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TEST YOURSELF: ANSWER

Test Yourself Answer: A 36-year-old male presenting with a painful swollen right thigh for several months—no previous history of trauma

Sherif Elsobky[1] · Ramanan Rajakulasingam[2] · Monika Madrova[3] · Daniel Lindsay[3] · Asif Saifuddin[2]

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Answer

Extraskeletal osteosarcoma (ESO)

Discussion

The plain radiograph (Fig. 1) demonstrates a calcified soft tissue mass in the right distal thigh. MRI (Fig. 2) confirms an isointense T1-weighted and hyperintense STIR signal intensity (SI) lesion within rectus femoris with haemorrhagic and necrotic areas. There is mild peri-lesional oedema with hypointense foci scattered throughout, better appreciated on CT (Fig. 3). Follow-up CT showed increased size of both the soft tissue component and amorphous calcific foci. Ultrasound-guided needle biopsy revealed tumour composed of pleomorphic epithelioid-to-spindle cells with lace-like osteoid deposition, focal chondroblastic differentiation and areas of necrosis, consistent with grade 3 ESO (Fig. 4). Despite local tumour excision, the patient developed osteoblastic lung metastases on follow-up and passed away 2 years after initial presentation.

ESO is a mesenchymal extra-osseous soft tissue tumour accounting for < 1% of all soft tissue sarcomas [1]. It is commonly located in the lower extremity deep to the fascia, and affects patients aged 47–61 years, being very rare in teenagers unlike conventional intra-medullary OS [1]. There is no clear link to trauma, but previous radiation exposure has been reported in up to 13% of cases [2]. ESO is composed of neoplastic cells that produce osteoid, neoplastic bone or chondroid matrix. It is histologically indistinguishable from conventional intra-medullary OS [1].

Plain radiographs show a soft tissue density mass with varying degrees of matrix mineralisation, which may be absent in 50% of cases [1]. Central calcification is best visualised on CT but without predilection for a peripheral 'zoning pattern'. ESO usually displays isointense and heterogeneously hyperintense SI on T1- and T2-weighted sequences respectively, commonly with haemorrhage and necrosis [2]. Rare features include large cystic components within a solid area at the lesion periphery [3]. High-grade lesions are larger with prominent central necrosis compared to low-grade lesions, the latter being very rare with only a handful of low-grade ESO lesions published in literature.

ESO is treated with wide local excision, with most patients having radiotherapy. Currently, there is no definitive role for chemotherapy. The overall prognosis is poor, with most literature quoting > 50% of patients dying within 2–3 years [1]. No definite prognostic factors have been identified, other than a single study citing a 5-year survival ranging from 30 to 65% for lesions over 5 cm [4]. Metastases and local recurrence are high, with both occurring in up to 90% of cases. Unlike conventional intra-medullary OS, ESO lung metastases may not show mineralisation [1].

ESO is a rare sarcoma which may be mistaken for other soft tissue osteogenic lesions. Central calcification, necrosis, haemorrhage and variable enhancing soft tissue components

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✉ Ramanan Rajakulasingam
ramanan.rajakulasingam1@nhs.net

Sherif Elsobky
sherif.elsobky@nhs.net

Monika Madrova
monika.madrova@nhs.net

Daniel Lindsay
daniel.lindsay@nhs.net

Asif Saifuddin
asif.saifuddin@nhs.net

[1] Department of Radiology, Royal Free NHS Foundation Trust, Pond Street, London NW3 2QG, UK

[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

on imaging favour ESO [2]. Crucially, the ossification is randomly distributed and not related to the peripheral capsule as in myositis ossificans. Synovial sarcoma can usually be differentiated by the absence of central osteoid production, but variants with extensive osteoid/bone formation have been reported [5]. The typical biphasic pattern of synovial sarcoma on MRI may also be absent in smaller lesions. If there is initial diagnostic uncertainty, close serial CT and MR imaging is recommended to assess lesion evolution, which should prompt biopsy.

Declarations

Conflict of interest The authors declare no competing interests.

References

1. Mc Auley G, Jagannathan J, O'Regan K, Krajewski KM, Hornick JL, Butrynski J, Ramaiya N. Extraskeletal osteosarcoma: spectrum of imaging findings. AJR Am J Roentgenol. 2012;198(1):W31–7.
2. Roller LA, Chebib I, Bredella MA, Chang CY. Clinical, radiological, and pathological features of extraskeletal osteosarcoma. Skeletal Radiol. 2018;47(9):1213–20.
3. Araki Y, Yamamoto N, Hayashi K, Takeuchi A, Miwa S, Igarashi K, Taniguchi Y, Yonezawa H, Morinaga S, Nojima T, Nojima T, et al. Cystic extraskeletal osteosarcoma: three case reports and review of the literature. Mol Clin Oncol. 2020;12:468–74.
4. Bane BL, Evans HL, Ro JY, Carrasco CH, Grignon DJ, Benjamin RS, Ayala AG. Extraskeletal osteosarcoma. A clinicopathologic review of 26 cases. Cancer. 1990; 65(12):2762–70
5. Milchgrub S, Ghandur-Mnaymneh L, Dorfman HD, Albores-Saavedra J. Synovial sarcoma with extensive osteoid and bone formation. Am J Surg Pathol. 1993;17(4):357–63.