Fibrosarcoma of Bone

29

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Abstract

Fibrosarcoma of bone is a malignant spindle cell tumor characterized by a herringbone or fascicular disposition of atypical, monomorphic fibroblasts and is always negative for any specific marker, which makes this tumor a diagnosis of exclusion. It can be primary or, less frequently, secondary to radiation or other pathologic conditions. It represents from 2 % to 4 % of primary bone tumors and is reported to occur from the second to the seventh decades of life. Fibrosarcoma has a preference for the metaphyses of long bones, especially the distal femur. The more important prognostic feature is histological grade. Wide resection or amputation is the indicated treatment.

Definition

- A malignant spindle cell tumor of the bone, characterized by a herringbone or fascicular disposition of atypical, monomorphic fibroblasts.
- It can be primary or, less frequently, secondary to radiation or other pathologic conditions.

Etiology

- There are no known causes.
- Fibrosarcoma may appear secondarily on numerous conditions like fibrous dysplasia, Paget's disease, Ollier disease, osteochondromatosis, giant cell tumor, bone infarct, dedifferentiated chondrosarcoma, dedifferentiated parosteal osteosarcoma, and others.

Epidemiology

- Represents from 2 % to 4 % of primary bone tumors

Sex

- There is no clear gender predominance.

Age

- It is reported to occur from the second to the seventh decades of life.
- These historical figures, however, can be challenged by the recent introduction of ancillary techniques that exclude cases morphologically similar to fibrosarcoma but which can now be classified under other neoplastic categories.

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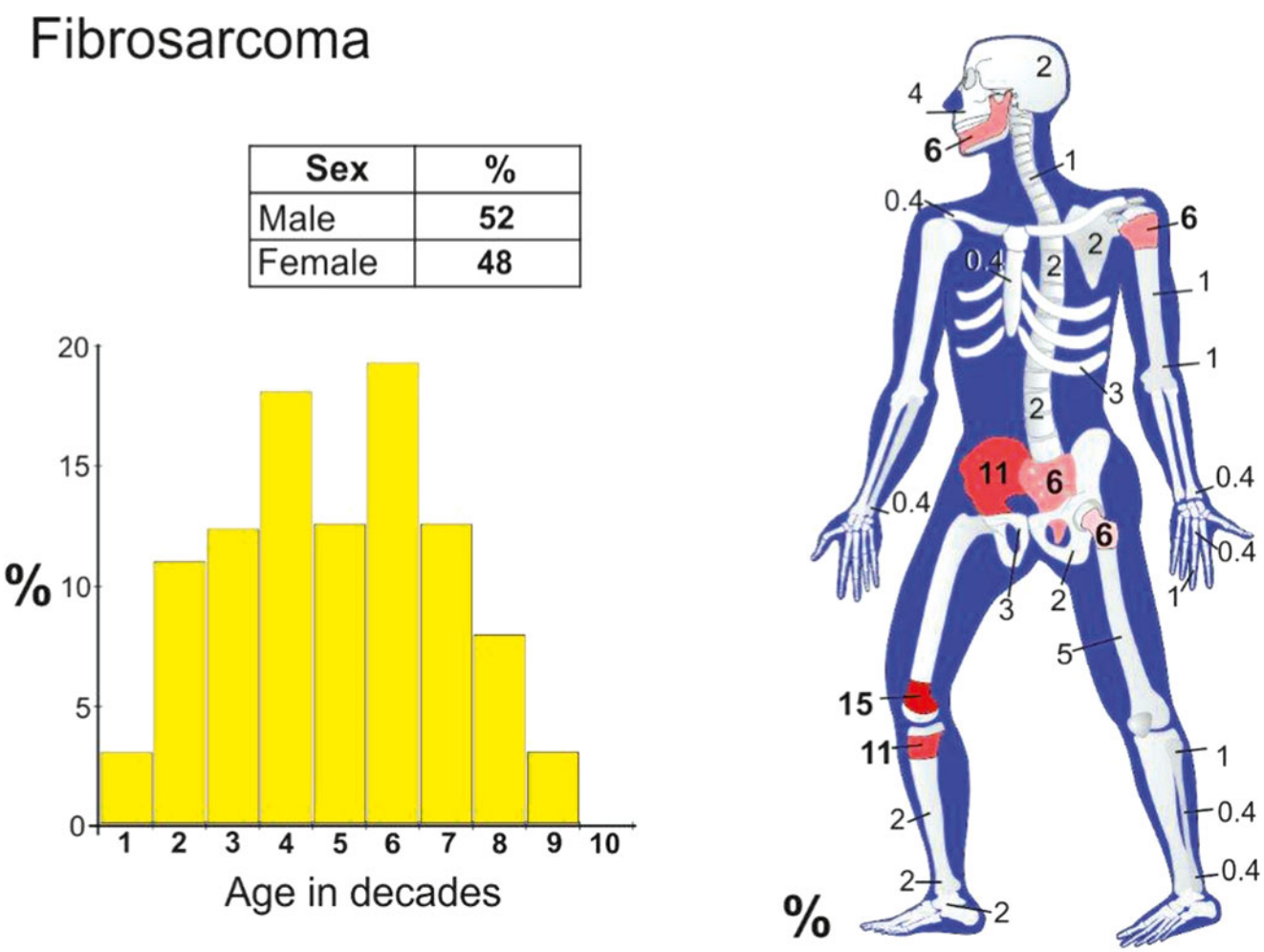

