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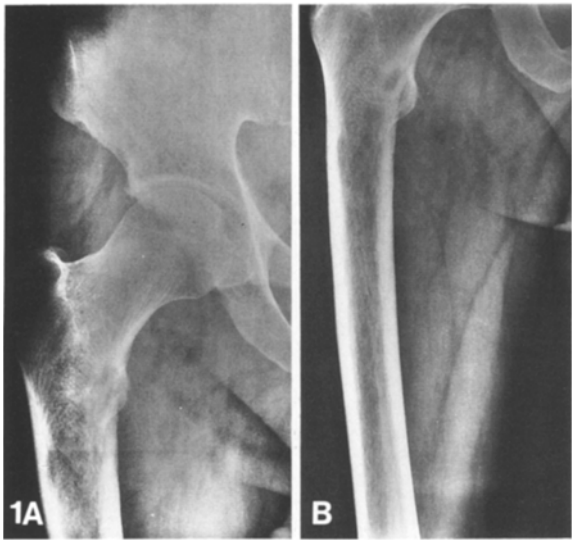
Case report 794

Ibrahim Fikry Abdelwahab, M.D.[1], George Hermann, M.D.[1], Samuel Kenan, M.D.[2], Michael J. Klein, M.D.[3], Michael M. Lewis, M.D.[2]

[1] Department of Radiology of the Mount Sinai Medical Center of the City University of New York, New York, USA
[2] Department of Orthopedic Surgery of the Mount Sinai Medical Center of the City University of New York, New York, USA
[3] Department of Pathology of the Mount Sinai Medical Center of the City University of New York, New York, USA



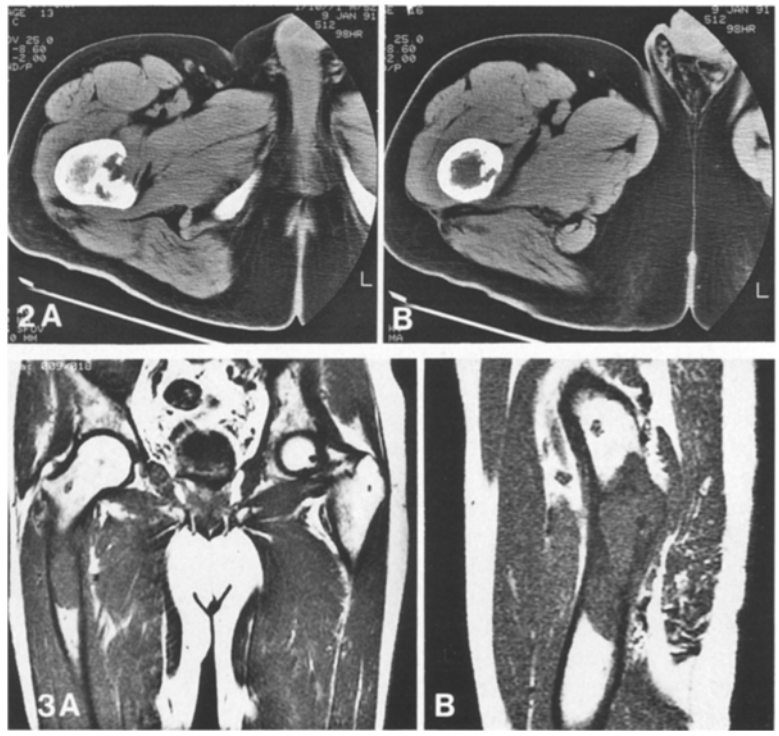


Fig. 1. **A** Anteroposterior view of the right hip. **B** Anteroposterior view of the intertrochanteric region and the proximal two-thirds of the shaft of the right femur. Both views demonstrate destruction in the region of the lesser trochanter and minimal endosteal scalloping of the cortex of the proximal end of the shaft of the femur

Fig. 2. **A** CT section at the level of the lesser trochanter of the right femur. **B** CT section at a lower level in the region of the proximal end of the shaft of the right femur. Both show destruction, endosteal scalloping, and interruption of the cortex. No soft tissue extension is noted

Fig. 3. **A** Coronal MRI of the proximal end of the right femur (TR350/TE35). **B** Sagittal MRI of the proximal end of the right femur (TR350/TE35). These two sections demonstrate vividly the extend of the involvement of the bone marrow by the tumorous lesion

Clinical information

A 52-year-old man presented with a 3-year history of intermittent pain in the right hip. The pain had become worse in the last few months, especially at night. When he presented, he was limping. Physical examination revealed a well-developed man with a mildly antalgic gait. He had tenderness over the region of the greater trochanter and right hip which was aggravated by external rotation. There was normal range of motion of the right and left hip and the neurovascular bundle was intact.

