Pediatric Vascular Anomalies

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Disclosure

• I have no financial disclosures

• I will discuss off-label use of gadolinium contrast
Objectives

• review classification scheme and appropriate terminology of vascular anomalies

• review imaging characteristics of common anomalies
# Classification

1996 International Society for the Study of Vascular Anomalies

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<th>Vascular Tumors</th>
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**Mixed**


- Vascular Tumors: plump, proliferative endothelium
- Vascular Malformations: thin, dysplastic endothelium
Imaging findings are most consistent with which anomaly?

- A. Arteriovenous malformation
- B. Congenital hemangioma
- C. Venous malformation
- D. Infantile Hemangioma
Venous Malformation

US Findings

- tangle of tubular hypoechoic structures
- often monophasic slow flow on Doppler
- occasionally (~15%) no detected flow due to stasis
Venous Malformation

MR Findings

- serpigenous T2 hyperintense, T1 intermediate intensity venous channels
- venous phase enhancement on TR-MRA
Venous Malformation

- **Pathology:** cluster of abnormal venous channels with dysplastic endothelium, variable smooth muscle, and absent valves

- **Time Course:** present at birth, but may not become clinically apparent until later in life

- **Notable:** repeated cycles of thrombosis and thrombolysis can eventually calcify, resulting in phleboliths

- **Treatment:** sclerotherapy
Case

STIR

T1 + GAD
Often associated with Down, Turner, and Noonan syndromes, the demonstrated lesion is a/an ...

- A. Lymphatic malformation
- B. Infantile Hemangioma
- C. Venous malformation
- D. AVM
Lymphatic Malformation

MR Findings

Macrocytic
- cysts >2 cm
- no central enhancement
- thin septal enhancement

Microcytic
- cysts <2 cm
- conglomeration of enhancing septa can give the appearance of an infiltrative mass
Lymphatic Malformation

US Findings

Macrocystic
- discreet anechoic cyst-like spaces without color flow
- separated by thin septae

Microcystic
- infiltrative soft tissue mass
- multiple tiny unresolvable cysts result in diffusely hyperechoic tissue

Mixed Micro- and Macro cystic Lymphatic Malformation
Lymphatic Malformation

- **Pathology:** cluster of abnormal, dilated, lymphatic channels

- **Time Course:** present at birth and typically enlarges proportionally with child

- **Notable:** can come to clinical attention due to rapid enlargement with hemorrhage or infection

- **Treatment:** sclerotherapy or surgery

Thursday, August 23, 12
Disparity between STIR signal abnormality and venous enhancement suggests the macrocystic lymphatic portion of a mixed venolymphatic malformation.
Case

STIR

T1 + GAD

Time-resolved MRA
Question - not SAM

Capillary malformation plus demonstrated findings are consistent with which syndrome (note lack of high flow component)?

- A. Parks-Weber
- B. Klippel-Trenaunay
- C. Sturge-Weber
- D. PHACE
Klippel Trenaunay Syndrome

Left lower extremity hypertrophy with mixed venous and lymphatic malformation and large lateral draining vein (arrow)
Klippel Trenaunay Syndrome

• **Pathology:** low flow vascular malformations and limb hypertrophy with cutaneous capillary malformation

• **Time Course:** presents in the newborn

• **Distribution:** typically involves a single extremity, most often a lower extremity

• **Notable:** often a persistent large lateral marginal vein (vein of Servelle) which is incompetent
Parkes Weber Syndrome

Similar to Klippel Trenaunay syndrome, but with high flow components as well.
Mild left lower extremity hypertrophy
Subtle STIR hyperintensity represents mixed vascular malformation

Similar to Klippel Trenaunay syndrome, but with high flow components as well
Case

Time-resolved MRA
Question - not SAM

The demonstrated lesion likely represents...

- A. Infantile Hemangioma
- B. AVM
- C. AVF
- D. Venous malformation
Arteriovenous Malformation

Time-resolved MRA

Angiographic Findings:

- coiled nest of vessels with arterial phase enhancement (central nidus)
- early opacification of draining veins
Angiographic Findings:

- coiled nest of vessels with arterial phase enhancement (central nidus)
- early opacification of draining veins (arrow)
Arteriovenous Malformation

• **Pathology:** tangle of vessels without intervening capillary bed, characterized by a central nidus

• **Time Course:** present from birth

• **Notable:** arteriovenous shunting results in spin echo T2 flow voids and arterialization of draining vein on US (or high peak venous velocities)

• **Treatment:** embolization of feeding arteries (coils) or across the nidus (glue); surgery may be required
Case
The demonstrated lesion likely represents...

