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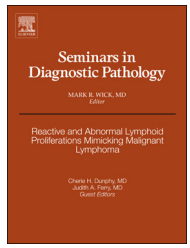

Vascular tumors of bone

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ABSTRACT

Vascular tumors of the bone represent a variety of neoplasms, ranging from benign hemangiomas and epithelioid hemangiomas to intermediate grade hemangioendotheliomas to frankly malignant angiosarcomas. Over the years, there has been considerable debate concerning the aggressivity, nomenclature, and mere existence of various nosologic entities, due to morphologic similarities and uncertainty regarding biologic behavior. Such debate has led to confusion among pathologists and clinicians, thus diminishing the prognostic implications in the diagnosis of these lesions. Here we review the current knowledge concerning the primary vascular neoplasms of the bone and correlate clinicopathologic features with tumor behavior.

Introduction

Vascular tumors of the bone comprise of a variety of neoplasms, in which constituent cells have undergone endothelial differentiation (Table). Similar to their soft tissue counterparts, these lesions range from common (usually asymptomatic) benign lesions to uncommon high-grade sarcomas with an aggressive clinical course. Here we review the primary vascular neoplasms of the bone.

Hemangioma

Primary osseous hemangioma is a fairly common neoplasm (affecting about 10% of the adult population), characterized by a well-demarcated proliferation of capillary-sized to cavernous vessels. Most cases are incidental findings, which are diagnosed on imaging alone based on their characteristic radiographic appearance, which includes coarse vertical trabecular thickening (sclerosis) within the otherwise osteopenic lesion, creating a "corduroy" pattern.[1] A minority of hemangiomas of the bone are symptomatic or have radiographic similarity to a more significant lesion, requiring surgical intervention or tissue diagnosis. These neoplasms are benign, and local recurrence is rare.

Primary osseous hemangioma may occur at any age, most frequently presenting in adults between 40 and 60 years of age, with a slight female predominance.[1] Vertebral bodies (thoracic and lumbar spine) are most commonly affected, followed by craniofacial and long bones. Lesions may be unifocal or multifocal, occasionally presenting in the setting of angiomatosis, in which multiple intra-osseous hemangiomas involve a large, localized skeletal area, or widespread bony lesions are present.[1] Involvement of multiple tissue planes adjacent to a vertebral hemangioma (e.g., skeletal muscle, epidural fat, skin, and even spinal cord) can suggest an aggressive tumor on imaging studies.

Microscopically, tumors are composed of capillary-sized to cavernous vessels set in a loose edematous stroma (Fig. 1A and B). Vessels surround sclerotic intra-lesional trabeculae; however, the lesion itself is well-demarcated, with respect to the adjacent medullary constituents. Vascular spaces are lined by a single layer of bland, mitotically inert endothelial cells. Occasional cases contain blood-filled cavities and

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Table – Typical histologic features of vascular tumors of bone.

Tumor	Aggressive growth	Solid sheets	Vascular channels	Blister cells	Atypia	Mitoses	Necrosis	Positive IHC and molecular findings
H	No	No	Yes	No	No	Rare	No	CD31, CD34, and ERG
EH	Yes	±	Yes	±	Mild	<1/10	Rare	CD31, CD34, ERG, and CK±
ES-H	Yes	Yes	No	No	Mild	<1/10	<20%	ERG, FLI1, CK (AE1/3), INI1 (retained), CD31±, and CD34 negative
EHE	±	No	No	Yes	Mod[a]	<3/10[a]	Rare	CD31, CD34, ERG, FLI1, CK (30%), and t (1;3)
AS	Yes	Yes	Yes	±	Severe	Many	Yes	CD31, CD34, ERG, FLI1, and CK (65%)

IHC = immunohistochemistry; H = hemangioma; EH = epithelioid hemangioma; ES-H = epithelioid sarcoma-like hemangioendothelioma; EHE = epithelioid hemangioendothelioma; AS = angiosarcoma; CK = cytokeratins; EMA = epithelial membrane antigen; ± = variable; Mod = moderate.
[a]High-risk EHE may display marked atypia and increased mitoses.

intra-vascular thrombi, which may be associated with papillary endothelial hyperplasia. As hematopoietic elements are lost, the native adipose tissue becomes more prominent, occasionally dominating the lesion. Endothelial cells express CD31, CD34, ERG, and other vascular markers.

