Congenital Vascular Disorders

Cynthia K. Shortell, MD

Disclosures

• I have no relationships to disclose

• I will be discussing off label or unapproved uses of drugs or devices in my presentation.

Outline

• Cystic Adventitial Disease
• Popliteal Artery Entrapment Syndrome
• Vascular Malformations

Cystic Adventitial Disease

• Rare (< 0.1% population)
• 1st described 1947 by Atkins and Key
  - 40 yo M w/ thigh claudication
  - "myxomatous tissue arising from the posterior aspect of the external iliac artery"
• May involve any artery or vein, but popliteal artery most common (85%)


Cystic Adventitial Disease

Etiology: Theories

• Embryologic synovial or ganglial cells trapped in artery
  - Histology of CAD lesions resemble ganglia and synovial cells...
• Repeated stress from knee joint causes microtrauma and degeneration of arterial wall with separation of components

**Cystic Adventitial Disease**

**Clinical Presentation:**
- Healthy, non-smoking middle aged men
- Rapid onset of *unilateral* claudication
- Loss of distal pulse with knee flexion

**Diagnosis:**
- Abnormal ABI (resting and exercise)
- Duplex, stenosis + cystic lesion
- MRI
- Catheter based arteriography

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**Arteriography**

- Scimitar sign
- Concentric lesion
- Hourglass sign
- Eccentric lesion

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**Magnetic Resonance Imaging**

Sequential axial T-2 weighted images

Severe compression of popliteal artery

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**Treatment**

- Few studies to guide therapy
- Aspiration (u/s, CT guidance)
  - Minimally invasive
  - Extremely high recurrence rate
- Angioplasty
  - Cyst rupture, thrombosis, embolization
- Surgical repair
  - Bypass and excision of lesion
  - Cyst evacuation (high recurrence rate)

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**Outline**

- Cystic Adventitial Disease
- Popliteal Artery Entrapment Syndrome
- Vascular Malformations

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**Popliteal Artery Entrapment**

- Rare congenital disorder
- Abnormal relationship of the PA to the muscles in the popliteal fossa
- Usually medial head of the gastrocnemius muscle
- Less commonly fibrous bands or anomalies of the popliteus muscle

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*References*


Popliteal Artery Entrapment

- Estimated prevalence
  - 0.165% 1
  - 15:1 male predilection 3
- Bilateral in 22% to 67% of cases 4
- No genetic predilection


- Normal Anatomy:
  - SFA becomes PA after passing *adductor hiatus*

- Pathophysiology:
  - Abnormal embryologic development
  - Numerous possible anomalous relationships responsible for PAES
  - Proximity of the popliteal artery to the gastrocnemius creates susceptibility to compression

- Classification:
  - Based on anatomic variations there are SIX types of PAES:
    - Type I: Popliteal artery running medial to the medial head of gastrocnemius
    - Type II: Accessory slip of gastrocnemius
    - Type III: Apex of popliteal artery lying below popliteal muscle
    - Type IV: Proximal venous entrapment
    - Type V: Functional entrapment

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Levien, Veler, JVS 30 (4): 587-98, 1999
Popliteal Artery Entrapment

- **Type II**
  - Medial head of gastrocnemius inserts *abnormally in a lateral position*
  - Popliteal artery descends normally but trapped medial to and beneath the muscle

Levien, Veller, JVS 30 (4); 587-98, 1999

Popliteal Artery Entrapment

- **Type III**
  - The popliteal artery is compressed by an *abnormal slip* of gastrocnemius muscle

Popliteal Artery Entrapment

- **Type IV**
  - Popliteal artery is entrapped by a *fibrous band* or by the *popliteus muscle*

Levien, Veller, JVS 30 (4); 587-98, 1999

Popliteal Artery Entrapment

- **Type V**
  - Any of the four anatomic variations that include the *popliteal vein*

