Acute Musculoskeletal Pain (Sickle Cell Crisis)

Greg Gaskey

November 13, 2019

Radiology 4001

Dr. Manickam Kumaravel

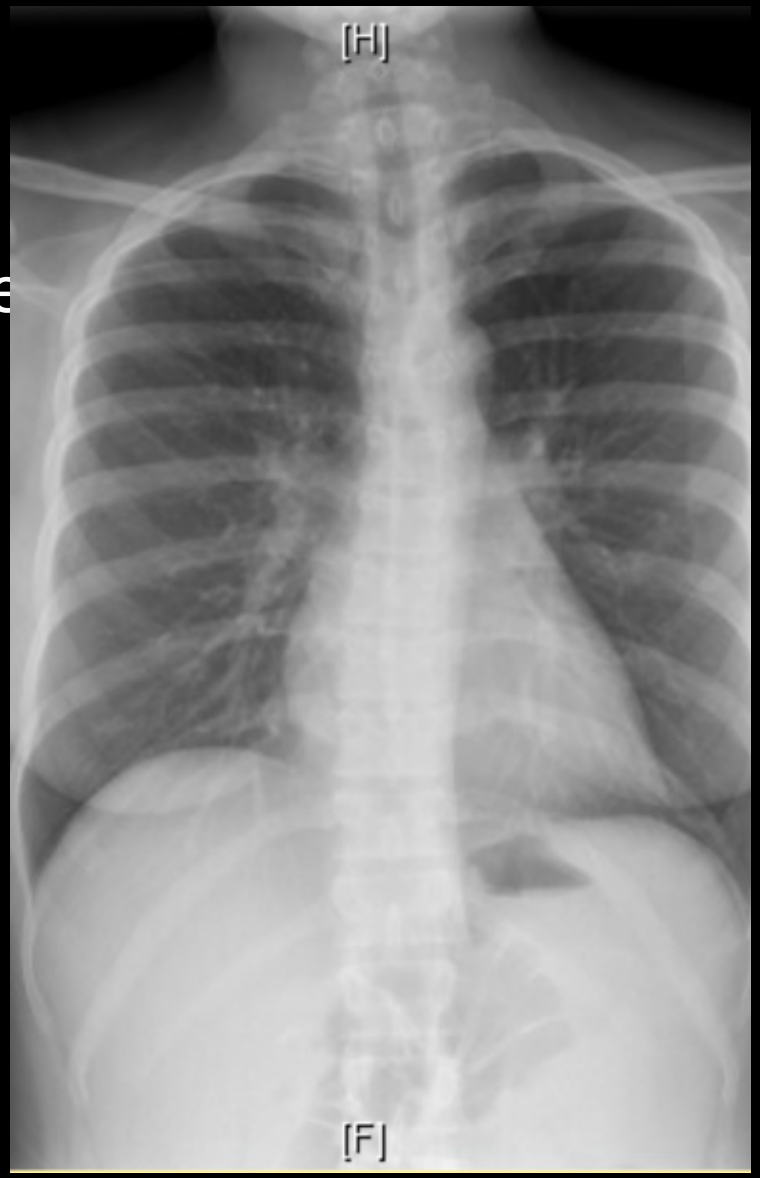
UTHealth®
The University of Texas Health Science Center at Houston
McGovern Medical School

