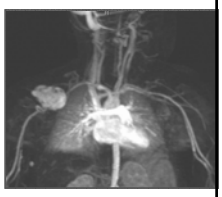
A Systematic Approach to MR imaging of Vascular Anomalies



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Case Based Learning
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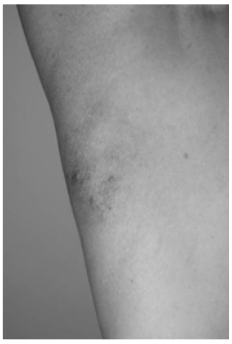
Objectives

- Understand the role of MRA is assessment of vascular anomalies
- Become familiar with the classification system of anomalies
- Describe a systematic approach to differentiation of anomalies
- Review characteristic imaging features of the more common entities

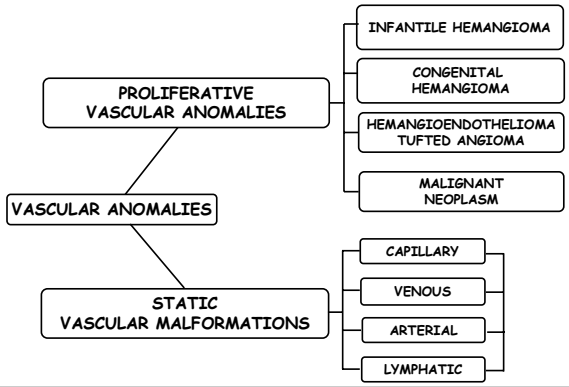
Field of Vascular Anomalies

- Complex, widely misunderstood
  - Generically "vascular birthmarks"
  - Fairly common
    - Hemangiomas 3-10%
    - Malformations ~ 0.5 %
- 1982 Mulliken & Glowacki
  - Classification scheme based on:
    - Biological differences
    - Pathologic differences
- Further refined,1996 ISSVA



Garzon et al J Ac Ped Derm 2007; Haggstrom et al J Ped 2007

Classification of Vascular Anomalies



MRI of Vascular Anomalies

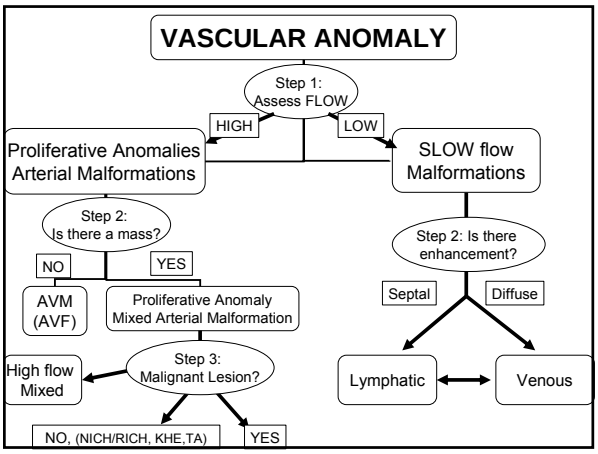
- CE-MRI is the ***ideal*** radiologic exam
  - Non-invasive, no iodinated contrast, no radiation
  - Cons: sedation, $
- Why MRI?
  - Define lesion ***extent***
  - Evaluate ***flow characteristics anatomic and temporal***
  - ***Classify*** anomaly into a category based on flow characteristics and imaging appearance
    - Keep in mind → clinical context is very important

Standard MR Protocol

- Localizer
- T2 FSE FS fat-suppressed sequence
  - At least 2 planes, coverage large field of view
  - IR may be used as an alternative if poor Freq. Selective FS
- SSFSE, single plane look for flow voids
- +/- non FS T1 SE, single plane (axial)
- 3D T1 fat-suppressed GRE pre, axial and/or cor
- Time-resolved contrast-enhanced MRA
  - Appropriate plane, < 6 second TA if possible
  - Parallel imaging + echo-sharing to improve temporal resolution
- 3D T1 fat-suppressed GRE post, axial and/or coronal

Typical Imaging Parameters

- Time-resolved MRA:
  - May use parallel imaging + echo sharing techniques to keep acquisition time down to ideally < 6 sec
  - 10-15 data sets acquired consecutively
- Initial unenhanced mask used for subtractions
- No timing run needed
  - Empiric 5-10 second delay useful to reduce number of unenhanced data sets but in very young patient with rapid circulation time start injection after mask obtained



