Leiomyosarcoma of Bone

43

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43.1 Definition

- A rare malignant intraosseous neoplasm composed of spindle cells showing smooth muscle differentiation and devoid of matrix production.

43.2 Etiology

- Mostly unknown.
- Some cases secondary to radiation therapy or associated with Epstein-Barr virus (EBV) infection have been reported.

43.3 Epidemiology

- Slight male predominance.
- More common in adults.

43.4 Sites of Involvement

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

43.5 Clinical Symptoms and Signs

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

43.6 Imaging Features

43.6.1 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.

43.6.2 CT and MRI Features

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

43.7 Imaging Differential Diagnosis

43.7.1 Lymphoma of Bone

- May present similar imaging features.

43.7.2 Other Malignancies

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

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43.8 Pathology

43.8.1 Gross Features

- The cut surface of the tumor is fleshy and gray or pinkish white, with areas of necrosis (Fig. 43.1).
- Margins may be well-defined or poorly defined.

43.8.2 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 (Figs. 43.2 and 43.3). "Schoolfish" pattern.
- Osteoid is always absent.
- Epithelioid, myxoid, and pleomorphic variants have been reported.

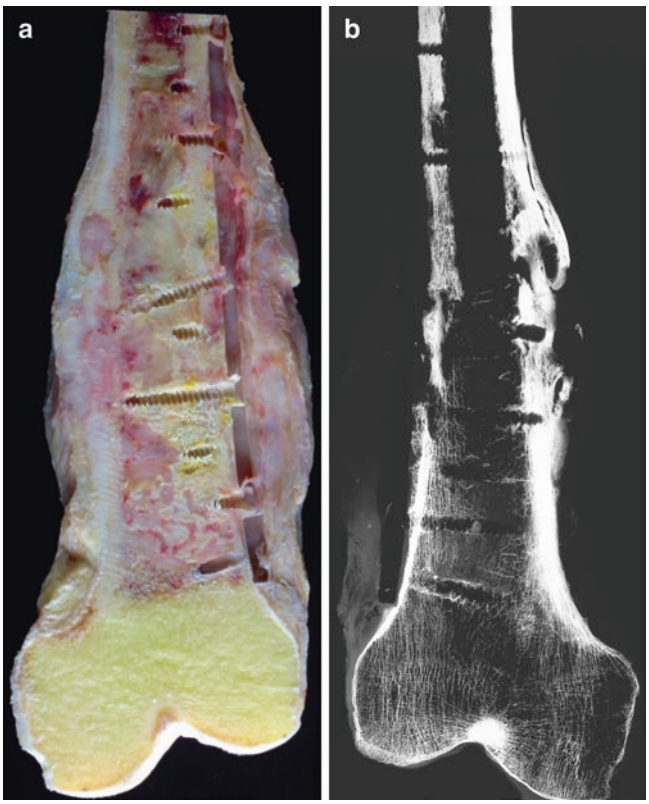

Fig. 43.1 Leiomyosarcoma of the metaphysis and diaphysis of a femur, shown in specimen photography and radiography. The lesion involved metallic implants from a previous surgical procedure. (**a**) Whitish-pink, firm and elastic lesion occupying the marrow space in a mostly longitudinal fashion, permeating the cortex and involving adjacent soft tissue. (**b**) Absence of mineral deposits in tumor tissue

43.9 Pathologic Differential Diagnosis

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

43.10 Ancillary Techniques

- Immunohistochemical techniques demonstrate positivity for muscle markers: desmin, h-caldesmon, and smooth muscle actin (SMA) (Figs. 43.4 and 43.5).

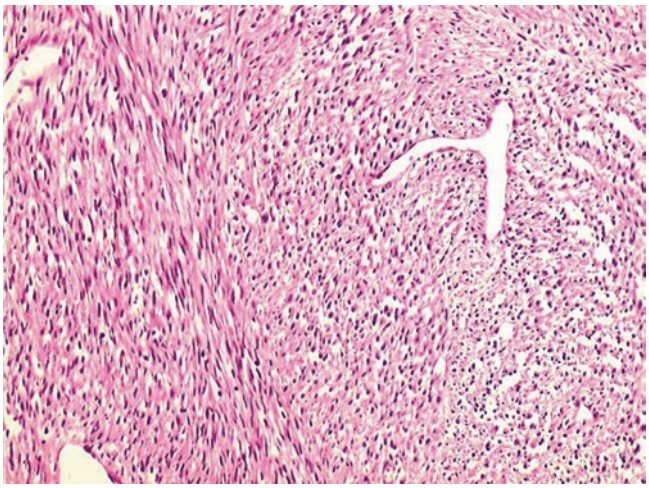

Fig. 43.2 Low-power microscopic view. Long and interwoven bundles of spindle cells

