AMSER Case of the Month:

30 yo F with knee pain





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Patient Presentation

- <u>HPI</u>: Patient is a 30 yo female presented to the ED with worsening of chronic right knee pain without swelling, effusion, redness, or warmth
- <u>PMH</u>: Hemoglobin SC disease, recurrent vaso-occlusive crises/pain episodes with multiple ED admissions, DVT, PE
- <u>PSH</u>: Indwelling central venous catheter (removed due to infection)
- <u>Meds</u>: Acetaminophen, cyclobenzaprine, diphenhydramine HCl, folic acid, hydromorphone, rivaroxaban
- <u>Allergies</u>: penicillin
- <u>Soc Hx</u>: Denies use of tobacco, ETOH, recreational drugs
- <u>Vitals</u>: T 36.8°C | BP 151/106 | Pulse 89 | RR 16 | SpO2 97%
- <u>Pertinent Labs</u>: Hgb 9.7 | HCT 28% | RBC 3.47 | MCV 81 | WBC 10.1 | PLT 322

What Imaging Should We Order?



Select the applicable ACR Appropriateness Criteria

<u>Variant 1:</u> Adult or child greater than or equal to 5 years of age. Chronic knee pain. Initial imaging.				
Procedure	Appropriateness Category	Relative Radiation Level		
Radiography knee	Usually Appropriate	÷ 🔶		
Aspiration knee	Usually Not Appropriate	Varies		
CT arthrography knee	Usually Not Appropriate	¢		
CT knee with IV contrast	Usually Not Appropriate	Ŷ		
CT knee without and with IV contrast	Usually Not Appropriate	Ŷ		
CT knee without IV contrast	Usually Not Appropriate	¢		
MR arthrography knee	Usually Not Appropriate	0		
MRI knee without and with IV contrast	Usually Not Appropriate	0		
MRI knee without IV contrast	Usually Not Appropriate	0		
Tc-99m bone scan knee	Usually Not Appropriate	\$ \$ \$		
US knee	Usually Not Appropriate	0		
Radiography hip ipsilateral	Usually Not Appropriate	* * *		

This imaging modality was ordered by the ER physician



Findings (unlabeled)







Findings (labeled)



AP and lateral radiographs of the right knee



Select the applicable ACR Appropriateness Criteria

<u>Variant 7:</u> Lesion on radiographs. Indeterminate for malignancy. Sclerotic or mixed lytic/sclerotic lesion.

Radiologic Procedure	Rating	Comments	RRL*
MRI area of interest without and with IV contrast	8		• (
MRI area of interest without IV contrast	7		О
CT area of interest without IV contrast	7		Varies
Tc-99m bone scan whole body	5		***
CT area of interest without and with IV contrast	4	Perform this procedure if MRI is contraindicated.	Varies
FDG-PET/CT whole body	3		***
X-ray skeletal survey	2		***
CT area of interest with IV contrast	2		Varies
US area of interest	1		0
<u>Rating Scale:</u> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			

This imaging modality was ordered by the admitting physician



Findings (unlabeled)





Findings (labeled)

Multifocal serpentine signal abnormalities in the distal femur and proximal tibia

Unenhanced coronal T2 STIR MRI sequence of the right knee



Final Dx:

Avascular necrosis of the distal femur and proximal tibia (in a patient with a sickle cell variant genotype)



Case Discussion

- Avascular necrosis (osteonecrosis) is a chronic vaso-occlusive complication in patients with sickle cell disease and sickle cell variants
- May be initially asymptomatic and found incidentally on imaging, but may progress to cause chronic pain and architectural destruction in bone



AP chest radiograph demonstrating an area of sclerosis in the right humeral head <u>found</u> <u>incidentally</u> in a patient with sickle cell disease imaged for acute chest syndrome



Sagital reconstruction of non-contrast CT of the spine showing multilevel H-shaped vertebrae with central height loss due to end-plate depression in a 28yo patient with sickle cell disease and acute-on-chronic back pain



Case Discussion

- Hemoglobin SC disease (HbSC) is a sickle cell variant in which individuals have compound heterozygous mutations resulting in one gene for HbS and one gene for HbC
- Presentation is typically milder than sickle cell disease (HbSS) with a later onset of complications, however, patients remain at risk for avascular necrosis <u>and other</u> <u>sequelae of chronic</u> <u>microvascular occlusion</u>



Contrast-enhanced CT, coronal reconstruction, maximum intensity projection through the left kidney during the renal excretory phase in a sickle cell patient presenting with hematuria and flank pain which demonstrates blunting of the calvces with filling defects in multiple poles of the left kidney and a "ball-on-tee" deformity. These findings are characteristic for renal papillary necrosis. Also seen is a tiny, calcified spleen consistent with splenic auto-infarction. Both are sequelae of chronic vaso-occlusive crises in sickle cell patients.



References:

- 1. ACR Appropriateness Criteria: https://acsearch.acr.org/docs/69432/Narrative/
- Vichinsky, E.P. (2019). Overview of variant sickle cell syndromes. Mahoney, D.H. & Tirnauer, J.S. (Eds.), UpToDate. Retrieved July 20, 2019 from https://www.uptodate.com/contents/overview-of-variant-sicklecellsyndromes?search=hemoglobin%20sc&source=search_result&selectedTitle=1~49&usage_type=default&di splay_rank=1#H2
- George, A. et al. (2019). Acute and chronic bone complications of sickle cell disease. Mahoney, D.H., Philips, W.A., Schrier, S.L. & Tirnauer, J.S. (Eds.), *UpToDate*. Retrieved July 20, 2019 from https://www.uptodate.com/contents/acute-and-chronic-bone-complications-of-sickle-cell-disease?search=sickle%20cell%20osteonecrosis§ionRank=1&usage_type=default&anchor=H16&source=machineLearning&selectedTitle=1~150&display_rank=1#H16
- 4. <u>https://radiopaedia.org/articles/avascular-necrosis?lang=us</u>
- 5. <u>https://radiopaedia.org/articles/sickle-cell-disease?lang=us</u>

