Case 14564

Eurorad

An unusual cause of bone lesions in paediatric age

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Section: Musculoskeletal system
Area of Interest: Haematologic Musculoskeletal bone Paediatric
Procedure: Contrast agent-intravenous
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Haematologic diseases Lymphoma Neoplasia Case Type: Clinical Cases
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Patient: 14 years, female

Clinical History:

A 14-year-old female patient was sent to our institution with a history of recurrent pain and swelling in the right knee.
Imaging Findings:

Plain radiography of the right femur showed an osteolytic lesion in the distal femoral metadiaphysis, with aggressive features, such as a permeative pattern and lamellated periosteal reaction (Fig. 1).

An MRI was performed in order to access the bone marrow and soft tissue extent of the lesion. It revealed multiple areas of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images within the distal femoral metadiaphysis, with physeal extension and interruption of the physeal scar. Cortical circumferential thickening, lamellated periosteal reaction and peri-tumoral oedema without evidence of soft-tissue mass was also present (Fig. 2 and 3). Sequences after contrast administration showed multiple areas of enhancement within the lesion (Fig. 4).

Surgical biopsy made the diagnosis of large B-cell lymphoma.

A bone scintigraphy was performed to exclude multifocal bone lesions. CT of the neck, chest, abdomen and pelvis excluded other sites of possible primary lesions and lymphadenopathies.
Discussion:

Primary bone lymphoma (PBL) was initially defined as lymphoma occurring in an osseous site with no evidence of disease elsewhere for at least 6 months after diagnosis [1].

PBL is a rare condition that accounts for less than 5% of all primary bone tumours [2].

PBL can occur in all ages, with a peak prevalence among patients in the 6th to 7th decade of life. It is rare in the paediatric group, especially in patients younger than 10 years [2]. It occurs more often in male children (ratio 6:1) [3]. The femoral metadiaphysis is the most common site of involvement [2]. Others sites include pelvis, humerus skull, tibia and vertebras [4].

Clinically, PBL manifests with insidious and intermittent bone pain that can persist for months [5]. Other symptoms

include local swelling and a palpable mass.

The radiographic appearance is not specific and varies from near normal appearing bone, to a wide spectrum of findings. The most common appearance is a lytic-destructive pattern (more than 70%), permeative or moth-eaten, and occasionally, with focal lytic areas with well-defined margins. Others findings: periosteal reaction (see in 60%), lamellated, layered or broken, cortical destruction, pathologic fractures, sequestra and soft tissues masses [2]. A mixed lytic/sclerotic pattern has also been described in lymphoma, particularly in Hodgkin disease [4].

MRI is useful to assess the extent of bone marrow, soft tissue involvement and in the follow-up after therapy. Sometimes, when symptomatic patients present with a “near normal” type of pattern on radiography, MRI is essential for further assessment as a more sensitive imaging modality [4]. Therefore, in the MRI we can see areas of bone marrow replacement (areas of low signal intensity on T1-weighted images) that enhance after contrast material administration. PBL may present a wide variability of intensities of T2-weighted images, but these areas are usually bright. Areas of hypointensity on T2 seem to be related to intralesional fibrosis [6]. Soft-tissue involvement is frequent.

In summary, although this is an uncommon cause of bone lesions and even rarer in the paediatric population, it should be on the differential when the clinical scenario of a solitary, permeative, metadiaphyseal lesion is present, particularly when a soft-tissue mass and marrow changes are associated with surprisingly little cortical destruction. The differences in treatment and the better prognosis compared with other primary malignant bone tumours emphasizes the need of an accurate and early diagnosis.
Differential Diagnosis List: Primary bone lymphoma - diffuse large B-cell lymphoma, Ewing sarcoma, Osteosarcoma, Osteomyelitis, Bone metastasis

Final Diagnosis: Primary bone lymphoma - diffuse large B-cell lymphoma

References:

