MRI Assessment of Abdominal Vascular Anomalies: Update

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Disclosure

• Lead author for Pediatrics in Amirsys/Elsevier
  – Fees/Royalties
Outline

• ISSVA classification

• MRI techniques

• Specific lesions
Background
# ISSVA classification for vascular anomalies

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

## Overview table

<table>
<thead>
<tr>
<th>Vascular tumors</th>
<th>Vascular malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Simple</td>
</tr>
<tr>
<td></td>
<td>Combined °</td>
</tr>
<tr>
<td></td>
<td>of major named vessels</td>
</tr>
<tr>
<td></td>
<td>associated with other anomalies</td>
</tr>
<tr>
<td>Benign</td>
<td>Capillary malformations</td>
</tr>
<tr>
<td></td>
<td>CVM, CLM</td>
</tr>
<tr>
<td>Locally aggressive or borderline</td>
<td>LVM, CLVM</td>
</tr>
<tr>
<td>Malignant</td>
<td>Arteriovenous malformations*</td>
</tr>
<tr>
<td></td>
<td>CAVM*</td>
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<tr>
<td></td>
<td>CLAVM*</td>
</tr>
<tr>
<td></td>
<td>others</td>
</tr>
<tr>
<td></td>
<td>See details</td>
</tr>
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<td></td>
<td>See list</td>
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</tbody>
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[http://issva.org](http://issva.org)

Vascular Anomalies

Vascular Neoplasms

- Hemangioma
  - Infantile
  - Congenital
    - RICH
    - NICH
- Hemangioendothelioma
- Kaposiform
- Others
- Angiosarcoma

Vascular Malformations

- Venous
- Lymphatic
- Arterial
- Capillary
- Combined
Body Manifestations

• Visceral &/or soft tissue masses
  – Solitary vs. multiple or confluent/diffuse
  – +/- Cutaneous lesions, serum anomalies

• Anomalies of named vessels

• Associations with distant malformations or tumors
MRI Techniques
What do I want from MRI?

- Determine internal lesion characteristics
- Define lesion extent & vascular relationships
- Detect additional smaller lesions
- Survey surrounding viscera
Targeted MRI: What sequences?

- Cor T1 FSE
- Ax & cor T2 FS
- Ax 2D TOF
- Ax DWI
- Axial fat sat pre & post LAVA/THRIVE/VIBE or Dixon
  - Dynamic > rapid time resolved MRA
- Cor fat sat T1 FSE

*Rough guidelines!
Targeted MRI: Other options?

- Ax T1 FSE (for body wall masses)
- STIR for fat sat inhomogeneity or spine
- SSFSE &/or SSFP for speed
- CUBE/SPACE/VISTA for reformatting
- Time resolved MRA for AVMs
- Other non-con MRA/MRV techniques
Whole Body MRI

- Cor T1
- Cor STIR
- Ax post LAVA/THRIVE/VIBE or Dixon
2D TOF
20-40 min

Immediate post
LAVA/THRIVE/VIBE
3 min
Specific Lesions: Neoplasms
Congenital Hemangioma

• Clinical
  – Rapidly (RICH) vs. non (NICH) vs. partially involuting (PICH)
  – Detected prenatally or at birth
  – Heart failure, mild anemia + thrombocytopenia, neonatal AFP levels
  – May have associated cutaneous lesions

• Histology
  – Benign endothelial channels, varying size; fibrosis, Ca $^{2+}$
  – GLUT1 negative on histology

Congenital Hemangioma
Congenital Hemangioma
Congenital Hemangioma

• Beware this diagnosis
  – After 1st week of life
  – Rising or steadily elevated alpha-fetoprotein (AFP)
  – Increasing size
  – If in doubt, biopsy (carefully)
Infantile Hemangioma

• Clinical
  • Characteristic cutaneous appearance
  • Number important
  • Predictable life cycle

• Histology
  • Benign neoplasms of capillaries
  • GLUT1 positive on histology
Multifocal Infantile Hemangiomas
Diffuse Infantile Hemangiomas
Infantile Hepatic Hemangiomas

• Medical Therapies
  – Propranolol
  – Steroids
  – Rarely Vincristine
  – Not Interferon

• Surgical Therapies
  – Transplant
Infantile Hepatic Hemangiomas

• Beware this diagnosis
  – If there are atypical or no cutaneous lesions
  – Dominant hepatic lesion or other abdominal masses
  – Life cycle deviation
## Pediatric Hepatic Hemangiomas

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Infantile</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present at birth</td>
<td>Appear within weeks-months</td>
</tr>
<tr>
<td>Solitary</td>
<td>Multifocal or diffuse</td>
</tr>
<tr>
<td>Large</td>
<td>Small to moderate</td>
</tr>
<tr>
<td>Heterogeneous</td>
<td>Homogeneous</td>
</tr>
<tr>
<td>Big vessels, shunting</td>
<td>+/- Vascularity, shunting</td>
</tr>
<tr>
<td>Broadening peripheral enhancement, not central</td>
<td>Early peripheral to delayed central enhancement</td>
</tr>
<tr>
<td>GLUT-1 negative</td>
<td>GLUT-1 positive</td>
</tr>
</tbody>
</table>
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
Specific Lesions: Malformations
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
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/compartment
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-Stout Disease

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

Specific Lesions: Uncertain
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)
Conclusions

- Familiarity with ISSVA classification + clinical & radiologic features → clinical impact

- Variations from expected patterns should drive further workup

- Variety of MR techniques may be employed but should be tailored to specific concerns