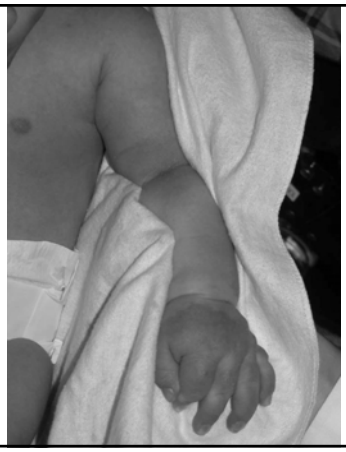
Approach to Vascular Anomalies: Q1

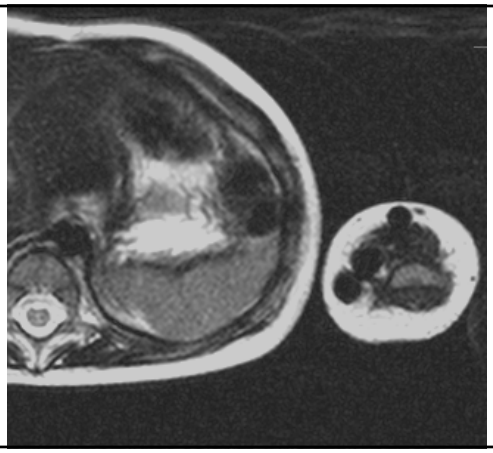
Is this a high-flow lesion?

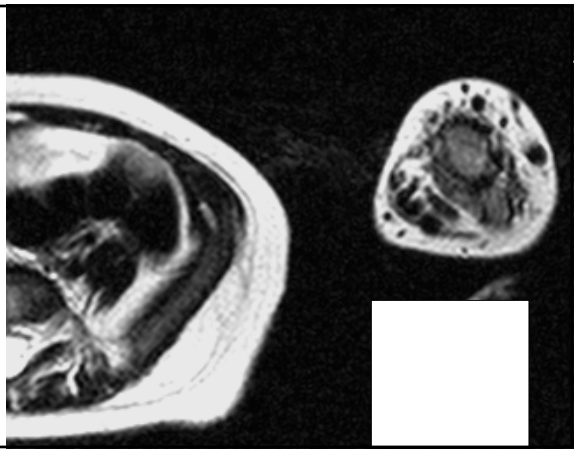
Time-resolved CE-MRA

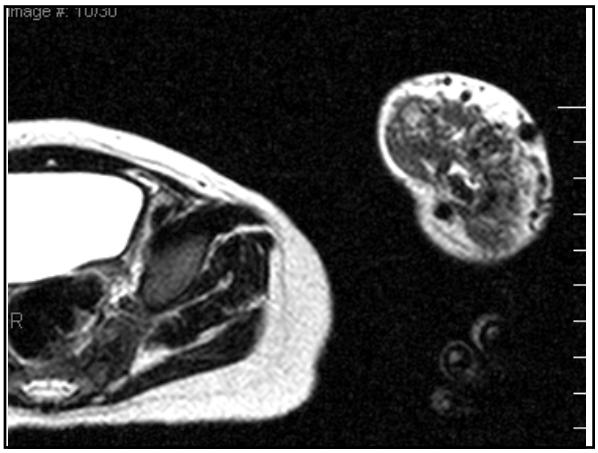
(< 6 sec, opacifies with contrast)

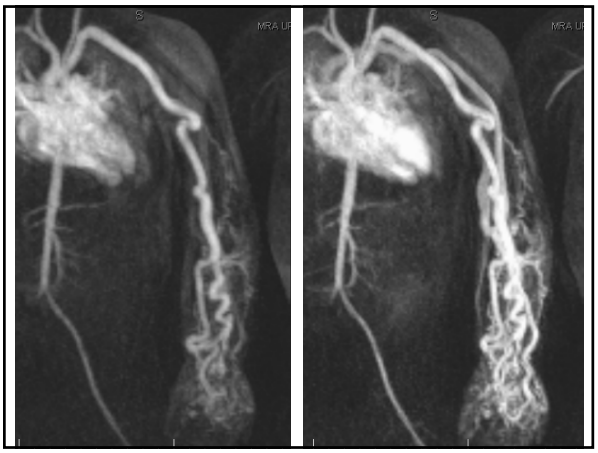
- Flow voids
- GRE: flow-related enhancement
- Comparison with contra-lateral side