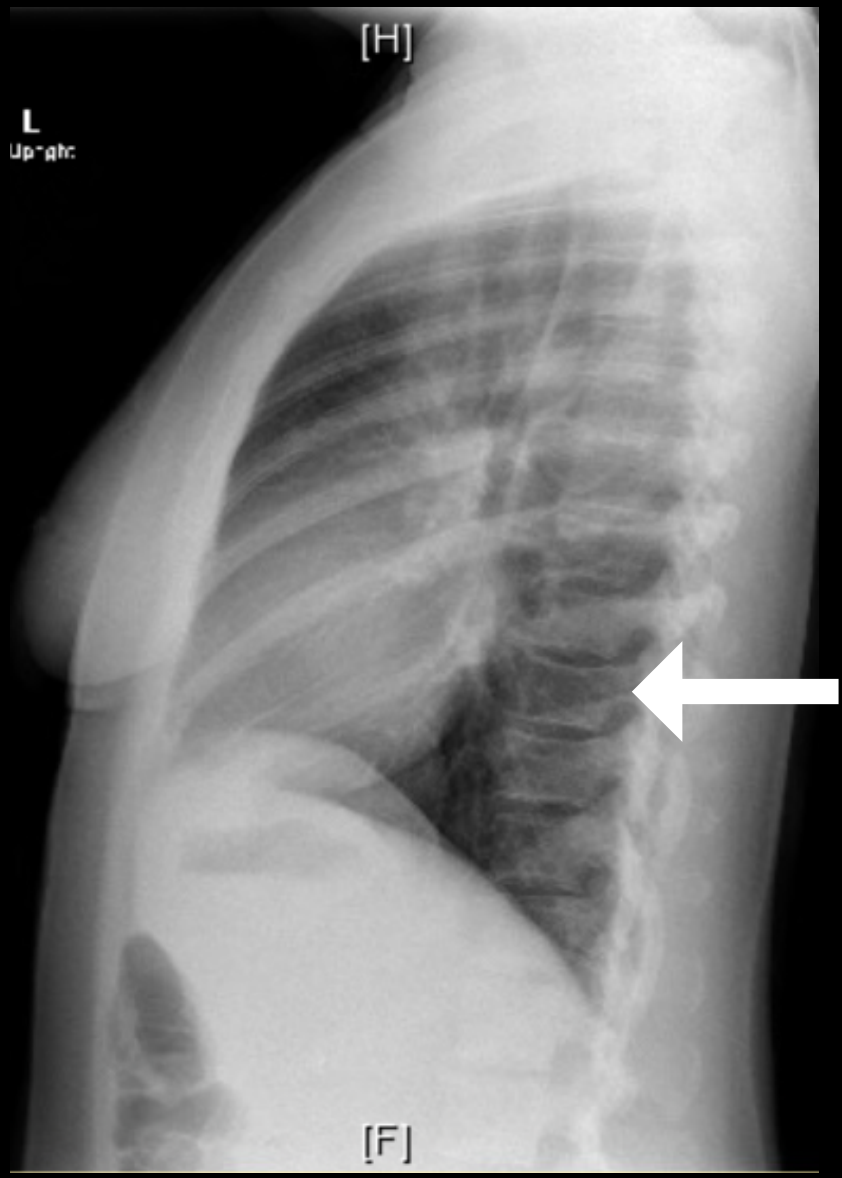
Clinical History

- A 35 year old F with PMH of sickle cell, scoliosis, DVT, and bigeminy presenting for worsening lower back and lower extremity pain
- HPI
  - Recent suspected viral URI with cough, N/V, diarrhea, and subjective fever
  - New onset worsening sharp stabbing pain in lower back, posterior knees, and anterior tibia
- Physical Exam
  - Vital signs: 98.0 F, HR 122, RR 18, BP 120/89, O2 97%
  - HR lowered to 89 after pain medication given in ED and stable during admission
  - MSK: tender to palpation in lower back, in popliteal fossa and superior anterior tibia bilaterally
- Workup
  - Known Hgb SC sickle cell disease followed by hematology with established home pain plan
  - CBC originally showed Hgb 11.8, MCV 78,RDW 14.8 , platelets at 98, retik 1.5, and LDH 261
  - Repeat CBC shows HGB 9.5 and retik 1.7
  - CMP wnl