Sites of Involvement

- Just as for osteosarcoma, fibrosarcoma has a preference for the metaphyses of long bones, especially the distal femur, with the proximal femur, distal humerus, proximal tibia, and pelvis following in frequency.

Clinical Symptoms and Signs

- Local pain and/or the presence of a mass is the usual initial symptom of primary fibrosarcoma of the bone.
- Limitation of motion of a neighboring joint and pathological fracture can occur in some cases at presentation.

Image Diagnosis

Radiographic and CT Features

- Imaging methods show a metaphyseal eccentric lytic lesion, with a predominantly permeative pattern, without peripheral or periosteal bone reaction even after cortical permeation and soft tissue extension.
- There is no sign of mineral deposits or of bone production by the tumor.
- Geographical, permeative, or "moth-eaten" patterns can probably be related to different histological grades.
- MRI, and bone scan do not contribute with further data for the diagnosis.

Image Differential Diagnosis

Fibroblastic Osteosarcoma and Low-Grade Central Osteosarcoma

- These tumors present very scarce or variable bone production that when and if identified by imaging methods can suggest the correct diagnosis.

Desmoplastic Fibroma of Bone

- Fibrosarcoma shows indistinct borders due to tumor permeation, and expansion of the cortices is unusual.

Malignant Lymphoma of Bone
- In long bones, it is preferentially situated in the diaphysis. Both lesions can share the "moth-eaten" pattern; other infiltration patterns are more suggestive of fibrosarcoma.

Undifferentiated Pleomorphic Sarcoma/MFH
- Has no imaging differences in relation to high-grade fibrosarcoma

Pathology

Gross Features

- Fibrosarcoma presents a white and firm cut surface, with a trabeculated pattern and circumscribed margins.
- Higher-grade tumors present a more fleshy appearance and focal friable necrotic or hemorrhagic areas.
- Margins tend to be indistinct.

Histological Features

- Fibrosarcoma is composed by a uniform spindle cell population arranged in "herringbone" or fascicular pattern.
- Collagen production is variable and related to tumor grade, high-grade lesions scarcely showing collagen matrix.
- By definition, there can be no other kind of matrix production except pure collagen to categorize a neoplasia as fibrosarcoma.
- High-grade lesions also present more frequent mitotic and atypical nuclei as well as occasional areas of necrosis.

Pathology Differential Diagnosis

Fibrous Dysplasia
- If the sample does not include immature bone trabeculae, it may be difficult to separate, on exclusively histological grounds, from well-differentiated fibrosarcoma.
- Attention to infiltrated margins on histology and radiological signs of aggressiveness can help in the diagnosis.

Fibroblastic Osteosarcoma
- Always contains neoplastic bone trabeculae that must be sought extensively in these predominantly fibroblastic tumors

Desmoplastic Fibroma of Bone
- Has elongated strands of well-differentiated active-appearing spindle cells and is seldom permeative, less cellular, and less atypical than fibrosarcoma; mitoses, when present, are always typical.

