CLINICAL ORTHOPAEDICS AND RELATED RESEARCH
Number 464, pp. 247–252

Orthopaedic • Radiology • Pathology Conference

Painful Tibial Lesion in a 16-year-old Girl

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HISTORY AND PHYSICAL EXAMINATION

A 16-year-old girl presented with swelling and a firm mass in the region of her right proximal tibia of several months' duration. She had intermittent pain in her right shin over a 2-year period. The pain occurred at night but did not keep her awake. She did experience slight pain with walking. Physical examination revealed swelling over the anterolateral aspect of her right proximal tibia, just distal to the tibial tuberosity, over an area measuring approximately 4 × 4 cm. She had no tenderness to palpation or erythema. She was afebrile. There was no lymphadenopathy, and she had full motion in all her joints. Her medical history was noncontributory.

Plain radiographs (Fig 1), bone scan (Fig 2), computed tomography (CT) (Fig 3), and magnetic resonance imaging (MRI) (Fig 4) of the right tibia were performed.

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

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DOI: 10.1097/BLO.0b013e31805444fa

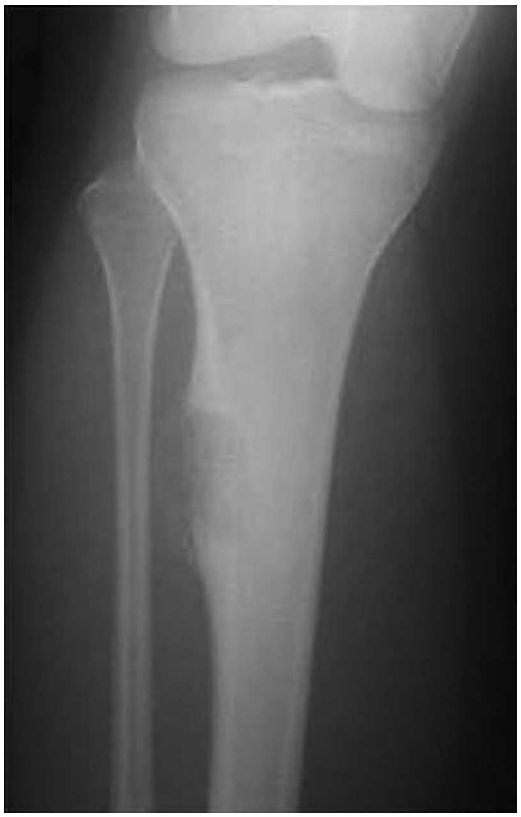

Fig 1. An anteroposterior radiograph of the right leg demonstrates an eccentrically located, cortically based expansile lytic lesion within the anterolateral aspect of the proximal tibial diaphysis. The lateral cortex is markedly thinned; however, no periosteal reaction is evident. There is no appreciable internal calcification.