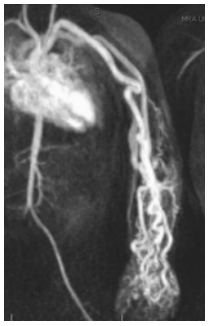






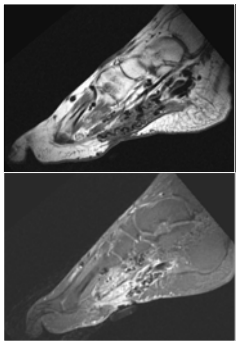
Approach to Vascular Anomalies: Q2

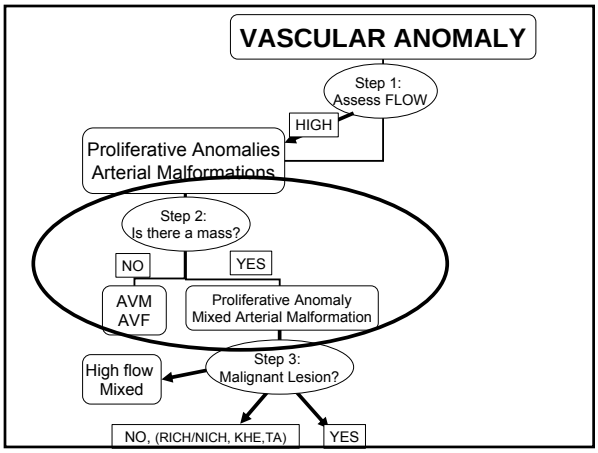
Is there a soft tissue mass?

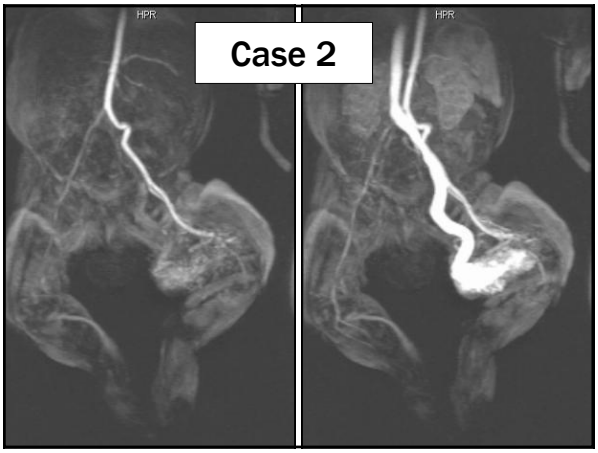


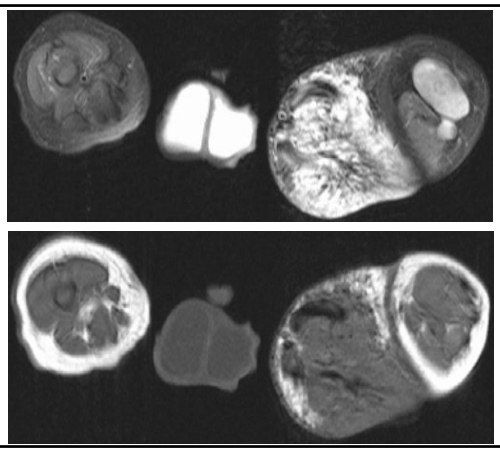
High flow, no mass = Arteriovenous Malformation

- Network of abnormal communications between arteries and veins
- MR
  - High flow enlarged vascular channels
  - Flow voids
  - Typically no associated soft tissue mass





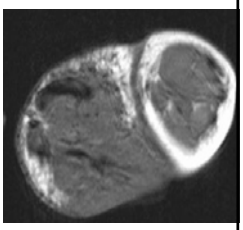




Approach to Vascular Anomalies: Q2

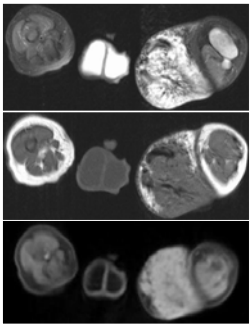
Is there a soft tissue mass?

