Case 13960

Eurorad

Extraskeletal Ewing sarcoma

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Section: Musculoskeletal system
Area of Interest: Musculoskeletal soft tissue
Procedure: Diagnostic procedure
Imaging Technique: Ultrasound
Imaging Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 40 years, female

Clinical History:

A 40-year-old woman with a known osteochondroma in the left distal tibial metaphysis consulted for a growing mass in the same ankle, with tenderness and posterior tibial nerve compression symptoms.

Imaging Findings:

Radiographs (Fig. 1) showed the osteochondroma and posterior noncalcified soft-tissue mass in the ankle. An ultrasound (Fig. 2) was performed revealing a solid soft-tissue mass, in close relationship with the posterior tibial nerve.
MRI (Fig. 3) was performed showing an oval lesion within the Kager fat pad with intermediate to high signal intensity on T1W images and heterogeneous signal intensity on T2W images, with areas of both intermediate and high signal intensity. No high-flow vascular channels were noted. The mass showed diffuse contrast-enhancement. The lesion was adjacent to the posterior tibial cortex but not to the osteochondroma. There were no signs of medullary invasion. The posterior tibial nerve could be seen apparently entering the mass. This last feature suggested the possible diagnosis of a neurogenic tumour.
Percutaneous biopsy of the lesion was performed and anatomopathological study revealed findings for an extraskeletal Ewing sarcoma.

Discussion:

Extraskeletal Ewing sarcoma (EES) is a highly malignant tumour, likely of neuroectodermal origin, included in the Ewing sarcoma family of tumours, which share a translocation of the long arms of chromosomes 11 and 22 (q24;q12) [1].
When compared with its osseous counterpart, EES is relatively rare and usually affects young patients (mean age is 20 years) [1].
Tumours are often large at diagnosis and rapidly growing. Clinical presentation depends on the location of the lesion, which most commonly affects the paravertebral region and the lower extremities. Metastases are common at presentation, lung and bone being the most frequent sites [2].
Histologically, the tumour consists of undifferentiated small round blue cells, that usually contain glycogen. The tumour can be distinguished from other small round cell tumours by immunohistochemical and cytogenetic or molecular analysis [3].
Imaging presentation is that of a nonspecific soft-tissue mass with prominent contrast enhancement. Large tumours usually show heterogeneous signal intensity on MR images, due to haemorrhage and necrosis. Tumours are usually

hyperintense on T2W images with or without intermediate signal intensity areas, as a result of the high cellularity. Fluid levels might also be seen due to haemorrhage. An imaging feature that is nonspecific but can assist in diagnosis is the presence of serpentine high-flow vascular channels [4]. Surface osseous changes (cortical erosion or periosteal reaction), without evidence of marrow space invasion, have also been described in the adjacent bone [4]. Calcification may be present but is not a typical feature [2].
Treatment usually combines chemotherapy and surgery, with or without radiation therapy [1].
In conclusion, EES usually affects young patients and reveals an imaging appearance of a nonspecific soft-tissue mass. Although nonspecific, the presence of serpentine high-flow vascular channels is a radiologic feature that can suggest this diagnosis.
Differential Diagnosis List: Extraskeletal Ewing sarcoma, Neurogenic tumour, Vascular lesion (haemangioma, haemangiopericytoma...), Synovial sarcoma, Lymphoma, Chondrosarcoma, Undifferentiated pleomorphic sarcoma

