Sonography of soft-tissue vascular lesions

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Declaration of Disclosure

- I have no actual or potential conflict of interest in relation to this presentation
Objectives

- Categorize vascular anomalies using the most recent classification
- Describe sonographic features of soft-tissue vascular anomalies
- Explain the importance of clinical information in the sonographic diagnosis of vascular anomalies
Indications for Imaging

- Superficial lesion but atypical presentation
- Deep lesion difficult to assess on physical examination
- Prior to therapy to define nature and extent and as a baseline
Imaging

- Ultrasound is first line modality for most cases
- MRI may be considered first line in
  - very large lesions
  - complex combined vascular malformations affecting entire extremity
  - assess deep extent in areas difficult to evaluate with US (orbit, intracranial, etc)
Ultrasound Technique

- Transducer selection determined by size & depth of lesion – higher frequency possible
- Larger & deeper lesions may require combination of transducers
- Color & Spectral Doppler are crucial but may be limited by patient motion and cooperation
- Compression
ISSVA Classification

- Updated in 2014
- Available at issva.org

- Vascular tumors
- Vascular malformations
Vascular tumors

- **Benign**
  - Infantile hemangioma
  - Congenital hemangiomas (RICH, PICH, NICH)

- **Locally aggressive or borderline**
  - Kaposiform hemangioendothelioma

- **Malignant**
  - Angiosarcoma
Infantile hemangioma

- Endothelial tumor
- Precursor lesion at birth
- Growth start few weeks after birth
- Proliferative phase until end of first year
- Slow involutive phase that takes years
Infantile hemangioma

- Location: head & neck (60%), trunk (25%), extremities (15%)
- Superficial: strawberry mark
- Deep: compressible, rubbery, bluish or normal skin, with visible draining veins
Infantile hemangioma

- Well-defined soft-tissue mass
- Variable echogenicity
  - Hypoechoic (65%)
  - Hyperechoic (19%)
  - Heterogeneous (16%)
- Calcifications are rare (8%)

Paltiel HJ et al (Radiology 2000; 214:747-754)
Infantile hemangioma

- Hypervascular with arterial and venous flow
  - ≥5 vessels/cm²
- High velocity arteries
  - Mean 28.4 ± 5 cm/s

Paltiel HJ et al (Radiology 2000; 214:747-754)

11w♀: Left breast mass
Infantile hemangioma

3m♀: Chest wall lump
Infantile hemangioma

8m♂: Chest wall lump
Infantile hemangioma

4m♀: Posterior neck mass

Courtesy Dr. Hameed, Evelina Children's Hospital, London, UK
Involved infantile hemangioma

9y♂: Hemangioma in forearm as a toddler with residual deformity ?resolution
Congenital hemangiomas

- Rapidly involuting (RICH)
- Non-involuting (NICH)
- Partially involuting (PICH)
Rapidly involuting congenital hemangioma (RICH)

- Hemangioma fully developed at birth
- Early involution in infancy, complete regression 12 to 18 months of age
Non-involuting congenital hemangioma (NICH)

- Present at birth
- No involution
- Proportional growth with child
Partially involuting congenital hemangioma (PICH)

- Hemangioma fully developed at birth
- Initial rapid involution in infancy, like RICH, but incomplete
- Evolves into NICH
Congenital hemangiomas

- Differentiation is based on clinical findings & evolution - not on imaging findings
- High vascular density similar to infantile hemangioma
- Distinct US features:
  - heterogeneous echogenicity (visible vessels on gray scale imaging)
  - calcifications

1d♀: Right chest wall mass diagnosed antenatally
8y♀: Lesion right mandibular area since birth, growing slowly with child, initially interpreted as venous malformation
Kaposiform hemangioendothelioma

- Intermediate (locally aggressive)
- Blue-red lesion
- Mostly presenting <1y
- Strong association with Kasabach-Merritt phenomenon (56-71%)
- Distinct histology, lymphatic abnormalities
- Mortality rate 12-24% (hemorrhage, local invasion)
Kaposiform hemangioendothelioma

- Ill-defined soft-tissue mass
- Involvement of multiple planes
- Heterogeneous
- Ca++ can be present
- Moderate to high vascular density

Dubois J et al (AJR 2002; 178:1541-1545)
Kaposiform hemangioendothelioma

2d♂: Left thigh mass diagnosed antenatally
Vascular malformations

- Congenital developmental anomalies
- Anomalous blood vessels/lymphatics without cellular hyperplasia
- Present at birth but not always evident
- Commensurate growth with child
- No tendency to involution
- Growth stimulated by trauma, clotting, hormones (puberty/pregnancy)
Vascular malformations

- AVM, AVF → High flow
- Capillary, venous, lymphatic → Low flow
- Combined
Arterio-venous malformation

- Fast-flow
- Complex network of primitive vessels directly connecting feeding arteries to draining veins
- Partial or complete absence of normal intervening capillary network
Arterio-venous malformation

- Multiple dilated, tortuous arteries & veins
- Sometimes no gray-scale abnormalities
- No discrete soft-tissue mass

13y♀: Pain and swelling, mostly with activities, over lateral malleolus for last 6 months
Venous malformation

- May present as varicosities
- Often as localized mass (solitary or multiple) – misnamed as cavernous or venous hemangioma
- If superficial: bluish, easily compressible, cold, enlarge with Valsalva maneuver, crying and when dependent
- If deep, difficult clinical diagnosis
Venous malformation

- Well-defined, hypoechoic, heterogeneous sponge-like mass
- Poorly marginated collection of veins
- Isoechoic thickening of subcutaneous tissues
- Compressibility
- Presence of phleboliths (16%)
- Low-velocity venous flow (84%)
- Absent flow (16%)

Trop I et al (Radiology 1999; 212:841-845)
Venous malformation

3y♂: Lump in left forearm
Venous malformation

3y♂: Swelling left posterior chest wall
Venous malformation

3y♂: Swelling left posterior chest wall
Lymphatic malformation

- Often referred to as lymphangioma
- 65-75% detected at birth
- May become larger and clinically evident due to hemorrhage or infection
- Common sites: neck, axilla, groin, chest wall
Lymphatic malformation

- Macrocystic (cystic hygroma)
- Microcystic
- Combined
Lymphatic malformation

Macrocystic

- Large, cystic cavities, separated by septa
- Debris may be present within cysts
- Arterial and venous flow only in septa

3y♀: Left arm mass
Lymphatic malformation

Microcystic
- Hyperechoic
- Ill-defined
- Small cysts <1 cm, may not be visible

5y♀: Pretibial swelling
Combined lymphatic malformation

8m♂: Left arm & chest wall mass
Summary

- US is useful for diagnosis of vascular anomalies
- US findings are better interpreted in conjunction with clinical findings
- Familiarity with ISSVA classification facilitates communication and collaboration with other clinicians