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Primary skeletal leiomyosarcoma

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Leiomyosarcoma typically occurs within the uterus, gastrointestinal tract, and mesentery. Primary skeletal leiomyosarcoma is exceedingly rare. The radiographic appearance is similar to that of other aggressive sarcomas of the bone. Definitive diagnosis is achieved through biopsy or excision and pathologic review.

Case report

A 28-year-old female presented with gradual onset of left-knee pain without a preceding traumatic event. Initially, her primary care physician diagnosed her with a meniscal tear based on physical exam. She was put into a brace, and the pain subsided after 2 weeks of conservative therapy. Two months later, the left-knee pain began increasing. She again saw her primary care doctor, who recommended an MRI, but because of insurance problems, she was unable to obtain one. A radiograph was not performed. She was given some narcotics, and again her pain improved.

She subsequently moved across the nation, and the left-knee pain returned. A new physician examined her and diagnosed a "muscle strain," subsequently referring her to physical therapy. Her physical therapist recommended radiographs. The radiographs demonstrated a poorly defined lytic lesion arising from the distal left femur with a wide zone of transition, cortical destruction, and a small anterior soft-tissue component (Fig. 1). At the time of repeat examination after imaging, she denied any pain in her left knee except at night and with full extension. She also denied any fevers or chills, or any pain in her tibia or her hip. An MRI was obtained to further characterize the distal femoral lesion. This demonstrated a heterogeneously enhancing intramedullary mass within the distal metaphysis of the left femur (Figs. 2 and 3). There was cortical breakthrough anteriorly, with an enhancing soft-tissue component. 18-fluorodeoxyglucose PET/CT and a Tc-99m MDP bone scan both showed marked radionuclide activity in the lesion (Figs. 4 and 5). Findings were most consistent with an osteosarcoma. At resection, the pathology demonstrated well-differentiated leiomyosarcoma that involved the medullary cavity, with focal extension through the cortex into the surrounding soft tissue (Fig. 6A). Histologically, the mass contained fascicles of highly mitotic spindle cells that

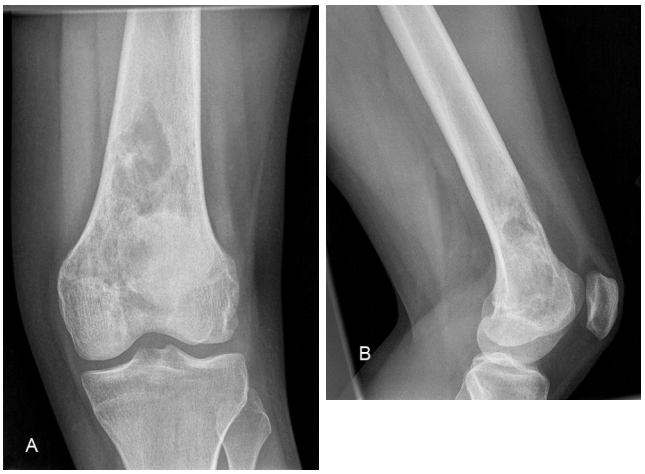

Figure 1. 28-year-old female with left-knee pain. A. Frontal radiograph demonstrated a permeative lytic lesion in the distal femur with wide zone of transition. No osseous matrix was identified. B. Lateral radiograph of the distal femoral permeative lytic lesion demonstrated anterior cortical breakthrough and associated small soft-tissue mass encroaching on the prefemoral fat pad.

