Update on Vascular Anomalies

A. Carl Merrow, MD
Corning Benton Chair for Radiology Education
Department of Radiology
Cincinnati Children’s Hospital Medical Center
Disclosures

• Pediatrics Lead Author for Amirsys-Elsevier
  – Royalties/Fees

• Will discuss off-label use of MRI contrast agents
  – No pharmaceutical financial ties

• NOT a dermatologist, pathologist, or interventionalist
Outline

• Background: Revised ISSVA classification

• Approach: Why/when/how to image

• Details:
  – MSK implications
  – Clues to specific lesions
  – Difficult cases
Background
Histology!

• 1982: Mulliken and Glowacki
  – Histology-based classification
  – Neoplasms vs. malformations
  – ISSVA adopts modified scheme in 1996

• Unfortunately...widespread misuse of terminology persists
Revised 2014 ISSVA Classification

- Attempt to create evolving scheme based on
  - Histopathology
  - Genetics
  - Clinical behavior of lesion

- Many fundamentals unchanged
  - A few new categories
  - Many new lesions
# ISSVA classification for vascular anomalies

(Approved at the 20th ISSVA Workshop, Melbourne, April 2014)

## Overview table

<table>
<thead>
<tr>
<th>Vascular anomalies</th>
<th>Vascular tumors</th>
<th>Vascular malformations</th>
<th>of major named vessels</th>
<th>associated with other anomalies</th>
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<tbody>
<tr>
<td>Simple</td>
<td>Combined °</td>
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<tr>
<td>Benign</td>
<td>Capillary malformations</td>
<td>CVM, CLM</td>
<td>See details</td>
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<tr>
<td>Locally aggressive or borderline</td>
<td>Lymphatic malformations</td>
<td>LVM, CLVM</td>
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<tr>
<td>Malignant</td>
<td>Venous malformations</td>
<td>CAVM*</td>
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<tr>
<td></td>
<td>Arteriovenous malformations*</td>
<td>CLAVM*</td>
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<td></td>
<td>Arteriovenous fistula*</td>
<td>others</td>
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### ISSVA classification of vascular tumors®

<table>
<thead>
<tr>
<th>Benign vascular tumors</th>
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<tbody>
<tr>
<td>Infantile hemangioma / Hemangioma of infancy</td>
<td><a href="#">see details</a></td>
</tr>
<tr>
<td>Congenital hemangioma</td>
<td></td>
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<tr>
<td>Rapidly involuting (RICH) *</td>
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<tr>
<td>Non-involuting (NICH)</td>
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<tr>
<td>Partially involuting (PICH)</td>
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<tr>
<td>Tufted angioma * °</td>
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<tr>
<td>Spindle-cell hemangioma</td>
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<tr>
<td>Epithelioid hemangioma</td>
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<tr>
<td>Pyogenic granuloma (aka lobular capillary hemangioma)</td>
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<tr>
<td>Others</td>
<td></td>
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<tr>
<td><strong>Locally aggressive or borderline vascular tumors</strong></td>
<td></td>
</tr>
<tr>
<td>Kaposiform hemangioendothelioma * °</td>
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<tr>
<td>Retiform hemangioendothelioma</td>
<td></td>
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<tr>
<td>Papillary intralymphatic angioendothelioma (PILA), Dabska</td>
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<tr>
<td>Composite hemangioendothelioma</td>
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<tr>
<td>Kaposi sarcoma</td>
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<tr>
<td>Others</td>
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<table>
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<th>Malignant vascular tumors</th>
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<tr>
<td>Angiosarcoma</td>
<td></td>
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<tr>
<td>Epithelioid hemangioendothelioma</td>
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<tr>
<td>Others</td>
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</table>

### Simple vascular malformations II

**Lymphatic malformations (LM)**

- Common (cystic) LM
- Macrocystic LM
- Microcystic LM
- Mixed cystic LM
- Generalized lymphatic anomaly (GLA)
- LM in Gorham-Stout disease
- Channel type LM
- Primary lymphedema ([different types](#))

Others

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Approach
Histopathology

- Neoplasms vs. Malformations vs. Secondary
Clinical/Imaging

• Discrete masses vs. Macroscopic channel abnormalities

• High flow vs. Low flow lesions

• Focal vs. Multifocal vs. Extensive/Diffuse

• Combinations in certain anomalies/syndromes
Imaging: When, why

• Newly detected mass(es)
  – Diagnosis (especially without skin involvement)
  – Extent of focal lesion
  – Therapy options/baseline prior to intervention
Imaging: When, why

• Cutaneous lesion(s) with specific implications
  – Local complications

  – Known associations of internal significance
    • Multifocal/extensive deep lesions
    • Other anomalies that may be remote/widespread
      – Congenital (visceral, vascular)
      – Tumor predisposition
Techniques: 
Limb vs. Joint
Limb assessment

