Hemangiomas and Other Vascular Tumors

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Disclosures

- No relevant financial disclosures

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

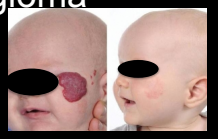
ISSVA Classification

- 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 angioma
- Spindle-cell hemangioma
- Epithelioid hemangioma
- Pyogenic granuloma
- Others

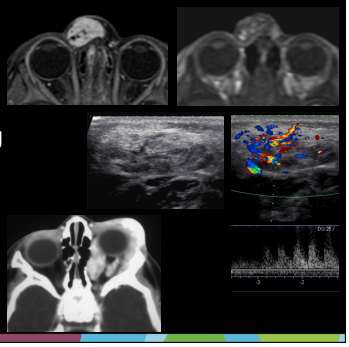
Infantile Hemangioma

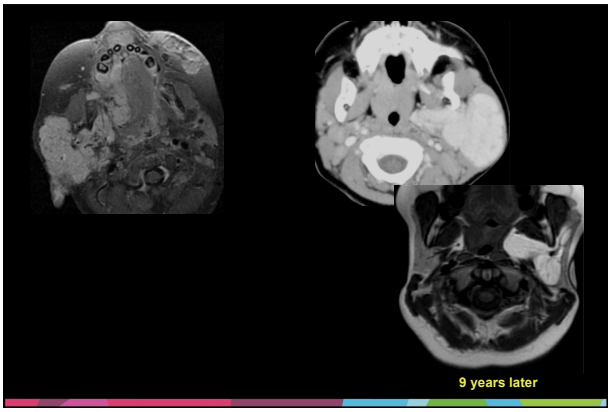


- Benign neoplasm
  - Proliferating endothelial cells
  - GLUT-1 positive in all phases
- Presents shortly after birth
  - Proliferative phase - enlarge up to 2 yrs
  - Involuting phase - spontaneous regression several yrs.
- 60% in H & N
  - Parotid, orbit, nasal, suglottic, anterior/posterior neck
- 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



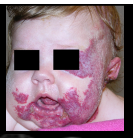
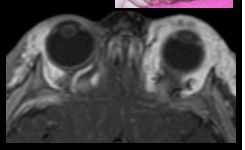
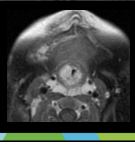


Infantile Hemangioma

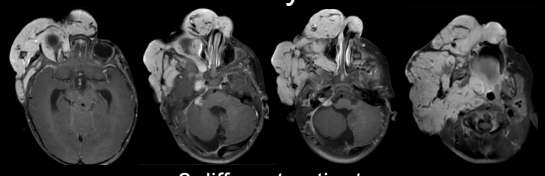
- Additional work up recommended
  - Segmental facial distribution IH (PHACE syndrome)
  - 5 or more cutaneous IHs
    - Associated with hepatic IH
      - If large &/or multiple, may result in liver failure, heart failure, abdominal compartment syndrome, hypothyroidism
  - Midline lumbosacral/perineal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

PHACE Syndrome

- Posterior fossa malformations
- Hemangiomas H&N
- Arterial
  - –Stenosis, occlusion, aneurysm
- Cardiovascular
  - –Coarctaion aorta, cardiac anomalies
- Eye
- Supra-umbilical & sternal clefts

PHACE Syndrome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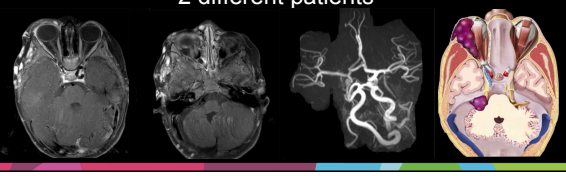
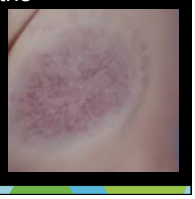


Congenital Hemangioma

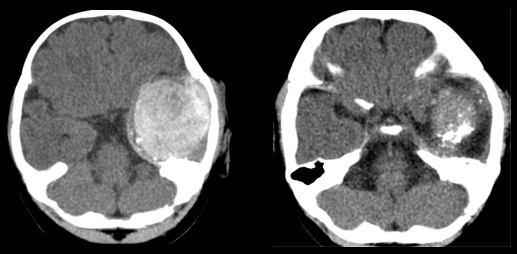
- Proliferation complete at/before birth
- GLUT-1 negative
- Rapidly involuting (RICH)
  - –Largely involuted by 12-15 months
- Noninvoluting (NICH)
  - –No change over time
- Partially involuting (PICH)