Chest X-ray

- No acute abnormalities
- Vertebral body endplate depressions



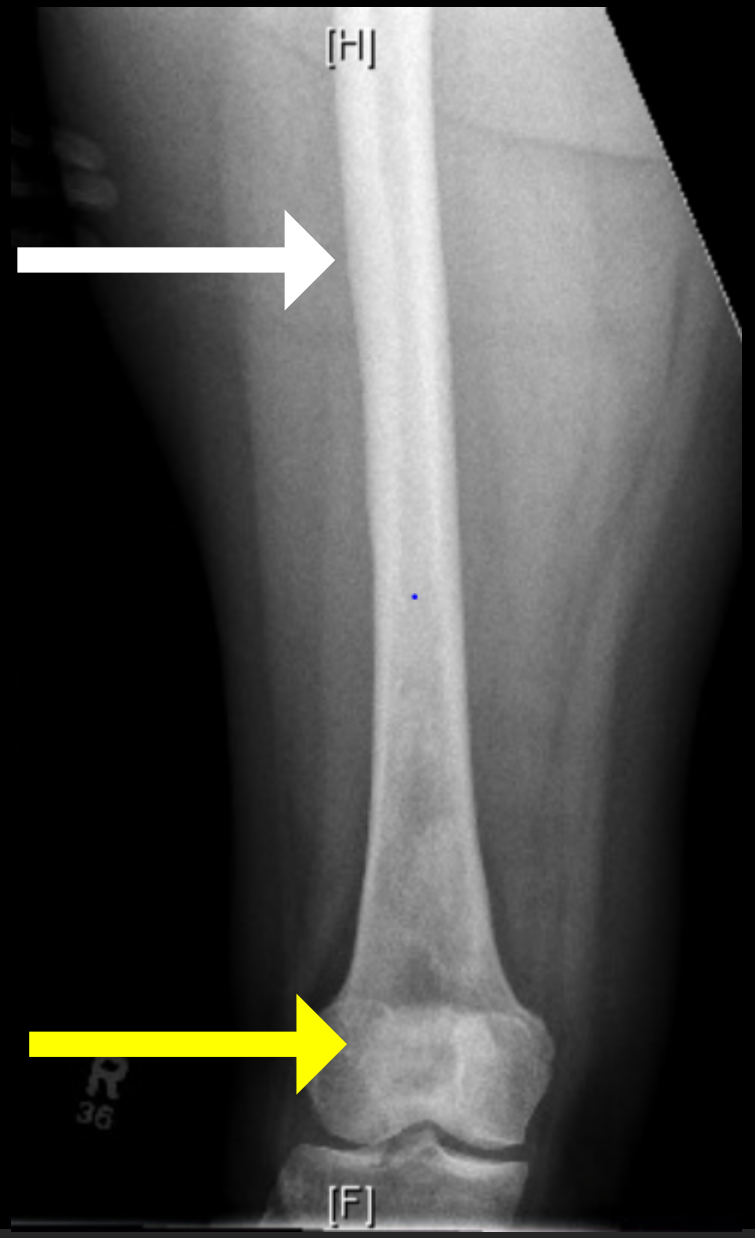


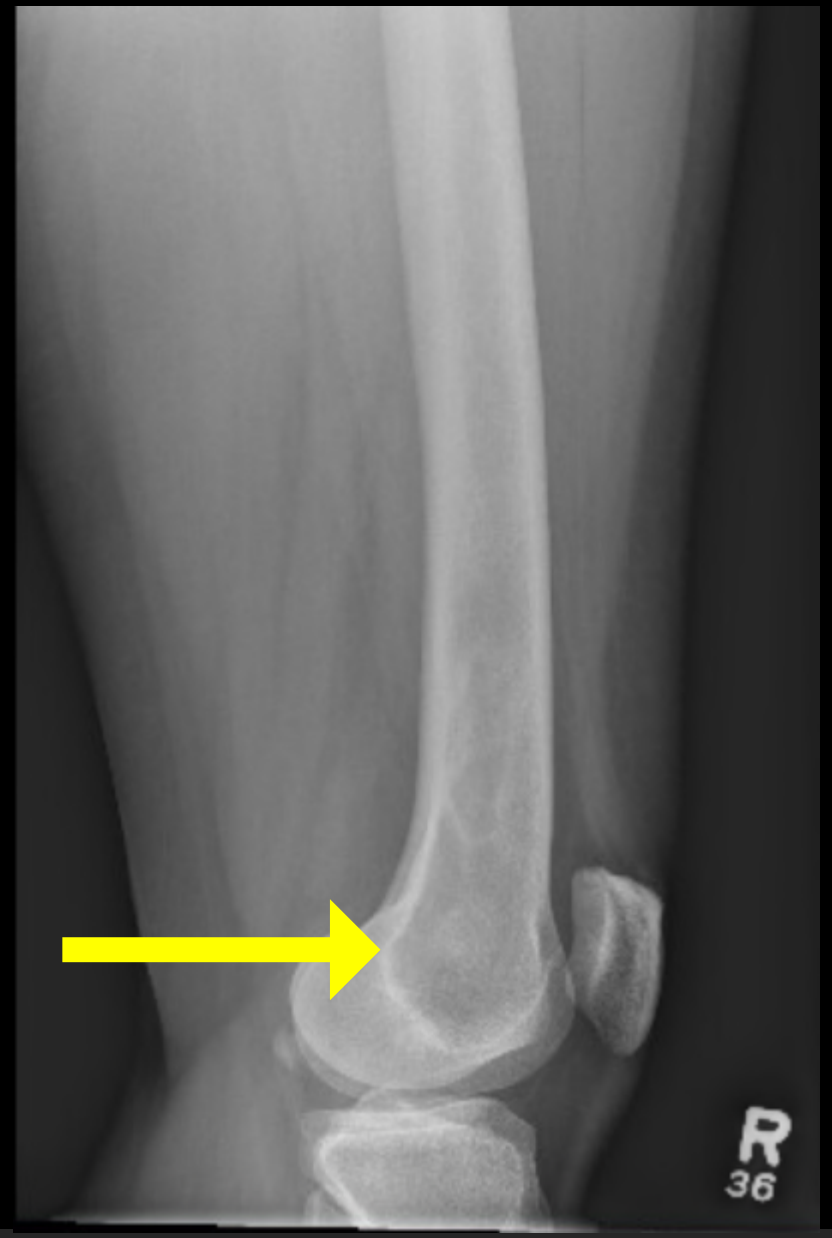
Bilateral Hip