If Yes, think proliferative anomaly rather than AVM



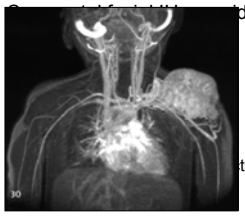
Diagnosis: Congenital Hemangioma

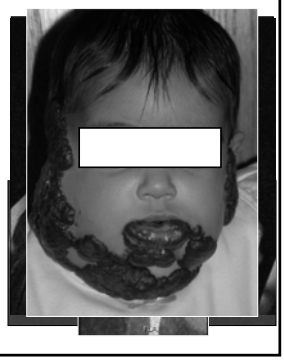
- Clinically and histologically distinct hemangiomas
- Fully developed at birth
- Undergo no further postnatal enlargement
- Two types (RICH>NICH):
  - Rapidly involuting congenital hemangioma (RICH)
  - Non-involuting congenital hemangioma (NICH)



Infantile Hemangioma

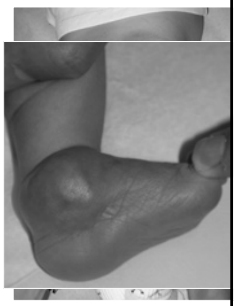
- Most common vascular tumor of infancy
- Rapid *postnatal* proliferation
- Variable stability
- Slow involution
- [illegible]



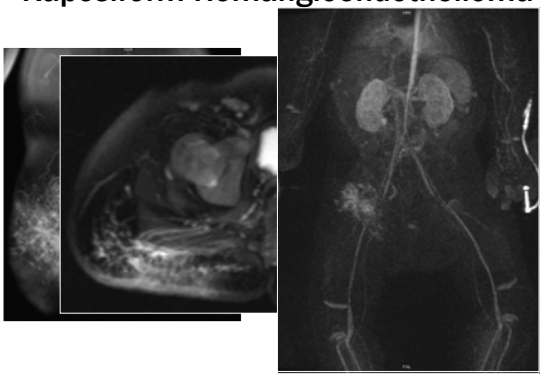


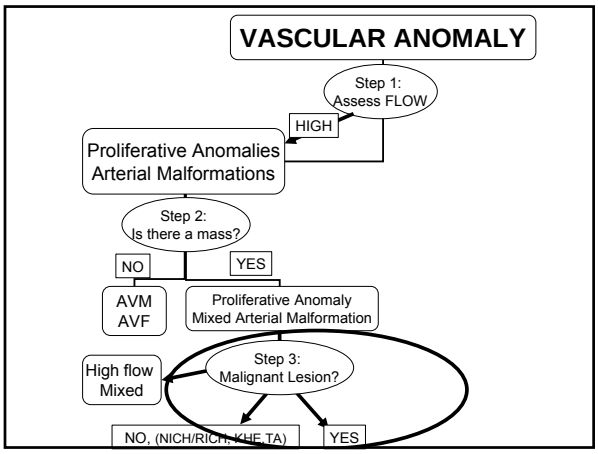
Kaposiform Hemangioendothelioma

- Rare, aggressive vascular neoplasm
- Can be present at birth or develop postnatally
  - Typically ill-defined red-purple indurated plaque
- Predilection for trunk, extremities, retroperitoneum
- Often associated with Kasabach-Merritt phenomenon
  - Severe coagulopathy due to platelet trapping



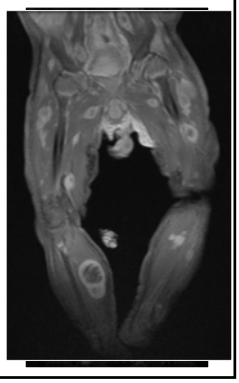
Kaposiform Hemangioendothelioma





Approach to Vascular Anomalies: Q3

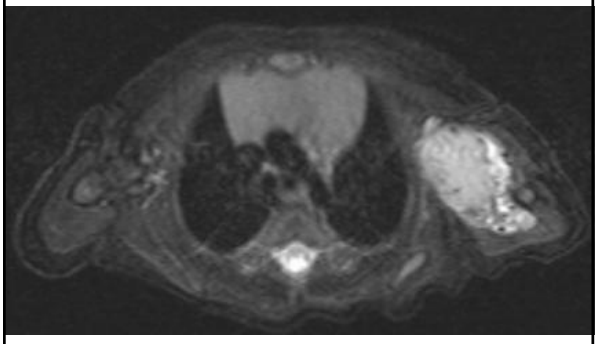
- Q3: Could this be a malignancy?
- Potential mimickers of vascular anomalies
  - Soft tissue sarcomas
    - Congenital infantile fibrosarcoma
    - Rhabdomyosarcoma
    - Malignant fibrous histiocytoma (MFH)
    - Synovial cell sarcoma
    - Angiosarcoma, hemangiosarcoma
  - Primitive neuroectodermal tumors
  - Neuroblastoma
  - Hemangiopericytoma
  - Fibromatosis/Myofibromatosis

