Vascular Lesions of the Head & Neck
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Lecture Outline
• Classification and nomenclature
• Highly simplified MRI-based differential algorithm
• Imaging appearance of commonly encountered lesions

“Hemangioma”
Orbital VM (aka “cavernous hemangioma”)
Intraosseous VM (aka “ossifying hemangioma”)
Facial nerve VM (aka “ossifying hemangioma”)
Cavernous malformation (aka “cavernous hemangioma”)
Cavernous sinus VM (aka “cavernous hemangioma”)

“Nomenclature has been the major obstacle to our understanding and management of vascular anomalies”*
• “Hemangioma/Angioma”: Wastebasket terms used to describe any vascular lesion
• 1982: Mulliken and Glowacki published seminal work on biological basis of vascular lesions (PRS 69: 412-422)

Biology a’ la Mulliken
• Vascular tumors (most commonly infantile hemangiomas) are true neoplasms and grow by endothelial hyperplasia
• Vascular malformations have a quiescent endothelium and are considered localized defects of vascular morphogenesis (“badly formed vessels”)

Immunohistochemical marker GLUT-1 accurately distinguishes

ISSVA Classification of Vascular Anomalies (2014)

Vascular Tumors
• Benign
  • Infantile hemangioma
  • Congenital hemangioma (RICH, NICH)
  • Pyogenic granuloma
• Borderline
  • Kaposiform hemangioendothelioma
  • Kaposi sarcoma
• Malignant
  • Angiosarcoma

Vascular Malformations
• Simple
  • Capillary malformation
  • Venous malformation
  • Lymphatic malformation
• Combined
  • CVM, CLM, LVM, CLVM, AVM-LM, CM-AVM
• Associated with other anomalies
  • Sturge-Weber
Nomenclature

<table>
<thead>
<tr>
<th>OLD</th>
<th>NEW</th>
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<tbody>
<tr>
<td>Capillary hemangioma</td>
<td>Hemangioma</td>
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<tr>
<td>Port wine stain</td>
<td>Capillary malformation</td>
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<tr>
<td>Strawberry hemangioma</td>
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<tr>
<td>Cavernous hemangioma</td>
<td>Venous malformation</td>
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<tr>
<td>Ossifying hemangioma</td>
<td>Lymphatic malformation</td>
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<tr>
<td>Lymphangioma</td>
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<td>Cystic hygroma</td>
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Imaging of H&N Vascular Lesions

- MRI appearance reflects histologic composition
- Hemangiomas = cellular tumors; avid enhancement, only moderate ↑ T2 SI
- The most commonly imaged vascular malformations are VM, LM (low flow) and AVM (high flow)
- MRI appearance reflects vascularity and specific vessel subtype

(Really) Simplified Differential Algorithm

<table>
<thead>
<tr>
<th>↑ T2 SI</th>
<th>(+) CE</th>
<th>(-) CE</th>
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<tr>
<td>Hemangioma</td>
<td>AVM</td>
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<tr>
<td>↑↑ T2 SI</td>
<td>Venous Malformation</td>
<td>Lymphatic Malformation</td>
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I. Hemangiomas (vascular tumors)
- Infantile hemangioma (common)
- Congenital hemangioma (rare)
- Kaposiform hemangioendothelioma

II. Vascular Malformations
- Low Flow:
  - Venous
  - Lymphatic
- High Flow:
  - Arterial-Venous (AVM)
  - Complex (e.g. LVM)

I. Infantile Hemangioma: Clinical

- Common benign endothelial tumor of infancy
- Not present at birth
- Appear a few weeks later with rapid growth – proliferative phase (Phase I)
- Regress slowly beginning at age 2-5 – involuting phase (Phase II)
- Typically completely regress by late childhood - involuted phase (Phase III)
- Glut-1 positive

Infantile Hemangioma (Phase I)

- Hypercellular and hypervascular tumors
- Large vessels
- Ectodermal location

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Infantile Hemangioma

- Intermediate T1 and T2 signal
- Prominent feeding and draining vessels reflect hypervascularity in Phase I