A bone scan showed increased uptake in the region of the lesser trochanter of the right femur. Plain radiography, computed tomography (CT), and magnetic resonance (MRI) were then performed and revealed a destructive bony lesion in the proximal end of the shaft of the right femur (Figs. 1–3). A biopsy was performed.

Correspondence to: Dr. I. Fikry Abdelwahab, Box 1234, Department of Radiology, Mount Sinai Medical Center, One Gustave L. Levy Place, New York, NY 10029-6574, USA

Diagnosis: Primary leiomyosarcoma of the right femur (Fig. 4)

The differential diagnosis included metastatic bone cancer, plasmacytoma, lymphoma, malignant fibrous histiocytoma, fibrosarcoma, and chondrosarcoma.

The patient underwent resection of the proximal third of the right femur with reconstruction by custom-made bipolar endoprosthesis. The surgical margins were free of tumor. The patient did not receive radiation treatment or chemotherapy and had an uneventful recovery. In the 1-year follow-up after the operation, the patient has had a good functional result and is disease-free.

Discussion

Primary leiomyosarcoma of bone (PLOB) is regarded as a rare form of bone sarcoma. So far, about 42 cases have been reported in the literature. Twenty-nine of them were located in the long bones of the extremities, involving mainly the metaphysis. Nine were located in the jaw bones [1–4]. The remaining four were located in the axial skeleton, one in the sacroiliac region, one in the acetabulum, and two in the ribs [3, 10].

PLOB is apparently more common than has been reported, since it is evident that it has not always been recognized and distinguished from other spindle-cell sarcomas of bone, particularly fibrosarcoma and malignant fibrous histiocytoma.

The age distribution is fairly even from the first to the eighth decades of life and the tumor seems to arise predominately in male subjects (ratio of 2:1 in a series of 16 cases [3]). However, in a recent series of five patients, four were women [10].

The origin of leiomyosarcoma of bone entails three possibilities: it may be (1) a primary soft tissue tumor which secondarily invades the bone, (2) a metastasis to bone from a leiomyosarcoma elsewhere, usually the uterus or gastrointestinal tract, or (3) it may be a leiomyosarcoma arising intrinsically in the bone [9]. In our case, there was no soft tissue mass invading the adjacent bone and there was no evidence of leiomyosarcoma elsewhere. Speculation must be aroused as to the origin of PLOB. The tumor most probably arises from the smooth muscle cells in the walls of the intraosseous blood vessels, although an origin in perivascular, multipotential, or mesenchymal cells cannot be ruled out [3, 14].

The diagnosis of PLOB still remains a light-microscopic diagnosis, and electron microscopy seems to make the most valuable contribution to the diagnosis. The cells are spindle-shaped with elongated, often cigar-shaped, dense nuclei with eosinophilic and picrinophilic cytoplasm. All the tumors contain varying amounts of collagen and are rich in reticulin fibers which encircle individual tumor cells. Focal hyalinized areas are a common finding, while necrosis is not a prominent feature [3, 14]. Clinically, PLOB presents as a painful bony lesion and may be accompanied by a palpable mass.