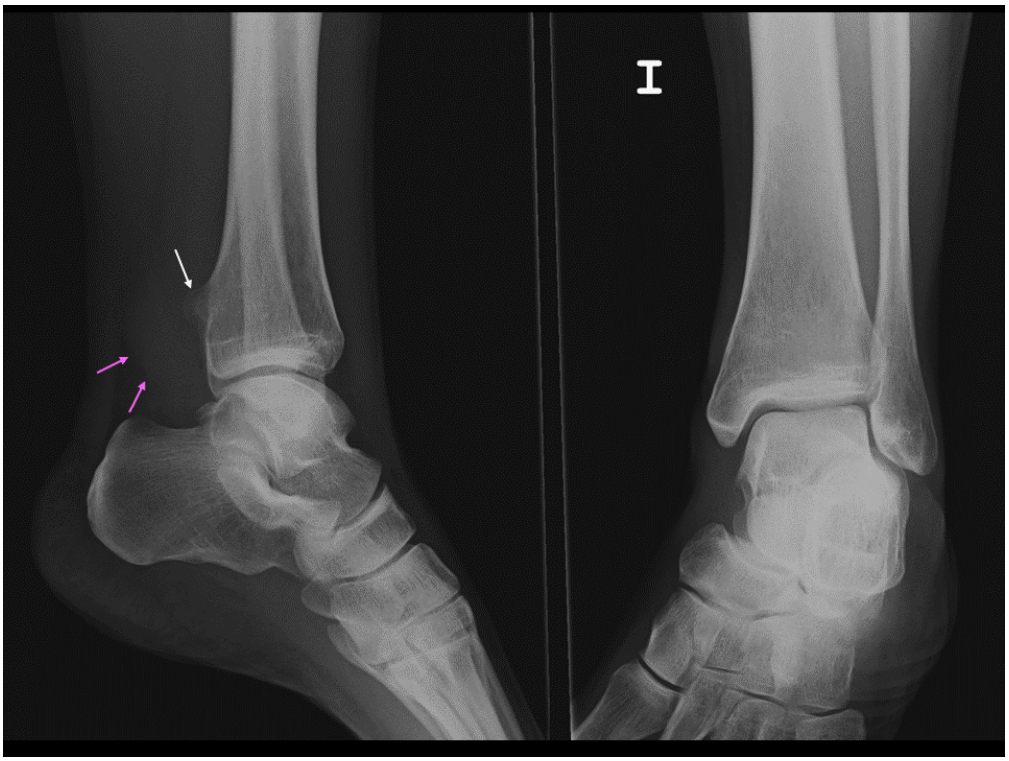
Final Diagnosis: Extraskeletal Ewing sarcoma

References:

Murphey MD, Senchak LT, Mambalam PK et-al (2013) From the radiologic pathology archives: ewing sarcoma family of tumors: radiologic-pathologic correlation. Radiographics 33 (3): 803-31 (PMID:23674776)
Javery O, Krajewski K, O'Regan K et-al (2011) A to Z of extraskeletal Ewing sarcoma family of tumors in adults: imaging features of primary disease, metastatic patterns, and treatment responses. AJR 197(6):W1015-22 (PMID: 22109315)
Tsokos M, Alaggio RD, Dehner LP, Dickman PS (2012) Ewing sarcoma/peripheral primitive neuroectoder¬mal tumor and related tumors. Pediatr Dev Pathol 15(1 suppl):108–126. (PMID:22420726)
Robbin MR, Murphey MD, Jelinek JS, Temple HT (1998) Imaging of soft tissue Ewing sarcoma and primitive neuroectodermal tumor [abstr]. Radiology 209(P):311

