A Systematic Approach to MR Imaging of Vascular Anomalies

Elizabeth M Hecht, MD
Rafael Rivera, MD
Assistant Professor
New York University
ISMRM 2009
Case Based Learning
Honolulu, Hawaii
April 23, 2009-11:50 am

Objectives
- Understand the role of MRA in 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

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

Classification of Vascular Anomalies

VASCULAR ANOMALIES

- INFANTILE HEMANGIOMA
- CONGENITAL HEMANGIOMA
- HEMANGIOENDOTHELIOMA
- Tufted Angioma
- MALIGNANT NEOPLASM

- PROLIFERATIVE VASCULAR ANOMALIES
- STATIC VASCULAR MALFORMATIONS

- CAPILLARY
- VENOUS
- ARTERIAL
- LYMPHATIC

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 coron
- 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 coron
**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

VASCULAR ANOMALY

Step 1: Assess FLOW

Step 2: Is there a mass?

Case 2
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
- Segmental facial IH - consider PHACES association
  - P - posterior fossa hemangioma of infancy
  - H - arterial abnormalities (Ao)
  - A - congenital heart disease
  - C - eye anomalies
  - E - sternal/supraumbilical defects

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
**VASCULAR ANOMALY**

- **Proliferative Anomalies**
  - Arterial Malformations

**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**
Hemangiopericytoma

**VASCULAR ANOMALY**

Proliferative Anomalies
Arterial Malformations

SLOW flow Malformations

Step 1: Is there enhancement?

- Diffuse
- Septal

Lymphatic
Venous

**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
**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

**Venous Malformations**

**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?