Leiomyosarcoma of Bone
- Spindle cell bundles are disposed at cross angles and present a "schoolfish" pattern different from the herring bone predominant pattern of fibrosarcoma; nuclei are elongated and blunt ended (cigar-shaped).
- Positive for smooth muscle markers and keratin.

Synovial Sarcoma, Monophasic
- Spindle cells are densely arranged in vague fascicles (so dense that no matter how thin the histological section, these areas will always have a thick appearance).
- Mast cells are an almost constant finding and may be abundant.
- Calcification or ossification areas may also be extensive.
- Immunohistochemical techniques may be necessary to rule out this diagnosis: epithelial markers and CD99.
- Translocation t(X;18)(p11:q11) is found exclusively in SS.
- RT-PCR detection of SYT-SSX1/2 fusion transcripts is diagnostic of SS.

Myxofibrosarcoma
- Shows frequent areas of myxoid matrix alongside areas of more dense, collagenic matrix
- Intratumoral heterogeneity of low-grade and high-grade juxtaposed areas
- Scattered positivity for SMA marker

Undifferentiated Pleomorphic Sarcoma/MFH
- This is always a high-grade and pleomorphic lesion.

Ancillary Techniques

- Fibrosarcoma is always negative for any specific marker, which makes this tumor a diagnosis of exclusion.

Genetics

- A number of apparently specific mutations in KIAA1432, CA9, TLN1, and MELK were found in a limited number of sarcomas, which includes fibrosarcoma, MFH, and Ewing family of tumors.

Prognosis

- The more important prognostic feature is histological grade.
- Five-year survival rate varies accordingly from 83 to 34 %.
- Ten-year survival rates can be as low as 28 %.

- Metastases, when present, usually spread to the lungs and other bones.
- Local recurrence is related to a poor long-term prognosis.

Treatment

- Wide resection or amputation is the usually the indicated treatment.
- There is no data in the literature regarding chemotherapy as a standard treatment procedure.

Images

See Figs. 29.1, 29.2, 29.3, 29.4, 29.5, and 29.6 for illustrations of malignant fibrosarcoma of the bone.

