Multimodality Imaging of Vascular Anomalies in Children

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I have no disclosures
### TUMORS (NEOPLASMS)
- Infantile hemangioma (IH)
- Congenital hemangioma:
  - Rapidly involuting congenital hemangioma (RICH)
  - Non-involuting congenital hemangioma (NICH)
- Kaposiform hemangioendothelioma
- Tufted angioma
- Hemangiopericytoma
- Pyogenic granuloma
- Spindle cell hemangioendothelioma

### VASCULAR MALFORMATIONS
- Simple
  - Venous
  - Lymphatic
  - Capillary
- Combined
  - Arterio-venous malformation
  - Arterio-venous fistula
  - Any other combination

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Classification Of Vascular Anomalies
International Society For The Study Of Vascular Malformations
INFANTILE HEMANGIOMA:
- Very common lesions
- Most present shortly after birth. 30% precursor lesion at birth
- GLUT-1 positive independent of stage
- Typical course: proliferation, involution, fibrosis
- Respond to PROPRANOLOL

CONGENITAL HEMANGIOMA:
- Rare lesions
- Fully developed at birth; even in utero
- GLUT-1 negative
- Two types – different course
  - RICH: Rapidly Involuting Congenital Hemangioma
  - NICH: Non Involuting Congenital Hemangioma
Infantile hemangioma: Precursor lesion

1 week

4 months

how did the lesion look at birth?

6 years
Infantile Hemangioma: types

**Focal**: more common, localized, more raised, tumor like

**Segmental**: flat, plaque-like, segmental distribution

**Indeterminate**
Not always that simple!

Hemangiomas or ........... mosquito bites?

not me!!!!!!

cutaneous hemangiomatosis

Purely subcutaneous hemangioma
Infantile hemangioma: Ultrasound imaging

- Multiple lesions up to 30%
- May coexist with CH
- Most imaging not for Dx
- IH do not calcify
Infantile hemangioma: MR imaging

T1:
+ fat

T2:
flow voids
hyperintense
< fluid

Rapid arterial enhancement

T1+
enhancement
Involuted infantile hemangioma

All infantile hemangiomas leave a scar of fibro-adipose tissue after involuting.
Congenital hemangioma: NICH-RICH

- Purple nodular masses
- Superficial telangiectasias
- Peripheral pale halo
- Head & neck, close to a joint

Can look identical to IH
- May calcify
- What makes it CH? History!
Vascular malformations

- **Slow flow:**
  - Lymphatic: cystic hygroma
  - Lymphangioma
  - Venous: cavernous hemangioma
  - Venous angioma
  - Capillary

- **High flow:**
  - Arterio-venous malformations
  - Arterio-venous fistulas
Lymphatic malformations

- Supple mass
- Trans-illuminate
- No bruit or warmth
- May change in size
- Intact skin

US:
- Cystic
- Most multilocular
- No vascularity
- F-F levels
Lymphatic malformations: MR

- variable T1 signal
- Cystic
- T2 Hyperintense
- +/- F-F levels
- Faint septal/peripheral enhancement unless inflamed
Lymphatic malformations: Types

Macrocystic LM: better tx response

Microcystic LM

Mixed LM
Lymphatic malformations: role of CT

Radiographs play **NO** role evaluating LM

Suboptimal internal characterization
Lymphatic Malformation: Infiltrative

- Does not respect boundaries
- Can infiltrate organs
Lymphangioma circumscriptum

- LM involving and infiltrating skin
- Small vesicles: “skin of a frog”
- May be the tip of the iceberg

- Not a contraindication for sclerotherapy
- May require laser

Clinical picture courtesy of Dr Teresa O
Venous Malformations (VM)

- Sporadic: >90%
  - Unifocal (93%)
  - Multifocal (1%)

- Inherited forms: 6%
  - Autosomal dominant
  
  - Ask for family history!

  - 1% part of cutaneo-mucosal VM
  - 5% multifocal glomovenous malf
Venous Malformations: Inherited

Cutaneo-mucosal VM
• Confined to superficial tissues
• Mucosal

Glomovenous malf.
• superficial
• involve dermis
• cobble-stoning
• painful
• non compressible

GVM pic courtesy of Dr Teresa O
Venous malformations: Sporadic

- Light to dark blue
- Empty by light compression
- No thrill/bruit, no warmth
- Painless on palpation unless thrombosis
- Can arise from any tissue including bone
VM: Radiographs

- Phleboliths
- Bone remodeling
- Focal gigantism
- Osteolysis-Gorham
Venous malformations: US

- Cystic to solid
- Variable borders
- Variable contents
- M/C no color flow/PD
- +/- augmentation
- YES, arteries can be seen!!
- Phleboliths
Venous malformations: MR

- T1: slightly hyperintense
- T2: fluid signal-”light bulb”
- Erratic enhancement
- +/- F-F levels
- Delayed Gad images helpful
- MRV for deep venous system
Venous malformations: the role of CT

CT has virtually no role in VM diagnosis
Venous malformations: a challenging dx

No skin discoloration!
Extensive pure venous limb malformations

- Extensive, diffuse
- May involve all limb components: always muscle/skin
- Normal limb length or slight undergrowth
- Joint destruction
Extensive pure venous limb malformations:

- Associated specific localized intravascular coagulopathy (LIC)
- Blood stagnation $\Rightarrow$ activation of coagulation, consumption of coag factors $\Rightarrow$ bleeding

Synovial hemangiomia

- LIC worsens with therapeutic procedures, fx
- Tx: stocking

Low molecular weight heparin
Intramuscular VM

- 1/3 diagnosed in adolescence!
- Confined to muscle tissue
- No growth/limb deformity

Path: small vessels with capillary structure and proliferative activity → capillary hemangiom

Masson phenomenon:
- Due to exuberant intraluminal clotting with subsequent ingrowth of capillaries: Masson’s papillary endothelial hyperplasia

Often confused with tumors!
Intramuscular VM: Imaging & Tx

- Tubular/channels follow long axis of muscle
- Phleboliths are common

Tx:
- conservative
- complete excision
- sclerotherapy
Capillary malformation (CM)

• Imaging plays no role in Dx
• Important to recognize due to associations: KTS, etc
• 30% cutaneous CM associated with AVM or AVF: RASA1 gene

AKA port wine stains
Arteriovenous malformations

- Area of discoloration
- Localized warmth to touch
- Associated thrill or murmur
- Tissue/limb hypertrophy
- Frequent late diagnosis

Not infrequently diagnosed later in childhood or adolescence.
Arteriovenous malformations: US-Doppler

- Area of abnormal mixed echogenicity
- **No discrete mass!**

- Hypervascularity
- Feeding arteries, increased diastolic flow
- Arterialized draining veins, pulsatile flow (different than hemangiomas)
Arteriovenous Malformations: MRI-MRA

- No discrete mass
- Tissue hypertrophy

MRA: vascular map feeding arteries draining veins nidus

Time resolved MRA: dynamic opacification of all components high temporal resol (4-D) sacrifices spatial resolution
Arteriovenous malformations: conventional arteriogram

- Little if any diagnostic role
- Needed for intervention, pre-surgical
- Selective catheterization of feeding arteries
Take home points:

1. Use the appropriate terminology
2. Exam the patient, talk to the parents, take clinical pictures
3. Imaging of vascular malformations and hemangiomas usually is not to make Dx.
4. In general, US and MRI are best modalities to image vascular anomalies:
   - Focal: US
   - More extensive: MRI
5. Vascular malformations always present at birth but not always apparent