**Soft tissue/marrow**
- Fat/marrow (T1)
- Fluid-sensitive (STIR/FS T2)
- Contrast or not? Subtraction?
- Other considerations
  - DWI
  - In/Out of phase
Limb assessment

Vascular

- Flow sensitive (2D SPGR, TOF)
- Dynamic enhanced techniques
  (Ultrafast 3D SPGR vs. 4D time-resolved)
- Traditional extracellular contrast vs. blood pool agent
- MR lymphangiography?
Soft tissue/marrow
Fat/marrow
(T1)
(STIR/FS T2)

2D TOF
20-30 min

Immediate post
3D T1 GRE
3 min
PreGad 2D TOF (11 min)

4 min post THRIVE (82 sec)

10 min post T1 FS SE (4.6 min)
Joint assessment

• Dedicated coil!
• Hemosiderin-sensitive (GRE)
• Cartilage-sensitive
  • Traditional 2D: GRE, PD/IW +/- FS
  • High resolution 3D: GRE, Cube/SPACE/VISTA PD/T2
  • Advanced: T2 map, T1 rho, dGEMRIC
• Dynamic vs. early post-contrast
Questions

- Actual size of coverage (cm)
- Deep venous vs. arterial assessment (vs. none)
- Dedicated joint imaging
- Follow-up exam
Potential Problems

STIR after Gadofosveset

STIR before contrast
MSK Implications
MSK Implications

- Growth disturbances
  - Overgrowth
  - Undergrowth
  - Deformities

- Hemarthrosis/degeneration

- Scoliosis

- Muscle dysfunction

Limb Length Discrepancy
Joint Involvement
Joint Involvement
Joint Involvement
Joint Involvement
Scoliosis
Specific Lesions
Benign tumors

Infantile hemangioma

Congenital hemangioma
- Rapidly involuting (RICH)
- Non-involuting (NICH)
- Partially involuting (PICH)

Tufted angioma

Spindle-cell hemangioma

Epithelioid hemangioma

Pyogenic granuloma

Others
Infantile Hemangioma

- Capillaries lined by plump endothelial cells
  - GLUT1+

- Age:
  - Proliferation: First weeks-months of life
  - Involution: Over years

- Coagulopathy: NO

- Therapy: Propranolol, steroids, rarely chemo
Infantile Hemangioma

- Rarely imaged unless deep or significant location
- Key imaging:
  - Solid, lobulated elongated or ovoid masses
  - Heterogeneous echogenicity
  - Highly vascular: Many low resistance arteries
  - Bright (not fluid) on FS T2; diffusely enhance early
  - Gradual fatty replacement

Infantile Hemangioma
Infantile Hemangioma
Congenital Hemangioma

- Capillaries intermixed with dilated vessels, hematopoiesis
  - GLUT1-
- Age: Perinatal detection; proliferation ceases by birth
  - RICH: Involution over 3-12 months
  - NICH: Stable
- Coagulopathy
  - Consumptive: Mild, transient
- Therapy: None, excision, embolization, ? steroids
- Key imaging: Variable, can be more heterogeneous

Locally aggressive/borderline tumors

- Kaposiform hemangioendothelioma
- Retiform hemangioendothelioma
- Composite hemangioendothelioma
- Papillary intralymphatic angioendothelioma/
  Dabska tumor
- Kaposi sarcoma
- Others
Kaposiform Hemangioendothelioma

- Aggressive, infiltrative lesion
- Nodules of spindled endothelial cells with abnormal lymphatics
- Age: Infants most common (>90%)
- Coagulopathy
  - Consumptive: Severe, sustained
    - Kasabach-Merritt phenomenon
- Therapy: Sirolimus, vincristine, steroids

Kaposiform Hemangioendothelioma

• Key imaging:
  – Solid, poorly defined, infiltrative mass
  – May have nodular components of low/intermediate T2
  – Diffusely enhances
  – +/- Surrounding edema
  – Few large vessels internally
Kaposiform Hemangioendothelioma
Kaposiform Hemangioendothelioma
Kaposiform Hemangioendothelioma
Kaposiform Hemangioendothelioma
Venous malformation

Isolated common VM
Blue Rubber Bleb Nevus syndrome (BRBNS)
Glomovenous
Cerebral cavernous malformation
Others
Venous Malformation

- Large dilated channels with muscularized walls
- Age: Congenital but presentation timing variable
- Coagulopathy
  - Localized intravascular
  - Rarely DIC
- Therapy: Compression, sclero, anticoagulation

- BRBNS
  - Numerous focal malformations, GI bleeding

Venous Malformation

• Key imaging:
  – Lobulated mass &/or numerous channels
    • Large intramuscular lesions often follow fiber orientation
  – Stagnant blood (fluid-fluid levels)
    • Compressible
  – Thrombi/phleboliths
  – +/- Prominent fat along margins
  – Patchy, gradual enhancement
Venous Malformation
Venous Malformation
Venous Malformation
Lymphatic malformation