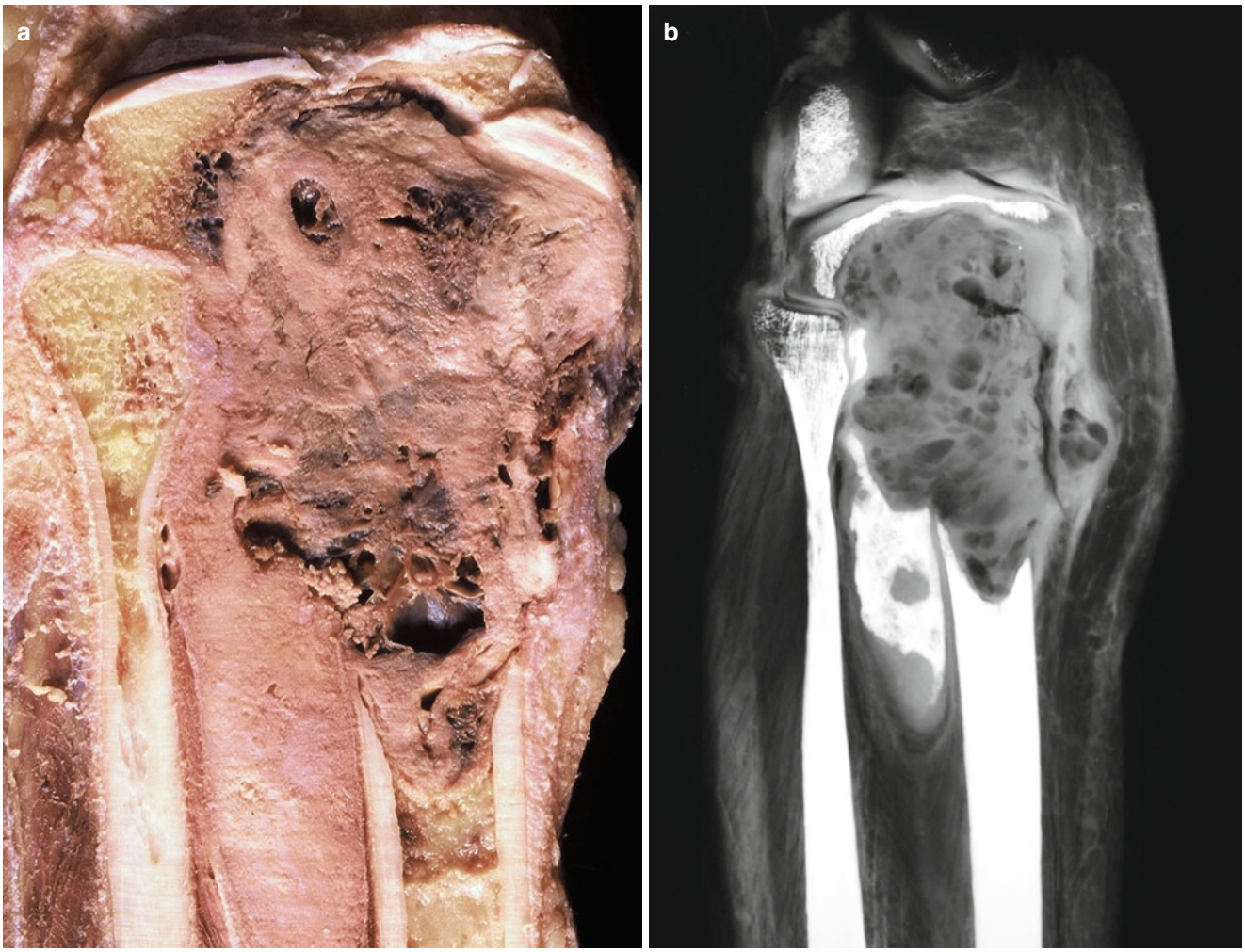

Fig. 29.1 (**a**) Macrophotography of a fibrosarcoma in the proximal end of the tibia. Lesion involves the entire width of the bone as well as the adjacent soft parts. *Pink-whitish* tissue with necrotic and hemorrhagic central areas. (**b**) Radiograph of the specimen in the previous picture depicting soft tissue lesion devoid of any bone or mineral deposits

