

Chondro-Osseous Lesions of Soft Tissue



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KEYWORDS

- Myositis ossificans • Ossifying fibromyxoid tumor • Osteosarcoma • Chondroma
- Synovial chondromatosis

ABSTRACT

Soft tissue lesions can contain bone or cartilage matrix as an incidental, often meta-plastic, phenomenon or as a diagnostic feature. The latter category includes a diverse group ranging from self-limited proliferations to benign neoplasms to aggressive malignancies. Correlating imaging findings with pathology is mandatory to confirm that a tumor producing bone or cartilage, in fact, originates from soft tissue rather than from the skeleton. The distinction can have dramatic diagnostic and therapeutic implications. This content focuses on the gross, histologic, radiographic, and clinical features of bone or cartilage-producing soft tissue lesions. Recent discoveries regarding tumor-specific genetics are discussed.

OVERVIEW

A diverse group of somatic soft tissue lesions, reactive and neoplastic, may contain skeletal matrix (osteoid, bone and/or cartilage) and can be classified into 2 main categories (**Box 1**). First, bone or cartilage may be a secondary, incidental feature but its presence is not definitional. Variants of some neoplasms (eg, melanoma,^{1,2} carcinoma,³ “dedifferentiated” liposarcoma,^{4,5} low-grade fibromyxoid sarcoma,⁶ and schwannoma⁷) can contain bone or cartilage produced by the neoplastic component or as a meta-plastic feature. The second category consists of soft tissue lesions that are *defined* by the presence of skeletal matrix and is the subject of this review. This group spans a wide clinical spectrum from self-limited mesenchymal proliferations to high-grade malignancies. Interpretation of gross, microscopic, clinical, and radiographic findings, together,

is critical for accurate diagnosis. The unraveling of the genetics of some of these tumors has refined classification schemes and provided diagnostic adjuncts.^{8–10}

Imaging correlation is indispensable if a soft tissue tumor contains bone or cartilage. Foremost, imaging confirms that the lesion, in fact, *arises from soft tissue*. Serious diagnostic errors can be avoided by reviewing the imaging firsthand or, at least, discussing the findings with the radiologist or treating clinician. For example, a cartilage tumor that originates in soft tissue is almost certainly benign, whereas a cartilage tumor of bone that *invades* soft tissue is probably malignant. If imaging is unavailable, the pathologist should render a descriptive diagnosis with a reasonable differential, refining with imaging correlation later, rather than a specific, but potentially incorrect diagnosis.

Osteoid is an organic extracellular matrix composed predominantly of type 1 collagen with smaller amounts of glycosaminoglycans and other proteins producing a waxy eosinophilic quality on routine hematoxylin-eosin histology. *Bone* contains both the organic osteoid component and a mineral component known as hydroxyapatite: $\text{Ca}_{10}(\text{PO}_4)_6(\text{OH})_2$. Soft tissue lesions, like their skeletal counterparts, can contain 2 histologic forms of bone: woven and lamellar. Polarized optics can be helpful to distinguish the 2 types of bone (**Fig. 1**). Lamellar bone is always produced by benign osteoblasts, although they may be a secondary component in other malignancies. Woven bone is produced by both benign and malignant osteoblasts and reflects less organized collagen and rapid synthesis. *Cartilage* is an extracellular matrix composed of predominantly water and smaller amounts of proteoglycan and collagen (type 2). Neoplasms often contain these components in

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Box 1**Categories of soft tissue lesions with skeletal matrix*****Skeletal matrix is not definitional***

Bone-forming melanoma
 “Dedifferentiated” sarcomas
 Liposarcoma
 Leiomyosarcoma
 Malignant peripheral nerve sheath tumor
 Tenosynovial giant cell tumor
 Lipoma (chondrolipoma)
 Fat necrosis
 Nuchal fibrocartilaginous pseudotumor
 Osteoma cutis
 Fibroma of tendon sheath
 Low-grade fibromyxoid sarcoma
 Carcinoma
 Schwannoma

Skeletal matrix is definitional

Myositis ossificans
 Soft tissue aneurysmal bone cyst
 Florid reactive periostitis
 Bizarre parosteal osteochondromatous proliferation
 Subungual exostosis
 Ossifying fibromyxoid tumor
 Soft tissue osteosarcoma
 Soft tissue chondroma
 Synovial chondromatosis
 Chest wall hamartoma
 Chondro-osseous loose body
 Chondroid syringoma
 Mesenchymal chondrosarcoma
 Extraskelatal myxoid chondrosarcoma

abnormal ratios, resulting in a “liquefied” or myxoid matrix. The cartilage in soft tissue neoplasms is typically the hyaline type; fibrocartilage and elastic cartilage are exceptionally rare in this setting.

Reactive and neoplastic soft tissue lesions can also contain amorphous or crystalline stromal calcifications that may raise the differential of skeletal matrix. Examples include calcifying aponeurotic fibroma, synovial sarcoma, crystal deposition diseases, and phosphaturic mesenchymal tumor.^{11–13} Space limitations preclude a full discussion of each of these entities.

MYOSITIS OSSIFICANS AND RELATED TUMORS

Myositis ossificans represents the prototypical example of a family of tumors that (1) are composed of a population of fibroblastic-myofibroblastic cells; (2) produce osteoid, bone, and, less frequently, cartilage, in a reactive pattern; (3) often, but not invariably, arise after trauma; and (4) have a self-limited clinical course.¹⁴ The group (summarized in **Table 1**) contains analogous tumors that differ

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