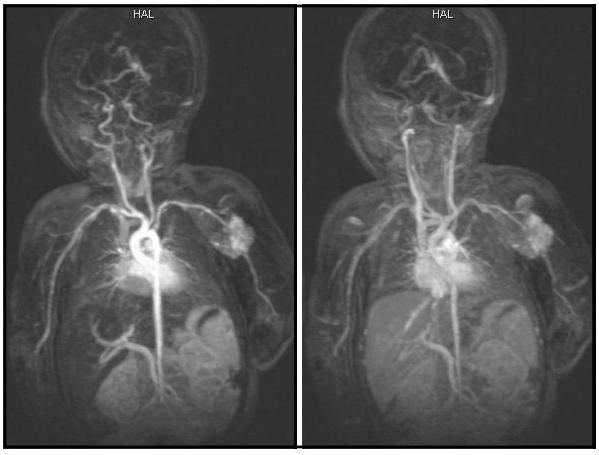


Differentiating Vascular Anomalies from Malignant Masses

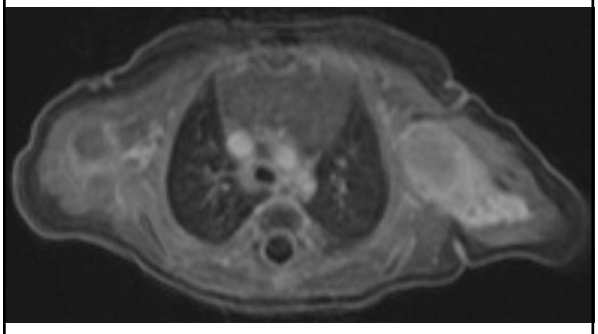
- Signal intensity, enhancement, and morphology
  - T1 signal intensity similar
  - T2 SI and contrast enhancement more uniform for hemangiomas
  - Lobulation, septation, and central low-signal intensity foci were all more common in hemangiomas
    - Presence of all three was specific
- Clinical context extremely important!
- Any doubt -> need tissue!

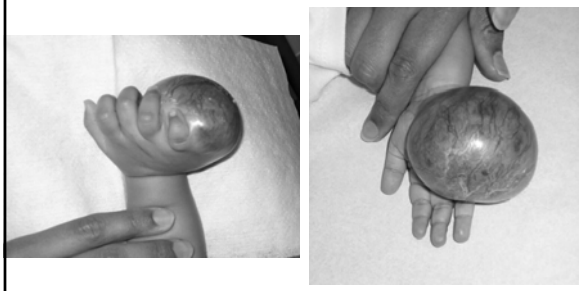
Teo et al (AJR 2000)

