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**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9	X-ray, CTA, and US are generally nonoverlapping and can be used sequentially.	☢
CTA coronary arteries with IV contrast	7	X-ray, CTA, and US are generally nonoverlapping and can be used sequentially.	☢☢☢
CTA chest with IV contrast	7	X-ray, CTA, and US are generally nonoverlapping and can be used sequentially.	☢☢☢

Variant 2: **Suspected pulmonary embolism. Intermediate probability with a positive D-dimer or high pretest probability.**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CTA chest with IV contrast	9	This procedure should be optimized for pulmonary circulation.	☢☢☢
CT chest with IV contrast	9	This procedure should be optimized for pulmonary circulation. This procedure may be an alternative to CTA, but both should not be performed.	☢☢☢
Tc-99m V/Q scan lung	7	This procedure may be an alternative to CTA, but both should not be performed.	☢☢☢

These imaging modalities were ordered by the ER physician

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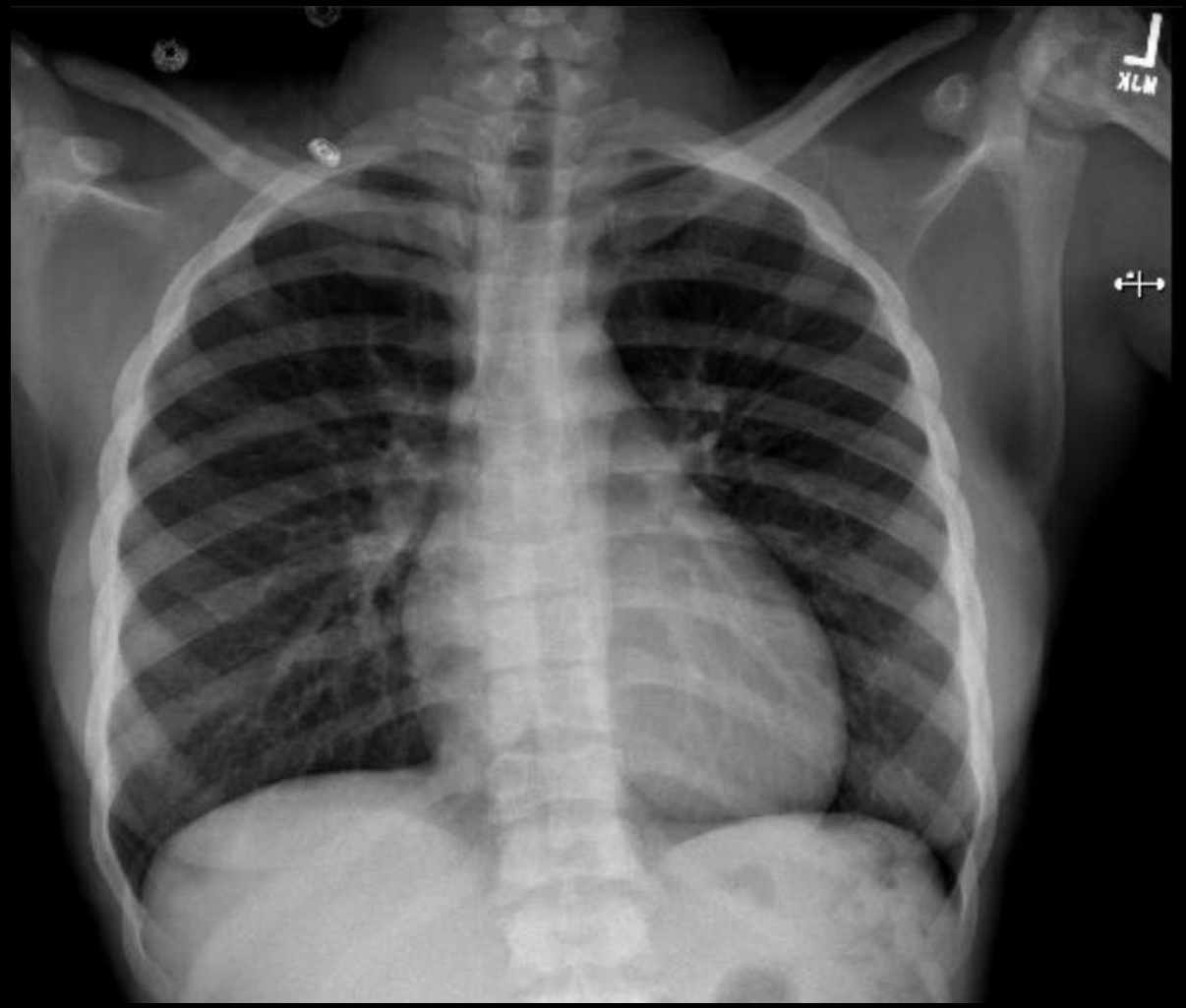
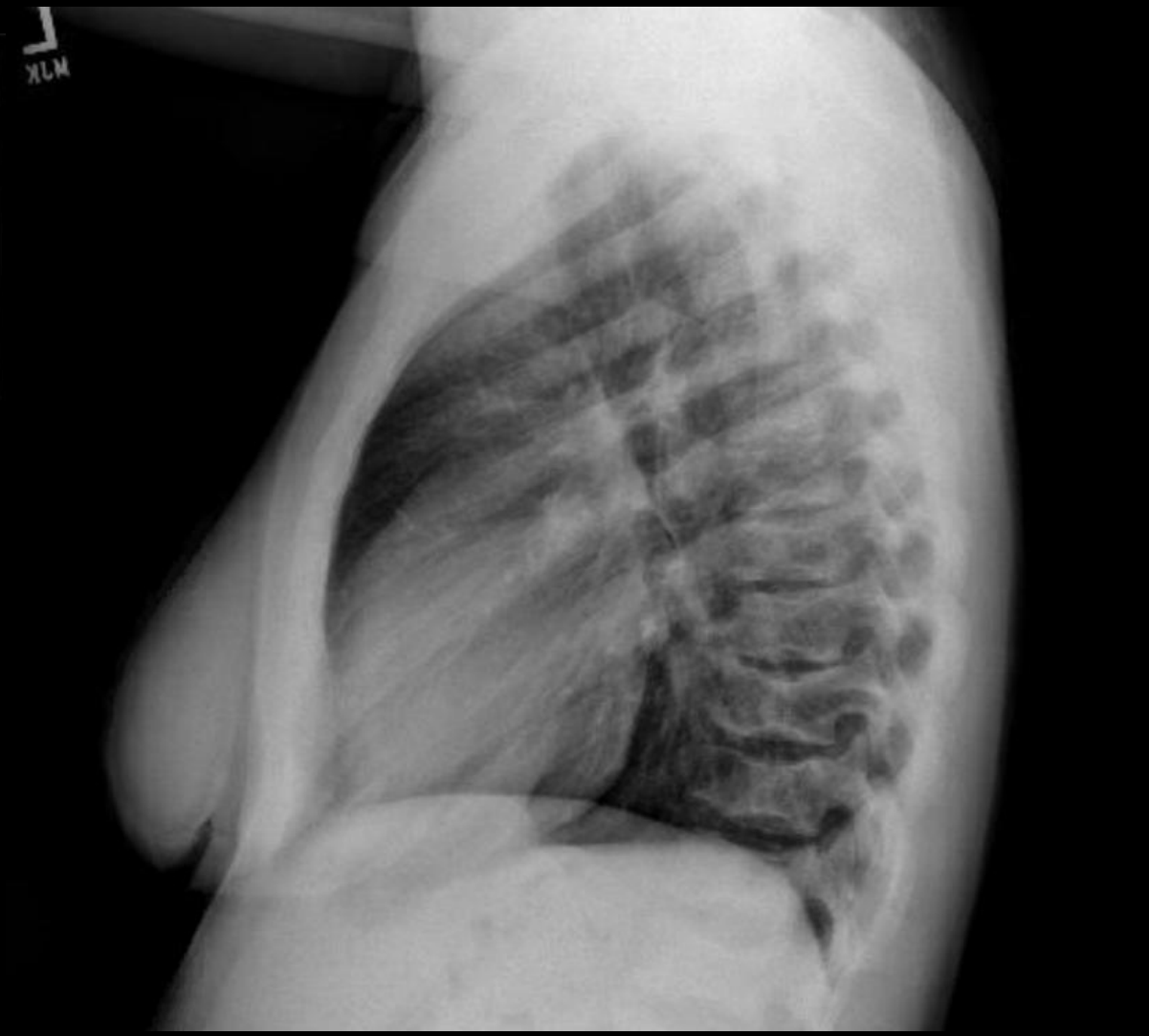
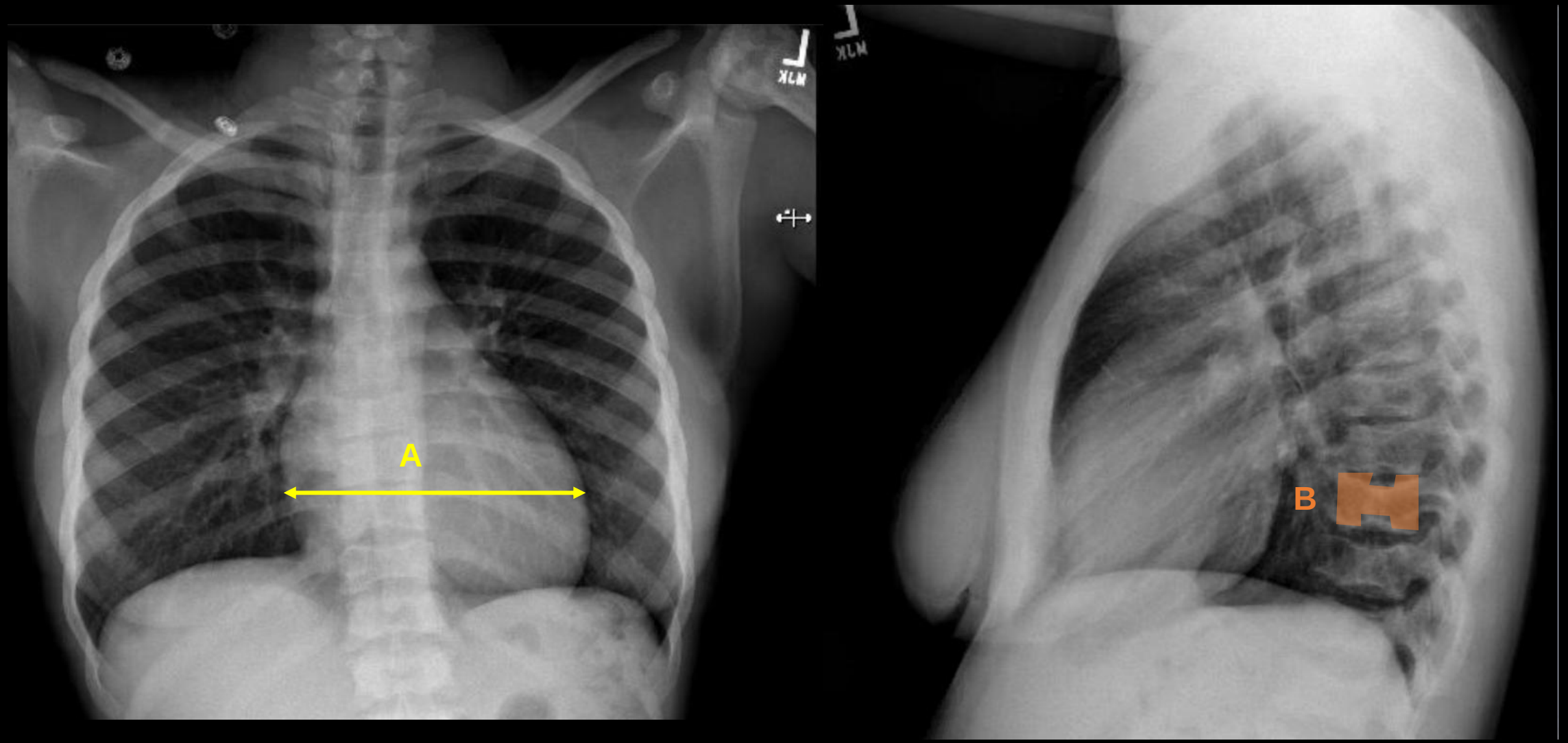
Findings (unlabeled)



