Musculoskeletal Angiomatous Lesions

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Angiomatous Lesions
- Hemangioma
- Lymphangioma
- Glomus tumor
- Angiomatosis and associated syndromes
- Hemangiendothelioma
- Hemangioendothelioma
- Angiosarcoma

Osseous Hemangioma
Clinical Characteristics
- Male > female = 2:1; fourth-fifth decade
- Majority asymptomatic
- May have soft tissue components
- Common sites: vertebral body (11% of spines; 25% multifocal), calvarium (30% of lesions)

Soft Tissue Hemangioma: Clinical Characteristics
- 7% of all benign soft tissue neoplasms
- 1%–4% of the general population; 10% children under 1 year old; 20% multiple
- Most frequent soft tissue neoplasm in children
- More common in young women – may increase in size with pregnancy
- Subcutaneous, intramuscular, or synovial

Hemangioma: Pathology
- Subtype based on predominant vascular component but usually mixed tumor
- Capillary: most common – first years of life – skin, subcutaneous, vertebrae (ISSVA – infantile hemangioma - high/low flow)
- Cavernous: childhood – larger and deeper (low flow)
- Arteriovenous: deep or superficial – persistent fetal capillary bed (high flow)
- Venous: adults – deep involvement (low flow) retroperitoneum, mesentery, or extremities
- Epithelioid: dermis/subcutis

Hemangioma or Vascular Malformation
- Vascular malformation: dysplastic vessels without cellular proliferation or regression and present at birth
- Hemangioma: rapid growth and cellular proliferation of endothelial cells in neonatal period followed by slow involution
- Hemangioea: benign nonreactive process with increased number of normal or abnormal appearing vessels

Osseous Hemangioma: Radiology
[Figures 1 to 5]
- Vertebrae: focal or diffuse – vertical striations (corduroy or polka dot) posterior element involvement more likely symptomatic
- Calvarium/mandible radiating weblike trabecular pattern
- Long bone: multifocal lytic “honeycomb” pattern, cortical lesions/erosions in diaphysis
- Bone overgrowth
- Arthritis from intraarticular bleeding

Figure 1 A & B
Vertebral hemangiomas with thickened vertical trabeculae (arrows show corduroy appearance) on radiograph and coronally sectioned gross specimen (different patients).

Figure 2 A, B, C & D
Vertebral hemangioma (asymptomatic) of lumbar spine with “polka dot” appearance and fat between trabeculae on CT.
Soft Tissue Hemangioma Radiology:

Soft Tissue Changes  
[Figure 6]  
- Calcification: curvilinear or amorphous, phlebolith (30%–50% of lesions)  
- Angiography: irregular enlarged feeding arteries, contrast pooling, arteriovenous shunting  
- Venous lesions seen only with venography

Soft Tissue Hemangioma: MR Imaging  
[Figures 7 to 12]  
- T1-weighted images – low to intermediate heterogeneous mass; look for fat overgrowth  
- Very high intensity T2-weighted images (low flow)  
- Serpentine vessels/cavernous spaces may help distinguish subtypes  
- Distinguish high- vs low- flow lesions  
- In our experience approximately 90% of lesions have a pathognomonic imaging appearance and do not require biopsy

Soft Tissue Hemangioma: Other Imaging  
[Figures 7 to 12]  
- Enhance with contrast  
- Phleboliths: CT > MRI  
- Bone scan: often only limited activity  
- Sonography: solid mass, Doppler may show low resistance flow

Figure 3 A, B & C  
Vertebral hemangioma (symptomatic patient) with “polka dot” appearance on CT (arrowheads) and soft tissue extension. Sagittal T1-weighted and T2-weighted MR images show vertebral fracture and spinal canal compromise caused by anterior epidural soft tissue component (arrows), but diagnostic trabecular thickening is difficult to appreciate.

Figure 4 A, B & C  
Calvarial hemangioma with “spoke wheel” pattern of trabecular thickening (arrows) on radiograph, CT and vascular channels/spaces on gross specimen (arrowheads).

Figure 5 A & B  
Femoral hemangioma on coronal T1-weighted MR image before and after contrast shows multifocal round areas of marrow replacement (arrowheads) representing vascular channels with enhancement and serpentine feeding vessels (arrows).
Figure 7
A & B
Soft tissue hemangioma of axilla (intramuscular and cavernous) on CT shows enhancing vascular channels (thick arrows), fat overgrowth (thin arrows), and phlebolith (arrowhead).

Figure 8 A, B & C
Soft tissue hemangioma of forearm (intramuscular and cavernous) on sagittal T1-weighted MR images before and after gadolinium showing intermediate signal intensity serpentine vascular channels and spaces (arrows) that enhance following contrast and fat overgrowth (arrowheads). Axial T2-weighted MR image reveals multiple circular high-signal areas corresponding to slow-flow cavernous spaces (*).