Popliteal Artery Entrapment

- **Type VI**
  - “Functional” PAES
  - In patients with *normal* mm attachments, where compression of the popliteal artery caused by *hypertrophic soleus muscle with extensive attachments of the soleus* to the fibula and tibia
  - Functional PAES is usually seen in young, healthy athletes
  - Provocative maneuvers must be done *against resistance*, unlike other PAES

Turnipseed et al, JVS 49(5), 1189-95, 2009
**Popliteal Artery Entrapment**

- **Diagnosis:**
  - Early Stages:
    - nl pulses/signals that disappear or decrease w/active plantar flexion or passive dorsiflexion of the foot (NB need to resist patient w/Type VI)
  - Later Stages:
    - pulses/ABIs may be reduced at rest d/t fixed stenosis or occlusion

  *Levien, Veller, JVS 30 (4); 587-98, 1999*

- **Imaging: Duplex**
  - Duplex in neutral position: normal triphasic waveform in popliteal artery

- **Imaging: Angiography**
  - In neutral position artery normal
  - The "classic" medial deviation of the PA is only seen in Type I, and is therefore not required for the dx

- **Imaging:**
  - Anatomy of muscular structures cannot be detected on US or conventional angiography
  - In addition, there may be a significant number of patients with false positive findings on arteriogram with provocative maneuvers

  *Therefore, MRI must be performed to demonstrate the underlying abnormal anatomic relationships in patients suspected of having PAES*

17 year old female with bilateral PAES
Popliteal Artery Entrapment

- **Imaging:**
  - MRI and MR Angiography demonstrate:
    - The vessel lumen
    - Presence/absence of anomalous anatomy and precise definition of muscular insertions for pre-operative planning

- **Imaging:**
  - abnormal slip of muscle from lateral head of the gastrocnemius muscle passing anterior to the popliteal artery

- **Imaging:**
  - MRA in neutral position shows normal arterial flow

- **Pathophysiology:**
  - Untreated PAES universally progresses to stenosis and occlusion of the popliteal artery due to repeated microtrauma to the vessel
  - Post-stenotic dilatation may also be seen

- **Treatment:**
  - Early stages/no arterial injury: Surgical release of the muscle or tendon is the definitive treatment
  - Late stages/stenosis: Patch angioplasty with SSV
  - Late stages/occlusion: GSV interposition graft
  - Functional PAES: Resection of the soleal attachments to fibula and tibia is performed to “release” the artery

Popliteal Artery Entrapment

• Operative Treatment:
  – Posterior approach (early):
    • Muscle release only
    • Preferable for stenoses or short occlusions (early stage):
      – Allows muscle release
      – Access to SSV for interposition grafting or patch

Popliteal Artery Entrapment

• Operative Treatment:
  – Medial approach (mid/late):
    • Option for moderate lesions requiring both bypass and muscle release
    • Required for long segment occlusion into the tibial (late stage)
      – Expose GSV, inflow and outflow vessels
      – Muscle/tendon release not helpful as vessel is already occluded

PAES vs. CAD

<table>
<thead>
<tr>
<th></th>
<th>Popliteal Entrapment</th>
<th>Cystic Adventitial</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Teens - 20s</td>
<td>30s-40s</td>
</tr>
<tr>
<td>Physical exam/pulse obliteration</td>
<td>Active plantar flexion, dorsiflexion</td>
<td>Knee flexion</td>
</tr>
<tr>
<td>Treat</td>
<td>Always</td>
<td>If symptoms/severe</td>
</tr>
</tbody>
</table>

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• Vascular Malformations

Vascular Malformations

• Definition
  – Embryologically developed, inborn errors of vascular morphogenesis leading to true structural anomalies

• Etiology:
  – Unknown, but genetic predilection
  • Spectrum of disorders ranging from minimal to fatal
  • Overall Incidence = 1.2-1.5%
Vascular Malformations

Archaic Terminology:
- hemangioma
- cavernous hemangioma
- birthmarks (naevi)
- port-wine stains
- cystic hygroma