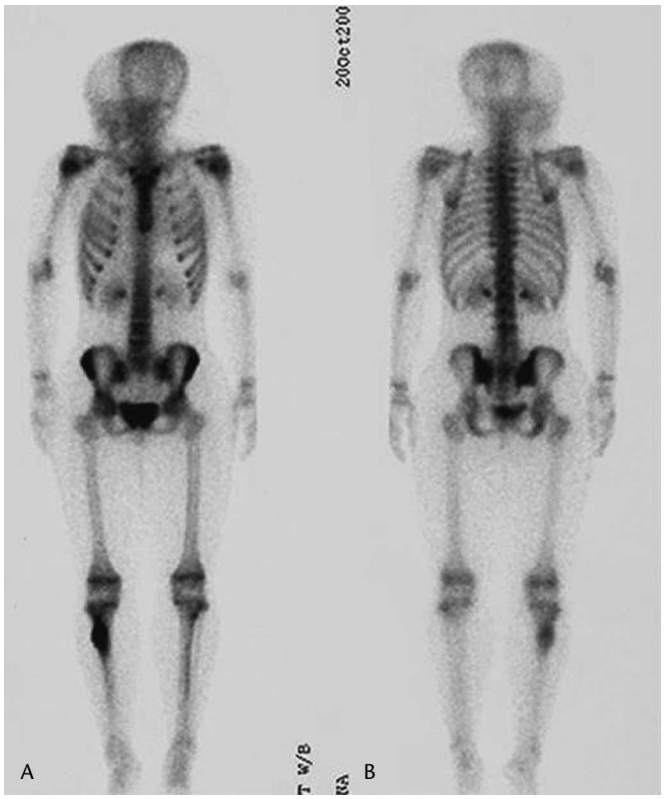

Fig 2A–B. (A) Anterior and (B) posterior views from a whole-body bone scan show significant radiopharmaceutical uptake within the right proximal tibia, corresponding to the radiographic abnormality. No other foci of abnormal uptake were evident.

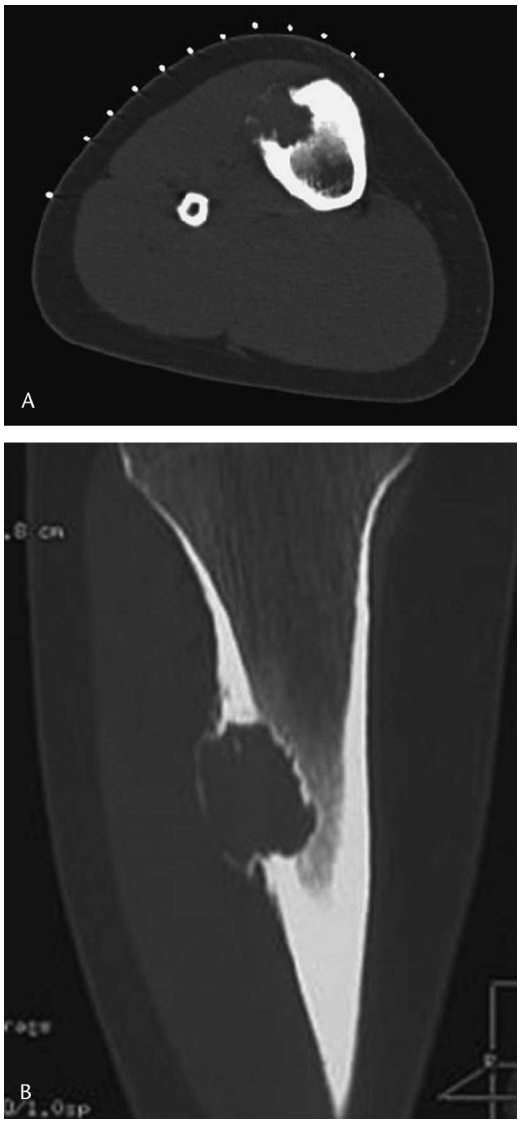

Fig 3A–B. Axial CT with (A) bone windows and (B) coronal reconstruction demonstrates the lesion is lytic in nature and is cortically based with extension into the medullary cavity. In addition, there is expansion laterally into the adjacent soft tissues associated with marked cortical thinning laterally; however, there is no appreciable soft tissue mass. No internal calcification is visualized.

