# Leiomyosarcoma of Bone

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**Abstract**

Leiomyosarcoma is a rare malignant intraosseous neoplasm, constituted by spindle cells showing smooth muscle differentiation and devoid of matrix production. More common in long bones of adults, especially the femur and tibia, craniofacial bones and the spine are other preferred sites. It is radiologically osteolytic and permeative. It is histologically similar to its soft tissue counterpart, with long and interwoven bundles of spindle cells with eosinophilic cytoplasm and characteristic blunt-ended nuclei. It is positive for smooth muscle immunohistochemical markers. Histologic grade correlates with prognosis. Surgical resection and chemotherapy are indicated.

## Definition

- A rare malignant intraosseous neoplasm, constituted by spindle cells showing smooth muscle differentiation and devoid of matrix production

## Etiology

- Mostly unknown.
- Some cases secondary to radiation therapy or associated to EBV infection were reported.

## Epidemiology

### Sex

- There is a slight male predominance.

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### Age

- More common in adults

## Sites of Involvement

- Most cases occur in long bones, especially the distal metaphysis of femur and the proximal metaphysis of the tibia.
- Craniofacial bones and the spine are other reported sites of occurrence.

## Clinical Symptoms and Signs

- Pain is the most frequent first symptom.
- Pathological fracture may occur.

## Image Diagnosis

### Radiographic Features

- Radiographs show a purely osteolytic mass centered in the medullary cavity with ill-defined margins. Cortical permeation and periosteal reaction suggestive of an aggressive lesion may be seen.

E. Santini-Araujo et al. (eds.), *Tumors and Tumor-Like Lesions of Bone: For Surgical Pathologists, Orthopedic Surgeons and Radiologists*, DOI 10.1007/978-1-4471-6578-1_40, © Springer-Verlag London 2015

### CT and MRI Features

- CT and MRI disclose an unusual high preference for this neoplasia to grow in length relative to a much less mediolateral expansion.
- MR images of the tumor are iso- or hypointense relative to the muscle on T1-weighted images, and iso- or hypointense areas relative to fat on T2-weighted SE images.

### Imaging Differential Diagnosis

#### Lymphoma of Bone

- May present similar imaging features

Other primary and secondary malignancies with lytic but otherwise nonspecific images may be considered in individual cases.

## Pathology

### Gross Features

- The cut surface of the tumor is fleshy and gray pinkish white, with areas of necrosis, with well or poorly defined margins.

### Histological Features

- Long and interwoven bundles of spindle cells with eosinophilic cytoplasm and characteristic blunt-ended nuclei, with variable pleomorphism, similar to its soft tissue counterpart.
- Osteoid is always absent.
- Epithelioid, myxoid, and pleomorphic variants have been reported.

### Pathology Differential Diagnosis

- **Metastasis from primary leiomyosarcoma from other sites** must be ruled out, especially from the uterus and bowel.

### Ancillary Techniques

- Immunohistochemical techniques demonstrate positivity for muscle markers: desmin, h-caldesmon, and smooth muscle actin.

### Genetics

- Genomic losses and absence of phosphorylated RB, similar to soft tissue leiomyosarcoma

## Prognosis

- Histologic grade correlates directly with recurrence and metastatic rates.
- Prognosis may be better than for other primary bone sarcomas of the same grade in nonmetastatic patients.

## Treatment

- Surgical resection and chemotherapy

## Images

See Figs. 40.1, 40.2, 40.3, 40.4, and 40.5 for illustrations of leiomyosarcoma of the bone.

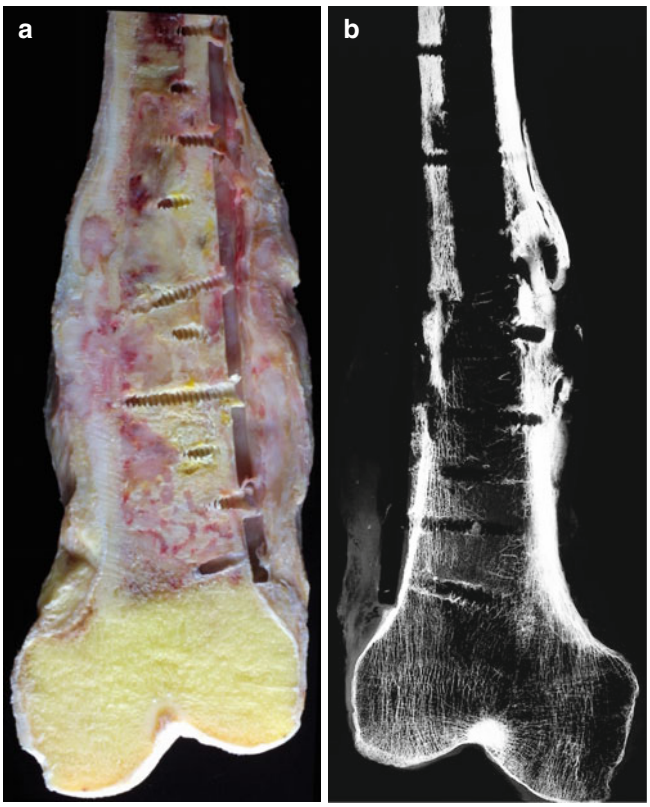


**Fig. 40.1** Leiomyosarcoma of the metaphysis and diaphysis of a femur. Specimen photography and radiography. The lesion involved metallic implants from a previous surgical procedure. (**a**) Whitish-pink firm and elastic lesion occupying the marrow space in a mostly longitudinal fashion, permeating the cortex and involving adjacent soft tissue. (**b**) Absence of mineral deposits in tumor tissue