Vascular Malformations

- **Hemangiomas** are true neoplastic disorders and pathohistologically they demonstrate increased endothelial cell turnover rate
- **Vascular malformations** arise by dysmorphogenesis without increased endothelial proliferation


Hemangioma

**Classification**

- **Vascular Anomalies**
  - Tumors
  - Malformations
    - High Flow (AVM)
    - Low Flow
      - Venous
      - Lymphatic
      - Combined

Hemangioma

- **Hemangioma:**
  - Proliferative phase during the first year of life
  - Spontaneous involution
  - Treatment often not needed
  - Therapy: usually medical (corticosteroids, propranolol)

**Vascular Malformations: Diagnosis**

- Determine extent of lesion
- Key decision point is high flow v low flow
- Treatment
  - High vs low: use of low flow tx in high flow lesions catastrophic
  - Focal vs extensive/invasive
- MRI: mainstay of Dx
  - Involvement of muscle, nerve, bone, etc
  - Can differentiate high vs low in 90% pts

  Litsky, Shortell, personal communication

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**Slow Flow Vascular Malformations**

- Capillary Malformation (CM)
- Venous Malformations
  - Simple venous malformations
  - Blue rubber bleb nevus syndrome
- Lymphatic Malformations
- Combined
  - Klippel-Trenaunay syndrome
  - Proteus syndrome
  - Maffucci syndrome

  As a group, Low Flow Malformations are less aggressive than High Flow Malformations, and grow with the patient (not progressive)


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**Venous Malformations**

- Pain, swelling
- Varicosities or bluish skin discoloration
- Soft and easily compressible lesions
- Drain with elevation
- Bleeding and/or thrombosis
- May be trivial or extensive
- Osteomuscular hypertrophy


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**Low Flow Vascular Malformations**

- Klippel-Trenaunay Syndrome Triad:
  - Capillary malformations ("port wine stain")
  - Venous/lymphatic lesion
  - Osteomuscular hypertrophy


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**Low Flow Malformations: Diagnosis**

- Aplasia or hypoplasia of deep venous trunks
  - Present in 8% of VM patients (with venous predominance)

  Prevalence of deep venous anomalies is even higher in patients with KTS

Low Flow Malformations: Treatment

- **Sclerotherapy:**
  - **Liquid**
    - Ethanol
    - Fluoroscopic guidance
  - **Foam**
    - Polidocanol (Asclera®)
    - Sodium Tetradecyl Sulfate (Sotradecol®)
    - U/S or visual guidance

**Ethanol sclerotherapy adverse effects:**
- EtOH toxicity
- Severe pain
- Ulceration and necrosis at injection site
- Ischemic bullae
- DVT and PE
- Peripheral nerve injury
- Pulmonary hypertension


Low Flow Malformations: Treatment

- **Foam sclerotherapy**
  - Fewer side effects, no need for GA
  - Sodium Tetradecyl Sulfate (USA)
    - Used widely in US and Europe in the treatment of varicose veins and superficial reflux
    - Not previously applied to the treatment of VMs
  - Polidocanol (Europe)
    - has shown benefit in the treatment of VM
    - Now FDA approved in US


High Flow Malformations: Diagnosis

**Duplex:**
Multi directional flow and high-amplitude arterial waveform with spectral broadening
High Flow Malformations: Diagnosis

- **MRI**
  Best to evaluate extent of AVM and relationship to adjacent structures

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Arteriogram

- If treatment planned
- Allows precise evaluation of feeding arteries and draining veins-feasibility of embolization

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High Flow Malformations: Treatment

**Coil embolization**

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**Glue embolization**

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9/12/2011
• The most common congenital vascular disorders are Cystic Adventitial Disease, Popliteal Artery Entrapment Syndrome, and Vascular Malformations

• PAES and CAD may be confused but can be distinguished by History, PEx and imaging

• Vascular Malformations are non-proliferative disorders that require multi-disciplinary care