Venous and lymphatic malformations and anomalies

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Disclosure Statement of Financial Interest

I, Robert Schainfeld, **DO NOT** have a financial interest/arrangement or affiliation with one or more organizations that could be perceived as a real or apparent conflict of interest in the context of the subject of this presentation.
Congenital Venous Malformations

- Rare complex lesions of malformed vessels occur due to maturational arrest during embryogenesis (weeks 4 – 10)
Congenital Venous Malformations

Clinical manifestations

- Port - wine stains
- Arteriovenous fistulas
Etiology of Congenital Venous Malformations

- **Stage 1** – syncytial network of undifferentiated capillary blood spaces
- **Stage 2** – retiform plexus develops as capillary network and organizes into central channels
- **Stage 3** – Development of axial vessels in limb bud occurs
Etiology of Congenital Venous Malformations

- **Stage 1** – port-wine stain & cutaneous nevi
- **Stage 2** – cavernous hemangiomas and venous lakes
- **Stage 3** – absence or hypoplasia of mature arteries and veins
Table 4  Updated ISSVA classification of vascular anomalies.

<table>
<thead>
<tr>
<th>Vascular tumors</th>
<th>Vascular malformations</th>
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<tbody>
<tr>
<td>Infantile hemangiomas</td>
<td><strong>Slow-flow vascular malformations:</strong></td>
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<tr>
<td>Congenital hemangiomas (RICH and NICH)</td>
<td>• Capillary malformation (CM)</td>
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<tr>
<td>Tufted angioma (with or without</td>
<td>• Port-wine stain</td>
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<td>Kasabach–Merritt syndrome)</td>
<td>• Telangiectasia</td>
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<td>Kaposiform hemangioendothelioma (with</td>
<td>• Angiokeratoma</td>
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<td>or without Kasabach–Merritt syndrome)</td>
<td>• Venous malformation (VM)</td>
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<td>Spindle cell hemangioendothelioma</td>
<td>• Common sporadic VM</td>
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<td>Other, rare hemangioendotheliomas (epi-</td>
<td>• Bean syndrome</td>
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<tr>
<td>thelioid, composite, retiform, polymor-</td>
<td>• Familial cutaneous and mucosal venous malformation (VMCM)</td>
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<td>phous, Dabska tumor, lymphangioendothe-</td>
<td>• Glomuvenous malformation (GVM)</td>
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<td>liomatosis, etc.)</td>
<td>(glomangioma)</td>
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<td>Dermatologic acquired vascular tumors</td>
<td>• Maffucci syndrome</td>
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<td>(pyogenic granuloma, targetoid hemangi-</td>
<td>• Lymphatic malformation (LM)</td>
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<td>oma, glomeruloid hemangioma, microven-</td>
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<td>ular hemangioma, etc.)</td>
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**Fast-flow vascular malformations:**

- Arterial malformation (AM)
- Arteriovenous fistula (AVF)
- Arteriovenous malformation (AVM)

**Complex-combined vascular malformations:**

- CVM, CLM, LVM, CLVM,
  AVM-LM, CM-AVM

C=capillary; V=venous; L=lymphatic; AV=arteriovenous; M=malformation. RICH=rapidly involuting congenital hemangioma; NICH=noninvoluting congenital hemangioma.
Clinical Presentation of CVM

• Depends on dominant feature
  - venous, arterial, capillary or lymphatic
• No gender predilection
• Incidence < 2% of population
• Venous (50%), extremities (67%)
• Arterial (< 1%)
• Mixed (15-30%)
Clinical Presentation of CVM

- Cutaneous vascular stain
- Palpable mass
- Limb edema
- Varicosities
- Thrombophlebitis
- Complications of venous hypertension
Clinical Manifestations of CVM

- Mass is firm, palpable, pulsatile, and non-compressible
- Continuous bruit over AVF & thrill
- Limb is warm, increased girth and heaviness
- Scoliosis and limb length disparities
Complications of CVM

• Extremity lesions may present with ischemic rest pain or ulcerations due to shunting
• Pelvic malformations
  - rectal pain
  - sexual dysfunction
  - uterine bleeding
  - ureteral outlet outlet (hydronephrosis)
• High - output CHF due to pelvic/abdominal high - flow lesions
Venous Aneurysms

- Jugular and superficial: soft, compressible, non-pulsatile and distend with Valsalva
- Vena caval: asx, incidentally found
- Popliteal: DVT/PE
- Rupture is rare
Diagnosis and Evaluation of CVM