Figure 11
Soft tissue hemangioma of forearm (intramuscular and cavernous) on sagittal T1-weighted MR images before and after gadolinium showing intermediate signal intensity serpentine vascular channels and spaces (arrows) that enhance following contrast and fat overgrowth (arrowheads). Axial T2-weighted MR image reveals multiple circular high-signal areas corresponding to slow-flow cavernous spaces (*).

Figure 10 A & B
Soft tissue hemangioma (arrows) with associated fat atrophy (arrowheads) in surrounding thigh muscle on gross specimen (left image). Histology (right image) reveals phleboliths (**) with calcification peripherally (arrows) and fat atrophy of muscle (arrowheads).

Figure 12
A, B & C
Capillary hemangioma on T2-weighted MR image (arrow) with nonspecific high-signal intensity in the face with typical extensive strawberry nevus clinically. No characteristic features of fat overgrowth or serpentine vascular structures are seen to suggest hemangioma as the vessels in this type of lesion are too small (capillary) to discern on imaging as demonstrated on the histology.
Hemangioma: Treatment
- Medical therapy – observation, propranol, steroids, alpha interferon, chemotherapy
- Surgical resection/laser therapy
- Sclerotherapy (low-flow lesions 90%)
- Embolization (high-flow lesions 10%)
- Radiation – symptomatic unresectable lesions – spine
- Vertebroplasty
- Recurrence (15%–30%) – large lesions

Lymphangioma (Lymphatic Malformation): Clinical Characteristics
- Rare lesion in bone, usually soft tissue
- Often present at birth (50%–65%)
- 90% apparent by 2 years old
- Head, neck, axilla – 75% of cases
- Soft fluctuant mass

Lymphangioma (Lymphatic Malformation): Pathology
- Sequestrated noncommunicating lymphoid tissue (macrocystic or microcystic)
- Large multiloculated cystic spaces
- Lined by lymphatic endothelium
- Filled with proteinaceous material

Lymphangioma (Lymphatic Malformation): Radiology
- Radiographs: soft tissue mass
- Imaging: large cystic spaces less common serpentine component may appear complex – solid components (high signal on T1 25%)
- Cystic hygroma: hydrops fetalis, Turner syndrome

Glomus Tumor
- Patients fourth to fifth decade
- Tumor of neuromyoarterial glomus
- Almost all terminal phalanx soft tissue
- Bone erosion/invasion (15%–65%)

Angiomatosis
- Multifocal or diffuse infiltration of bone by hemangiomatous or lymphangiomatous lesions with or without soft tissue involvement

Angiomatosis: Clinical Characteristics
- Young patients – first 3 decades
- Male > female = 2:1
- Osseous involvement only – benign course
- Visceral involvement – poor prognosis
- No malignant potential

Angiomatosis: Pathology
- Capillary or cavernous hemangioma
- Lymphangioma: lymphatic backflow
- Mixed vascular lesion difficult to distinguish
Angiomatosis: Radiology

- MR/CT imaging: same as solitary angiomatous lesions more extensive
- Imaging to evaluate visceral involvement/extent
- Lymphangioma: proven with lymphangiography and contrast in lesion
- Diffuse round/oval medullary lytic lesions
- May have sclerotic margins
- Location: femur, ribs, spine, pelvis, humerus, scapula, other long bones, clavicle

Angiomatous Syndromes

- Maffucci syndrome
- Osler-Weber-Rendu
- Klippel-Trenaunay-Weber
- Massive osteolysis of Gorham
- Associated osteomalacia and thrombocytopenia

Maffucci Syndrome

- Multiple enchondromata
- Cavernous soft tissue hemangioma
- Often hands/feet, unilateral predominance
- Malignant potential both lesions and viscera

Osler-Weber-Rendu Syndrome

- Hereditary hemorrhagic telangiectasia
- Dilated capillaries and veins
- Autosomal dominant
- GI, GU, lung, spinal; bone – rare

Klippel-Trenaunay-Weber Syndrome

- Nonhereditary, lower extremity (95%)
- Unilateral cutaneous capillary hemangioma
- Varicose veins and local gigantism
- Can have arteriovenous component

Massive Osteolysis of Gorham: “Vanishing Bone” Disease

- Patients < age 40 years old
- History of trauma (50%)
- Upper extremity favored, may extend across joint
- Progressive bone resorption and fragmentation (simulate neuropathic)
- Pathology: proliferating vascular channels

