VASCULAR BIRTHMARKS: HEMANGIOMAS AND VASCULAR MALFORMATIONS

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7 yo F with soft painless temple mass: Diagnosis? Therapy?

32 M, painless pulsatile cheek mass: Diagnosis? Therapy?
**Vascular Birthmark: What is it?**

Confusing Terminology

- Strawberry hemangioma
- Cavernous hemangioma
- AVM
- Cystic hygroma
- Angioma
- Port wine stain
- Naevus flammeus
- Eponymous syndromes

**Common Misconceptions (Dowd)**

- All vascular birthmarks are not “hemangiomas”!
- All vascular malformations are not “AVMs”!
- 50% of patients referred to the UCSF Vascular Anomalies Clinic carry an incorrect diagnosis!

**Vascular Birthmark: Classification**

**Vascular Tumor**
- Infantile Hemangioma
- Tumors producing KMP
  - KHE - Kaposiform Hemangioendothelioma
  - Tufted Angioma
- Congenital Hemangioma
  - RICH
  - NICH

**Vascular Malformation**
- Arterial (AVM)
- Venous
- Lymphatic
- Capillary
- (Combination)

(*Mulliken and Glowacki, 1982)

**Vascular Birthmark: Classification Clinical and Cellular Differences**

**Hemangioma (Infantile)**
- Tumor of blood vessels
- Not present at term birth
- Proliferation/involution
- F/M: 3/1
- ↑ endothelial turnover
- ↑ FGF
- GLUT 1 staining

**Vascular Malformation**
- Malformed blood vessels
- Present at birth
- Commensurate growth
- F/M: 1/1
- nl endothelial turnover
- nl FGF
- No GLUT 1 staining
Diagnosis of Vascular Birthmark

• More important:
  – age of patient
  – physical examination
  – history (esp. birth/childhood/recent activity)

• Less important:
  – imaging

-> Go look at patient and ask a few questions!

Hemangioma (Infantile)

• Benign tumor of blood vessel origin
• Endothelial cell proliferation (↑ bFGF)
• GLUT1: immunohistochemical stain
• Most common tumor of infancy (~10%)
• F/M : 3/1
• Share phenotype markers of placenta
• Clinical: classically appear at age ~2 wks
  – Proliferative phase: rapid growth to age 10-12 mo.
  – Involuting phase: slower involution -> fibrofatty scar
• Common mimic: venous malformation
• Focal (70%) vs. Segmental

Infantile Hemangioma: Imaging

• well-circumscribed
• T1: intermediate signal
• T2: high signal
• enhancement: homogeneous
• “salt and pepper” pattern of vessels within tumor
• [angio: vascular tumor blush, normal size feeding arteries, no A-V shunt]
Hemangioma: Therapy

- No therapy: preferred because hemangiomas involute!
- Medical therapy
  - Propranolol (2008)
  - steroids (systemic or intralesional)
- Surgery
  - early, when vital structures compromised
  - late, to treat residual fibrofatty scar
- Embolization:
  - Not necessary preoperatively
  - [Often used to Rx Kasabach-Merritt Syndrome]

Kasabach-Merritt Syndrome

- Severe thrombocytopenia (platelet trapping)
- Formerly thought to arise in aggressive variant of Infantile Hemangiomas
- Now known to arise in:
  - Kaposiform Hemangioendothelioma (KHE)
  - Tufted Angioma
- Clinical: “angry” red-purple vascular tumor
- Imaging: appear similar to infantile hemangioma with less discrete borders
- Rx: embo, prednisone, Sirolimus, VCR, surg, (α INF)
**Congenital Hemangiomas**

**RICH**: Rapidly-Involuting Congenital Hemangioma (limb, near joint, “soufflé”)
**NICH**: Non-Involuting Congenital Hemangioma (flat, pallid, “red-white-blue”)

- Fully grown *in utero*
- Present at birth
- F/M : 1/1
- No GLUT1 staining

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**RICH vs NICH vs IH: Clinical Behavior**

(from Nozaki et al. Radiographics 33:175-195, 2013)

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  - Tufted Angioma
- Congenital Hemangioma
  - RICH
  - NICH

**Vascular Malformation**
- Arterial (AVM)
- Venous
- Lymphatic
  - “low flow”
- Capillary
  - (Combination)

(*Mulliken and Glowacki, 1982*)
Arteriovenous Malformation (AVM)

- Vascular malformation characterized by a nidus of vessels lacking the normal capillary bed, causing high-flow arteriovenous shunting.
- F/M: 1/1
- Growth commensurate; also may be triggered by trauma, surgery, hormonal effects (puberty, pregnancy). Role of angiogenesis factors?
- Clinical: pulsatile mass with thrill/bruit; pain, swelling, bleeding (may be life-threatening); high-output CHF
- Difficult to eradicate fully