- Plain films (phleboliths)
- CT angiography – inhomogenous contrast enhancement
- Contrast venography
- Duplex ultrasound - hypoechogenic, septated masses
- MRI/MRV
  - Hyperintense on T2- images
  - No flow voids
  - Inhomogenous contrast enhancement
Diagnosis and Evaluation of CVM

- MRI is diagnostic imaging modality of choice
- Allows imaging in both axial and sagittal planes, in addition to ID extension into soft tissue and bone
Management of CVM

- Depends on lesion type, location, extent, and patient symptoms
- Venous anomalies treat conservatively with external support prn
- Jugular aneurysms benign, if large may be ligated
- Caval saccular aneurysms may be ligated at base and excised
Management of CVM

• Know deep venous physiology!!!
  - Superficial varicosities
  - Popliteal venous aneurysms high - risk for VTE (DVT / PE)
  - Compression stockings and sleeves
Management of CVM

• Multidisciplinary Team
  - vascular surgeons, vascular medicine, interventional radiologists, plastic and orthopedic surgeons
Indications for Treatment of CVM

• Interventional and/or surgical excision
  - CHF
  - ischemia
  - bleeding
  - ulceration
  - functional impairment
  - severe cosmetic deformity
Medical Treatment of CVM

• Sclerotherapy is primary treatment of CVM
• Larger lesions are treated with 95% ethanol
• Smaller cutaneous lesions Rx with sodium teradecyl sulfate (1%)
• Multiple sessions
• Propensity for recanalization and recurrence
• Alternative to sclerotherapy is foam sclerosant
Treatment Options of CVM

- Multiple, staged transcatheter embolizations
- Surgical excision
- Arterial embolization may decrease inflow sxss, venous changes do not respond, require lifelong compressive Rx
Treatment Algorithm for CVM

MRI → US → Low-flow malformation
  | Symptomatic
  | → Sclerotherapy
  | Follow-up with
  | US

Asymptomatic
  | Observation
  | → Surgical procedure
  | → Physical therapy
Complications of Endovascular Therapies

- Occur < 6% patients
- Edema, swelling
- Ischemia
- Distal tissue necrosis
- Contrast nephrotoxicity
- Allergic rxns
- Embolization of particulate matter via AVF (stroke / PE)
Timing of Hybrid Procedures

• Embolization should be done as close to surgery so as to decrease time available for collateral recruitment
Case Study

- 25 yo male with hx of right leg pain/edema
- Prior Hx of right small saphenous vein STP
Duplex Ultrasound of Right Leg
MRI of Right Calf

- T1 pre
MRI of Right Calf

- T2
MRI of Right Calf

- T1 post
MRA of Right Leg
MRA of Right Leg
Contrast Angiography of Right Leg
Contrast Angiography (Late)
Contrast Angiography (Later)
Contrast Angiography (Latest)
Factoids About Hemangiomas

• Most hemangiomas asymptomatic and small
• Muscle hemangiomas account for 10 of 570 hemangiomas
• Intramuscular hemangiomas occur most often in:
  - young people (ranges 2 mo to 66 yrs)
  - 80-90% presenting in persons younger than 30 YO
  - Males and females affected with equal frequency
  - In contrast to venous malformations, intramuscular hemangiomas have arterial feeders and in contrast to AVMs, these vascular lesions do not demonstrate arteriovenous shunting
Hemangiomas

• Birthmark appears as a rubbery, bright red nodule of extra blood vessels in the skin (AKA strawberry mark)
• Grows during 1st year of life, and recedes over time
• Hemangioma during infancy usually little visible trace of the growth by age 10
• Occurs anywhere on body, most commonly face, scalp, chest or back
Hemangiomas
Treatment of Hemangiomas

- Most **DO NOT** warrant treatment, resolve over time
- Corticosteroids can be injected into nodule, given orally or applied topically
- Lasers can arrest growth
  - treat sores on a lesion that won't heal.
  - side effects include pain, infection, bleeding, scarring and changes in skin color
MRI of Left Leg
MRI of Left Leg
MRI of Left Leg
MRI of Left Calf
Arterial Phase (No Filling)
Venous Phase (No Filling)
Delayed Phase (With Filling)
Delayed Phase (Filling)
Delayed Phase (Filling)
Clinical Outcomes

- Many authors reporting 80% or greater control or cure of venous anomalies
- Venous components nearly always can be closed (treated) with sclerotherapy
- The lymphatic components are more difficult
  - Sclerotherapy response rates of 40-50%
Conclusions

• Venous Malformations
• Diagnosis from
  – Clinical examination
  – Imaging, especially duplex ultrasound
• Treatment with laser ablation and sclerotherapy is effective