- No acute Abnormalities

R Femur X-ray

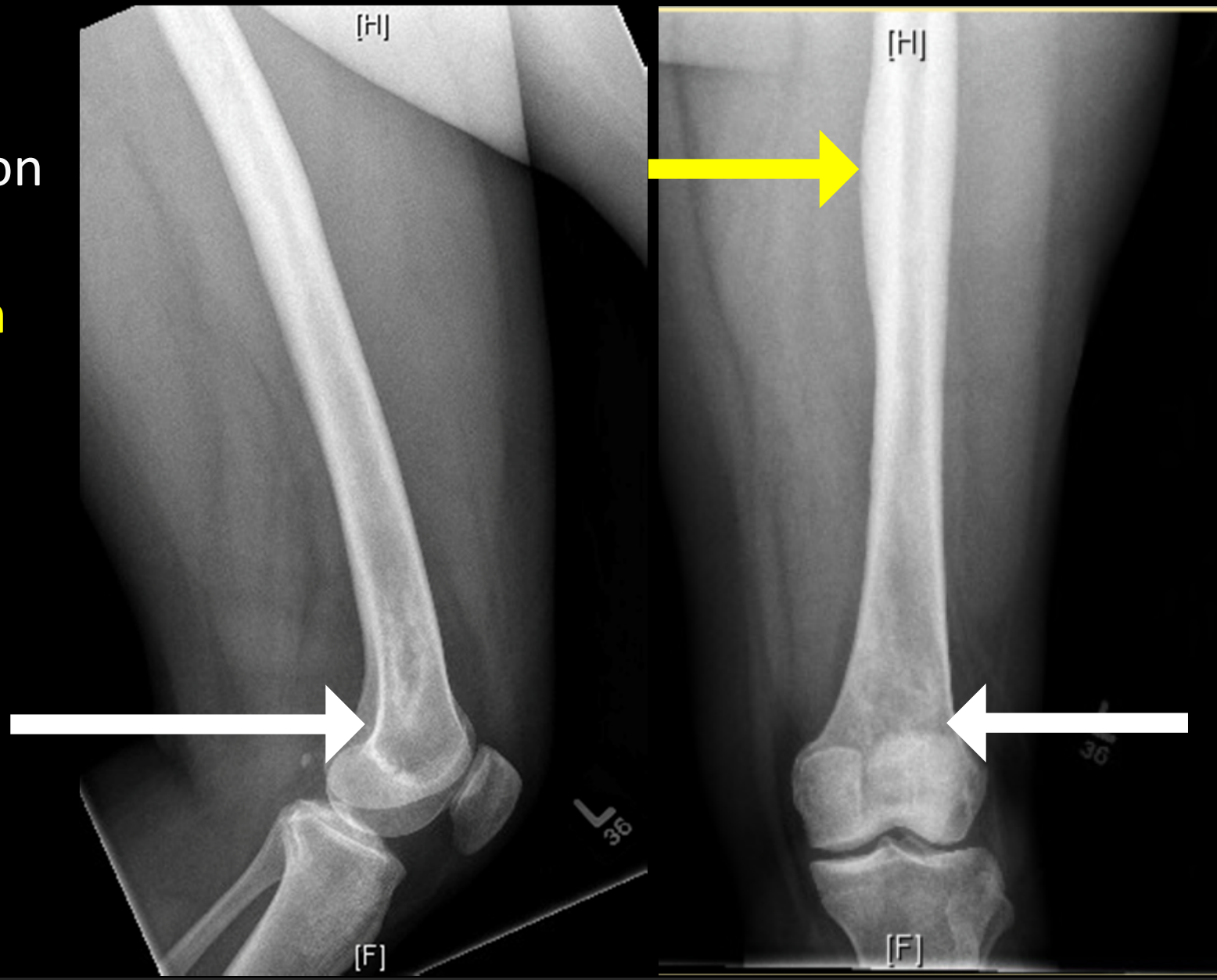
- Proximal Cortical thickening
- Distal sclerotic focus with internal lucency