- A. Hemangioma
- B. AVM
- **C. AVF**
- D. Venous malformation
Congential Arteriovenous Fistula

Similar to arteriovenous malformation, but without central nidus of vessels. Tend to embolize with coils.
Congenital Arteriovenous Fistula

Similar to arteriovenous malformation, but without central nidus of vessels. Tend to embolize with coils.

Arterial phase CT

Angiogram

Thursday, August 23, 12
High and low flow vascular malformations can commonly coexist
High and low flow vascular malformations can commonly coexist.

AVF with VM

Time-resolved MRA

Direct high flow component

Low flow venous malformation
Question - not SAM

The demonstrated lesion likely represents...

- A. Infantile Hemangioma
- B. AVM
- C. Lymphatic malformation
- D. Venous malformation
Infantile Hemangioma

**MR Findings:**
- well defined mass lesion
- hyperintense on T2, hypointense flow voids (arrows)
- homogeneous arterial phase enhancement
Infantile Hemangioma

US Findings

- High vessel density (>5/cm²)
- High peak arterial Doppler shift (>2kHz)
- Together findings suggest hemangioma
  - sensitivity: 84%
  - specificity: 98%

Dubois et al. 1998

- Solid tissue component distinguishes a hemangioma from vascular malformations
- No significant difference in vessel density or peak arterial velocity between hemangioma and AVM, but greater than low flow lesions

Paltiel et al. 2000
Infantile Hemangioma

- **Pathology:** benign vascular neoplasm, proliferative endothelium, GLUT-1 positive

- **Time Course:** typically absent at birth, with rapid growth (most in first 4-6 months) followed by involution (70% gone by 7 years)

- **Notable:** no perilesional edema, if atypical in imaging appearance, consider biopsy to evaluate for kaposiform hemangioendothelioma or angiosarcoma

- **Treatment:** typically untreated; can use beta-blockers, steroids, pulsed-dye laser or surgical resection if located in a critical area
Case

T1 + GAD At Birth
T1 + GAD At 11 months
T1 + GAD At 23 months
The demonstrated lesion likely represents...

- A. Infantile Hemangioma
- B. Rapidly involuting Congenital Hemangioma (RICH)
- C. Non-involuting Congenital Hemangioma (NICH)
- D. Venous malformation
Rapidly Involuting Congenital Hemangioma (RICH)

Similar imaging features as infantile hemangiomas, RICH are fully formed at birth and rapidly involute over the first two years of life.
Non-involuting Congenital Hemangioma (NICH)

16 year old female with left arm mass since birth

Though also fully formed at birth, NICH demonstrate little or no involution over time, in contrast to their rapidly involuting counterparts.
**Congenital Hemangioma**

- **Pathology:** benign vascular neoplasm, plump endothelium, GLUT-1 negative

- **Time Course:** fully formed at birth, will rapidly involute over the first two years of life (RICH) or never involute (NICH)

- **Notable:** vascular aneurysms, intravascular thrombi, and arteriovenous shunting are some features that can help to distinguish congenital from infantile hemangiomas by imaging (?)

- **Treatment:** none for involuting subtype, surgical resection for non-involuting subtype
Case

T1 + GAD

STIR

TR-MRA

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Kaposiform Hemangioendothelioma

MR Findings:

- Hyperintense STIR signal
- Early arterial enhancement
- Locally aggressive mass with infiltrative margins
Kaposiform Hemangioendothelioma

- **Pathology:** locally aggressive vascular neoplasm of intermediate malignant potential, rare nodal metastases

- **Time Course:** can be present at birth or develop within the first few months of life, rarely involutes

- **Notable:** associated platelet sequestration results in extremely low platelet counts and fibrinogen levels, known as Kasabach-Merritt phenomenon

- **Treatment:** resection and chemotherapy
Summary

Classification

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Imaging

- MRI with time-resolved MRA is the modality of choice
- US is useful for evaluation of flow characteristics
References