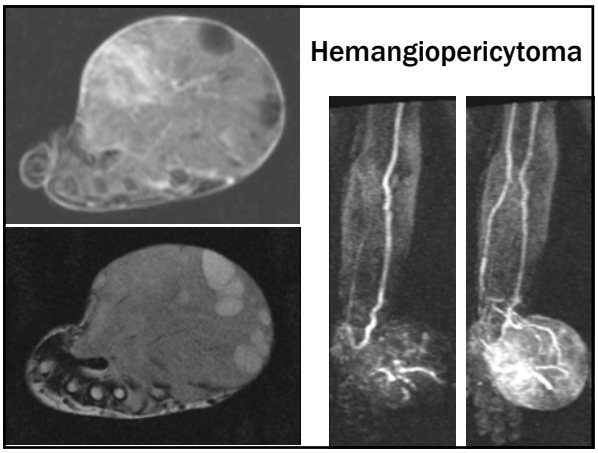


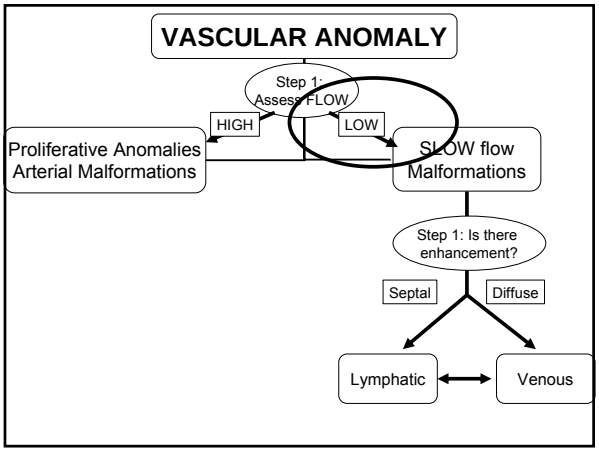


Congenital-Infantile Fibrosarcoma

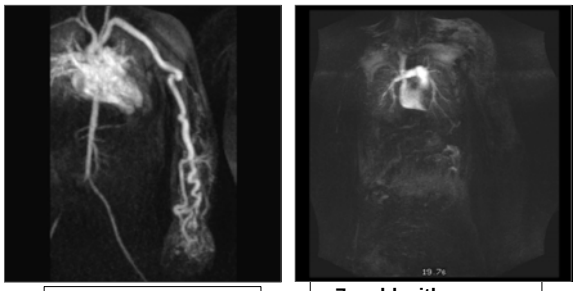








Pediatric Vascular Imaging

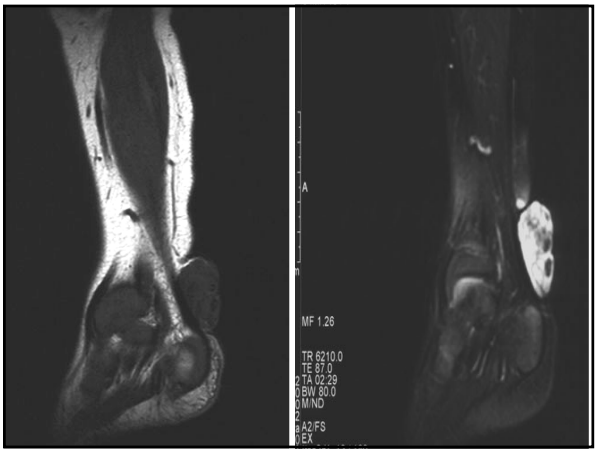


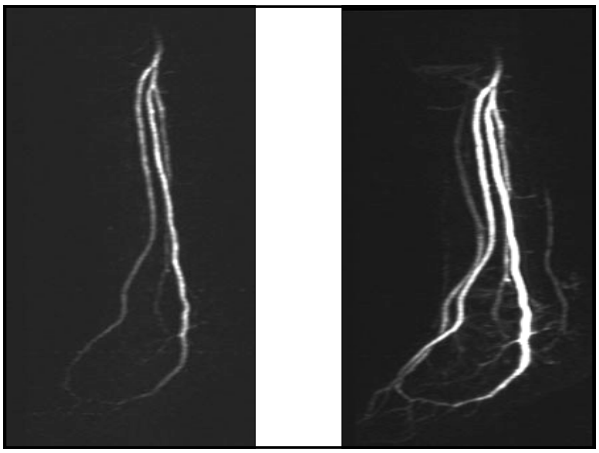
2 y old with AVM (high flow), TA=3 sec

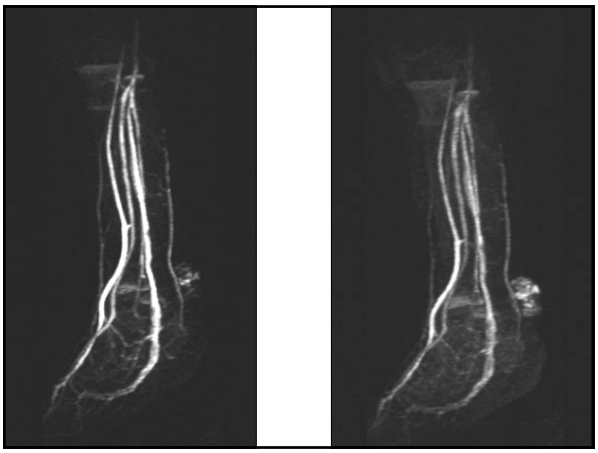
7 y old with venous malformation (low flow)