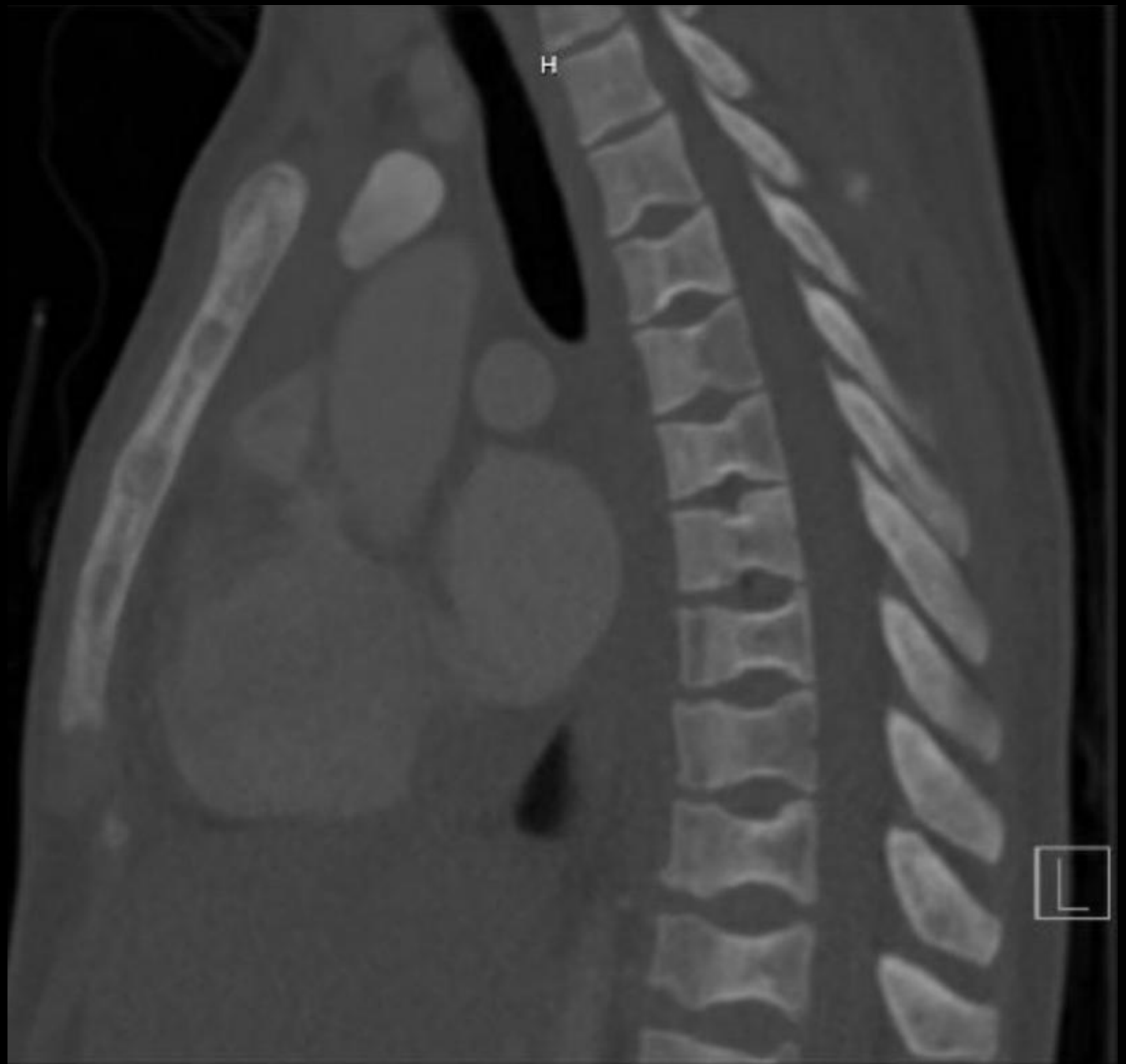


Findings (labeled)