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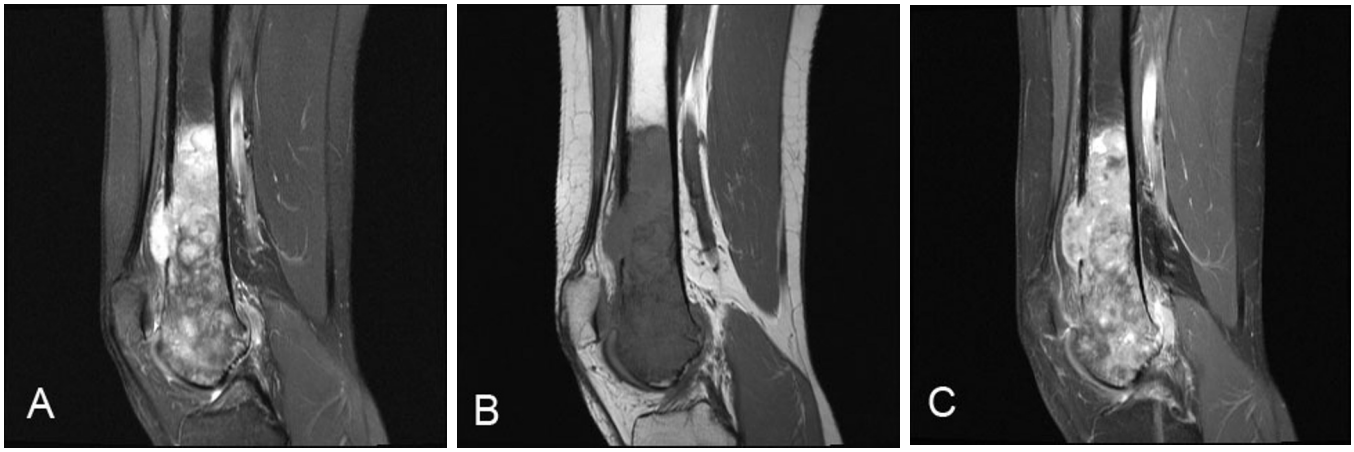
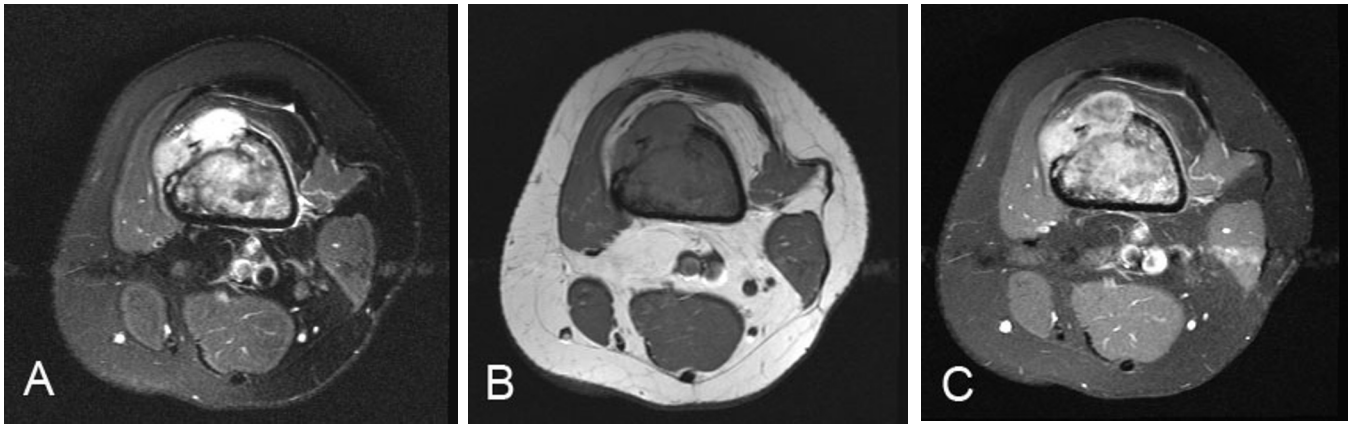


Figure 2. A. 28-year-old female with left-knee pain. Sagittal T2 fat-saturated MR image (TR = 5600, TE = 55) demonstrated heterogeneously hyperintense intramedullary mass with cortical breakthrough and anterior soft-tissue mass. B. Sagittal T1 precontrast MR image (TR = 750, TE = 10) demonstrated T1 hypointense mass replacing the distal femoral marrow. C. Sagittal T1 fat-saturated postcontrast MR image (TR = 786, TE = 11) demonstrated heterogeneously enhancing intramedullary mass with anterior extracortical extension and small soft-tissue mass.



