Pathological Radiological Findings In Sickle Cell Disease

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Diagnostic Radiology 4001

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Clinical History

- This is a 22 Y/O African American female with a PMH of Sickle Cell Disease (HBSS) complicated by avascular necrosis of the L hip, iron overload and hyperhemolytic transfusion syndrome who presented from an outside hospital with complaining of “excruciating” back pain and left hip pain she rated a 10/10 requiring pain medication, she denied any chills cough or chest pain
- Vitals: T: 97.8 F HR: 66 RR: 18 BP: 97/53 SpO2: 95%
- PE: General Appearance: awake, alert, in severe distress

  Lungs: clear to auscultation bilaterally, no wheezes rales and rhonchi

  Heart: Regular rate and rhythm and no murmurs

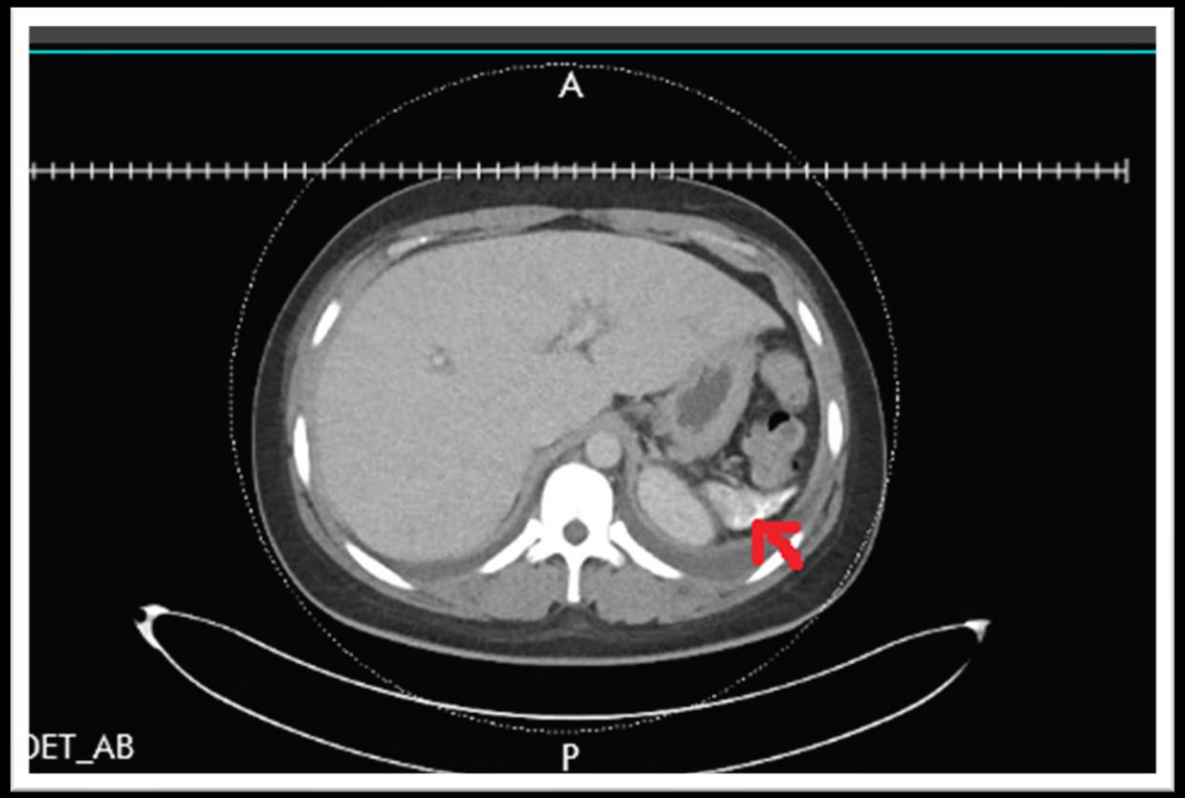
  Abdomen: soft non tender non distended no masses

  Extremities: Normal Range of motion, tenderness to palpation of left thigh and lower back