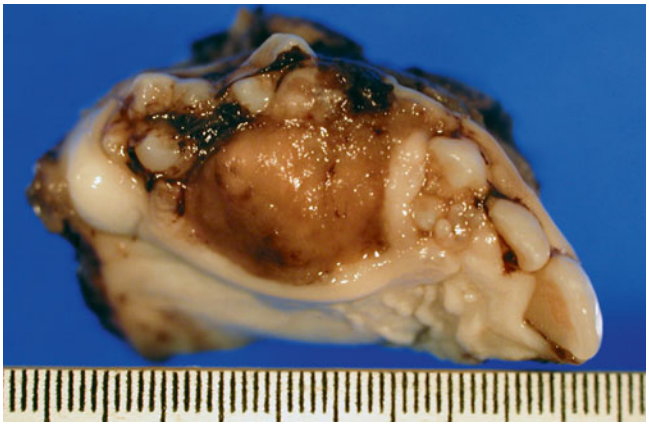

Fig. 29.2 Macrophotography of a fibrosarcoma of the maxilla, an infrequent site

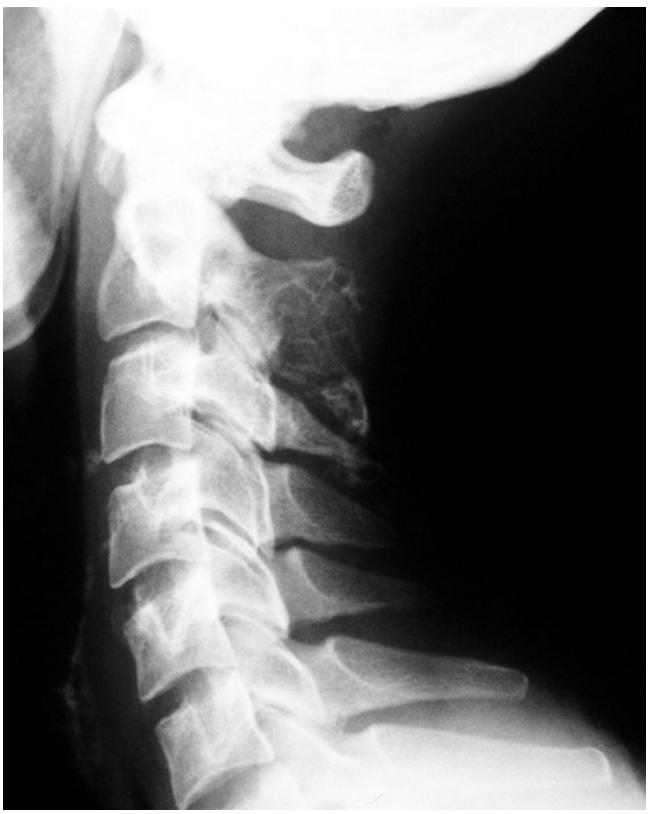

Fig. 29.3 Radiograph of a fibrosarcoma in the posterior vertebral elements, another rare site

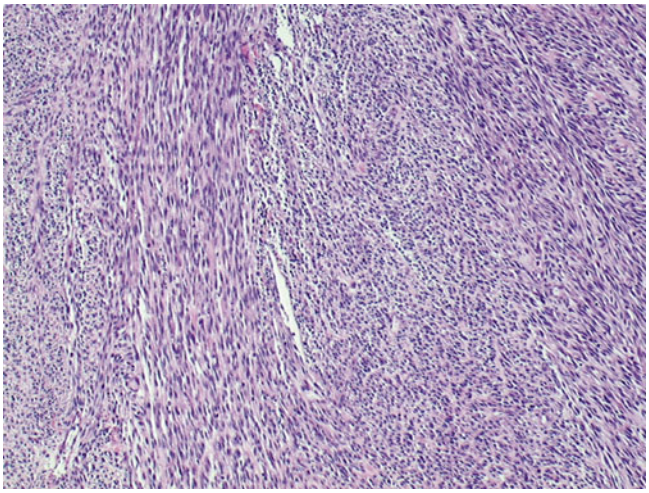

Fig. 29.4 Low-power microphotography. Grade 3 fibrosarcoma

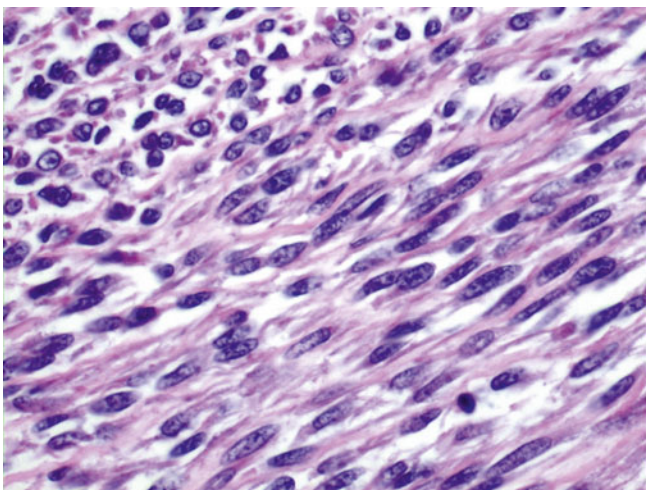

Fig. 29.5 High-power view of the same case as Fig. 29.4

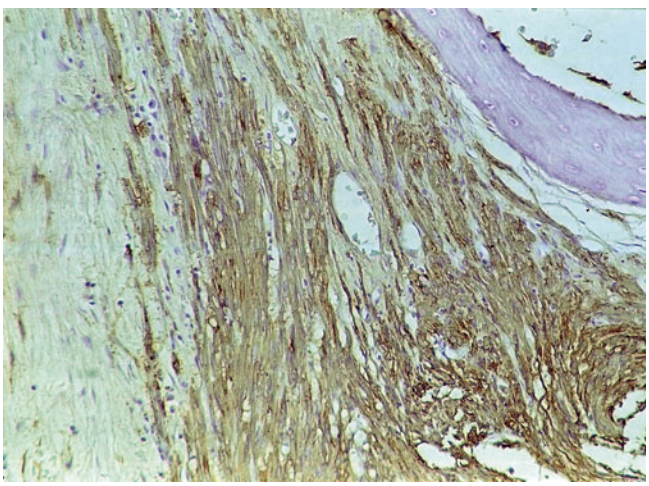

Fig. 29.6 Immunohistochemistry positivity for vimentin. No other marker was positive

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