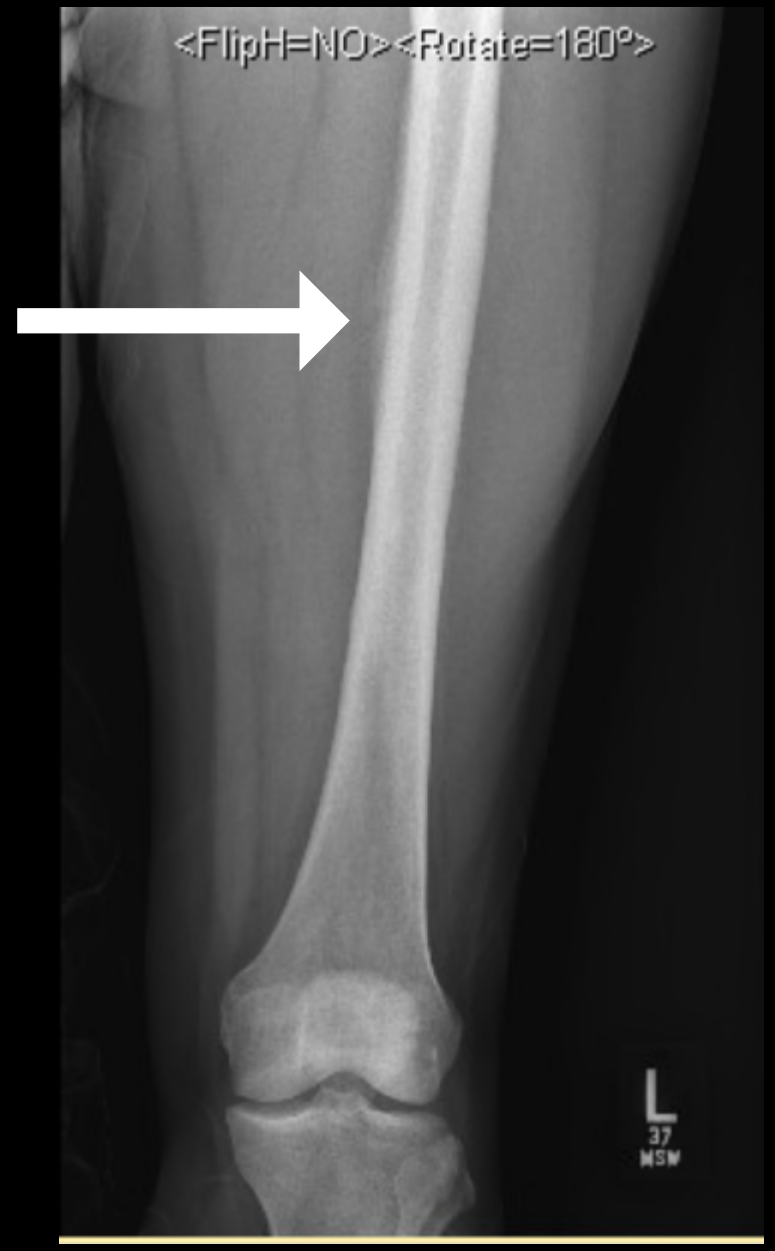
L Femur Xray

- Chronic Periosteal reaction and calcification
- Distal sclerotic focus with internal lucency



Prior L Femur Xray

- Small periosteal reaction



Key imaging findings

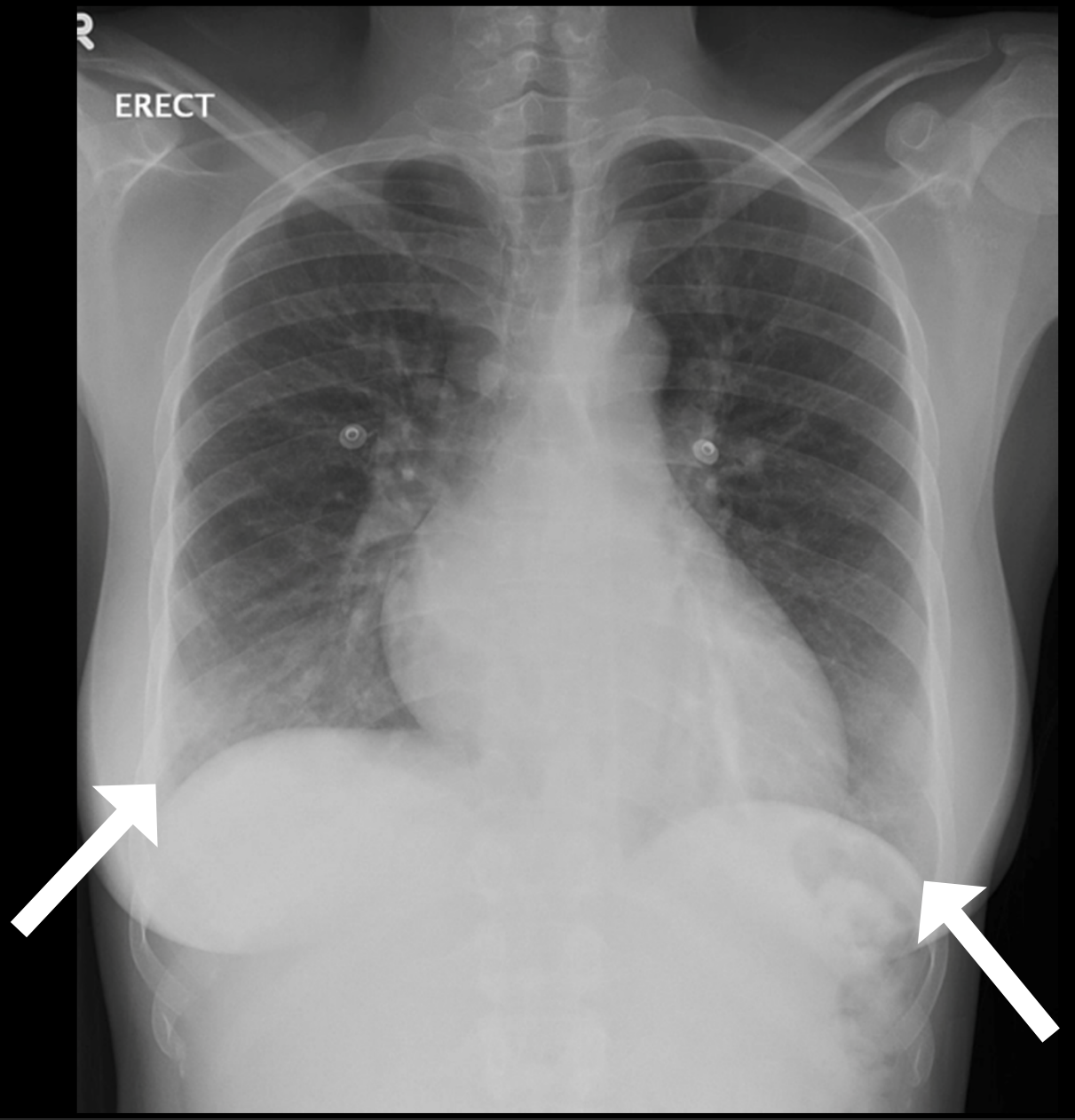
- Vertebral endplate depression
- Normal hip Xrays
- Sclerotic focus in distal femurs bilaterally
- Cortical thickening in R proximal femur
- Periosteal reaction and calcification in L femur