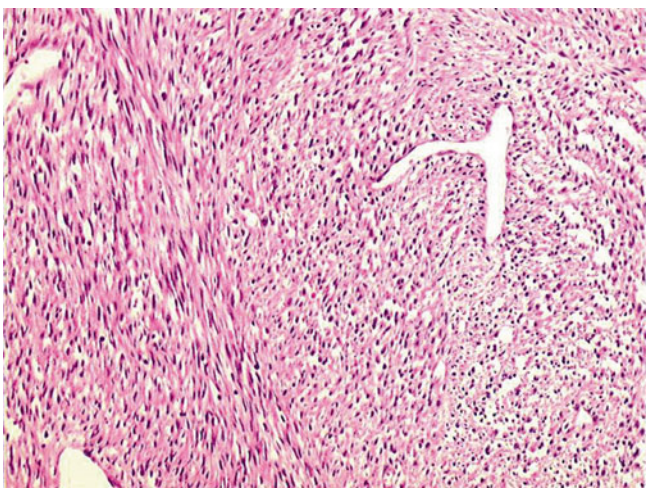

**Fig. 40.2** Low-power microscopic view. Long and interwoven bundles of spindle cells

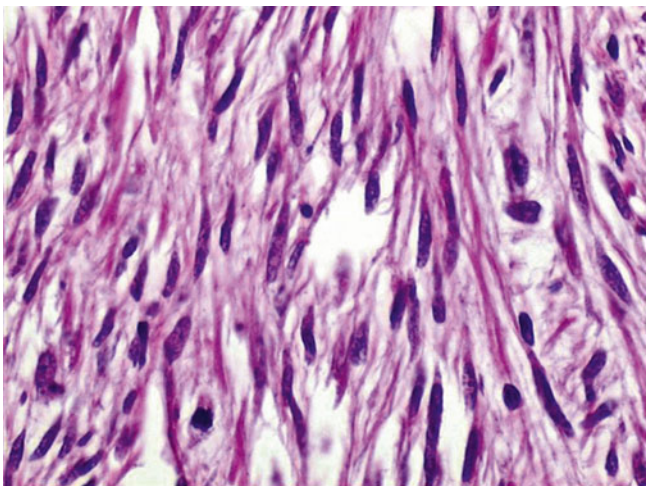

**Fig. 40.3** High-power microscopic view. Spindle cells with hyperchromatic nuclei, sometimes showing blunt ends. Atypical mitoses are frequent

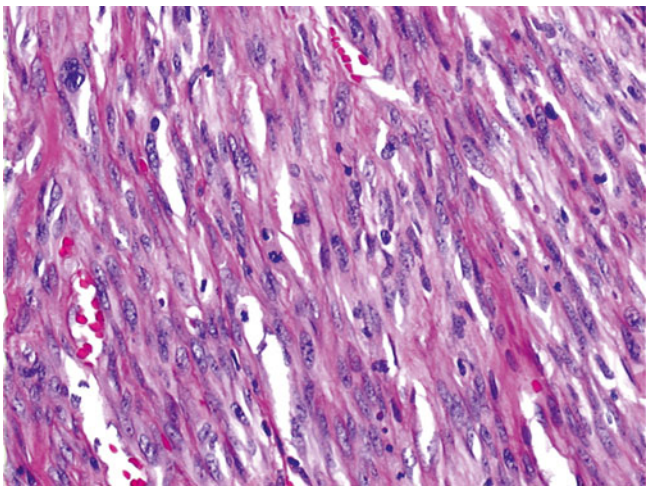

**Fig. 40.4** Medium-power microscopic view of a leiomyosarcoma of a higher grade than the previous figure. Immunohistochemistry may be needed to properly identify the neoplasia

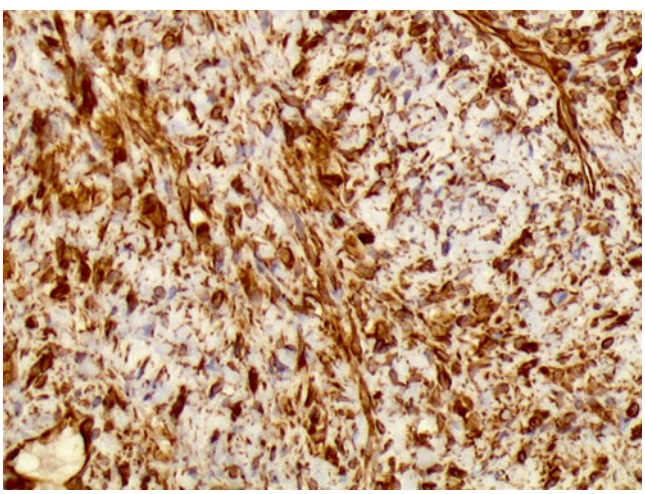

**Fig. 40.5** Immunohistochemistry – SMA positive in neoplastic cells

## Recommended Reading

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