Hemangiomas and Other Vascular Tumors

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Objectives

• Review clinical and imaging characteristics of infantile hemangiomas and other vascular tumors with particular attention to the revised ISSVA classification.

History

• 1982 – Mulliken & Glowacki – histology & behavioral characteristics described
• 1992 ISSVA formed, 1996 classification created
  – Increasing # of vascular lesions recognized as histologically distinct entities
  – Interval advances in understanding genetics and behavior of some lesions
  – Updated classification 2014 to guide appropriate therapies

Misuse of nomenclature remains widespread in the literature

• Risk of inappropriate therapy
• Best approach is multidisciplinary vascular anomalies clinic
  – Hematologist-oncologist
  – Surgeon
  – Dermatologist
  – Pathologist
  – Radiologist/interventional radiologist

New ISSVA Classification

• Fundamental classification remains
  – Vascular tumors vs malformations
    • True neoplasms with cellular (endothelial) proliferation vs congenital errors of vessel formation
    • Lesions grow independent of patient size vs lesions grow commensurate with the child
    • Malformations grow rapidly if hemorrhage, infection, or during periods of hormonal stimulation (puberty, pregnancy)
• Addition of evolving category of provisionally unclassified vascular anomalies

Disclosures

• No relevant financial disclosures
ISSVA Classification

- Subdivisions and lesion assignment/nomenclature modified to be more histologically precise
- Vascular tumors
  - Benign
  - Locally aggressive or borderline
  - Malignant
- Vascular malformations
  - Simple
  - Combined
  - Anomalies of major named vessel
  - Malformations associated with other anomalies

Vascular Tumors, Benign

- Infantile hemangioma
- Congenital hemangioma
  - RICH, NICH, PICH
- Tufted angiomatous
- Spindle-cell hemangioma
- Epithelioid hemangioma
- Pyogenic granuloma
- Others

Infantile Hemangioma

- Benign neoplasm
- Proliferating endothelial cells
  - GLUT-1 positive in all phases
- Presents shortly after birth
  - Proliferative phase - enlarge up to 2 yrs
  - Involuting phase - spontaneous regression several yrs.
- 60% in H & N
  - Parotid, orbit, nasal, suglottic, anterior/posterior neck
- Majority single in subQ tissue
  - No imaging required
- Occasionally multiple, trans-spatial, deep
- Further workup if segmental facial distribution, ≥ 5 subQ lesions, midline lumbosacral

Infantile Hemangioma (IH)

- Intense enhancement
- High flow vessels during proliferative phase
- Fatty infiltration during involuting phase
- Tx - expectant waiting, oral propranalol, steroids, laser tx, Rapamycin, excision

Infantile Hemangioma

- Additional work up recommended
  - Segmental facial distribution IH (PHACE syndrome)
  - 5 or more cutaneous IHs
    - Associated with hepatic IH
      - If large & multiple, may result in liver failure, heart failure, abdominal compartment syndrome, hypothyroidism
    - Midline lumbosacral/perineal IH
      - Associate with tethered cord/spinal abnormalities

Infantile Hemangioma

- 9 years later
**PHACE Syndrome**
- Posterior fossa malformations
- Hemangiomas H&N
- Arterial
  - Stenosis, occlusion, aneurysm
- Cardiovascular
  - Coarctation aorta, cardiac anomalies
- Eye
- Supra-umbilical & sternal clefts

**Imaging Congenital Hemangioma**
- More heterogeneous than IH
  - Calcifications, hemorrhage, necrosis
  - Less T2 hyperintense vs. IH
  - Vessels more frequently visible on grayscale US than IH
  - High flow periphery +/- large feeding arteries/draining veins, especially in liver
  - No fibrofatty residua

**Congenital Hemangioma**
- Proliferation complete at/before birth
- GLUT-1 negative
- Rapidly involuting (RICH)
  - Largely involuted by 12-15 months
- Noninvoluting (NICH)
  - No change over time
- Partially involuting (PICH)

**Rapidly Involuting Congenital Hemangioma**
Rapidly Involuting Congenital Hemangioma

Pyogenic granuloma

• "Lobular capillary hemangioma" - Misnomer
• Vascular tumor - ? at sites of prior trauma
• H&N > trunk & extremities
  - Most common in children & pregnant women
• Small erythematous papules
  - Fissile with tendency to bleed
• Most removed without imaging
• Hypointense T1/hyperintense T2
• Contrast enhancing
  +/- iso/hypointenuating cap on CECT
• +/- bone displacement or erosion
• Inferior turbinate >> nasal septum

Locally aggressive or borderline vascular tumors

• Kaposiform hemangioendothelioma
• Retiform hemangioendothelioma
• Papillary intralymphatic angioendothelioma (PILA), Dabska tumor
• Composite hemangioendothelioma
• Kaposi sarcoma
• Other

Kaposiform hemangioendothelioma (KHE)

• Locally aggressive vascular tumor primarily found in infants
• Kasabach-Merritt phenomenon
  - Sustained, profound consumptive coagulopathy (thrombocytopenia, hypofibrinogenemia) due to intrallesional trapping
    • KHE, tufted angiomia
  - Occurs in 70% of patients with KHE
  - Up to 30% mortality from hemorrhage
• Retroperitoneum > skin > H&N, mediastinum, extremities

Imaging KHE

• Poorly defined, infiltrative
• Heterogeneous enhancement
• Cutaneous/subcutaneous vs. deep visceral/muscular
• +/- edema, esp. if KMP
• +/- prominent vessels

Kaposiform hemangioendothelioma (KHE)
Kaposiform hemangioendothelioma (KHE)

- Associated with human herpesvirus 8 (HHV-8)
- In H&N: skin, mucosa, lymph nodes
- Nodular enhancing mass +/- skin thickening and subcutaneous edema
- Hyper-attenuating adenopathy with heterogeneous enhancement
- Adenoid enlargement

Kaposi sarcoma

Vascular tumors, malignant

- Angiosarcoma
- Epithelioid hemangioendothelioma
- Others

Angiosarcoma

- 60% occur in H&N
- Skin of scalp, face, neck > sinonasal, oral cavity, thyroid
- Overall 5-year survival in adults < 30%
- Nodal recurrence and hypervascular distal mets common
  - Lung, liver, bone

Imaging Angiosarcoma

- Contrast enhancing scalp or soft tissue mass
- +/- underlying bone erosion
- Intermediate T1, hyperintense T2 +/- flow voids
- FDG PET: high FDG uptake
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Others