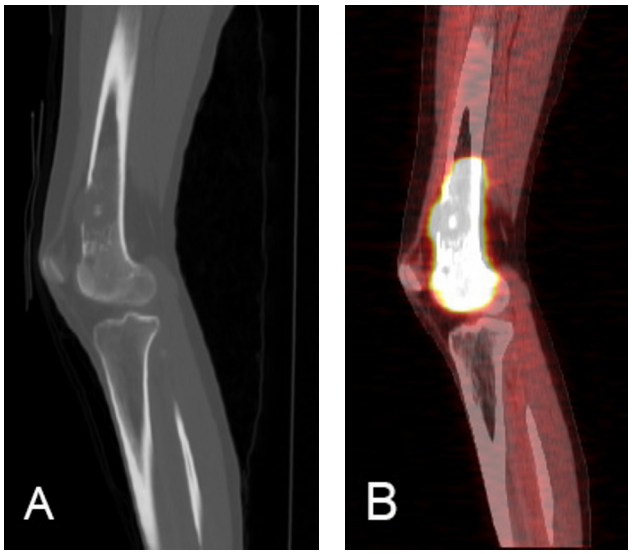


Figure 3. A. 28-year-old female with left-knee pain. Axial T2 fat-saturated MR image (TR = 5600, TE = 55) demonstrated heterogeneously hyperintense intramedullary mass with anterior cortical breakthrough and small soft-tissue mass. B. Axial T1 precontrast MR image (TR = 750, TE = 10) demonstrated T1 hypointense mass replacing the femoral marrow with anterior cortical breakthrough and small soft-tissue mass. C. Axial T1 fat-saturated postcontrast MR image (TR = 786, TE = 11) demonstrates a heterogeneously enhancing intramedullary mass with anterior extracortical breakthrough and small soft-tissue mass.

Figure 4. 28-year-old female with left-knee pain. A. Sagittal computed tomography image demonstrated a lytic intramedullary lesion of the distal left femur with an anterior cortical breach. B. Sagittal positron emission tomography-computed tomography image demonstrated copious fluorodeoxyglucose avidity within the lytic intramedullary lesion of the distal left femur (maximum standardized uptake value measured 10.1).

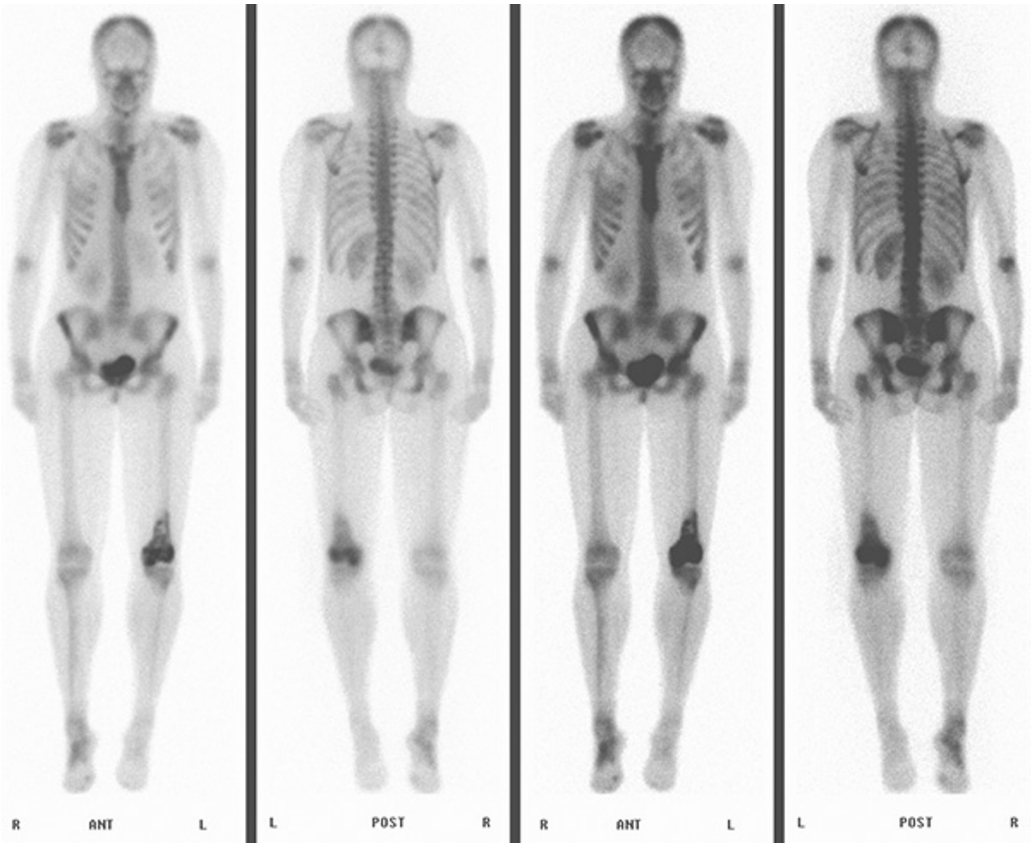


Figure 5. 28-year-old female with left-knee pain. MDP bone scan demonstrated intense radiotracer deposition within the distal femur. No lesions suspicious for metastasis were present. Mild tracer deposition of the right ankle and foot was likely inflammatory.

