Cases on the Coast

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“Anybody Want FAST Food ?”
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• 25-year-old male presented to our outpatient clinic
  • With a three-week history of pain in his right index finger with bluish discoloration.
  • He currently states his finger is tender to touch.
  • Denies any fever or chills.
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- Family history: Mother has Raynaud's Phenomenon
- Social history: one pack a day smoker, 15 year history
- No past Medical or Surgical history
- Works at McDonald’s
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- Review of systems: no shortness of breath, chest pain, weight loss, or fever/chills noted
- No history prior to 3 weeks ago
- History of trauma to the right index finger 3 weeks prior when he dropped a box of frozen hamburgers on his right hand
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• Physical Exam
  • Vital signs normal
  • No skin changes or adenopathy in the lower extremities
  • Right index finger discolored to the mid finger with a tender punctate ulcer at the tip of right index finger
  • Slow capillary refill in the right hand, right hand was cool to touch
• Partially positive Allen Test, with reduced blood return in radial artery distribution
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• Chem 19, complete blood count PT/INR and urinalysis all within normal limits
• ESR 18, CRP 0.93 blood cultures negative ×2
• Any further studies required?
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- ANA, RF, SCL-70, and complements were normal
- Hypercoagulable panel was within normal limits
- Urine toxicology screen negative
- 2-D echocardiogram within normal limits
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- Differential diagnosis?
- Crush injury with secondary superinfection
- Vasculitis
  - Systemic lupus erythematosus
  - Mixed Connective Tissue Disorder
  - Scleroderma
  - CREST
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- Peripheral vascular disease
- Thrombophilia/hypercoagulable state
- Buerger’s disease (thromboangitis obliterans)
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• Arterial duplex ultrasound of the upper extremities
  • Revealed normal blood flow of right radial and right ulnar artery to the level of the wrist.
  • Decreased distal flow noted to the second and third distal phalanx
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Normal Brachial Artery
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Distal Involvement with “Corkscrew” appearance
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“Ratty” appearing Ulnar Artery
“Anybody Want FAST Food?”

“Ratty” appearing Ulnar Artery

Note: Lack of Radial Artery distribution
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Lack of Radial Artery distribution
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• This is a diagnosis of exclusion, biopsy not performed

• Criteria:
  • Age less than 45 years old
  • Current tobacco abuse
  • Distal extremity ischemia
  • Typical arteriogram findings of TA / Buerger’s
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• Pathogenesis
  • Segmental inflammatory, non-atherosclerotic occlusive vascular disease of small and medium sized arteries and veins
  • Intraluminal thrombus with relative sparing of vessel wall and internal elastic lamina
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• Exclusion:
  • Autoimmune
  • Diabetes
  • Hypercoagulable state

• Biopsy is rarely needed
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• Treatment

• Immediate smoking cessation

• Iloprost: Ventavis, Remodulin

• Lancet article (1990), placed iloprost versus aspirin treatment, would benefit with iloprost, as it pertains to pain relief, healing, and amputation prevention
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• Other treatments include calcium channel blockers, phosphodiesterase inhibitors, sympathectomy, stem cell transfusion, and HBO T

• Patient was treated in traditional manner, smoking cessation, wound care and hyperbaric, with 80 percent resolution of symptoms in digital ischemia within 5 weeks
“Not Your Usual College 20”
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- 19-year-old college freshman presented to emergency department with 6 week history of dysphagia with solid food during his first semester in college, he lost 20 pounds. Started to eat slower and switch to more liquid diet which has helped. No heartburn noted

- Social: nonsmoker no alcohol

- Unremarkable past medical or surgical history
“Not Your Usual College 20”

• Physical exam was unremarkable
• Chem-7, complete blood count and differential within normal limits
• Modified barium swallow revealed normal upper esophagus and normal swallow without aspiration
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• Differential diagnosis

• Dysphagia
  Oropharyngeal dysphagia—difficulty initiating the swallowing mechanism
  VS.
  Esophageal dysphagia—difficulty swallowing several seconds after starting the swallowing process
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• Differential diagnosis of esophageal dysphasia
• Intraluminal-mostly consistent with food impaction
• Intrinsic
• Stricture
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• Peptic stricture, acid or caustic exposure, systemic sclerosis, Zollinger - Ellison syndrome, history of NG tube placement

• Esophageal webs and rings often associated with iron deficiency anemia (Plummer-Vinson syndrome)

• Carcinoma
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- Infectious esophagitis
  - HSV, CMV, fungal - more common in immunocompromised patients
- Previous radiation exposure
- Eosinophilic esophagitis
“Not Your Usual College 20”

• Extrinsic causes-cardiovascular abnormalities with aberrant vessels forming rings around the esophagus

• Motility disorders including achalasia or spastic motility “nutcracker”
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• This patient had no radiation, ingestion, history of symptoms consistent with carcinoma, previous surgery, or childhood problems with swallowing.