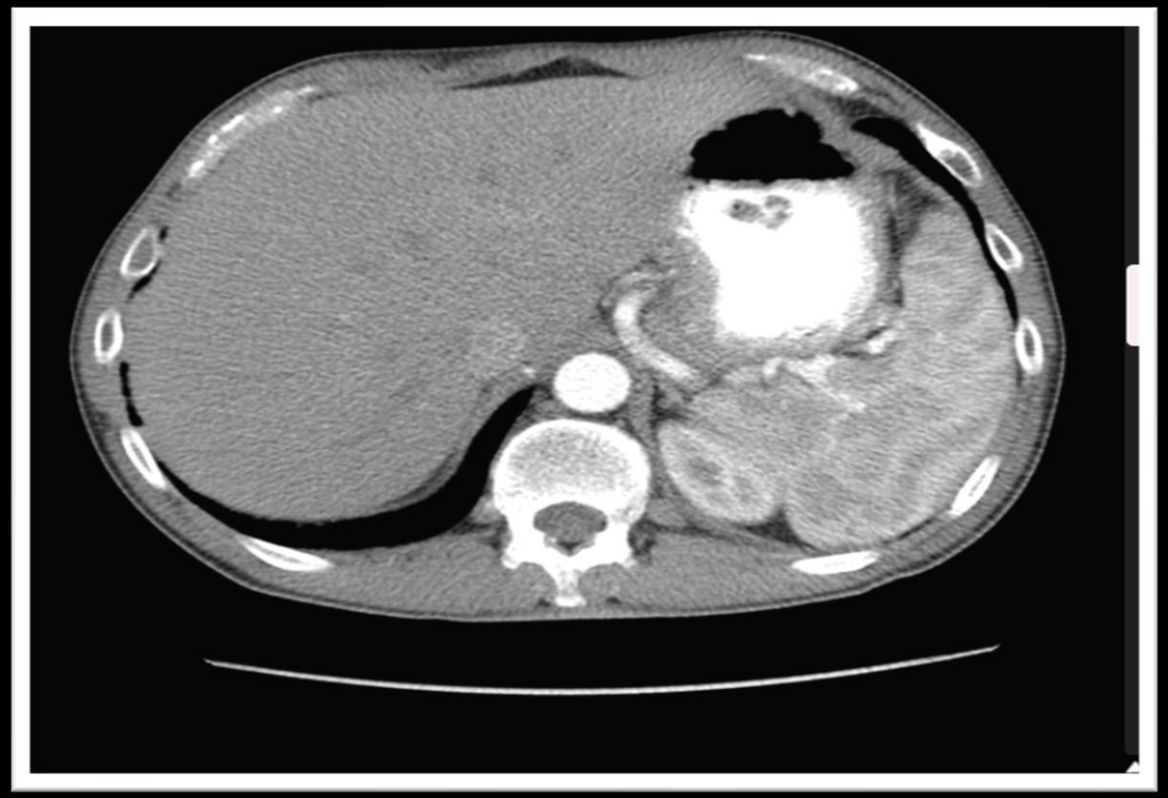
Clinical History Cont.

- Labs were remarkable for:
  - -Elevated Leukocytosis: 16.9
  - -LD: 40
  - -Hg/Hct: 7.4/21.1%
  - -Platelets: 507
  - -RDW:17.8
- The patient has had acute chest syndrome, avascular necrosis of the L hip S/P hip replacement and hyper-hemolysis syndrome
- The patient was originally worked up for Acute Sickle cell crisis:
  - -0.45% NS 100mL/hr IV
  - -MS Contin 30 mg q12h scheduled/ Dilaudid 1 mg IV to q4h prn
- The patient's course was complicated by epigastric pain that progressed to diffuse abdominal pain and SOB warranting US of RUQ with subsequent CT w/wo contrast of the abdomen and Pelvis and CTA chest with contrast for PE protocol