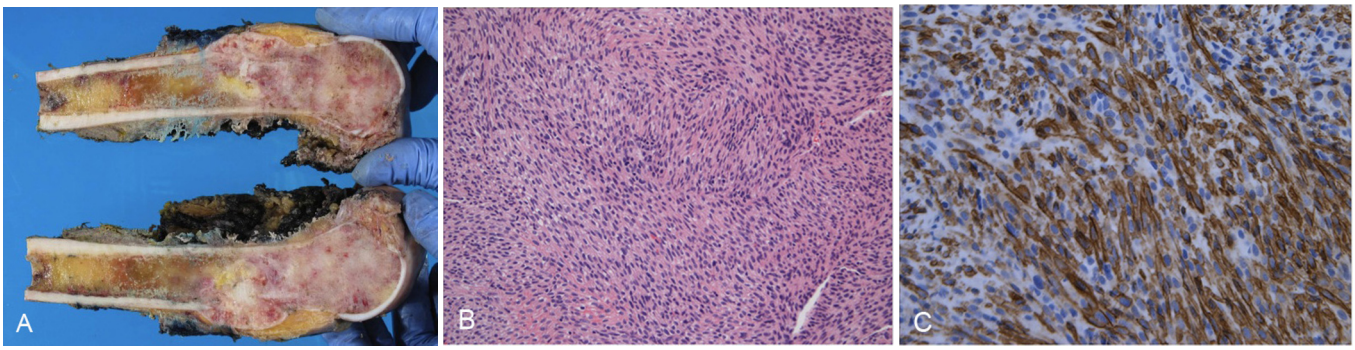


Figure 6. A. 28-year-old-female with left-knee pain. Gross pathologic photograph of the distal femur mass with extracortical extension. B. Hematoxylin- and eosin-stain micrograph demonstrated fascicles of highly mitotic spindle cells. C. Smooth-muscle-actin, immunohistochemical-receptor-labeled micrograph demonstrated positive staining for smooth-muscle receptors.

stained positive for smooth-muscle actin immunohistochemical receptor, also consistent with a well-differentiated leiomyosarcoma (Fig. 6C)

Discussion

Sarcomas are a heterogeneous group of malignant tumors with mesenchymal cell origin and are named for their predominant tissue type. Leiomyosarcomas are malignant smooth-muscle tumors composed of spindle cells. The majority of leiomyosarcomas arise in the uterus, gastrointestinal tract, mesentery, and omentum (1). Primary leiomyosarcoma of bone is exceedingly rare (2, 3); as of 2010, only 107 well-documented cases had been reported (4). When primarily located within bone, these lesions are reported to occur predominantly in large bones, most commonly in the distal femur (5). First reported by Evans and Sanarkin in 1965 (3), skeletal leiomyosarcomas are thought to arise from the vascular smooth-muscle cells within the bone (5).

Radiologically, primary skeletal leiomyosarcoma cannot be differentiated from other aggressive sarcomas of the bone. The differential diagnosis should include destructive infections and metastasis. The characteristics of primary skeletal leiomyosarcoma include permeative lysis of the bone, often with a soft-tissue component. Calcified matrix is often not seen, but when it is present, it can make differentiation from osteosarcoma challenging. Cortical breakthrough can be seen in up to 86% of cases (6). Pathologic fractures are also possible. Primary soft-tissue leiomyosarcoma with osseous metastasis can be excluded by lack of primary tumor on computed tomography, with or without positron-emission tomography (7). Definitive diagnosis is made by pathologic evaluation after biopsy or resection.

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1. Figure 6C. 48-year-old male with pheochromocytoma. Fused coronal I-123 MIBG SPECT/CT showed avid uptake of radiotracer within the left adrenal pheochro-