AVM: Imaging

- Flow voids
- May have “soft tissue” element (Type I vs II)
- Best-seen on gradient echo
- Involved ST may be swollen
- May involve bone
- Partition images helpful
- MRA not helpful [Dowd]
- Angio: A-V shunt with nidus, enlarged feeding arteries and draining veins

AVM: Therapy

- No therapy
- Embolization (agent, route)
  - Palliative
  - Preoperative
  - Curative
- Surgery (you’d better get it all!)
29F, popliteal AVM
Rx: ETOH embolization

24M, “neurofibromatosis”, increasing arm pain, “please biopsy”
Venous Malformation

- Vascular malformation characterized by dilated veins, limited mural smooth muscle (allows gradual expansion)
- Non-proliferating: no endothelial turnover, no ↑ bFGF
- Outdated name: “cavernous hemangioma”
- F/M : 1/1
- Clinical: dilated venous channels, usu. boggy/compressible, no thrill, enlarge w/ gravity-dependence or Valsalva, firmer w/ thrombosis
- Spongiform (“cave-like”) vs. Phlebectatic (“tubular”) phenotypes
- Commensurate growth
- Large/IM VMs -> low-grade consumptive coagulopathy (“LIC”)

Venous Malformation: Pathology

- Large serpiginous channels
- Single-layer endothelial lining
- Very little smooth muscle

Mulliken and Young 1988
Venous Malformation: Imaging

- discrete/scattered soft tissue.
- T1: intermediate signal
- T2: high signal
- enhancement: homogeneous
- no flow voids
- uni/multilocular
- can involve muscle/bone
- + phleboliths (CT, plain)
- angio: normal art. phase, may have venous puddling
- direct px: irregular venous pouches

Venous Malformation: Therapy

- No therapy
- ASA
- Compressive stocking
- Laser
- Sclerotherapy
- Surgery

Sclerosing Agents: venous malformation

- Sotradecol (sodium tetradecyl sulfate 3%)
- Ethanol (~pure)
- Bleomycin
- Sodium morrhuate
- Ethanolamine oleate

Mix with contrast for visualization
Avitene slurry to close puncture site(s)

39F, venous malformation, sclerotherapy

U/S and fluoro guidance
**Capillary Malformation**

- Vascular malformation characterized by ectatic vessels within the upper dermis
- Non-proliferating: no endothelial turnover, no ↑ bFGF
- Aka: “port-wine stain”, “naevus flammeus”
- Trigeminal (V1) lesions assoc. w/ Sturge-Weber Syndrome
- F/M : 1/1
- Clinical: sharply-demarcated flat pink-red stain; grows proportionately; no involution; hue deepens with crying, warmth, fever; color darkens w/age (pink->red->purple); texture more nodular w/age.
- Usually no imaging in isolated cases

**Capillary Malformation: Therapy**

- No treatment
- Laser therapy to remove color (temporary)
Lymphatic Malformation

- Vascular malformation characterized by malformed lymphatic cavities lined by flattened endothelium
- Non-proliferating: no endothelial turnover, no ↑ bFGF
- Aka: “cystic hygroma”, “lymphangioma”
- F/M : 1/1
- Microcystic vs. Macrocystic
- Clinical: Neck/face/axilla common; diverse morphology (enlarged limb, nodular vesicles, translucent cysts)
- Commensurate growth
- Enlargement with infection (bacterial/viral), hemorrhage

Lymphatic Malformation: Imaging

- macro- vs. micro-cystic
- multiple cysts
- T1: low (water) signal, unless prior hemorrhage, Rx
- T2: high (water) signal
- fluid-fluid layers typical
- enhancement: rim only
- angio: normal
- direct px: well-defined cysts, +/- intercommunicate

7 yo F with macrocystic lymphatic malformation

6wkM, combined macro- and microcystic lymphatic malformation
**Lymphatic Malformation: Therapy**

- No therapy
- Compressive stocking
- Manual lymphatic drainage
- Antibiotics
- Sclerotherapy
- Surgery
- Medical (Sildenafil, Sirolimus)

**Sclerosing Agents: lymphatic malformation**

- Doxycycline
- Sotradecol (sodium tetradecyl sulfate 3%)
- Ethanol (~pure)
- Bleomycin
- OK 432

*Mix with contrast for visualization*

*Avitene slurry to close puncture site(s)*

**Vascular Birthmark: Classification Old and New Nomenclature**

**Vascular Tumor**
- Hemangioma
  - Strawberry hemangioma
  - Capillary hemangioma

**Vascular Malformation**
- Arterial (AVM)
  - Angioma
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- Capillary
  - Poar wine stain
  - Naevus flammeus
- Lymphatic
  - Lymphangioma
  - Cystic hygroma
Summary

- Vascular birthmark classification useful for predicting behavior, proposing therapy
- History, physical examination are most important elements for proper diagnosis
- Imaging helpful to differentiate types of vascular lesions, determine tissue involved
- Transarterial or direct-puncture techniques provide definitive, palliative, or preoperative therapy
- Collaborative “Vascular Anomalies Clinic” is a good model for patient evaluation