Relevant Imaging

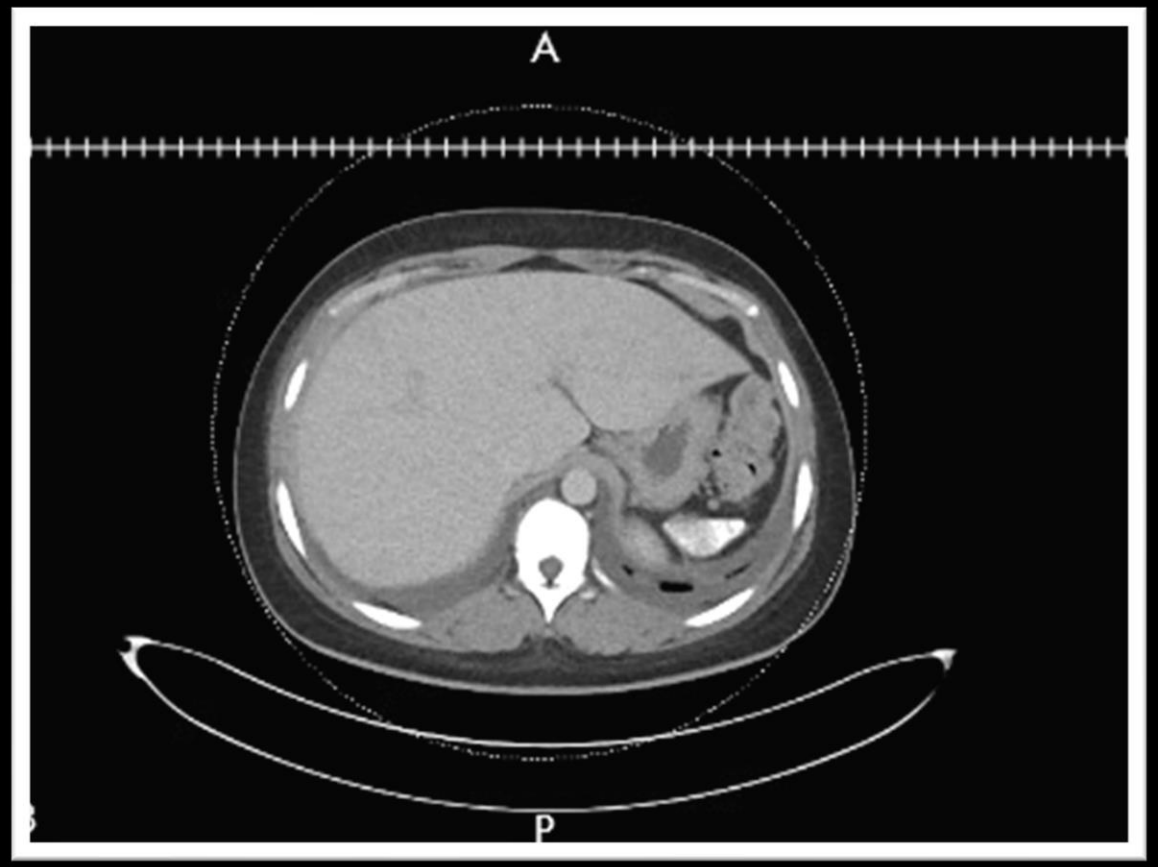


CT abdomen and Pelvis with contrast
Axial Orientation
Portal venous phase
08/23/2020

