Soft Tissue Tumors Rarely Presenting Primary in Bone; Diagnostic Pitfalls

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KEYWORDS

• Primary sarcoma of bone • Malignant fibrous histiocytoma • Hemangiopericytoma • Leiomyosarcoma of bone • Synovial sarcoma of bone

Key points

- Primary bone sarcomas (nonosteosarcoma, nonchondrosarcoma, and non-Ewing sarcoma) rank among the rarest malignancies and represent a major diagnostic challenge.
- The reappraisal of now obsolete entities, such as malignant fibrous histiocytoma (MFH) and hemangiopericytoma (HPC), has led to recognition of specific tumor entities.
- Leiomyosarcoma (LMS) represents the most common lesion; however, a broader variety of soft tissue sarcomas may rarely occur in bone.
- Diagnosis relies on integration of clinical, radiologic, immunomorphologic, and molecular findings, ideally in context of expert centers.

ABSTRACT

Primary bone sarcomas represent extremely rare entities. The use of now abolished labels, such as malignant fibrous histiocytoma and hemangiopericytoma, has significantly hampered the chance of identifying specific entities. It is now accepted that a broad variety of mesenchymal malignancies most often arising on the soft tissue may actually present as primary bone lesions. A more accurate morphologic partition is justified based on availability of distinct therapeutic options. An integrated diagnostic approach represents the only way to achieve a correct classification. In consideration of the significant complexity, primary bone sarcomas should ideally be handled in the context of expert centers.

OVERVIEW

When dealing with primary bone malignant mesenchymal tumors, pathologists are offered a limited number of diagnostic options, including mainly osteosarcoma, Ewing sarcoma, and chondrosarcoma. Aside from the intrinsic diagnostic challenges that these entities generate, a broader variety of lesions that arise predominantly in the soft tissues may present as a primary bone malignancy. In the past, most of these cases have been lumped within 2 main categories; MFH and

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fibrosarcoma. In the past 2 decades both MFH and fibrosarcoma underwent a profound conceptual reappraisal. The latest *WHO Classification of Tumours of Soft Bone and Soft Tissue Tumours*[1] has ultimately denied to MFH the dignity of a specific entity. This decision has been taken based on MFH at best representing a collection of unrelated pleomorphic sarcomas (ie, pleomorphic variants of rhabdomyosarcoma [RMS], LMS, and liposarcoma), even including nonmesenchymal lesions, such as metastatic sarcomatoid melanoma and carcinoma. The term, MFH, is currently replaced by the label, undifferentiated pleomorphic sarcoma (UPS), to designate those rare pleomorphic sarcomas in which no specific line of differentiation can be demonstrated. Fibrosarcoma, as defined by the presence of a spindle cell malignancy featuring a herringbone pattern of growth, in the current view corresponds to the fibrosarcomatous variant of dermatofibrosarcoma protuberans. There exist, however, a variety of fibroblastic sarcomas that includes distinctive entities, such as infantile fibrosarcoma, low-grade fibromyxoid sarcoma (LGFMS), sclerosing epithelioid fibrosarcoma (SEF), and myxoinflammatory fibroblastic sarcoma, that have also contributed to replacing fibrosarcoma as a defined clinicopathologic entity.

The process of reclassification was mostly generated within the soft tissue sarcoma community but, unavoidably, has also contaminated the bone sarcoma one. As a consequence, both MFH and fibrosarcoma in bone have also disappeared, replaced by morphologically distinct tumor entities.[2–5]

Sarcomas in general are known to represent one of the most challenging areas of surgical pathology; however, the situation in which a rare tumor arises in an exceptional location like bone generates even greater difficulty. This review is aimed at discussing the main clinicopathologic features of a selected group of malignant and locally aggressive and/or rarely metastasizing soft tissue lesions that can rarely occur primarily in the bone. Together they do not represent more than 5% of all primary bone malignancies.

LEIOMYOSARCOMA

Primary LMS of bone accounts for a majority of primary bone (nonosteogenic/chondrogenic and non-Ewing) sarcomas. They tend to occur in adult or elderly patients and the vast majority occurs in the long bones.[6–8] The femur is the most frequently affected site followed by the tibia and the pelvic bones.

Radiologically, LMS most often presents as a large, mostly lytic lesion, almost always breaking the cortex and extending into the surrounding soft tissue (**Fig. 1**).

Macroscopically, LMS exhibits a gray to white cut surface, associated in most cases with foci of necrosis and hemorrhage (**Fig. 2**). The lesion tends to be well demarcated, most often featuring extension in the soft tissues.

The microscopic features of LMS, similarly to its soft tissue counterpart, vary according to the degree of differentiation. Well-differentiated lesions are characterized by fascicles composed of spindle cells containing oval, blunt-ended nuclei and distinctive eosinophilic fibrillary cytoplasm

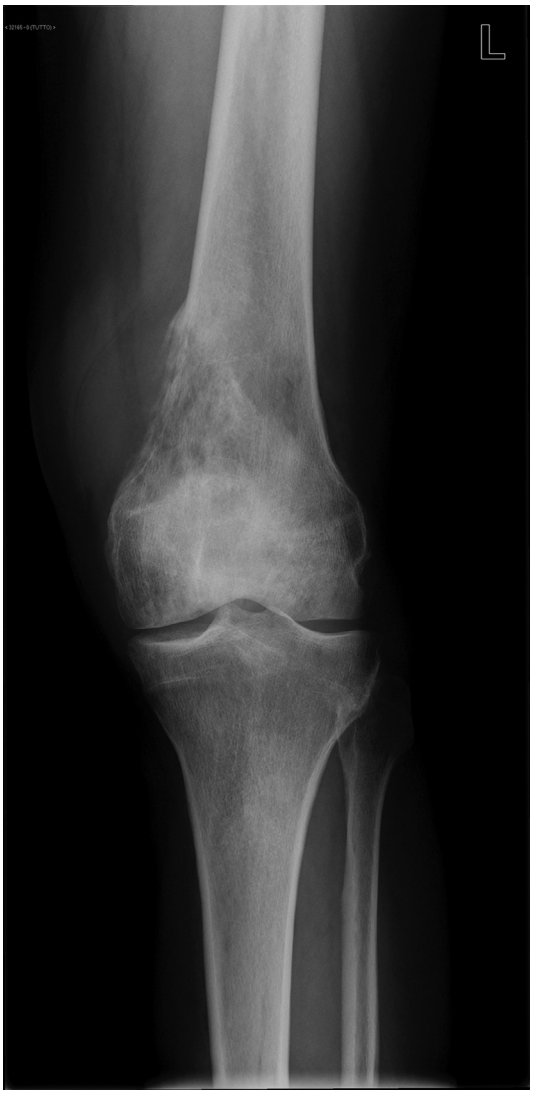

Fig. 1. LMS of bone: radiograph demonstrates a lytic lesion with cortical destruction and soft tissue infiltration.