AMSER Case of the Month:
September 2018

24 y/o female presenting with bilateral lower chest pain, radiating to the back

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### Patient Presentation

#### History
- 3 weeks of bilateral lower chest pain
- Pain radiating to back; mild dyspnea
- PMH: Sickle Cell Disease, Anemia, GERD, Depression
- SxH: Marijuana 1x/month, No EtOH or tobacco; unemployed
- FamHx: Mother: Sickle Cell Trait; Father: Diabetes, Sickle Cell Trait
- Meds: Ibuprofen, Morphine, Oxycodone, Nortriptyline

#### Pertinent Labs
- WBC: 9,200/μL
- RBC: 3.29
- Hgb/Hct: 7.9 g/dL/ 23%
- MCV: 71.7
- Target Cells: >5/HPF
- Howell Jolly Bodies: Present
- Sickle Cells: Present
- D-Dimer: 1.95 mg/L FEU
What Imaging Should We Order?
# ACR Appropriateness Criteria

## American College of Radiology
### ACR Appropriateness Criteria®

#### Clinical Condition:
**Acute Nonspecific Chest Pain—Low Probability of Coronary Artery Disease**

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL.*</th>
</tr>
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<tbody>
<tr>
<td>X-ray chest</td>
<td>9</td>
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<td>CTA coronary arteries with IV contrast</td>
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#### Variant 2:
**Suspected pulmonary embolism. Intermediate probability with a positive D-dimer or high pretest probability.**

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These imaging modalities were ordered by the ER physician.
Findings (unlabeled)
Findings (labeled)

A = mild cardiomegaly
B = H-shaped vertebrae
Findings (labeled)

A = H-shaped vertebrae
Final Dx:

Acute on Chronic Sickle Cell Pain with mild cardiomegaly and chronic bone infarcts (H-shaped vertebrae)
Sickle Cell Disease

• Autosomal Recessive
  • Substitution mutation in the Beta-globin gene: Glutamic Acid $\rightarrow$ Valine

• Sickle Cell Anemia vs. Disease vs. Trait
  • Anemia = homozygous gene mutations – two HbS beta-chains
  • Disease = two abnormal beta-chains (HbS + either HbC or thalassemia)
  • Trait = one HbS beta-chain and one normal HbS beta-chain

• Abnormal RBCs sickle in deoxygenated states $\rightarrow$ microvascular occlusions & premature hemolysis.

• Clinical Complications
  • Bone Infarcts
  • Vaso-occlusive Crises
  • Osteomyelitis
  • Osteonecrosis
  • Osteoporosis
H-Shaped Vertebrae

• Pathognomonic radiographic finding for Sickle Cell Anemia
  • Seen in approximately 10% of patient with Sickle Cell Anemia

• Description: Lincoln log deformity, central square-shaped endplate depression

• Cause: Avascular necrosis of vertebral body endplate
  • Microvascular occlusion of endplate → deficient endochondral ossification → overgrowth of surrounding area
  • Endplates have low-flow terminal vasculature and are very thin, particularly in the center (prone to occlusion with sickled cells)

• Sequela: Adjacent vertebrae lengthen to compensate and support spine.
References:


3. Lonergan G; Cline D; Abbondanzo S. Sickle Cell Anemia. Radiographics 2001; 21 (4).