Differential Diagnosis

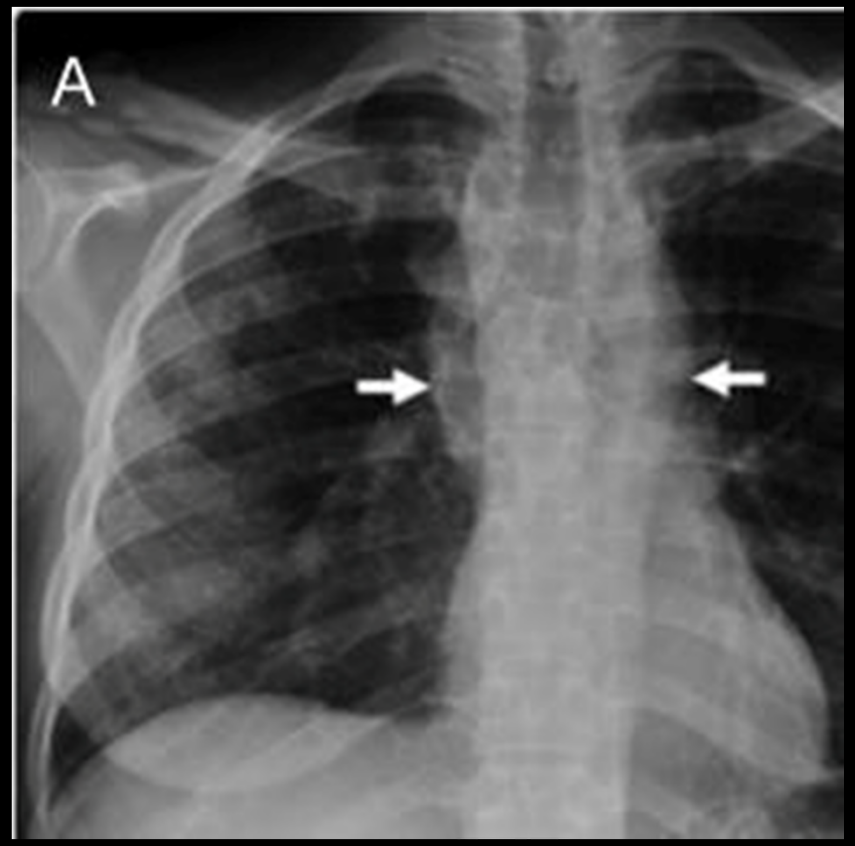
- Vaso-occlusive sickle cell pain episode
- Viral infection
- Bony mass/tumor
- Acute chest syndrome

Acute Chest Syndrome

- Caused by vaso - occlusion in pulmonary microvasculature
- Can be triggered by many causes
- Presents as cough, fever, hypoxia, respiratory distress, or chest pain
- Must have radiographic findings of new infiltrate/consolidation or pleural effusion
- Other CXR findings of sickle cell disease include bone infarcts, rib enlargement, or cardiomegaly