• He went to upper endoscopy on hospital day 2.
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Stacked Circular Rings
“Feline Rings”

Whitish Papules: representing eosinophil microabscesses
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• At the time of EGD and review of pathology patient diagnosed with eosinophilic esophagitis

• Eosinophilic esophagitis

• At time of endoscopy, often see active circular rings or strictures, whitish papules, and a small caliber esophagus
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• Biopsy reveals greater than 15 eosinophils per high-powered field

• AND see peripheral eosinophilia (5%)
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• Plans-treat with 2 months of proton pump inhibitor, with referral to allergy / immunology

• Plan to repeat endoscopic survey, and see if patient is a proton pump inhibitor responsive versus nonresponsive patient

• Patient was seen back by gastrointestinal 2 months after PPI therapy, and endoscopy with biopsy was still consistent with eosinophilic esophagitis with no real change with PPI therapy
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• Patient started on topical Flonase, 220 µg twice a day, and was able to have a esophageal dilatation at the time of the second endoscopy

• Allergy immunology workup was negative
“Not Your Usual College 20”

- Eosinophilic Esophagitis
  - Other treatment options include oral systemic steroids if severe disease is refractory to other treatments.
  - May be a role for prostaglandin inhibitors
  - Antihistamines and cromolyn sulfate show little if any benefit
  - Several small uncontrolled trials are ongoing with numerous monoclonal antibodies with mixed results.
“Chest Pain in Room 14”
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• 54-year-old white female awoken from sleep this morning at 0300 with 10/10 chest pain with radiation to left arm and neck.
  • It was associated with shortness of breath, without pleurisy, nausea, vomiting or diaphoresis.
  • No history of coronary disease angina or anginal equivalents.
• Past medical history of hypothyroidism and hypertension
“Chest Pain in Room 14”

• Medications include Prinivil, Synthroid, Prozac, and Estrace

• Family history father with coronary artery disease at 48, died of an myocardial infarction at 50

• Social history nonsmoker social alcohol
“Chest Pain in Room 14”

- Review of systems 10 pound weight loss (unintentional)
- Left arm pain noted with exertion, myalgias, and overall fatigue for the last 2-3 years.
“Chest Pain in Room 14”

• Physical Exam
  • Moderate distress, clinically ill-appearing
  • Blood pressure 123/70 on the right 110/60 in the left
    afebrile, heart rate 60 in sinus rhythm
  • Clear to auscultation
  • Regular rate and rhythm, reproducible chest wall pain to
    palpation, left carotid bruit
“Chest Pain in Room 14”

• Physical Exam
  • Nontender, nondistended, no HSM no rebound tenderness
  • Left upper extremity pulses 1+ distally, pulses 2+ on the right both upper and lower extremities
  • No skin lesions or adenopathy noted
  • Neurologic exam nonfocal
“Chest Pain in Room 14”

• Further evaluation chest x-ray and electrocardiogram within normal limits
• Pertinent positives include albumin of 3.0, ESR 81, CRP of 2.4, d-dimer 2.19
• Troponin negative
• CT A rule out pulmonary embolism ordered
“Chest Pain in Room 14”

Thick walled ascending aorta.

Normal caliber descending aorta.
“Chest Pain in Room 14”

Involvement of Innominate Artery

Involvement of left Common Carotid
“Chest Pain in Room 14”

Involvement of ascending aorta to the level of the aortic valve
Involvement left Common Carotid
Involvement of Ascending Aorta, Left Common Carotid, and Left Subclavian
“Chest Pain in Room 14”

• Possible differential diagnosis based on CT findings, presentation, and history?

• Aortitis
  • Fibromuscular dysplasia - usually more focal with no systemic symptoms
  • Ergotamine excess not consistent with this history
“Chest Pain in Room 14”

• **Aortitis**
  • Ehlers-Danlos associated usually with multiple aneurysms, also not a systemic process
  • Large vessel arteritis - Takayasu Arteritis versus giant cell
  • Infectious aortitis is rare and usually associated with thoracic aorta and previous aortic implementation or instrumentation
“Chest Pain in Room 14”

• Further testing revealed
  • Blood and urine cultures were negative
  • ANCA, ANA, complements, rheumatoid factor were all negative
  • MRA of the chest is consistent with CT findings with no evidence of dissection, false lumen, or aneurysmal formation
“Chest Pain in Room 14”

- This was not a straightforward diagnosis, but historically, and imaging-wise, most consistent with Takayasu Arteritis
  - Predominantly female
  - Age 20-40 years of age
  - Primarily affects aorta in the primary branches
  - Consider giant cell arteritis, but not classically consistent with the age, or arterial circulation involvement.
“Chest Pain in Room 14”

• Treatment Plan
  • Steroids: 45-60 mg of prednisone, and to assess response to therapy, tapering based on this response
  • May be role for follow-up imaging
  • Steroid resistant disease - possible role for PO methotrexate, azathioprine, and other anti-tumor necrosis factor drugs may have a role, but studies are small and mainly uncontrolled
“Chest Pain in Room 14”

Our patient responded well to high-dose steroids, repeat CT showed decreasing aortic wall changes, and was referred to UAB for further rheumatologic and CV surgery evaluation.