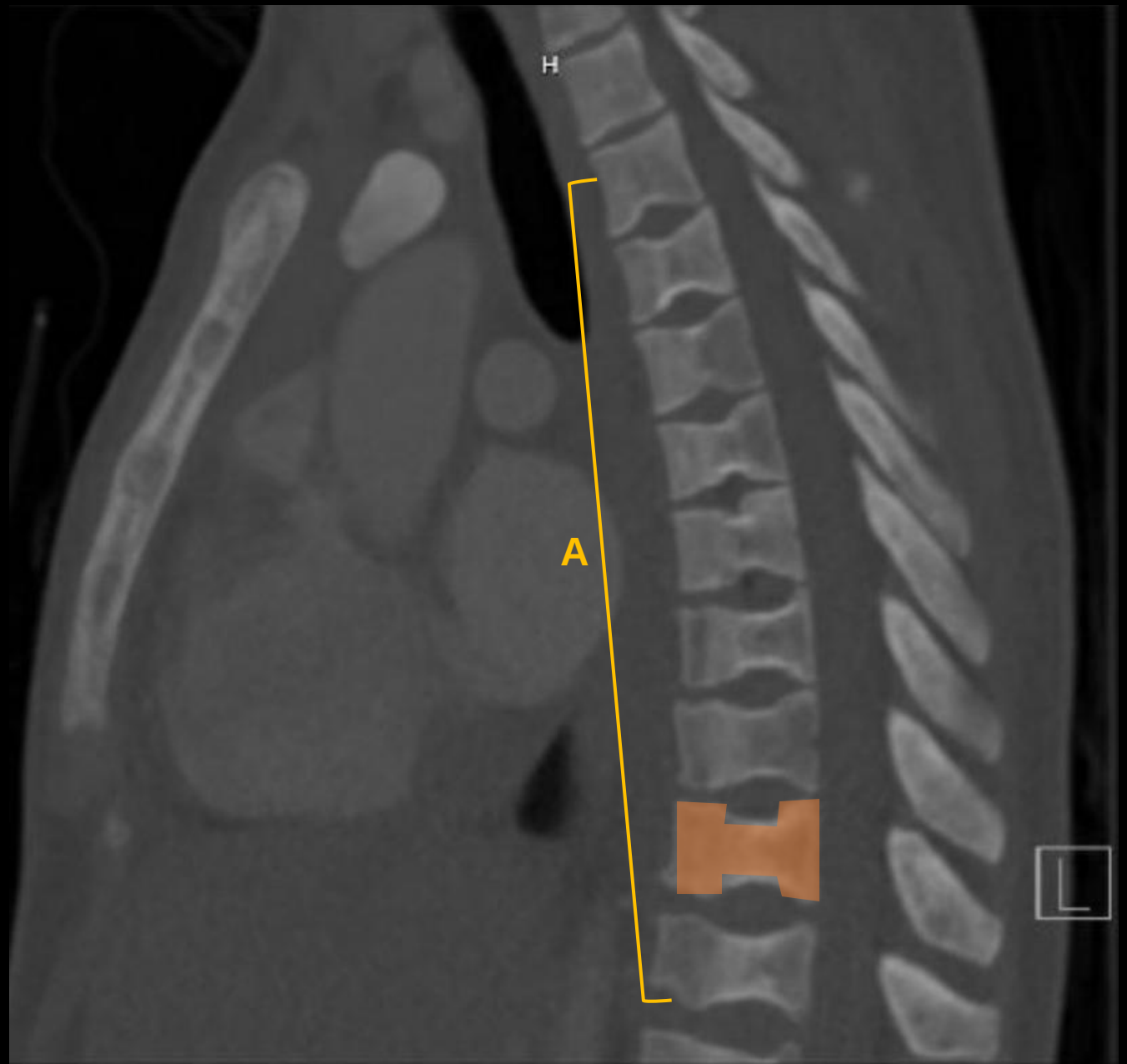
A= mild cardiomegaly
B = H-shaped vertebrae

Findings (unlabeled)



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 → 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 → 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:

1. Ganguly A; Boswell W; Aniq H. Muskuloskeletal Manifestations of Sickle Cell Anaemia: A Pictoral Review. Anemia 2010; 2011: 794283.
2. Kosaraju V; et. al. Imaging of musculoskeletal manifestation in sickle cell disease patients. Br J Radiol 2017; 90: 20160130.
3. Lonergan G; Cline D; Abbondanzo S. Sickle Cell Anemia. Radiographics 2001; 21 (4).
4. Kartikyeyan R; et. al. Characteristic Vertebral Imaging in Sickle Cell Disease. J Neurosci Rural Pract 2017; 8(2): 270-271.
5. Hansen G. Central Depression of Multiple Vertebral End-plates: A “Pathognomonic” sign of Sickle Hemoglobinopathy in Gaucher’s Disease. Am J Roentgenol 1977; 129: 343-344.