Other CXR findings

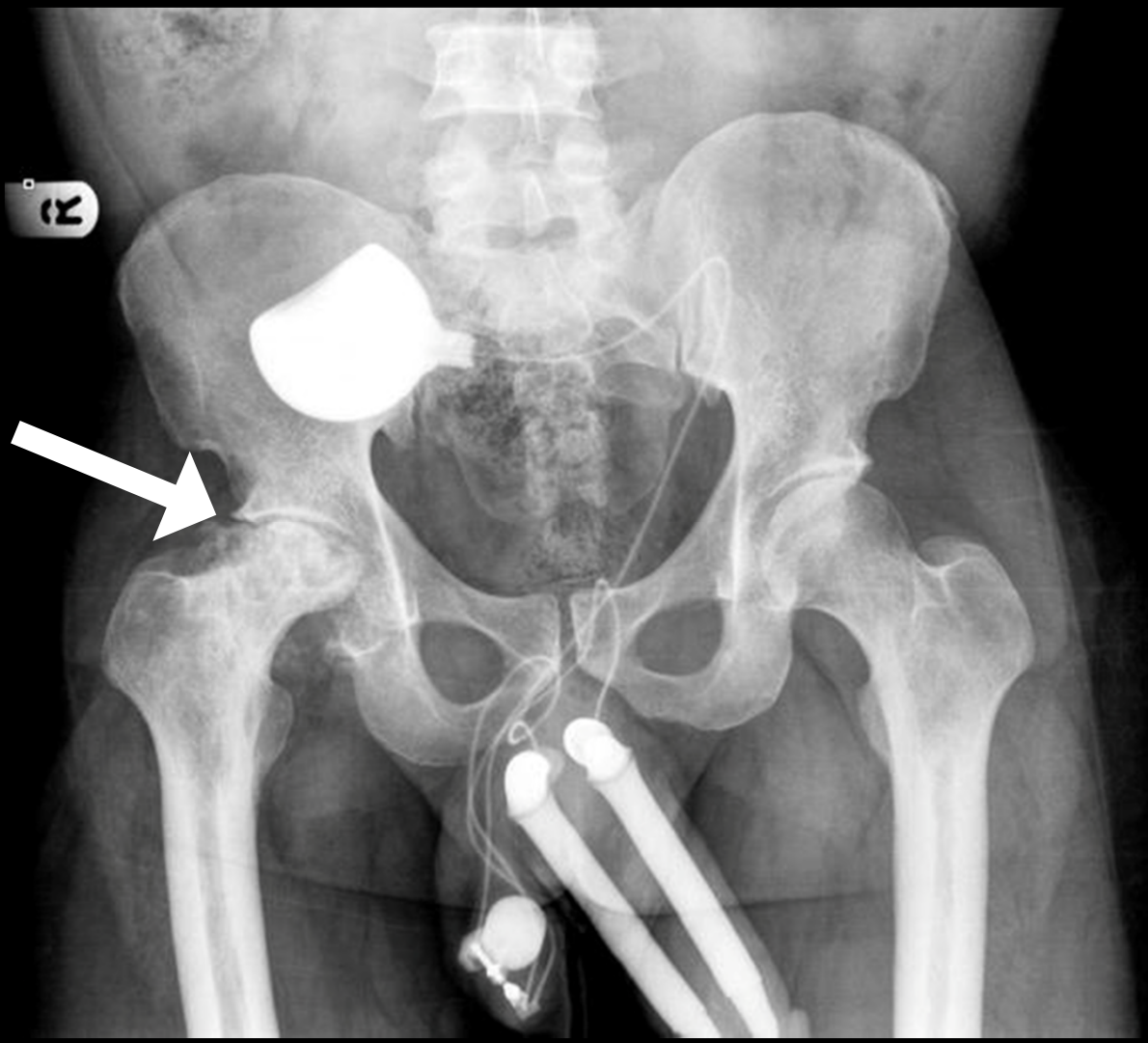


Extra-medullary hematopoiesis

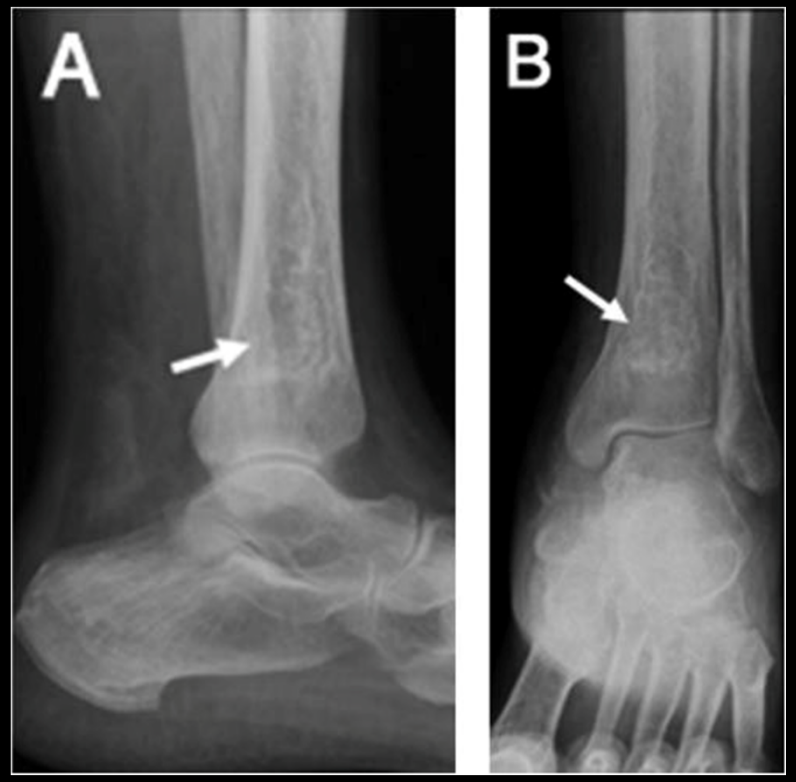
Vertebral Body infarcts (Hshaped)

