AMSER Case of the Month: October 2020

56-year-old male exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea

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AMSER

Patient Presentation

- HPI: 56-year-old male with exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea, weight gain (20 pounds over last 4 months), leg swelling, cough
- Medical History: Hypertension, hyperlipidemia, chronic kidney disease
- Surgical History: None
- Medications: Albuterol inhaler, cholecalciferol, fexofenadine, fluticasone nasal spray
- Physical Exam: Tachycardic, bibasilar crackles, abdomen mildly distended, +1 bilateral peripheral edema up to midshins bilaterally
- Labs: Cr 1.27 (unknown baseline), BNP 1,501

What Imaging Should We Order?

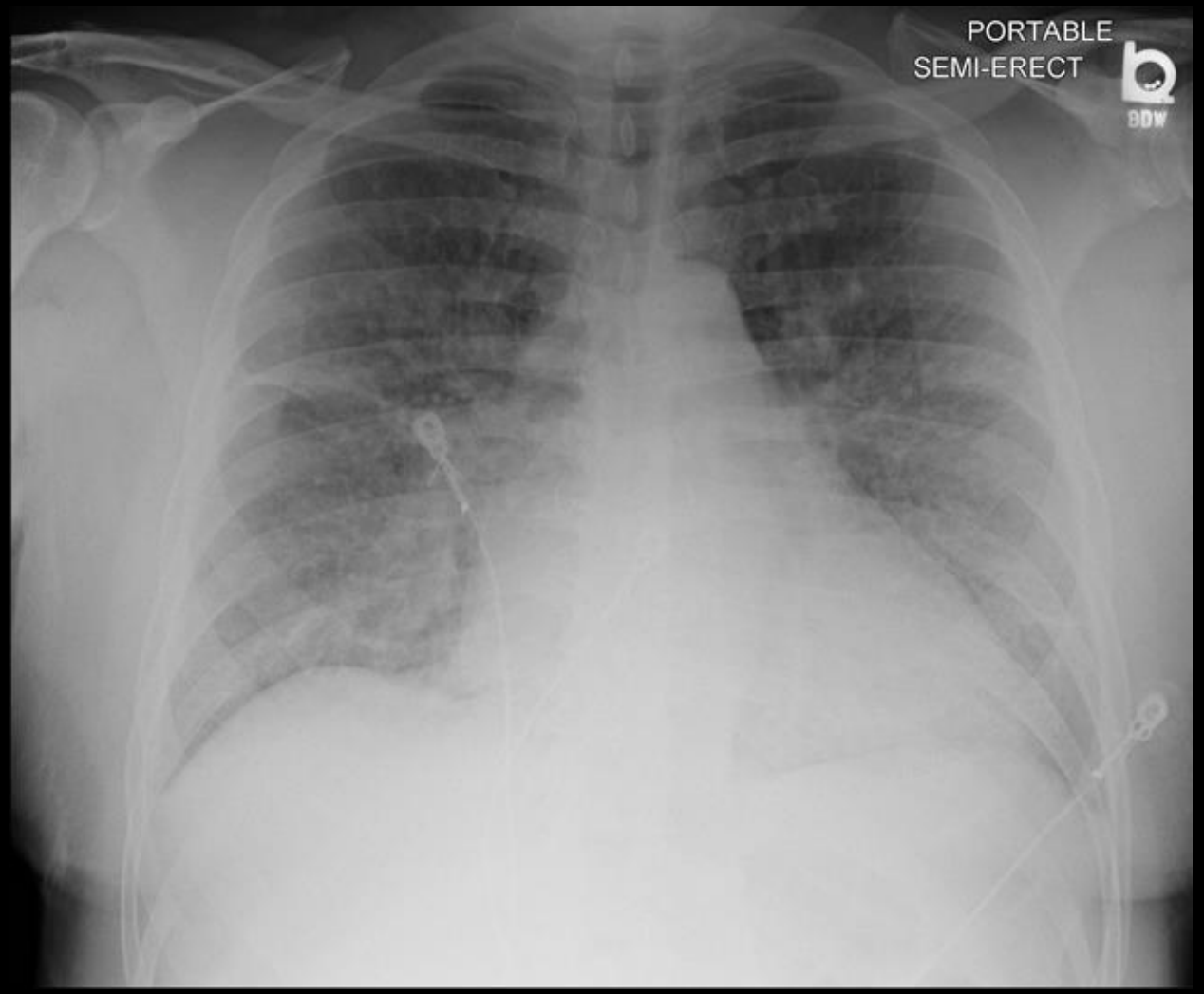
Select the applicable ACR Appropriateness Criteria

Variant 1: Dyspnea due to heart failure. Ischemia not excluded.

