Desmoplastic fibroma of the femur
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Section: Musculoskeletal system
Area of Interest: Musculoskeletal bone
Procedure: Diagnostic procedure
Imaging Technique: Conventional radiography
Imaging Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
Authors: Blockx Laurent
Patient: 21 years, female

Clinical History:

The patient is a 21-year-old woman who presented with persistent left knee problems with flexion-extension and going up stairs after a fall on the left knee 4 months earlier. Radiographs were taken and showed a lesion in the distal left femur. Additionally, an MR examination was performed for further evaluation.

Imaging Findings:

Radiographs showed a metaepiphyseal osteolytic lesion in the distal left femur with partial ill-defined outline. There was cortical breakthrough posteriorly with periostal reaction and associated soft-tissue mass.

On MR images, the tumour showed a rather homogeneous signal intensity iso-intense to muscle on T1-weighted images. Sagittal TIR images showed an inhomogeneous mass in the distal left femur containing several remarkably low signal intensity areas. There was cortical destruction of the distal left femur posteriorly with the mass extending into the adjacent soft tissues mainly periostally contained. Gadolinium-enhanced T1-weighted MR images demonstrated inhomogeneous enhancement.

Discussion:

Desmoplastic fibroma (DF) is a rare benign primary bone tumour and is thought to be the intra-osseous counterpart to soft tissue fibromatoses, also known as desmoid tumour. The clinical presentation is nonspecific. The commonest presenting symptoms are pain, swelling or a palpable mass. However, some patients are asymptomatic and the lesion may be detected as an incidental finding on imaging taken for other reasons.

It can affect almost any bone but DF is most often found in the mandible, the femur, the pelvic bones, the radius and the tibia [1]. The mean age of the patients with DF at the time of diagnosis is reported to be 23 years [1], while the issue of sex predilection remains unclear. The tumour is usually found in the metaphysis or metadiaphysis, although it can extend into the epiphysis in adults after closure of the growth plate.

On radiographs, DF appears as a non-mineralized osteolytic lesion. In most cases, the lesion is well-defined with variably expressed marginal sclerosis. Periosteal reaction is exceptional but cortical thinning is often present and in fewer cases cortical breakthrough can be seen with extension into the adjacent soft tissues. Such destructive pattern with soft tissue invasion may mimic a malignant bone tumour.

On MR, T1W sequences show signal intensities within the lesion that are iso- to hypointense to adjacent normal muscles while T2W sequences demonstrate large areas of low to intermediate signal intensities compared to normal
muscles [2]. Being histologically similar to the soft tissue desmoid tumour, these areas of low signal intensity on T2W images in DF may be explained by the relative low cellularity and the presence of dense fibrous matrix [4]. After intravenous gadolinium contrast administration, somewhat heterogeneous enhancement of the lesion is seen [2, 3].

The radiographic differential diagnosis of desmoplastic fibroma is very broad. However, few bone tumours display features of a predominant osteolytic lesion with prominent T2 shortening not corresponding to calcifications on MR images, permitting to include desmoplastic fibroma in a relatively short differential diagnosis list.

DF behaves in a locally aggressive manner without capacity to metastasize. The preferred therapy is en bloc resection since curettage alone has a higher rate of recurrence. However, when resection is not possible, curettage and grafting may be an adequate treatment modality with close follow-up.

**Differential Diagnosis List:** Desmoplastic fibroma of the femur, Lymphoma, Primary leiomyosarcoma of the bone, Fibrous dysplasia, Non-ossifying fibroma, Giant cell tumour

**Final Diagnosis:** Desmoplastic fibroma of the femur

**References:**


Description: A metaepiphyseal osteolytic lesion is present in the distal left femur. The lesion contains some areas of more ill-defined outline (black arrow). Subtle periostal reaction can be noticed laterally (white arrow). Origin: Blockx L., Department of Radiology, University Hospital, Leuven, Belgium
**Description:** Cortical breakthrough posteriorly in the distal left femur is present with soft tissue extension (black arrow) of the lesion. There is also evidence of mineralization most consistent with periostal reaction (white arrow). **Origin:** Blockx L., Department of Radiology, University Hospital, Leuven, Belgium
Description: The lesion contains several remarkably low signal intensity areas (red arrow). There is cortical destruction of the distal left femur posteriorly with the mass extending postero cranially, mainly periostally contained (white arrow). **Origin:** Blockx L., Department of Radiology, University Hospital, Leuven, Belgium
Description: The mass in the distal portion of the left femur shows a rather homogeneous signal intensity iso-intense to muscle. Origin: Blockx L., Department of Radiology, University Hospital, Leuven, Belgium
Figure 5

Description: The tumour in the distal left femur demonstrates inhomogeneous enhancement. Origin: Blockx L., Department of Radiology, University Hospital, Leuven, Belgium