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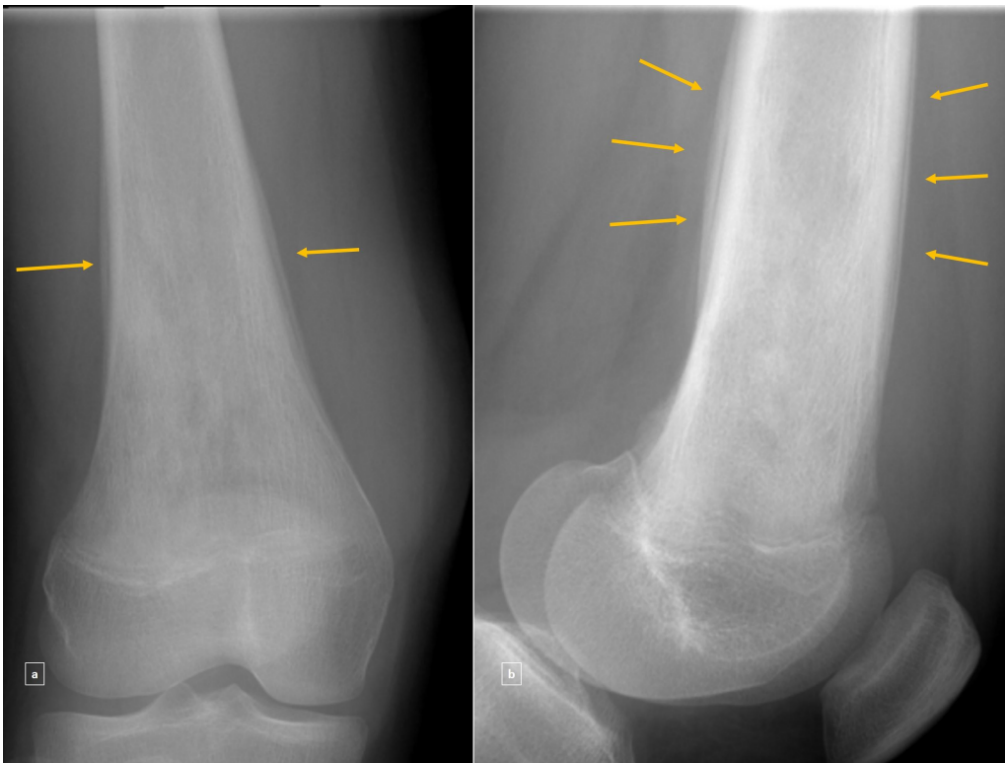
Figure 1

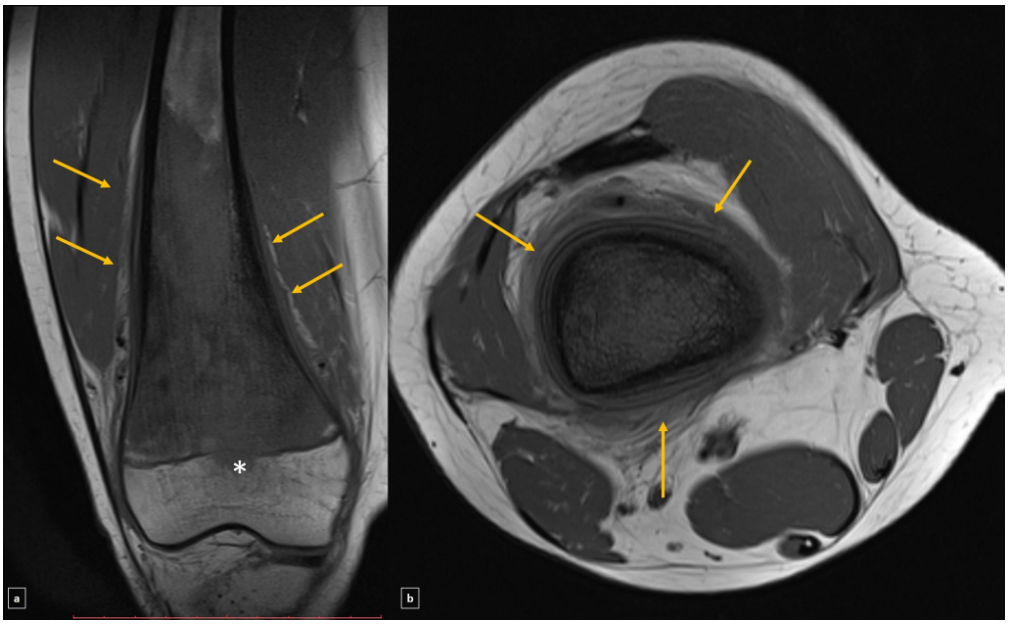
a



Description: Frontal (a) and lateral (b) radiographs show lytic permeative changes in the distal metadiaphysis of the right femur with a lamellated periosteal reaction (arrows). **Origin:** Coelho P, Coimbra Hospital and University Centre, Coimbra Pediatric Hospital