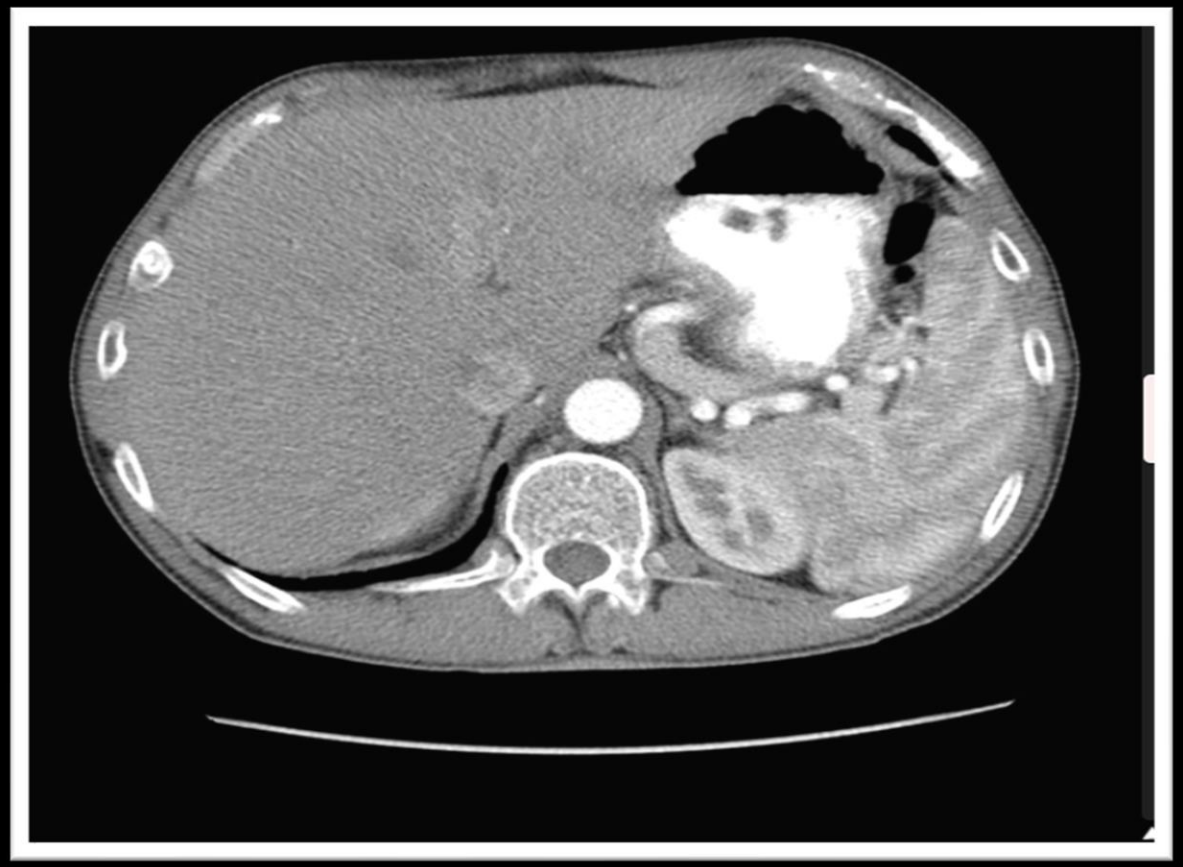


Normal Spleen: arterial phase
https://radiopaedia.org/cases/normal-arterial-phase-ct-appearance-of-the-spleen?lang=us

Relevant imaging cont.

