Disclosure

- Consultant/Author for Amirsys/Elsevier
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**

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### ISSVA classification of vascular tumors

#### Benign vascular tumors

- Infantile hemangioma / Hemangioma of infancy
- Congenital hemangioma
  - Rapidly involuting (RICH) *
  - Non-involuting (NICH)
  - Partially involuting (PICH)
- Tufted angioma *
- Spindle-cell hemangioma
- Epithelioid hemangioma
- Pyogenic granuloma (aka lobular capillary hemangioma)
- Others

#### Locally aggressive or borderline vascular tumors

- Kaposiform hemangioendothelioma *
- Retiform hemangioendothelioma
- Papillary intralymphatic angioendothelioma (PILA), Dabska tumor
- Composite hemangioendothelioma
- Kaposi sarcoma
- Others

#### Malignant vascular tumors

- Angiosarcoma
- Epithelioid hemangioendothelioma
- Others

### 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
MSK Implications
MSK Implications

• Growth disturbances
  – Overgrowth
  – Undergrowth
  – Deformities
• Hemarthrosis/degeneration
• Scoliosis
• Muscle dysfunction

Limb Length Discrepancy
Joint Involvement
Joint Involvement
Joint Involvement
Scoliosis
Scoliosis
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 (T1)
Fat/marrow (STIR/FS T2)

2D TOF
20-40 min

Immediate post 3D T1 GRE
3 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
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
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
Congenital Hemangioma
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
Blue Rubber Bleb Nevus Syndrome
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

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)

**Combined**
- Capillary-venous malformations
- Capillary-lymphatic malformations
- Capillary-veno-lymphatic malformations
- Capillary-arterio-veno-lymphatic malformations
- Others

**Syndromic malformations**
- Klippel-Trenaunay
- Parkes-Weber
- Servelle-Martorell
- Sturge-Weber
- Maffuci
- CLOVES
- Proteus
- Bannayan-Riley-Ruvalcaba
Klippel-Trenaunay

- CM + VM +/- LM
  - No “high flow” components
- Limb overgrowth
  - Fat, bones, vessels
- Large lateral primitive veins
  - Thromboembolism
  - Thrombophlebitis
- Abnormal deep venous system
Klippel-Trenaunay
Maffucci

- Widespread enchondromas
- Soft tissue vascular anomalies with phleboliths
  - Spindle cell hemangioiomas
- Risk of malignancy
  - Enchondroma → Chondrosarcoma
  - Vascular → Angiosarcoma
  - Ovarian, GI, glial
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)
Kaposiform Lymphangiomatosis (KLA)
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
  - Macroccephaly, penile freckling, developmental delay
  - No coagulopathy

PTEN Hamartoma (PHOST)

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
MSK Specifics

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

- Depend on diagnosis, location, extent
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!
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!
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