Figure 2

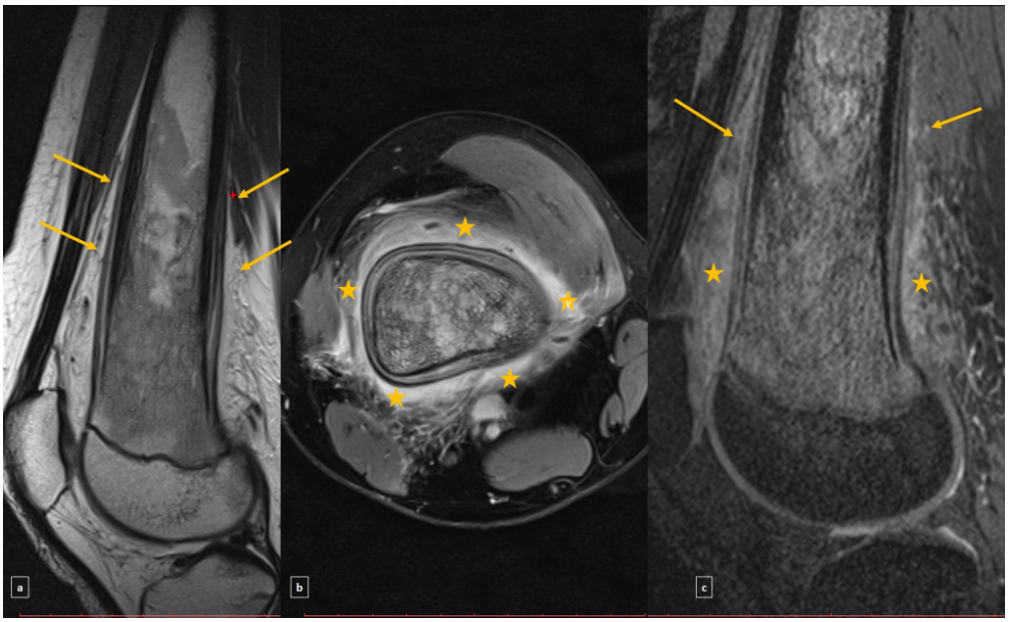
a



Description: Multiple areas of bone marrow replacement in the distal femoral metadiaphysis (low signal areas), with physeal extension (asterisk). Cortical circumferencial thickening and lamellated periosteal reaction (arrows) is also present. **Origin:** Coelho P, Coimbra Hospital and University Centre, Coimbra Pediatric Hospital

Figure 3

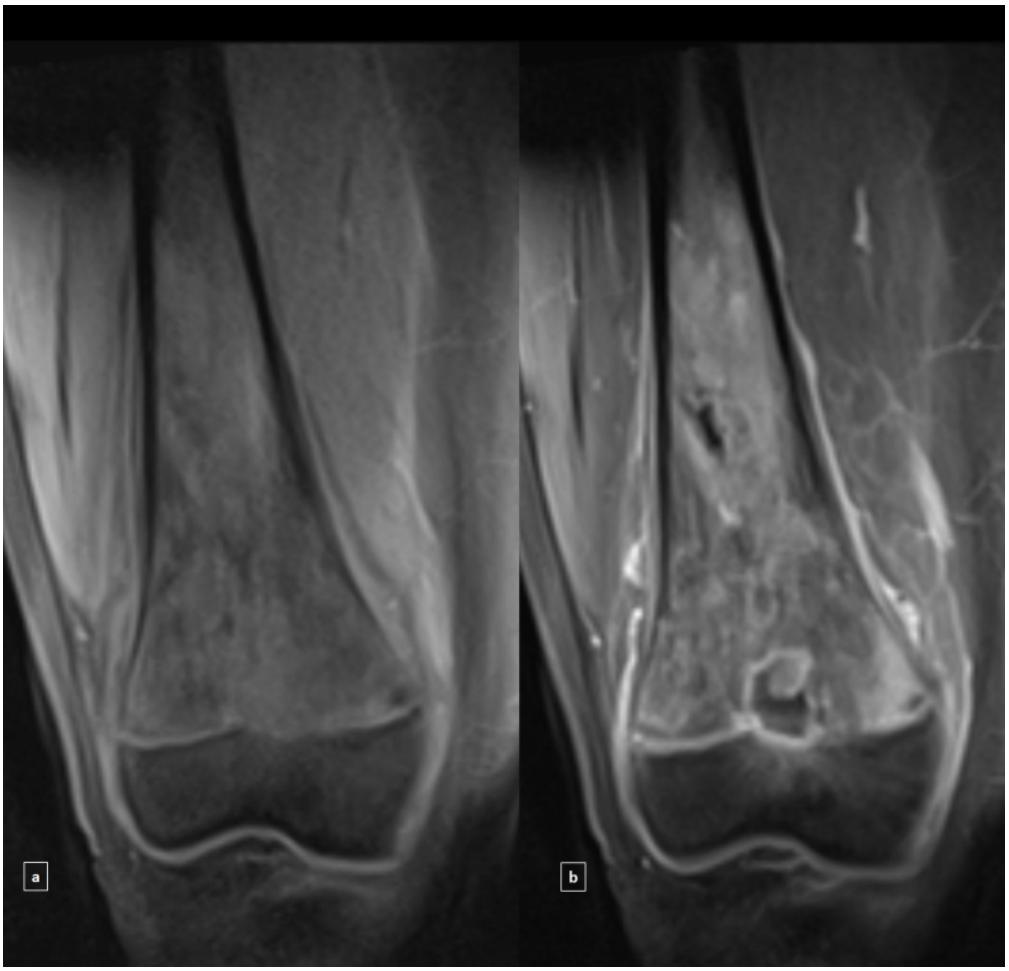
a



Description: Multiple areas low/intermediate signal in the distal femoral metadiaphysis. Cortical circumferencial thickening and lamellated periosteal reaction is also evident (arrows). Note the peri-tumoral oedema (stars) without evidence of soft-tissue mass. **Origin:** Coelho P, Coimbra Hospital and University Centre, Coimbra Pediatric Hospital

Figure 4

a



Description: Fat-suppressed T1 weighted images before (a) and after (b) administration of contrast material shows areas of enhancement within the intramedullary lesion, without evidence of soft-tissue mass. **Origin:** Coelho P, Coimbra Hospital and University Centre, Coimbra Pediatric Hospital