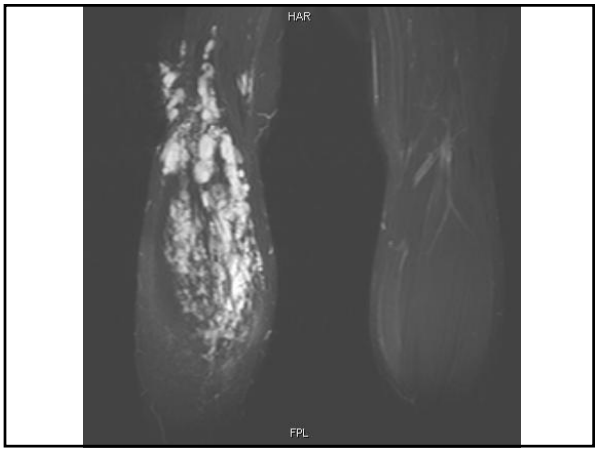
Approach to Vascular Anomalies: Q4

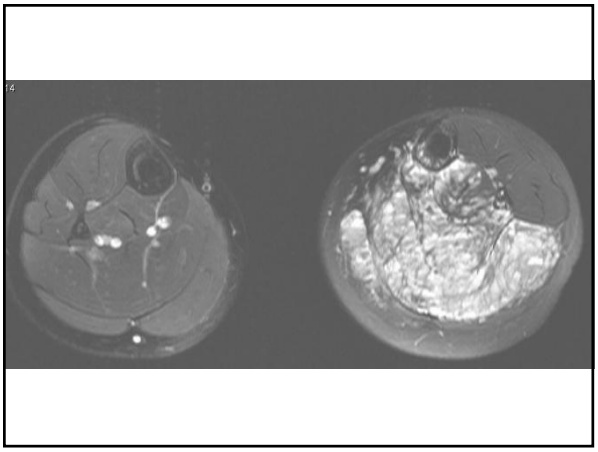
- Question Four: Okay, we're left with a low-flow lesion. Is it primarily a venous malformation or lymphatic malformation?
- Enhancement pattern:
  - Septal vs diffuse progressive enhancement





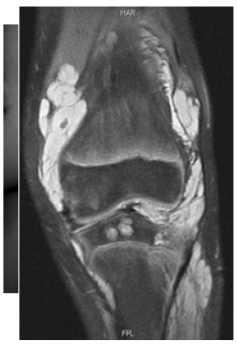






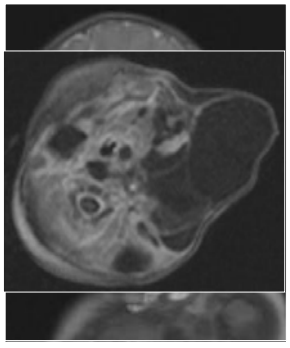
Venous Malformations

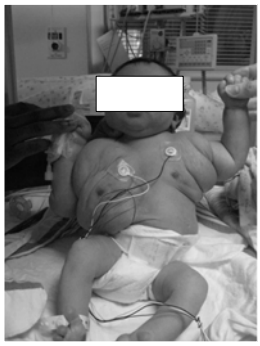
- MC asymptomatic vascular lesion
- Present at birth, may not be seen till years later
- Slow steady enlargement
- Superficial or deep, determines appearance
- Can be painful to the touch, vague congestive pain
- Cx: thrombosis/embolism, hemorrhage