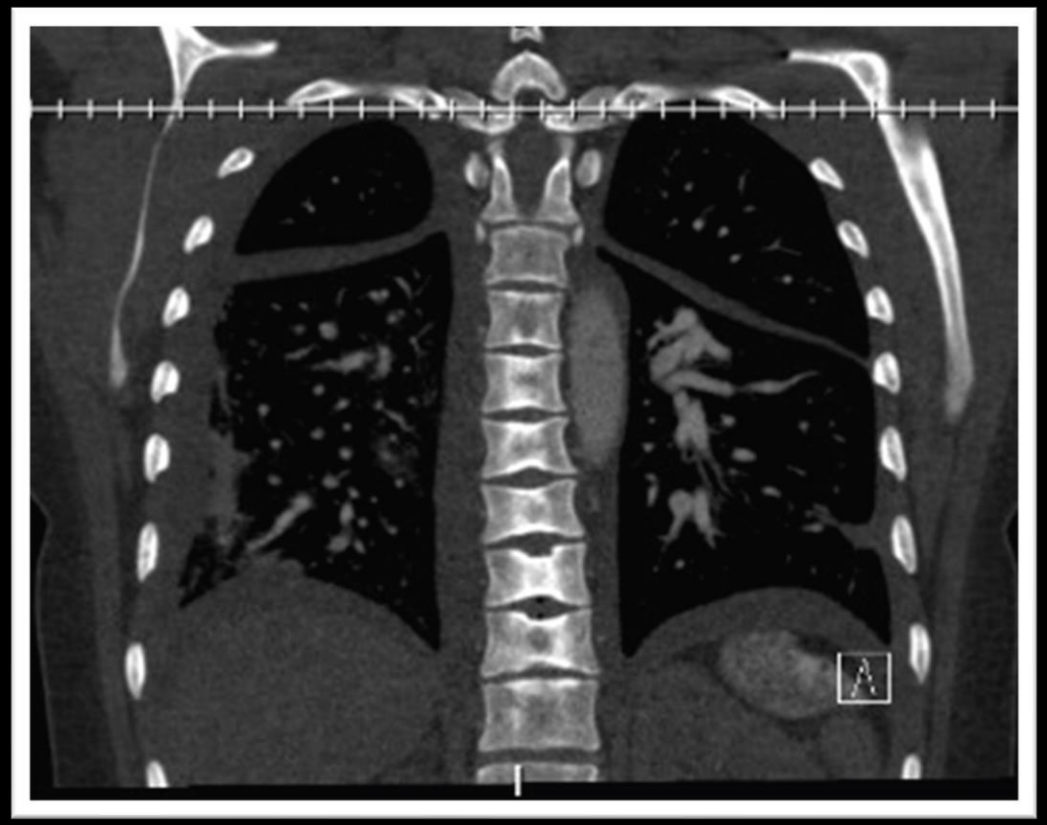


CT abdomen and pelvis
Axial orientation
Portal Venous Phase
08/23/2020

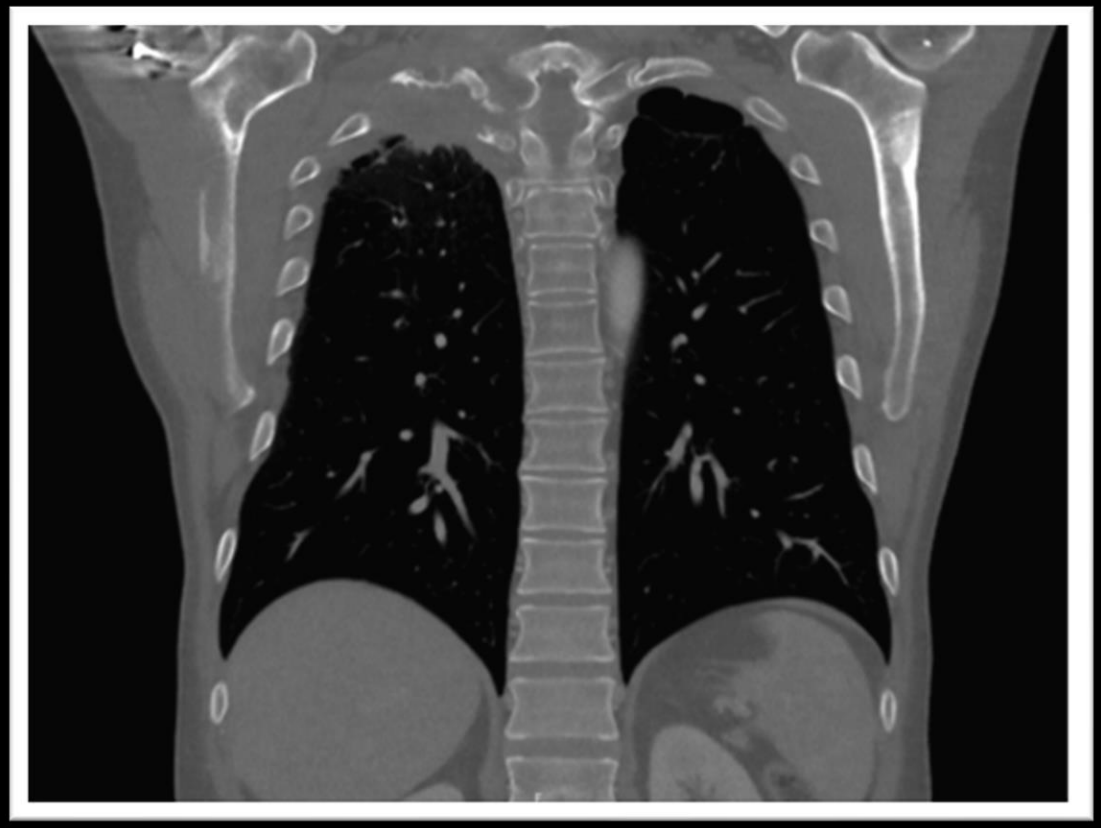


Normal spleen arterial phase
https://radiopaedia.org/cases/normal-arterial-phase-ct-appearance-of-the-spleen?lang=us