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

Rapidly Involuting Congenital Hemangioma



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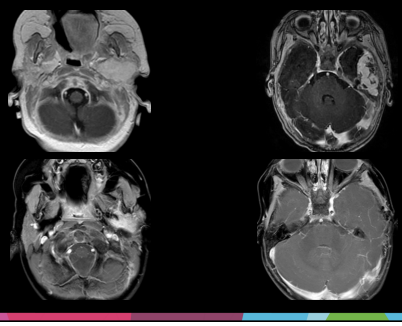

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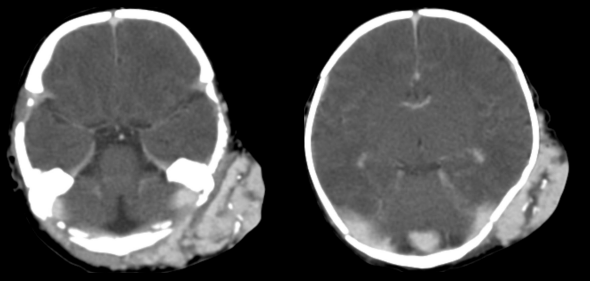
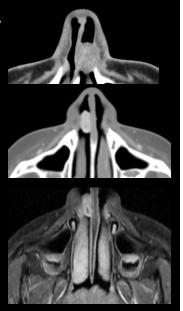


Pyogenic granuloma

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



Lee et al. AJNR Am J Neuroradiol. 2010 Apr;31(4):749-54

Locally aggressive or borderline vascular tumors

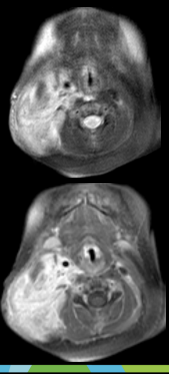
- Kaposiform hemangioendothelioma
- Retiform hemangioendothelioma
- Papillary intralymphatic angioendothelioma (PILA), Dabska tumor
- Composite hemangioendothelioma
- Kaposi sarcoma
- Other

Kaposiform hemangioendothelioma (KHE)

- Locally aggressive vascular tumor primarily found in infants
- Kasabach-Merritt phenomenon
  - Sustained, profound consumptive coagulopathy (thrombocytopenia, hypofibrinogenemia) due to intralesional trapping
    - KHE, tufted angioma
  - 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



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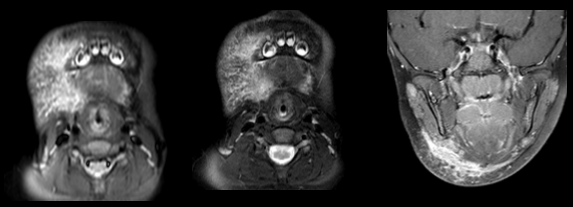

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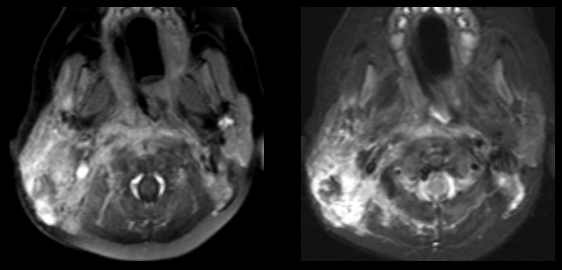
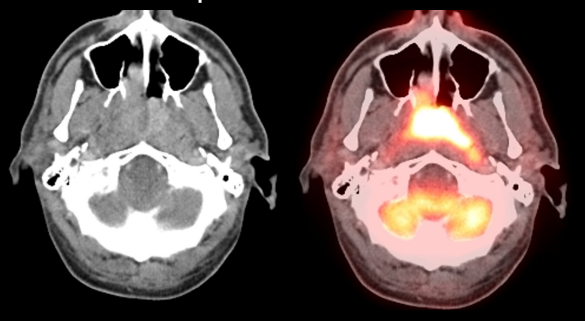


Kaposi sarcoma

- 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



Courtesy Dr. Shatzkes

Vascular tumors, malignant

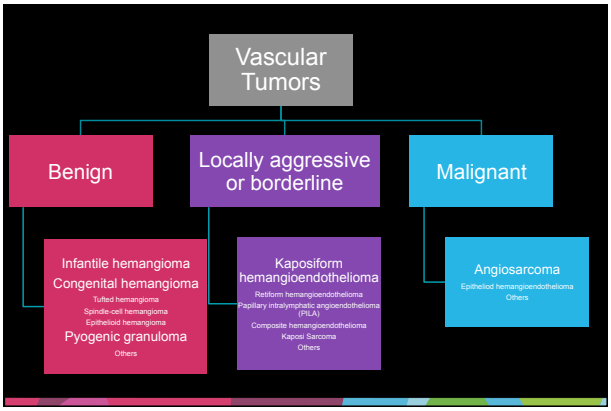
- Angiosarcoma
- Epithelioid hemangioendothelioma
- Others

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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