Figure 1

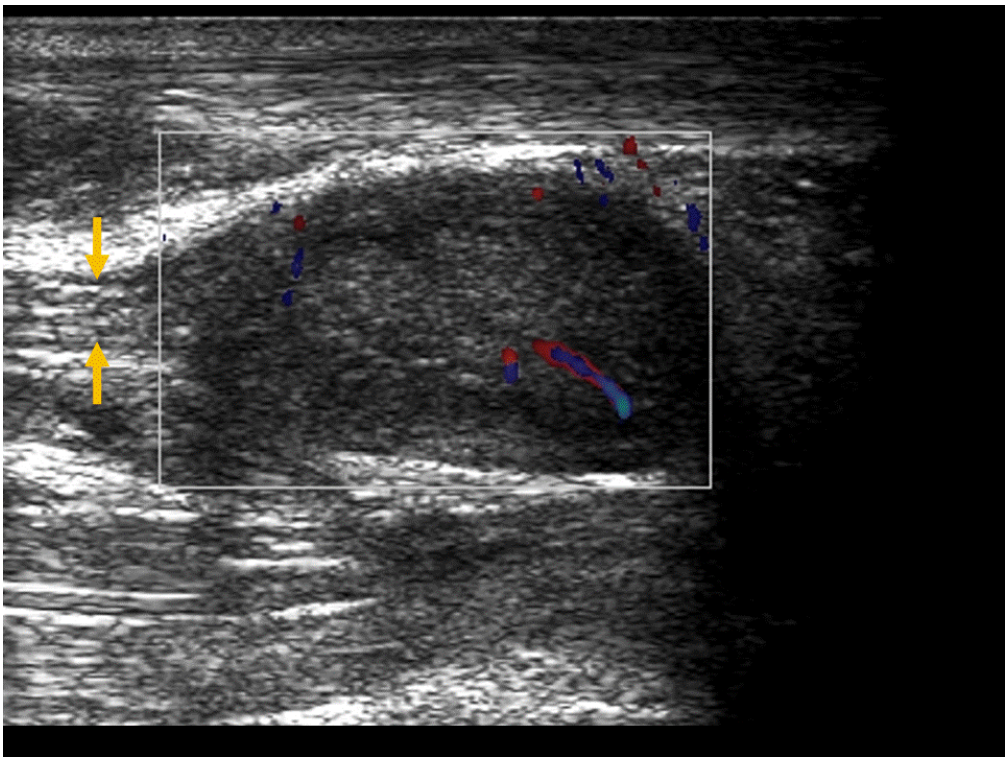
a



Description: Radiographs show an osteochondroma (white arrow) and a soft-tissue mass, not seen in the AP projection, obliterating the Kager fat pad (pink arrows). **Origin:** Complejo Hospitalario de Navarra

Figure 2

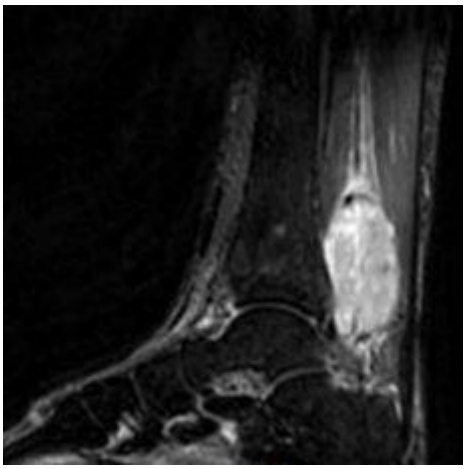
a



Description: US reveals an hypoechoic oval soft-tissue mass with internal vascularization at Colour Doppler US. Posterior tibial nerve can be seen apparently entering the lesion (arrows). **Origin:** Complejo Hospitalario de Navarra