Radiographically, the tumors are usually osteolytic and may exhibit

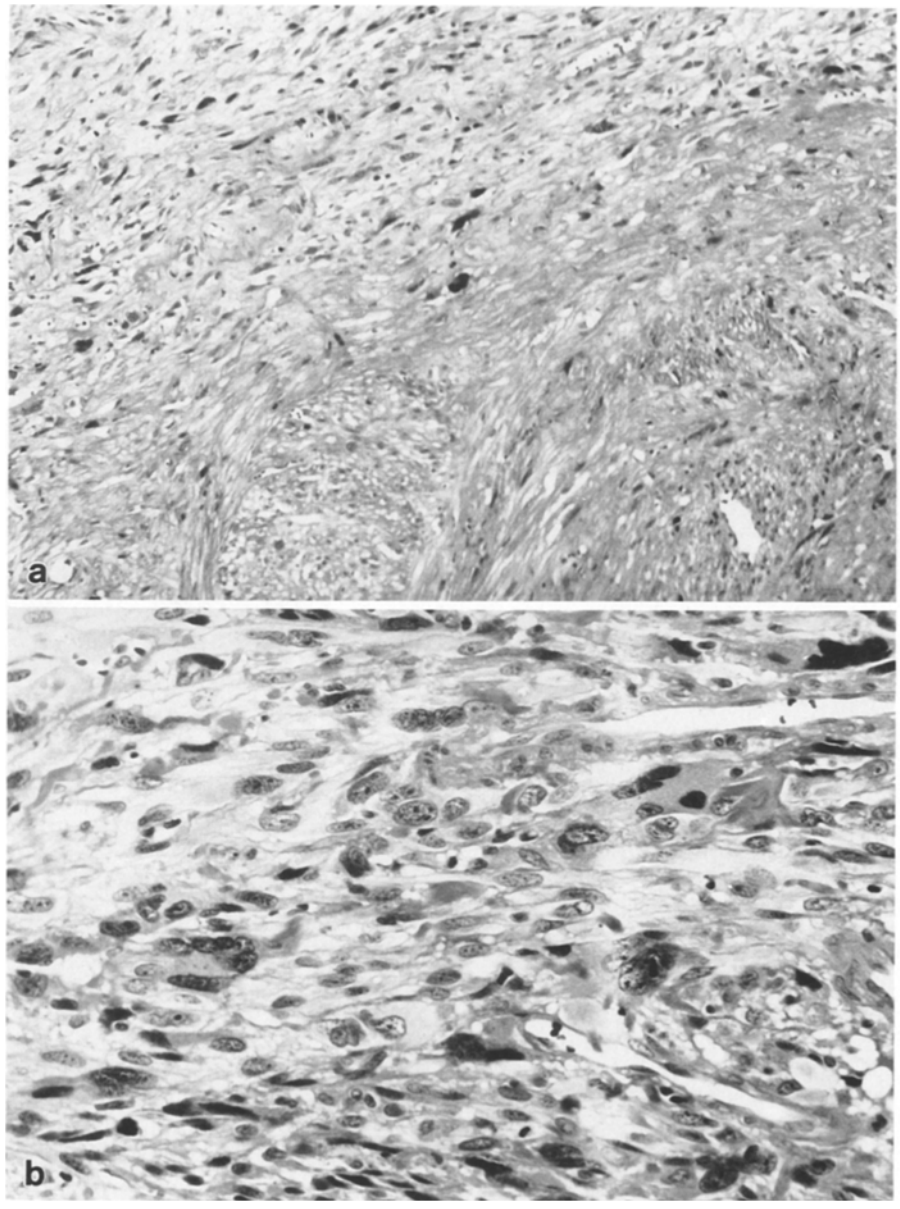


Fig. 4. A Low-power photomicrograph demonstrating sweeping fascicles typical of smooth muscle running in parallel directions. Note the large numbers of bizarre, atypical nuclei at *upper left*. (×100). **B** High-power photomicrograph reveals that the cells have bizarre, hyperchromatic, blunt-ended nuclei with a considerable mitotic rate. (×400)

aggressive characteristics with permeation of the cortical bone. In a group of 16 cases of PLOB, a pathological fracture developed in 3 before diagnosis [3].

PLOB may rarely cause cortical expansion. Therefore, the lesion has no typical radiographic appearance and can mimic other primary and secondary malignant tumors. CT is helpful in determining the cortical invasion and the extent of bone destruction, while MRI is the most exact modality in evaluating the extent of the bone marrow invasion and soft tissue extension. In our case the cortex was invaded but the tumor was contained within the bone.

The prognosis of such tumors is poor even if they are treated by a wide excision and chemotherapy. In a series of 16 PLOB, pulmonary metastases had occurred in 8 cases, indicating that PLOB is a highly malignant tumor [3, 14]. Our patient has been followed-up for more than 1 year without evidence of metastases or recurrence.

In *summary,* a rare case of primary leiomyosarcoma of the proximal end of the shaft of the right femur in a 52-year-old man has been reported. Proof was obtained by an open biopsy. The origin, pathological features, and clinical presentation have been discussed. The radiographic appearance, the differential diagnosis, and the poor prognosis of this tumor have also been indicated.

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