Mesenteric Arteriopathies: Pathologies and Mimickers

Aditya Sharma
Medical Director, Vascular Medicine
Associate Professor of Medicine
@adsh01

Median arcuate ligament syndrome / Celiac Artery Compression Syndrome / Dunbar syndrome
- Anatomical compression of the celiac axis and/or celiac ganglion by the MAL and diaphragmatic crura
  - More in women (ratio of 4:1)
  - Ages of 30 to 50 years & thin body habitus
- Symptoms: chronic postprandial abdominal pain (>80%), nausea, vomiting (55%), diarrhea, and unintentional weight loss (50%)
- Pathophysiology: Theories
  - Compressed celiac artery causes foregut ischemia resulting in epigastric pain ( collateral flow
  - Pain is neuropathic from a combination of chronic compression & overstimulation of the celiac ganglion leading to direct irritation of sympathetic pain fibers and/or splanchnic vasoconstriction and ischemia.
- Radiographic studies: 10-24% have MAL physiology
  - Symptomatic MALS: 2/100,000

Expiration
Inspiration

Median arcuate ligament syndrome: Diagnosis
- Duplex abdominal ultrasonography during inspiration & deep expiration
  - Compression noted in B-mode or color images
  - Increased velocities in expiration and lowering of elevated velocities in inspiration
  - Reliable indicator: Maximum expiratory peak velocity >350 cm/s & deflection-angles >50°
  - Dynamic maneuvers are performed in CTA/ MRA/ conventional angiogram
Symptom relief:
Complete: 50%
Partial: 73-82%

Open Surgery:
- Decompression alone
- Decompression with celiac dilatation
- Decompression with reconstruction and bypass of the stenosed artery
- Ganglionectomy: pain resolution

Laparoscopic: small incisions & less postop morbidity
- Difficulty in controlling hemorrhage, potential for incomplete release, & higher risk of aortic injury

Angioplasty alone has failed as 1st line Rx

Recurrent symptoms and stenosis:
- 1st line Angioplasty +/- stenting
- Mesenteric bypass

Pancreaticoduodenal Artery Aneurysm Associated with Celiac Trunk Stenosis
- Pancreaticoduodenal arcades and the dorsal pancreatic artery form the main collateral pathways between the celiac axis and the SMA
- Not usually visible unless there is celiac or SMA stenosis

Pathogenesis of PDA aneurysms:
- Increased flow in the small caliber vessels results in local arterial hypertension which then causes focal arterial wall weakening and true aneurysm formation

Treatment:
- Symptomatic i.e. bleeding aneurysm ➔ Coil embolization +/- MALS repair
- Asymptomatic aneurysm ➔ follow up imaging vs. Coil embolization with MALS repair vs. MALS repair

CT angiogram shows isolated pancreaticoduodenal artery aneurysms (white arrowhead) with peri-pancreatic hematoma between the hepatic artery (white arrow) and the superior mesenteric artery (black arrow).

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### Congenital Vascular / Connective Tissue Disorders

**When to suspect?**
- Presence of vascular abnormalities in other locations – dissections, aneurysms, tortuosity
- Family history of aneurysms, sudden death, dissections
- Physical exam: Hyperflexibility, high-arched palate, high brighton score, bifid uvula, easy bruising, fragile skin, scoliosis, pectus abnormalities etc
- Lack of abnormal inflammatory markers

#### Mutations in Gene

<table>
<thead>
<tr>
<th>Conditions</th>
<th>Mutations in Gene</th>
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<tbody>
<tr>
<td>Ehlers-Danlos syndrome</td>
<td>COL3A1</td>
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<tr>
<td>Loeys-Dietz syndrome</td>
<td>TGFBR1, TGFBR2, SMAD3, TGFBR2, TGFBR3</td>
</tr>
<tr>
<td>Familial TAAD</td>
<td>ACTA2, TGFBR2</td>
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<tr>
<td>Marfan syndrome</td>
<td>FBN1</td>
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</tbody>
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### Isolated Spontaneous Visceral Artery Dissection

**When to suspect?**
- Often can be associated with connective tissue disorders such as Ehlers-Danlos syndrome, Loeys-Dietz syndrome, fibromuscular dysplasia
- Often have traditional cardiovascular risk factors: HTN, HLD & Smoking