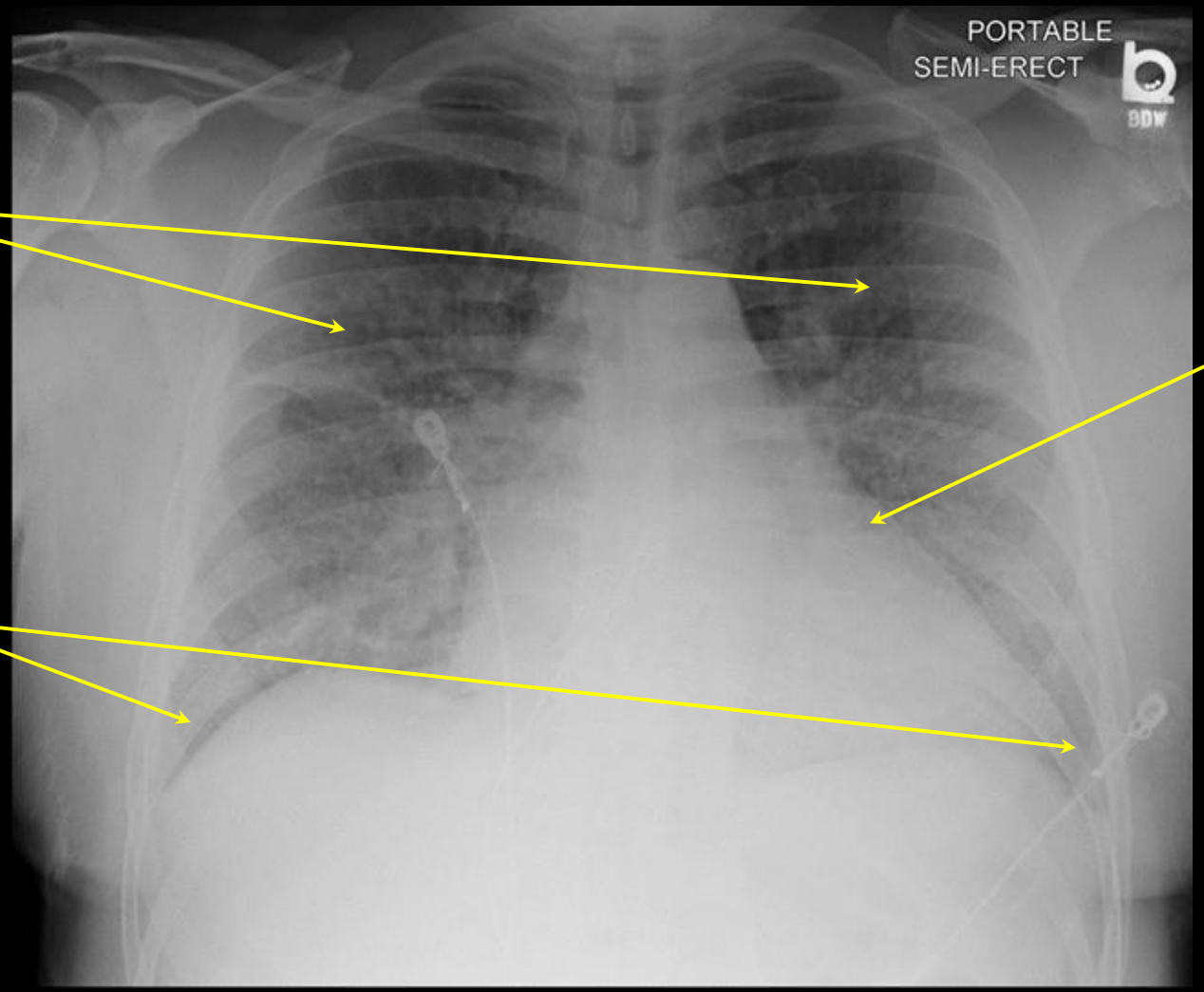
Procedure	Appropriateness Category	SOE	Adult RRL	Peds RRL	Rating	Median	Final Tabulations								
							1	2	3	4	5	6	7	8	9
US echocardiography transthoracic resting	Usually appropriate		O 0 mSv	O 0 mSv [ped]	9	n/a	0	0	0	0	0	0	0	0	0
US echocardiography transthoracic stress	Usually appropriate		O 0 mSv	O 0 mSv [ped]	9	n/a	0	0	0	0	0	0	0	0	0
Radiography chest	Usually appropriate		☢ <0.1 mSv	☢ <0.03 mSv [ped]	9	n/a	0	0	0	0	0	0	0	0	0
Arteriography coronary with ventriculography	Usually appropriate		☢☢☢ 1-10 mSv		8	n/a	0	0	0	0	0	0	0	0	0
MRI heart function and morphology without and with IV contrast	Usually appropriate		O 0 mSv	O 0 mSv [ped]	8	n/a	0	0	0	0	0	0	0	0	0
MRI heart with function and inotropic stress without and with IV contrast	Usually appropriate		O 0 mSv	O 0 mSv [ped]	7	n/a	0	0	0	0	0	0	0	0	0