Gorham “vanishing bone” disease involving the foot with radiograph and gross specimen showing extensive sharply defined bone resorption (arrowheads and arrow).
Musculoskeletal Radiology

**Musculoskeletal Angiomatous Lesions**

- Tumor-induced osteomalacia – most frequent vascular lesions – phosphaturic mesenchymal tumor
- Hemangioma/hemangiopericytoma
- Kasabach-Merritt syndrome – hemangioendothelioma/hemangioma/hemangiopericytoma associated with thrombocytopenia and purpura

**Intermediate to Malignant Musculoskeletal Angiomatous Lesions**

- Hemangioendothelioma
- Hemangiopericytoma
- Angiosarcoma

**Hemangioendothelioma**

- Intermediate: benign or malignant
- Composed of vascular endothelial cells
- Often in young patients
- Bone or soft tissue
- Locally aggressive, unusual to metastasize

**Hemangiopericytoma**

- Intermediate: benign or malignant (more likely in large lesions >10 cm)
- Same as extrapleural solitary fibrous tumor
- Tumor of cells around vessels – pericytes
- Tumor of middle-aged adults; male = female
- Sites: soft tissue of lower extremity (35%); head/neck, pelvis, and retroperitoneum (25%)
- Rare in bone

**Angiosarcoma**

- Malignant; male > female = 2:1
- Composed of hemangiosarcoma or lymphangiosarcoma cellular elements
- Location: skin, muscular, viscera, bone
- Associated with lymphedema post-mastectomy (Stewart-Treves syndrome)

**Osseous Hemangioendothelioma, Hemangiopericytoma, and Angiosarcoma: Skeletal Location**

- Hemangioendothelioma: skull, vertebrae, lower extremity
- Hemangiopericytoma (rare): pelvis, proximal long bones, vertebrae, mandible
- Angiosarcoma: long tubular bone lower extremity

**Osseous Hemangioendothelioma, Hemangiopericytoma, and Angiosarcoma: Radiographic Findings**

*Figure 19*

- Multifocal lytic lesions, “honeycomb” appearance
- Aggressive bone destruction with expansion and soft tissue mass

**Radiology of Hemangioendothelioma, Hemangiopericytoma, and Angiosarcoma: Advanced Imaging**

*Figures 20 to 22*

- Angiography: intensely vascular with peripheral vessels displaced by tumor early; dense blush late
- Sonography: hypo- or hyperechoic mass
- Doppler: arteriovenous shunting
- MRI
  - T1-weighted: usually similar to muscle
  - Can be high-intensity hemorrhage
  - Feeding vascular pedicle (35% hemangiopericytoma)
- Look for prominent serpentine high-flow vessels and solid regions (biopsy with caution)
- Fluid levels, contrast enhancement
- Dominant skin mass in chronic lymphedema (angiosarcoma)
Musculoskeletal Radiology

Musculoskeletal Angiomatous Lesions

Cannot distinguish hemangioendothelioma, hemangiopericytoma, or angiosarcoma from other soft tissue masses if prominent serpentine vessels are not recognized. Hemangioendothelioma, hemangiopericytoma, and angiosarcoma cannot be differentiated from each other radiologically.

Distinction of Hemangioendothelioma, Hemangiopericytoma, and Angiosarcoma from Hemangioma

- Large masses
- Aggressive characteristics with infiltration
- No fat overgrowth

Treatment and Prognosis:
Hemangioendothelioma, Hemangiopericytoma, and Angiosarcoma

- Surgical resection
- Malignant lesions: radiation and chemotherapy
- Local recurrence common
- Metastases common to lung in angiosarcoma

Figure 21 A, B & C

Hemangiopericytoma in the thigh showing high-flow vessels (arrows) in the soft tissue mass (*) and feeding the lesion on both axial T1-weighted and coronal T2-weighted MR images. Photograph of the sectioned gross specimen also shows the soft tissue mass (*) and the high-flow vessels (arrowheads).

Figure 20 A, B, C & D

Angiosarcoma developing in a patient with chronic leg lymphedema. Axial T1-weighted, T1-weighted postcontrast, and T2-weighted MR images show the enlarged leg with subcutaneous edema (arrows) and dominant skin mass (arrowheads) representing the angiosarcoma. The superficial angiosarcoma is also seen on the clinical photograph (black arrows).
Summary:
Musculoskeletal Angiomatous Lesions

- Osseous – multifocal bone lysis
  - “Honeycomb” appearance
- Soft tissue – MRI
  - Look for serpentine vascular pattern
  - Overgrowth of fat
- Multiple associated syndromes and angiomatosis
- Higher grade lesions – hemangioendothelioma, hemangiopericytoma, angiosarcoma
  - Larger aggressive lesions
  - Infiltrative characteristics

References