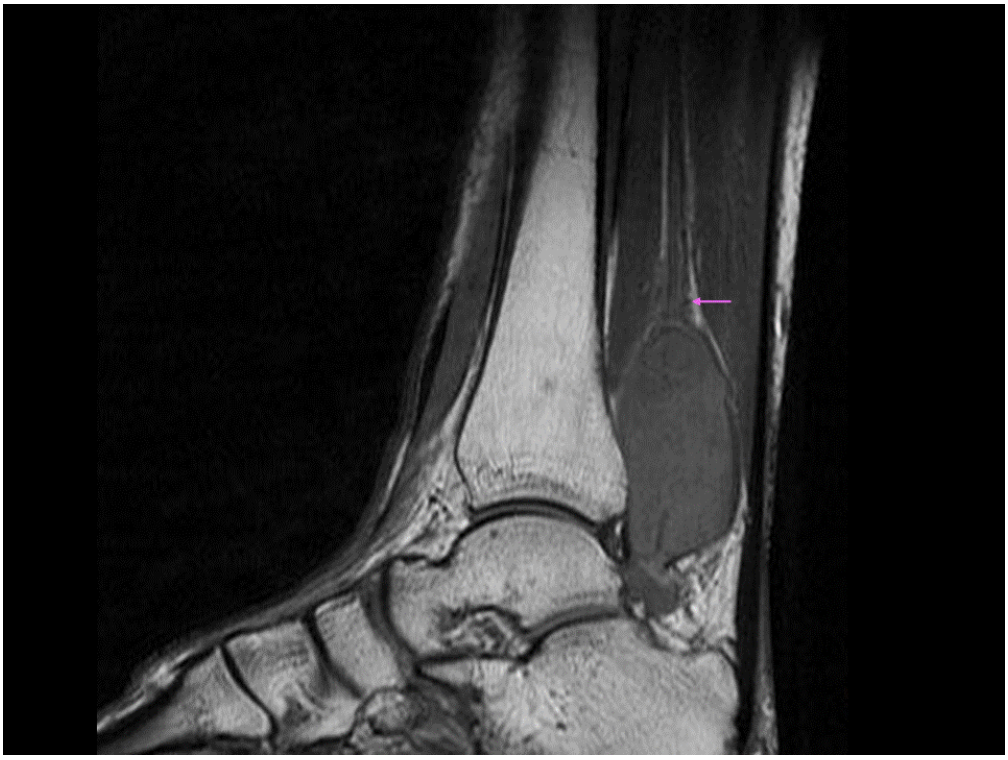
Figure 3

a



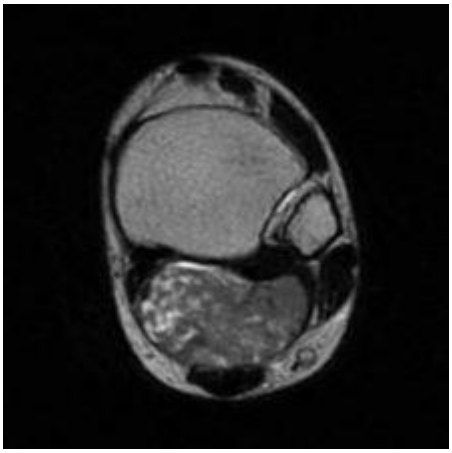
Description: STIR image demonstrates a hyperintense lesion. **Origin:** Complejo Hospitalario de Navarra

b



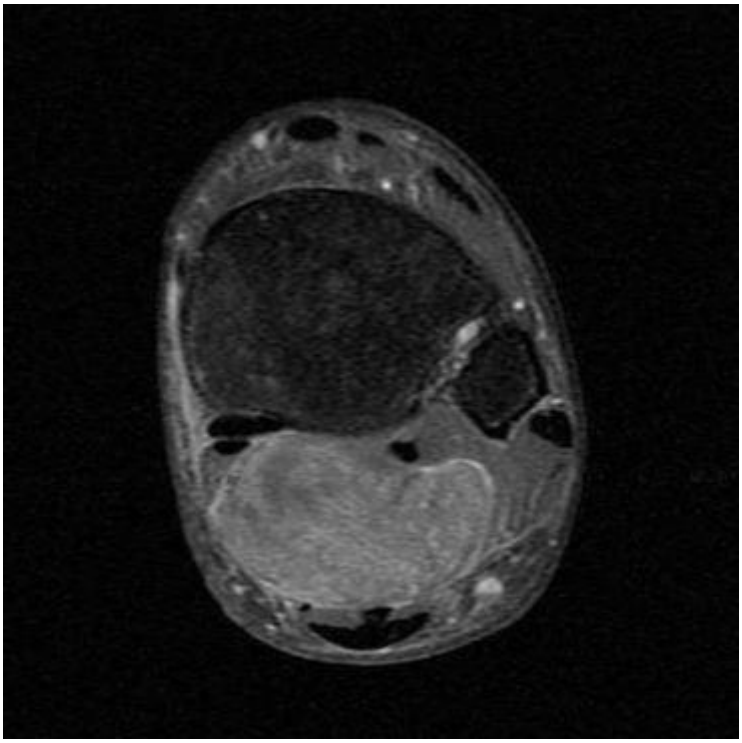
Description: T1W image reveals an oval mass obliterating the Kager fat pad. The mass demonstrates intermediate to high signal intensity. The posterior tibial nerve can be seen apparently entering the lesion (arrow). **Origin:** Complejo Hospitalario de Navarra

c



Description: T2W image reveals a heterogeneous signal intensity mass with areas of both intermediate and high signal intensity. **Origin:** Complejo Hospitalario de Navarra

d



Description: T1FSW image after contrast administration: the mass demonstrates diffuse enhancement, slightly heterogeneous. **Origin:** Complejo Hospitalario de Navarra