This imaging modality was ordered by the ER physician

Chest Radiograph (Unlabeled)



Chest Radiograph (Labeled)



Bilateral reticulonodular opacities

Enlarged cardiac silhouette

Small bilateral pleural effusions

Select the applicable ACR Appropriateness Criteria

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

Procedure	Appropriateness Category	SOE	Adult RRL	Peds RRL	Rating	Median	Final Tabulations								
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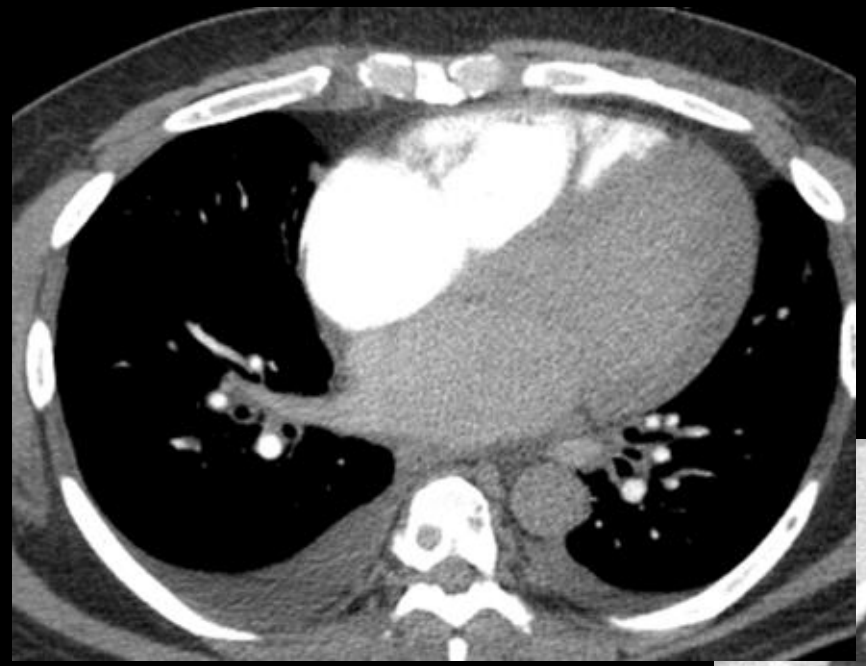
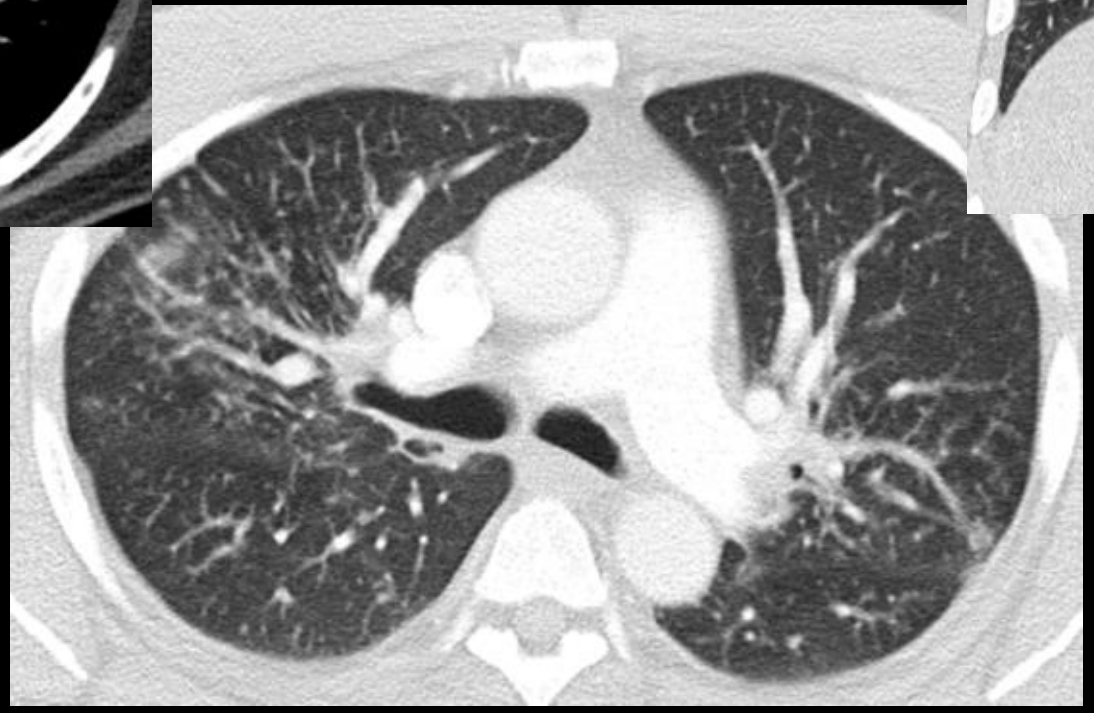
Radiography chest	Usually appropriate		☢ <0.1 mSv	☢ <0.03 mSv [ped]	9	n/a	0	0	0	0	0	0	0	0	0