Common (cystic) LM
- Macrocystic
- Microcystic
- Mixed

Generalized lymphatic anomaly (GLA)

Gorham-Stout Disease

Channel type LM

Others
Lymphatic Malformation

• Macro/microcysts with characteristic endothelium
  – Prox1+
  – D240+

• Age: Congenital but presentation timing variable

• Coagulopathy
  – Localized intravascular
  – Rarely DIC

• Therapy: Compression, sclero, surgery, sirolimus

Lymphatic Malformation

• Key imaging
  – Multicystic mass
    • Varying fluid complexities in different cysts
      – Fluid-fluid levels
      – May show T1 shortening pre-contrast
    • Thin septations
      – +/- Rim enhancement
  – Extends across tissue planes/compartments

Lymphatic Malformation

Lymphatic Malformation

Generalized Lymphatic Anomaly

- Macrocystic LM
- Pleural effusions
- Numerous noncontiguous cystic lesions
  - Bone (+/- expansion, no osteolysis)
    - Additional osseous fatty infiltration often present
  - Spleen

Generalized Lymphatic Anomaly
Gorham

- Microcystic LM
- Aggressive local osteolysis
- Visceral involvement much less common
- Characteristic imaging: Gradual destruction of multiple adjacent bones (beyond one joint)
Gorham

Syndromic malformations

- Klippel-Trenaunay
- Parkes-Weber
- Servellle-Martorell
- Sturge-Weber
- Maffuci
- CLOVES
- Proteus
- Bannayan-Riley-Ruvalcaba

Combined malformations

- Capillary-venous malformations
- Capillary-lymphatic malformations
- Capillary-veno-lymphatic malformations
- Capillary-arterio-veno-lymphatic malformations
- Others
Klippel-Trenaunay

- CM + VM +/- LM
  - No “high flow” components
- Limb overgrowth
  - Fat, bones, vessels
- Large lateral primitive veins
  - Thromboembolism
  - Thrombophlebitis
- Abnormal deep venous system
• Widespread enchondromas
• Soft tissue vascular anomalies with phleboliths
  – Spindle cell hemangiomas
• Risk of malignancy
  – Enchondroma → Chondrosarcoma
  – Vascular → Angiosarcoma
  – Ovarian, GI, glial
Maffucci
CLOVES

Congenital Lipomatous Overgrowth Vascular malformations Epidermal nevi Spinal/Skeletal anomalies
Unclassified anomalies

Verrucous hemangioma
Multifocal lymphangioendotheliomatosis with thrombocytopenia (MALT)/Cutaneovisceralangiomatosis with thrombocytopenia (CAT)
Kaposiform lymphangiomatosis (KLA)
PTEN hamartoma of soft tissue (PHOST)
Kaposiform Lymphangiomatosis (KLA)

- Lesion of spindle cells, abnormal lymphatics
- Age: Wide range (median 6.5 years)
- Coagulopathy:
  - Mild/moderate thrombocytopenia, hypofibrinogenemia
- Key imaging: Many GLA-type features, PLUS
  - Infiltrative microcystic-appearing disease of
    - Mediastinum, pleura, pericardium
    - Perihilar & peripheral pulmonary interstitium

Kaposiform Lymphangiomatosis (KLA)
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Kaposiform Lymphangiomatosis (KLA)
**PTEN** Hamartoma of Soft Tissue (PHOST)

- Variety of vascular and fatty hamartomatous lesions, including high and low flow masses

- Variable age; clinical characteristics include
  - Macrocephaly, penile freckling, developmental delay
  - No coagulopathy

PTEN Hamartoma (PHOST)

Difficult Cases
2-month-old with growing foot mass
2-month-old with growing foot mass
Infantile Fibrosarcoma
5-year-old with shoulder mass
5-year-old with shoulder mass
Extraskeletal Ewing sarcoma
4-month-old with facial mass
Rhabdomyosarcoma
Conclusions
Vascular Anomalies

• Initial clinical/imaging approach
  – Clinical: Age, cutaneous appearance, firmness, coagulopathy
  – Imaging:
    • Masses vs. vessels or combination
    • High or low flow
    • Focal or extensive

• Histology-based classification ultimately key for
  – Diagnosis
  – Prognosis
  – Treatment
How to Image

• Tailor protocols to specific needs
  – Coverage: Limb vs. Joint
  – Tissues: Osseous, soft tissues vs. synovium, cartilage
  – Vessels: Arteries, veins, or lymphatics

• Dig for more information from clinicians!
MSK Specifics

- Many implications
  - Hemarthrosis/degeneration
  - Limb deformities
  - Scoliosis

- Depend on diagnosis, location, extent
Final Diagnosis

- Many suggestive clinical/imaging features of specific vascular anomalies

- For a solid or mixed mass without clear clinical &/or imaging findings of a specific vascular anomaly, get tissue!
Thank you!

carl.merrow@cchmc.org