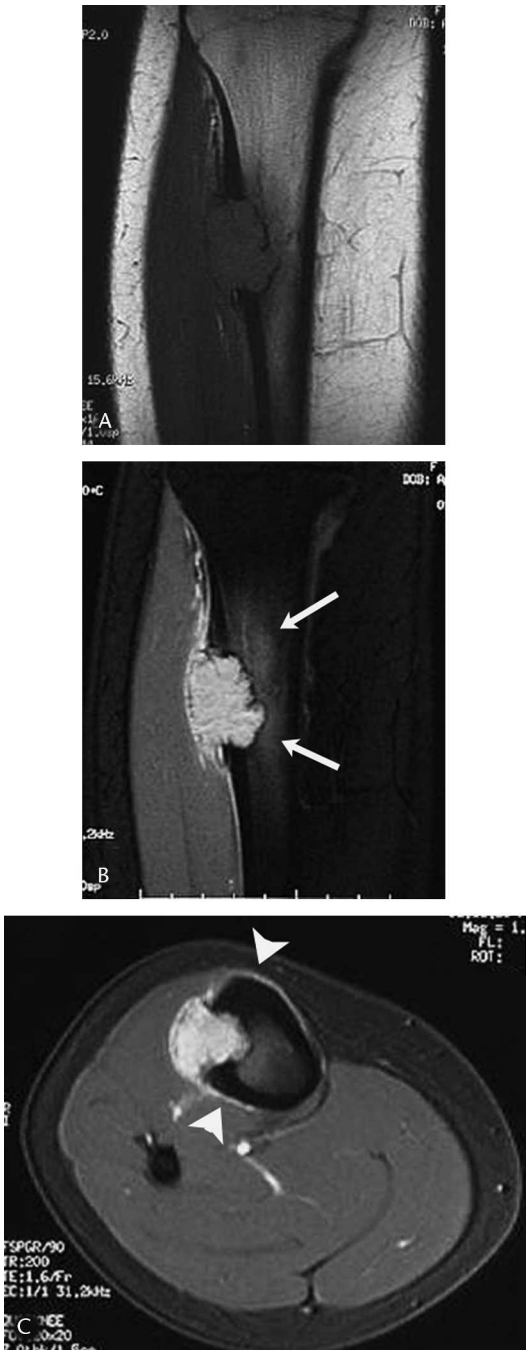

Fig 4A–C. (A) A coronal T1-weighted MR image and (B) a coronal T2-weighted fat-saturated MR image demonstrate the lesion is predominantly hyperintense on T2-weighted images and low signal on T1-weighted images with a somewhat lobular configuration and suggestion of thin internal septa. There is a well-defined low signal intensity border medially; however, there is increased T2 signal within the adjacent bone marrow, suggestive of reactive edema (arrows). (C) An axial T1-weighted fat-saturated MR image after intravenous gadolinium administration shows relatively homogeneous enhancement of the lesion. Enhancement is also noted involving the periosteum along the lateral and anterior margin of the proximal tibia (arrowheads).

IMAGING INTERPRETATION

Plain radiographs of the right lower leg revealed an eccentrically located, cortically based expansile, geographic, lytic lesion within the anterolateral aspect of the proximal tibial diaphysis (Fig 1). No internal calcification was seen and no periosteal reaction was noted.

Whole-body bone scan demonstrated significant radiopharmaceutical uptake within the right proximal tibia (Fig 2). No other foci of abnormal uptake were evident.

CT confirmed the cortical origin of the lytic lesion with extension into the medullary cavity and expansion laterally into the adjacent soft tissues (Fig 3). Reactive periosteum remained intact and surrounded the overlying soft tissue component of the lesion. No internal calcification was seen. The medullary border of the lesion was circumscribed by a thick sclerotic rim.

On MRI, the lesion was low signal on T1-weighted images (Fig 4A) and predominantly hyperintense on T2-weighted images (Fig 4B), with a somewhat lobular configuration and suggestion of thin internal septa. There was a well-defined low signal intensity border medially; however, we noted mild T2 signal hyperintensity in the adjacent marrow, suggestive of reactive edema. There was relatively homogeneous enhancement on postcontrast imaging. Enhancement was also noted involving the periosteum along the lateral and anterior margin of the proximal tibia (Fig 4C).

DIFFERENTIAL DIAGNOSIS

Osteoblastoma
Brodie's abscess
Periosteal chondroma
Chondromyxoid fibroma
Aneurysmal bone cyst
Osteofibrous dysplasia

A CT-guided biopsy of the lesion was performed. Subsequently, the lesion was excised surgically and the histology was studied (Fig 5).

Based on the history, physical findings, imaging studies, and histology, what is the diagnosis and how should the lesion be treated?