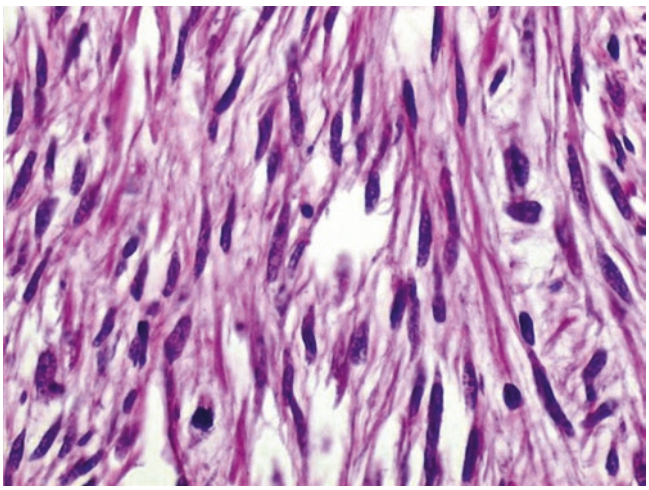

Fig. 43.3 High-power microscopic view. Spindle cells with hyperchromatic nuclei, sometimes showing blunt ends. Atypical mitoses are frequent

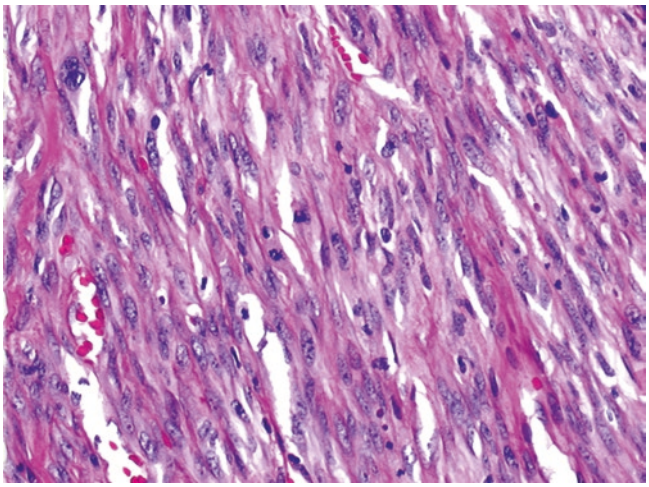

Fig. 43.4 Medium-power microscopic view of a leiomyosarcoma of a higher grade than the previous figure. Immunohistochemistry may be needed to properly identify the neoplasia

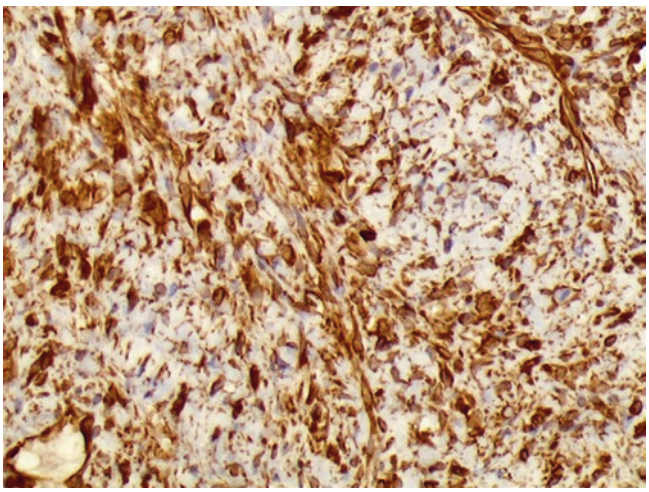

Fig. 43.5 Immunohistochemistry: SMA positive in neoplastic cells

43.10.1 Genetics

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

43.11 Prognosis

- Histologic grade correlates directly with rates of recurrence and metastasis.
- Metastasis at diagnosis and less than wide resection with free margins are adverse prognostic factors.
- Prognosis is statistically better than for other primary bone sarcomas of the same grade in patients without metastasis.

43.12 Treatment

- Surgical resection with wide margins combined with chemotherapy is the treatment of choice.

Suggested Reading

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