CTA chest with IV contrast	May be appropriate		☢☢☢ 1-10 mSv	☢☢☢☢ 3-10 mSv [ped]	5	n/a	0	0	0	0	0	0	0	0	0
US duplex Doppler lower extremity	Usually not appropriate		O 0 mSv	O 0 mSv [ped]	3	n/a	0	0	0	0	0	0	0	0	0
US echocardiography transesophageal	Usually not appropriate		O 0 mSv	O 0 mSv [ped]	1	n/a	0	0	0	0	0	0	0	0	0
US echocardiography transthoracic resting	Usually not appropriate		O 0 mSv	O 0 mSv [ped]	2	n/a	0	0	0	0	0	0	0	0	0
Arteriography pulmonary with right heart catheterization	Usually not appropriate		☢☢☢☢ 10-30 mSv		1	n/a	0	0	0	0	0	0	0	0	0

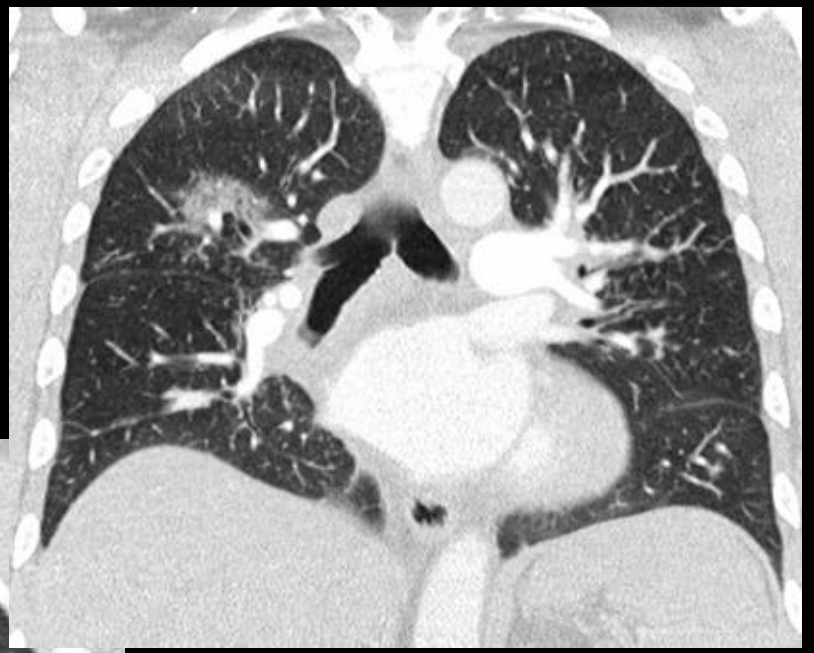
Ordered to rule out PE!