Hip Osteonecrosis

- Caused by infarcts (most commonly see bony infarcts in medullary cavity and epiphysis)
- Most commonly see necrosis in proximal femur, proximal humerus, and vertebral body



Bony Infarcts



Prior infarct leading to Sclerosis

Periosteal Reaction

Infarct causing osteonecrosis

Treatment and Prognosis of Acute Pain Crisis

- Treatment
  - Multimodal Pain management – hydromorphone, acetaminophen-hydrocodone, gabapentin, and ibuprofen
  - Fluid resuscitation – D5 ½ NS 100 cc/h
  - Folic Acid – 1 mg daily
  - Incentive spirometry
  - DVT prophylaxis - eliquis
  - Blood Transfusion and antibiotics if required
- Prognosis
  - Generally resolves with pain management and fluids
  - Long term common causes of mortality include renal failure, pulmonary hypertension, and infection

ACR appropriateness Criteria and Cost

Clinical Condition: Osteonecrosis of the Hip

Variant 1: Adult or Child. Clinically suspected osteonecrosis. First study.

Radiologic Procedure	Rating	Comments	RRL*
X-ray pelvis and hips	9	This procedure includes the frog-leg view. The RRL for the adult procedure is ☢☢☢.	☢☢☢☢
CT hips without IV contrast	1	The RRL for the adult procedure is ☢☢☢.	☢☢☢☢
CT hips with IV contrast	1	The RRL for the adult procedure is ☢☢☢.	☢☢☢☢
CT hips without and with IV contrast	1	The RRL for the adult procedure is ☢☢☢☢.	☢☢☢☢☢
Tc-99m bone scan with SPECT hips	1	The RRL for the adult procedure is ☢☢☢.	☢☢☢☢
MRI hips without IV contrast	1		O
MRI hips without and with IV contrast	1		O

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate — *Relative Radiation Level

Variant 2: Suspicion of acute or potentially unstable chronic cardiopulmonary disease by history or physical examination.

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest routine admission	9		☢
X-ray chest routine preoperative	8		☢
X-ray chest routine outpatient	8		☢

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate — *Relative Radiation Level

- Cost
  - Femur series – insured $164, uninsured $231
  - Bilateral Hip series – insured $60, uninsured $310
  - Tibia/Fibula series – $167 insured, uninsured $311
  - 2 view Chest Xray – insured $261, uninsured $274

Take Home Points

- Sickle Cell Disease can lead to chronic musculoskeletal changes including
  - Osteonecrosis
  - Periosteal Reactions
  - Extra-medullary hematopoiesis
- Acute sickle cell crisis patients should be monitored for acute chest syndrome and diagnosis requires CXR findings
- Acute Pain Crisis is managed with pain management and fluids

References

- 1) uptodate https://www.uptodate.com/contents/overview-of-the-management-and-prognosis-of-sickle-cell-disease
- 2) uptodate https://www.uptodate.com/contents/acute-chest-syndrome-in-adults-with-sickle-cell-disease
- 3) radiopedia https://radiopaedia.org/articles/sickle-cell-disease?lang=us
- 4) radiopedia https://radiopaedia.org/articles/sickle-cell-disease-acute-chest-syndrome-1?lang=us
- 5) Ejindu VC, Hine AL, Mashayekhi M, Shorvon PJ, Misra RR. Musculoskeletal Manifestations of Sickle Cell Disease. RadioGraphics. 2007;27(4):1005-1021. doi:10.1148/rg.274065142.
- 6) Kosaraju V, Harwani A, Partovi S, et al. Imaging of musculoskeletal manifestations in sickle cell disease patients. The British Journal of Radiology. 2017;90(1073):20160130. doi:10.1259/bjr.20160130.
- 7) Allareddy V, Roy A, Lee MK, et al. Outcomes of Acute Chest Syndrome in Adult Patients with Sickle Cell Disease: Predictors of Mortality. PLoS ONE. 2014;9(4). doi:10.1371/journal.pone.0094387.
- 8) ACR Appropriateness Criteria. https://acsearch.acr.org/list?_ga=2.207061973.79406764.1573535160-1601025735.1572372435. Accessed November 12, 2019.
- 9) Memorial Hermann Pricing. How Do I Get a Pricing Estimate? memorialhermann. https://www.memorialhermann.org/patients-caregivers/pricing-estimates-and-information/. Published February 25, 2019. Accessed November 12, 2019.
- 10) Other Pricing Estimates. https://www.sparrow.org/upload/docs/MySparrow/Radiology%20Pricing.pdf


Questions?