Parotid Hemangioma

- Parotid derived from ectoderm
- “Light bulb” enhancing parotid mass in infant virtually pathognomonic

Involuting Hemangioma (Phase II)

- Decrease in tumor volume
- Increase in interstices and fatty content

Involuted Hemangioma (Phase III)

- Minimally enhancing fatty soft tissue mass
- Excess fat and bony overgrowth may result in persistent cosmetic deformity

PHACES Syndrome

- Posterior fossa
- Hemangioma
- Arterial
- Cardiac
- Eye
- Sternal

PHACES

- Posterior fossa
- Hemangioma
- Arterial
- Cardiac
- Eye
- Sternal
**Congenital Hemangioma**
- Rare
- **Present and fully formed at birth**
- GLUT-1 negative
- **RICH**: Rapidly Involuting Cong. Hemangioma
- **NICH**: Non Involuting Cong. Hemangioma
- **PICH**: Partially Involuting Cong. Hemangioma
- Clinical diagnosis without specific imaging features

**Kaposiform Hemangioendothelioma (KHE)**
- “Borderline” vascular tumor
- Ill-defined, invasive, subQ fat stranding
- >50% associated with Kassabach-Merritt syndrome ("hemangioma" with thrombocytopenia)

**II. Vascular Malformations - Clinical**
- **Present at birth**
- GLUT-1 negative
- Grow in conjunction with somatic growth in a linear fashion
- Will not regress spontaneously
- **High flow** (AVM) and **low flow** (VM, LM)
- Complex variants demonstrate mixed imaging features

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**II. Vascular Malformations**
- Venous
- Lymphatic
- Arterial-Venous (AVM)
- Complex (e.g. LVM)

**Venous Malformation (VM)**
- Malformed venous channels with sluggish flow
- Bright T2, avid enhancement
- Phleboliths common

**NICH**
- 2 years
- 7 years
Left Cheek and Lip Mass

- Well demarcated though infiltrative
- Clinically evident lesion may represent “tip of the iceberg”

Venous Malformation

- Frequently trans-spatial, with common masticator space (masseter) and palatal involvement

Venous Malformation

- Initial patchy contrast enhancement “fills in” on more delayed imaging
- Orbital VM, aka “cavernous hemangioma” is most common orbital mass in adults

Venous Malformation

- Phleboliths result from sluggish venous flow and are highly specific

30 y/o woman noticed intermittent left cheek fullness

- Venous malformation

Intraosseous VM (“Ossifying Hemangioma”)

- Facial skeleton, skull, temporal bone (“facial n. ossifying hemangioma”)
- Internal bone spicules
**Lymphatic Malformation (LM)**
- Macrocyts or microcyts
- Well-defined or infiltrative
- Enhancement capsule/septae
- “Soft” lesions
- Prone to internal hemorrhage

**Lymphatic Malformation**
- Macrocyst forms image like cysts
- “Cystic Hygroma” = macrocystic LM
- Common at the root of the neck

**Lymphatic Malformation**
- Microcyst variants more infiltrative
- Less fluid = lower T2 signal
- “Beard” distribution common

**Orbital LM**
- Orbit is common location
- Soft lesions may be deformed by optic nerve
- Present when hemorrhage leads to proptosis
  (fluid levels in orbital mass = LM)

**Lymphovenous Malformation (LVM)**
- Form of combined vascular malformation
- Shares features of LM and VM

**AVM**
- Large vessels
- No discrete mass
- Tissue edema
- Bony lysis and overgrowth
Facial AVM

- Ill-defined; no discrete mass
- Look for flow voids and large feeding and draining vessels

Nasal AVM

- Findings of tissue edema (T2 hyperintensity, mild enhancement) may be subtle
- Look for flow voids!

AVM

- Bony overgrowth and/or lytic destruction

Vascular Lesions of H&N

- Reserve term “hemangioma” for vascular tumors of infancy
- History is important (present at birth?)
- Enhancement pattern is good first step in distinguishing lesions
- Phleboliths (VM), Flow voids (AVM) and fluid levels (LM) are imaging clues

Thank you!