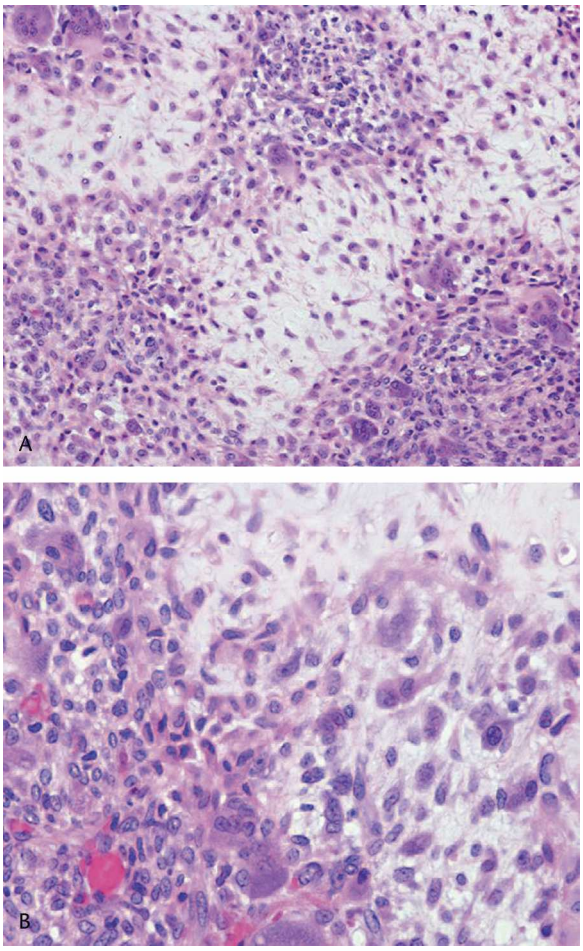

Fig 5A–B. (A) A low-power photomicrograph shows a lobular architecture with central hypocellular areas and peripheral hypercellular areas with osteoclastic giant cells (Stain, hematoxylin and eosin; original magnification, ×100). (B) A high-power photomicrograph shows areas of oval and stellate cells in myxoid/chondroid matrix (right upper) and areas of hypercellularity with osteoclastic giant cells (lower left) (Stain, hematoxylin and eosin; original magnification, ×200).

See page 251 for diagnosis and treatment.

Continuation of ORP conference from page 250.

HISTOLOGY INTERPRETATION

The pathologic specimen from the intraregional excision demonstrated a lobular configuration on the gross level. The central portion of the lobules was composed of hypocellular chondroid areas (Fig 5). The cells were oval to stellate and were separated by extracellular myxoid- and chondroid-like matrix. The peripheries of the lobules were hypercellular with oval to spindle-shaped mononuclear cells and osteoclastic giant cells. The large amount of reactive bone at the edge of the lesion may have been due to the subperiosteal location of the tumor.

DIAGNOSIS

Chondromyxoid fibroma

DISCUSSION AND TREATMENT

Chondromyxoid fibroma (CMF) is a rare benign cartilage neoplasm accounting for 1% of primary bone tumors.[16] Eighty percent of patients are younger than 36 years, with a smaller peak between 50 and 70 years.[9] There is a slight male predominance.[4,5,17,18,23] Symptoms include slowly progressive pain and tenderness with or without restriction of motion.[16,23]

The differential diagnosis in this case included aneurysmal bone cyst, osteofibrous dysplasia, Brodie's abscess, periosteal chondroma, and osteoblastoma. Clinical presentation and imaging proved useful in prioritizing the differential diagnosis. The patient's age, lesion location, and appearance on both plain radiographs and CT were compatible with an aneurysmal bone cyst. However, the absence of internal fluid-fluid levels and the homogeneous enhancement pattern were unusual for this diagnosis.[16] Osteofibrous dysplasia or ossifying fibroma is characteristically found in the anterior tibia in the first or second decades of life. However, the lesions are typically found in the middle third of the tibia and are commonly accompanied by mild anterior or anterolateral bowing of the tibia, features missing in this case. In addition, it is not unusual to have concomitant involvement of the fibula in ossifying fibroma and distinguishing between this lesion and both fibrous dysplasia and adamantinoma can be difficult.[15] Cortically based infection with Brodie's abscess is possible; however, these lesions typically extend in the long axis of the bone, are often described as "channel-like," and usually provoke surrounding sclerosis and periosteal reaction. Thus, they can be confused with osteoid osteoma. The absence of these imaging features, as well as the absence of a history of infection, fever, or other signs of systemic toxicity, made this diagnosis less likely. Periosteal chondromas can be found in this location and age group; however, they are surface-based lesions that tend to cause erosion or saucerization of the cortical surface rather than expansion and extension into the medullary cavity, as was the case here. Calcification is seen in up to 50% of periosteal chondromas, which was also missing in our case. Osteoblastomas tend to be eccentric, lytic lesions based within the medullary cavity or cortex often containing areas of internal calcification and ossification. They are more common in the spine or flat bones and have a male predilection (2:1).[15,16] Although these features were missing in our case, given the imaging appearance and history of night pain, this entity was considered. However, the final diagnosis was compatible with subperiosteal CMF.