Epithelioid hemangioma

Epithelioid hemangioma (EH) is a benign, but locally aggressive vascular neoplasm, which may demonstrate bony expansion with cortical erosion and soft tissue extension, albeit a broad, pushing border (Fig. 1C and D). EH has an approximately 10% local recurrence risk, and rare cases reportedly may develop regional lymph node metastases[3,4]; widespread metastasis or death from disease has not been reported, hence its designation as a benign neoplasm. EH of the bone shows certain histologic differences from its soft tissue counterpart including a more solid growth pattern and lack of association with a damaged blood vessel.

EH is an uncommon tumor, with most cases occurring in the third to sixth decades, although it may present at any age. The long tubular bones are most commonly involved, followed by the distal lower extremities, flat bones, vertebrae, and hand bones.[3] Approximately 20% of cases are multifocal and rare cases present with concomitant extra-osseous EHs.[3]

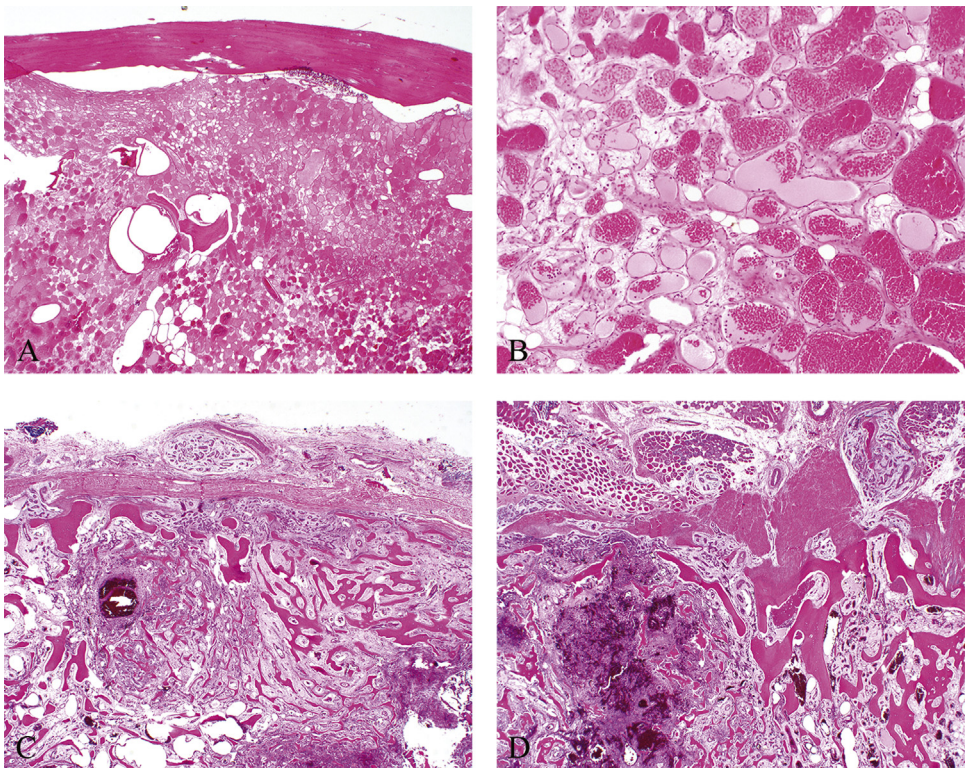

Fig. 1 – Osseous hemangioma (A and B) does not infiltrate through the overlying cortical bone. It is composed of well-formed blood vessels lined by bland endothelial cells similar to its soft tissue counterpart. Epithelioid hemangioma of bone (C and D) encases the native bone trabeculae and often displays locally aggressive behavior including erosion through the cortical bone and invasion into adjacent soft tissue.