Unusual Presentation/Complications Of Undiagnosed Thrombophilia

42nd Veith Symposium
New York City, NY
Friday, November 20, 2015
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Unusual Presentation of Antiphospholipid Antibody Syndrome

• 66 year old presents to ER
• 3 day history of pain, nausea and vomiting
• PMH: HTN, bipolar disorder, COPD and TIA
• Surgical: Iliac stent, appendectomy
• Smoking: currently 0.5 packs per day, 70 packs/year history
• P2G2
• No personal or family history of DVT

Current Medications

• Restoril
• Lipitor
• Seroquel
• Colace
• Amlodipine
• Sinement
• Wellbutin XL
• Zyrtec
• Cymbalta
• Aricept
• ASA 81 mg
• Zantac
• No antibiotics in the last 6 months

Warning: Not for diagnostic use

Presentation

PE:
- hypotensive,
- tachycardia,
- abdomen
- distended, cool,
- clammy skin,
- mottling
- bilateral LE

Nothing to Disclose

I have no relevant financial relationship(s) with any proprietary entity producing health care goods or services related to the content of my talk.

Good Samaritan Hospital, Cincinnati, OH
Procedures
• Exploratory laparotomy – Arterial vessels patent, no venous thrombi
• Subtotal abdominal colectomy
• Brooke ileostomy
• Pop #1 – C diff negative
  – Dusky ostomy - heparinized
• Pathology
  – Complete transmural devitalization
  – Dilatation features acute transmural ischemic colitis
  – Toxic megacolon
  – Rare mesenteric intravascular thrombosis
  – Medium sized vessels
  – Proximal margin free inflammation
  – Distal margin involved acute ischemia and transmural inflammation

Hypercoaguable Profile
• + Lupus anticoagulant (nl <49.5)56.1
• Anticoagulated x 12 months
• Endoscopy of ileostomy and rectum negative biopsies
• Attempted takedown – complete stricture anus 6 cm
• Residual sigmoid resected – pathology showed acute patchy inflammation and acute mucosal ulceration

Antiphospholipid Antibody Syndrome (aPL)
• Arterial and/or venous thrombosis
• Pregnancy morbidity associated
  – Anticardiolipin antibodies
  – Anti beta 2 glycoprotein I antibodies
  – Positive lupus anticoagulant test
• Potentially affects any organ system including the skin
• Falsely elevates PT/INR testing

Cutaneous Lesions
• Livedo reticularis (most frequent), livedo racemosa
• Ulcerations
• Digital gangrene
• Subungual splinter hemorrhages
• Superficial venous thrombosis
• Thrombocytopenic purpura
• Pseudovasculitic manifestations
• Extensive cutaneous necrosis
• Primary anetoderma
• Skin lesions more frequently observed (70%) in catastrophic antiphospholipid syndrome characterized by widespread microvascular occlusions involving multiple organs simultaneously
**Organ Involvement aPL**
- Renal 70%
- Pulmonary 66%
- Brain 60%
- Heart 52%
- Skin 47%
- Those with cardiac and pulmonary complications most likely to be associated poorer prognosis and death
- Neurologic complication frequent

**The Catastrophic Antiphospholipid (Asherson’s) Syndrome**
- Rapidly progressive
- Often fatal
- Rapid onset resulting multiple organ system dysfunction
- Small vessel occlusive disease
- Pathologic evidence of thrombotic microangiopathy
- Fulminant tissue necrosis particularly involving GI tract
- SIRS
- Unusual organ involvement—reproductive organ infarctions, bone marrow necrosis, acalculous, cholecytitis, polyneuropathy, splenic, hepatic and adrenal infarctions
- Serologic evidence DIC present in a significant proportion of patients
- ICU multiple physicians often miss diagnosis

**Treatment for Patients with Antiphospholipid Thrombosis**
- Long-term oral anticoagulants (intensity guided by nature of thrombosis) and ASA
- aPL associated pregnancy morbidity should be treated with aspirin plus heparin with close monitoring
- INR target 3.0 – 4.0 for those with arterial thrombi
- Recurrent thrombosis common

**Treatment Catastrophic aPL**
- Unsatisfactory
- High dose intravenous steroids
- Parental anticoagulation supplemented with intravenous gamma globulins and repeated plasma exchanges using FFP early in course of syndrome
- Recognition early/prevention
- Smoking cessation
- Cessation estrogen containing products
- Avoid “triggering” factors (60%)