#### Types of dissection: Sakamoto classification
- Type I: Patent false lumen
- Type II: ‘Cul-de-sac’ blind-ending false lumen without re-entry
- Type III: Thrombosed false lumen with ulcer-like projection
- Type IV: Thrombosed false lumen without ulcer-like projection

### Vasculitides

**When to suspect?**
- Constitutional symptoms: fever, chills, arthralgias, generalized symptoms
- Elevated inflammatory markers
  - ESR/CRP
  - RF, ANA, ANCA etc
- Skin lesions: purpura, livedo reticularis
- Bowel thickening
- Vessel wall thickening: chronic → long segment stenosis
- Microaneurysms / aneurysms in mesenterics and renal
- Purpuric erythema within the GI mucosa

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Vasculitis | Imaging | Other features
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Polyarteritis Nodosa | Microaneurysms w/ stenosis (renals, mesenteric & hepatic) | Mononeuritis multiplex, skin lesions, orchitis, absent ANCA or glomerulonephritis
Localized vasculitis of GI tract | Wall thickening w/ stenosis | Incidentally on surgical pathology
IgA vasculitis | Bowel wall thickening & dilatation, intussusception, endoscopy: purpuric erythema of GI tract mucosa | Skin purpura, arthritis, hematuria, colicky abdominal pain
ANCA-associated vasculitis | Bowel ulceration & ischemia | + ANCA, glomerulonephritis, eosinophilia
Takayasu arteritis | Circumferential vessel wall thickening, stenosis, occlusion | Extremity pulses, bruits, age<40, women, absent upper extremity pulses, bruits,
Lupus vasculitis | Bowel ulcers & bowel wall ulcers | Arthritis, skin lesions, oral ulcers, low complements
RA associated vasculitis | Bowel infarcts / edema / thickening | + RF / CCP, arthritis
Behcet's disease | Ileocecal ulcerations, VTE esp portal vein, pulmonary artery aneurysms | Oral / genital ulcers, skin lesions, pathergy test, PE, pulmonary artery aneurysms, ocular disease
Hypocomplementemic urticarial vasculitis | Bowel edema | Angioedema, purpura, + anti-C1q antibody

Treatment:
Immuno suppressive therapy
Ischemia: Endovascular or surgical therapy

Standing/Stationary Waves

Case: 61-year old female patient with abdominal pain

Mimics
left colic artery biopsy: Fibromuscular dysplasia

- Multiple medial tears and dissections with aneurysmal formation without inflammation or atherosclerosis
- Secondary fibrointimal thickening with focal deficient internal elastic membrane in aneurysmal areas and poorly defined external elastic laminae
- The medial layer is mostly absent in areas of intimal proliferation, with focal preserved areas
- There is adventitial expansion
- An overlying organized thrombus is observed in one section, secondary to the medial tear and dissection observed

Right Renal Artery

Inferior Mesenteric Artery
Superior Mesenteric Artery

Approach to Evaluation for Mesenteric Arteriopathies

CT / MR findings

- No evidence of atherosclerosis or arterial thrombosis or embolism
- Circumferential wall thickening
- Intimal flap, intramural hematoma
- Vasculitis
- Dissection
- Dynamic celiac compression +/- PDA aneurysms
- Median arcuate ligament syndrome
- Beaded appearance
- Fibromuscular dysplasia: Likely FMD in other locations
- Regular beads: Standing waves
- Isolated multiple mesenteric aneurysms +/- GI bleed
- Segmental arterial mediolysis
- Connective tissue diseases: Other findings of CTD and aneurysms
- Trauma or procedural
- Spontaneous
- FMD with mesenteric dissection

Clinical hx: emergent presentation, constitutional symptoms, other symptoms - PT, migraines, hx of RA / SLE etc
Physical exam: hyperflexibility, skin lesions, livedo reticularis, arthritis
Family hx: sudden deaths, aneurysms
Imaging: aneurysms, dissections, MALS, wall thickening
Labs: Inflammatory markers, rheum labs, genetic testing – COL3A1, TGFBR2 etc