Relevant Imaging cont

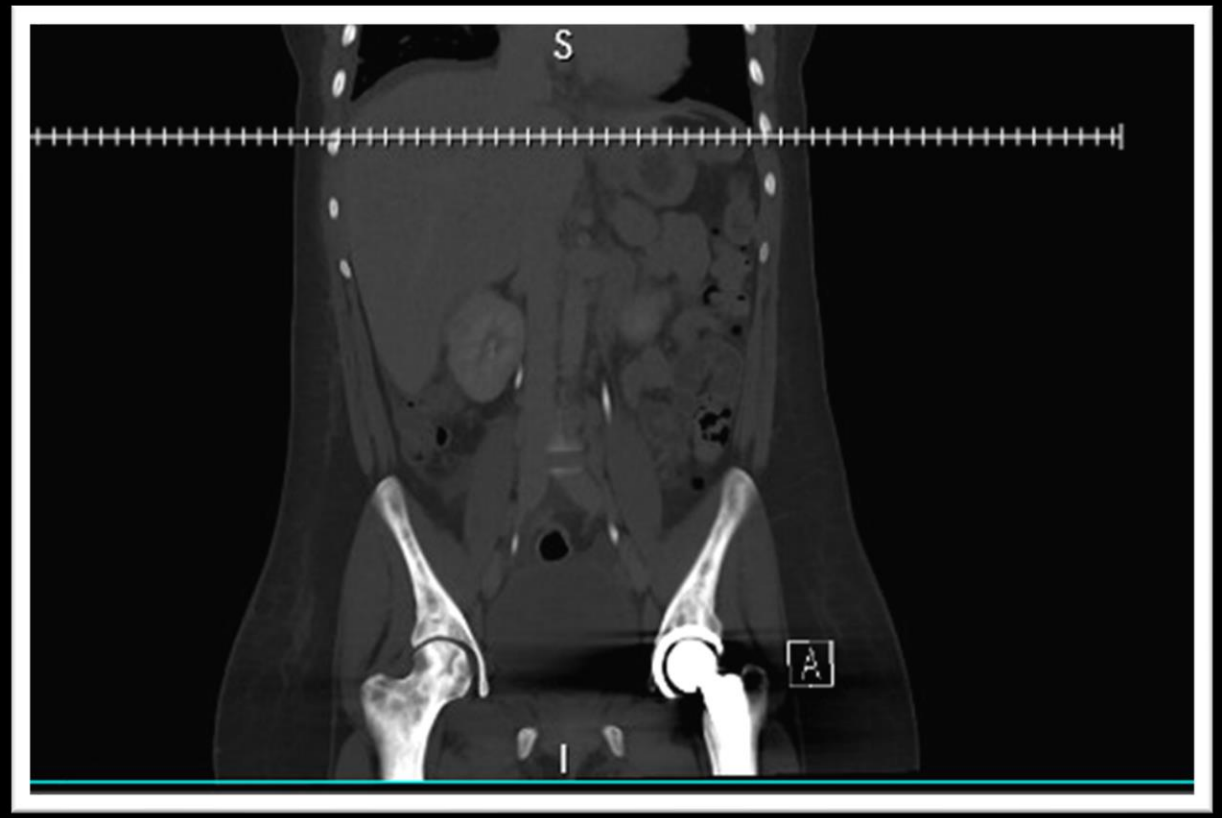


CTA chest w/contrast coronal reconstruction bone window (H sign)
08/23/2020

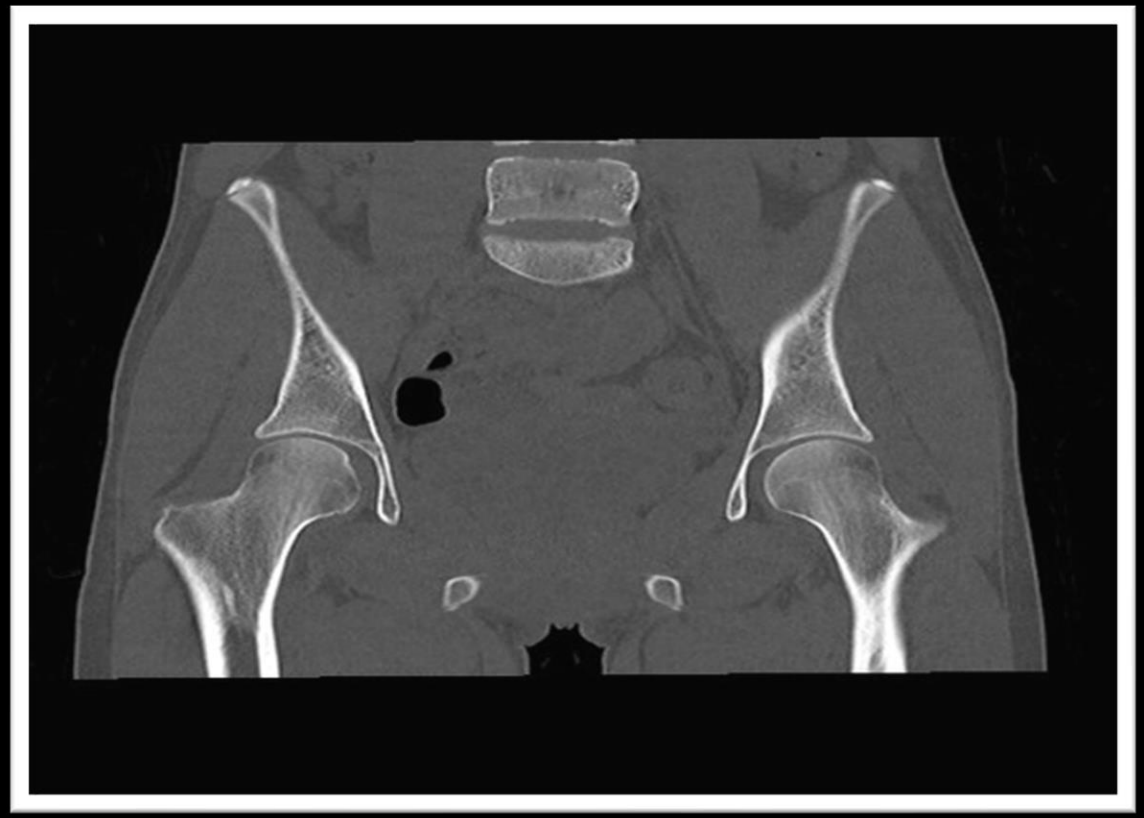


Normal thoracic vertebra: reconstructed coronal bone window
https://erj.ersjournals.com/content/50/3/1700611.figures-only

Relevant imaging cont.



CT w/wo Contrast of abdomen and pelvis coronal view bone window
08/23/2020



Normal Pelvis: coronal view bone window
https://radiopaedia.org/cases/normal-pelvis-ct

Findings and Impressions

- Due to the patient's given PMH of sickle cell disease and progression of symptoms including abdominal pain and SOB patient indicated aCT abdomen and pelvis w/wo contrast, CTA chest with contrast for PE protocol
- CT abdomen and pelvis was correlated with an RUQ US performed the day before for abdominal pain (not shown)
- Findings:
    1. Calcified spleen likely secondary to prior infarctions from repeated sickle cell crisis
    2. Bones sequalae of Sickle cell disease:
       -Multiple H shaped vertebrae
       -Patchy sclerosis of the pelvis and proximal femur

Differential Diagnosis of Findings:

- *Atrophic Spleen/Splenic infarction*:

  -CT abdomen with contrast is highly sensitive and specific to splenic infarction and when images are optimal differential diagnosis is minimal

  -Differential Diagnosis: infarction secondary to hematologic disorders (sickle cell disease)

  normal inhomogeneous splenic infarction (seen on arterial phase), lymphoma and splenic abscess

- *H shaped Vertebrae (Lincoln Vertebare)*:

  -results from microvascular end plate infarction

  -Differential diagnosis: seen in 10% of patients with sickle cell anemia and occasionally in Gaucher's disease

- *Sclerosis of the Hip:*

  -sclerosis of generalized increase in bone density

  -Differential diagnosis: hematologic disorders (Sickle cell disease), metabolic (renal osteodystrophy), Paget disease

