton, AB T6G 2R3, Canada

[2] Department of Radiology, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore HA7 4LP, UK

[3] Department of Histopathology, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore HA7 4LP, UK

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.

References

1. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation. Radiographics. 2000;20(6):1721–49.
2. Ferrante L, Mastronardi L, Celli P, Lunardi P, Acqui M, Fortuna A. Intramedullary spinal cord ependymomas? A study of 45 cases with long-term follow-up. Acta Neurochir. 1992;119(1–4):74–9.
3. Zaidi SN, AlKhalidi H, Al-Rikabi AC. Myxopapillary ependymoma masquerading as subcutaneous saccroccygeal non-healing ulcer: case report. Ann Saudi Med. 2014;34(3):262–4.
4. Chung JY, Lee SK, Yang KH, Song MK. Subcutaneous sacrococcygeal myxopapillary ependymoma. Am J Neuroradiol. 1999;20(2):344.
5. Schiavello E, Biassoni V, Antonelli M, Modena P, Cesaro S, Pierani P, et al. Pediatric extraspinal sacrococcygeal ependymoma (ESE): an Italian AIEOP experience of six cases and literature review. Childs Nerv Syst. 2018;34(7):1291–8.
6. Helwig EB, Stern JB. Subcutaneous sacrococcygeal myxopapillary ependymoma: a clinicopathologic study of 32 cases. Am J Clin Pathol. 1984;81(2):156–61.
7. Biagini R, Demitri S, Orsini U, Bibiloni J, Briccoli A, Bertoni F. Osteolytic extra-axial sacral myxopapillary. Skelet Radiol. 1999;28:584–9.
8. Wippold F, Smirniotopoulos JG, Moran CJ, Suojanen JN, Vollmer DG. MR imaging of myxopapillary ependymoma: findings and value to determine extent of tumor and its relation to intraspinal structures. Am J Roentgenol. 1995;165(5):1263–7.