CMF typically occurs in the metaphysis of long bones, with the tibia and femur involved in 55% of cases. Less commonly, it may involve the pelvis, vertebral bodies, skull, sternum, metatarsals, calcaneus, and phalanges of the feet.[3,8,19,20,26] Primary diaphyseal or epiphyseal locations are rare. Our case demonstrated some classic radiographic features of CMF, but its diaphyseal subperiosteal location was unusual.

CMF is overwhelmingly monostotic.[2,4] It is benign, although there have been some reports of malignant transformation in 1% to 2% of cases.[1,2,10,15] Five percent of cases report an accompanying pathologic fracture.[9]

CMFs are eccentric, radiolucent lesions resulting in cortical expansion and endosteal sclerosis.[12,14] Larger lesions may penetrate the cortex and result in a characteristic "cortical bite." Significant periosteal reaction is unusual; however, rare cases have demonstrated thick reactive bone formation with a central lucency.[21] Cartilage formation on plain radiographs is uncommon.[22]

CT can identify calcifications not visible on plain radiographs and delineates the extent of cortical expansion.[25] MRI is nonspecific for cartilaginous lesions and demonstrates low signal on T1-weighted images and high signal on T2-weighted sequences. T2-weighted images demonstrate surrounding bone marrow edema and reactive periostitis. There is generally diffuse enhancement postcontrast.

Histologically, the components of the tumor, being chondroid, myxomatous, or fibrous tissue, can occur in different proportions.[4,6,17,18,24,25,27] It may appear similar to chondrosarcoma and correlation with radiographic studies helps distinguish the two.[7] The surface CMFs (juxtacortical, periosteal, subperiosteal, intracortical) are extremely rare with only 20 cases reported in the English literature.

Differential diagnosis includes giant cell tumor, chondroblastoma, osteoblastoma, Brodie's abscess, hemangio-

ma, aneurysmal bone cyst, and fibrous dysplasia. Periosteal chondroma, periosteal myxoma, and subperiosteal hemorrhage should be considered in the differential diagnosis with surface CMFs.

CMFs are benign but aggressive tumors. Untreated, they continue to progress, destroy more bone, and result in local complications. Treatment is entirely surgical. Most tumors are treated with an intralesional curettage followed by patching with bone graft or polymethylmethacrylate.[11] Recurrence may occur in up to 25% of cases. Lersundi et al[11] reported a local recurrence rate of 45% in patients treated with curettage alone. Cryosurgery may be utilized as a local adjuvant for eradication of microscopic disease and reducing the rate of local recurrence, after a thorough curettage, to less than 3%.[13]

Our patient was treated with thorough curettage with hand curettes. The tumor cavity was shaved with a high-speed burr to accomplish a burr-down resection. Following this, two cycles of cryosurgery were performed utilizing liquid nitrogen as described by Malawer et al.[13] Iliac crest bone graft was harvested from the ipsilateral iliac crest and packed into the defect. The patient remained nonweightbearing with crutches in a hinged knee brace for 12 weeks. There were no postoperative complications. The defect appeared radiographically to be healed within 12 weeks. Walking was subsequently progressed. The patient is now 2.5 years posttreatment with no local recurrence.

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