Discussion of Findings based on Clinical Hx

- These radiological findings are all secondary to the pathophysiology seen within Sickle cell disease:
  - Repeated vaso-occlusive crises leading to end organ/ bone infarctions
  - chronic anemia (from RBC lysis) resulting in expansion of the medullary spaces as well as sequestration syndrome (specific to spleen)
  - Infection (i.e osteomyelitis)
- Auto infarction, H shaped Vertebrae and Sclerosis of pelvis and proximal femur are classical radiological findings of a patient with sickle cell disease secondary to microvascular infarction
- Further management includes:
  - oral Abx as PPX for increased infection 2/2 encapsulated bacteria
  - Surveillance for bone changes and pathological fractures and infection (Osteomyelitis)
  - Total hip replacement 2/2 auto infarction (as this patient received)

Final Diagnosis

- Radiological findings overwhelming demonstrate pathologies of microvascular infarctions
- Based on the patient's demographics such as age and ethnicity a congenital hematological pathology such as Sickle Cell disease should be worked up for most probable diagnosis
- Diagnosis of Sickle Cell Disease is made by Hg Electrophoresis/ genetic studies demonstrating point mutation resulting in abnormal hemoglobin
- Management is primarily on pain management for acute sickle cell crisis/ anemia management and Abx prophylaxis for functional asplenia, patients often receive hydroxyurea to increase fetal hemoglobin concentration
- leading causes of death included pulmonary hypertension, sudden death, renal failure, and infection

ACR appropriateness Criteria

Based on patient's presentation and positive ROS remarkable for SOB and Diffuse abdominal pain CT abdomen and pelvis and CTA of the chest for PE protocol (in the setting of Sickle cell disease) are appropriate based on ACR criteria:

Variant 1: **Suspected pulmonary embolism. Intermediate probability with a negative D-dimer or low pretest probability.**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CTA chest with IV contrast	5	This procedure should be optimized for pulmonary arterial enhancement. This procedure may be appropriate but there was disagreement among panel members on the appropriateness rating as defined by the panel's median rating.	☢☢☢
CT chest with IV contrast	3	This procedure should be optimized for pulmonary arterial enhancement.	☢☢☢
US duplex Doppler lower extremity	3	This procedure has a low yield in the absence of symptoms of DVT.	O
CT chest without IV contrast	2		☢☢☢
Tc-99m V/Q scan lung	2		☢☢☢
CTA chest with IV contrast with CT venography lower extremities	2		☢☢☢
MRA chest without and with IV contrast	2		O
US echocardiography transthoracic resting	2		O
CT chest without and with IV contrast	1		☢☢☢
Arteriography pulmonary with right heart catheterization	1		☢☢☢☢
MRA chest without IV contrast	1		O
US echocardiography transesophageal	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 1: **Acute nonlocalized abdominal pain and fever. No recent surgery. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	O
US abdomen	May Be Appropriate	O
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
MRI abdomen and pelvis without IV contrast	May Be Appropriate	O
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	☢☢☢☢
Radiography abdomen	May Be Appropriate	☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
WBC scan abdomen and pelvis	Usually Not Appropriate	☢☢☢☢
Nuclear medicine scan gallbladder	Usually Not Appropriate	☢☢
Fluoroscopy contrast enema	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	☢☢☢

Cost of imaging: CT Abdomen w& w/o contrast: $1,294; CTA Chest: $1,160; US: $ 187

CXRAY: 300$; Total= $2,941

https://www.newchoicehealth.com/places/texas/houston/x-ray

Take Home Points / Teaching points

- Sickle cell disease is genetic hematological disease due to a point mutation disrupting the normal morphology of Hemoglobin resulting sickling of RBCs causing Vaso occlusion and end organ damage
- Although diagnosis is made through Hg electrophoresis, many radiological findings can suggest end organ damage/infarction highly suspicious of highly sickle cell disease given the clinical history
- These radiological findings include but not limited to: Splenic infarction (most commonly seen), bone changes (H shaped vertebrae and Sclerosis of the bone) as seen in the patient

References

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- .Fondi C, Franchi A. Definition of bone necrosis by the pathologist. Clinical cases in Mineral and Bone metabolism. 2007 Jan; 4(1): 21-6. Available at http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2781178/#R11
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Questions?