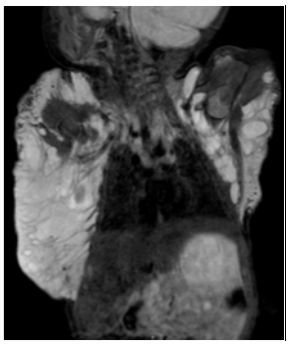


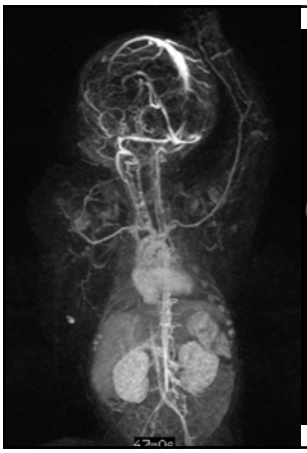
Lymphatic Malformation

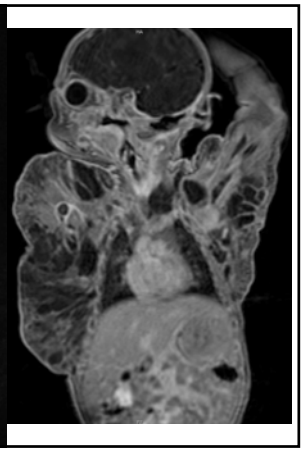
- Sponge-like collections of abnormal lymphatic channels/spaces
  - Macrocystic vs microcystic
- Neck, axilla predilection
- Steadily increase in size
- Lymphangioma, cystic hygroma (poorer names)





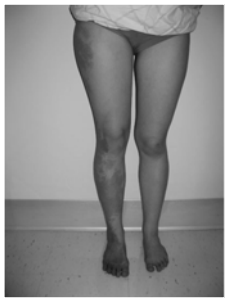


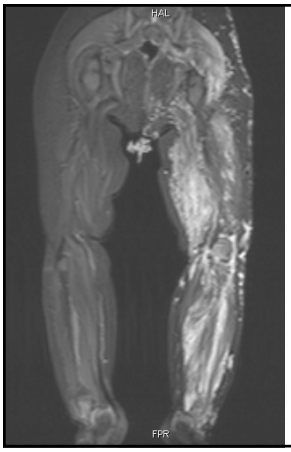


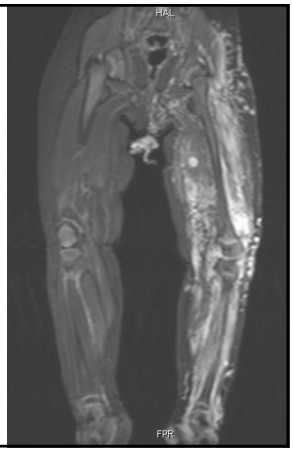


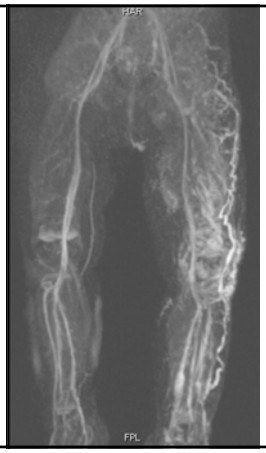
Klippel Trenaunay Syndrome

- Classic triad:
  1. Slow-flow vascular malformations
     - Cutaneous capillary malformation
     - Underlying slow flow malformation
  2. Bone and/or soft tissue hypertrophy
  3. Venous varicosities/deep venous system anomalies



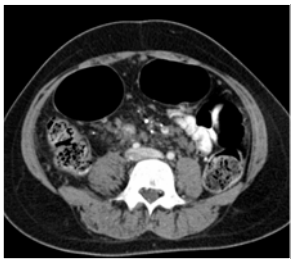






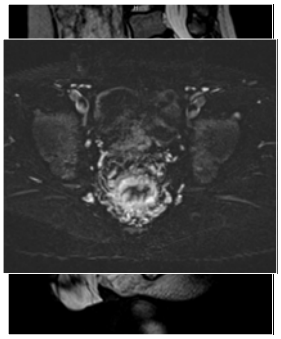
KT: Complications

- Skin/subcutaneous tissue:
  - Cellulitis
  - Chronic ulceration
- Limb length discrepancy
- Thrombophlebitis
  - Phleboliths
- Clotting abnormalities
  - Bleeding and/or thrombosis
  - Thromboembolism
  - Localized disseminated intravascular coagulation
    - Can lead to systemic coagulopathy



KT: Visceral Involvement

- Not uncommon
  - At NYU, pelvis gets imaged along with LE
- Pelvic extension fairly common
- Pay attention to GI/GU involvement
  - Can be source of life-threatening hemorrhage



Summary

- MR is the single best imaging test
- Lesions best diagnosed on basis of both clinical & imaging findings
- Remember the 4 key questions
  - High or low flow lesion?
  - Soft tissue mass?
  - Enhancement pattern?
  - Could this be a neoplasm?