This imaging modality was ordered by the ER physician

CTA Chest (Unlabeled)

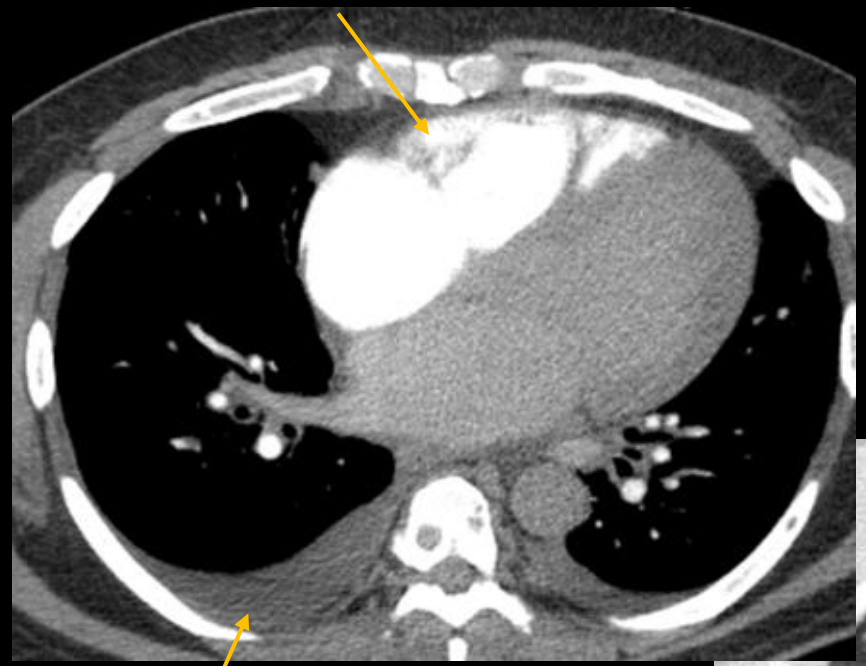




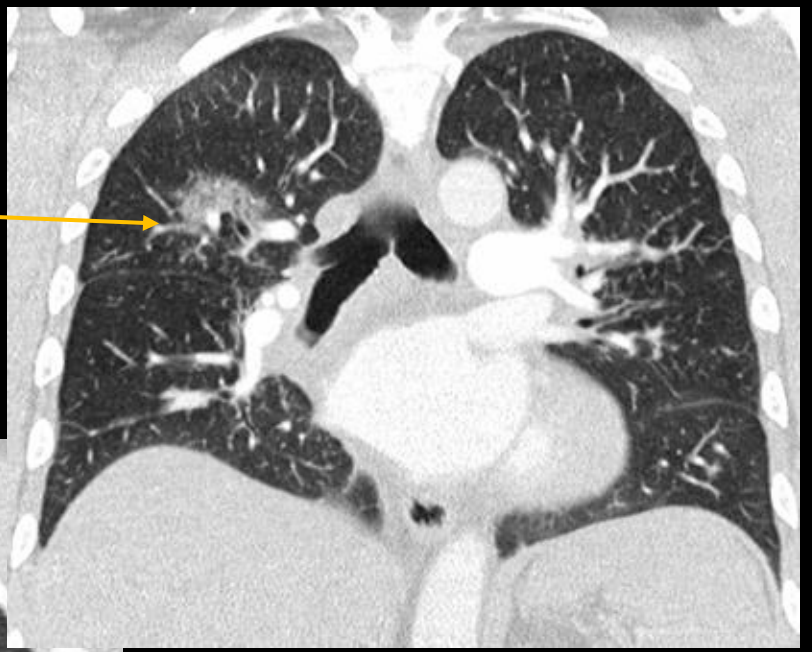


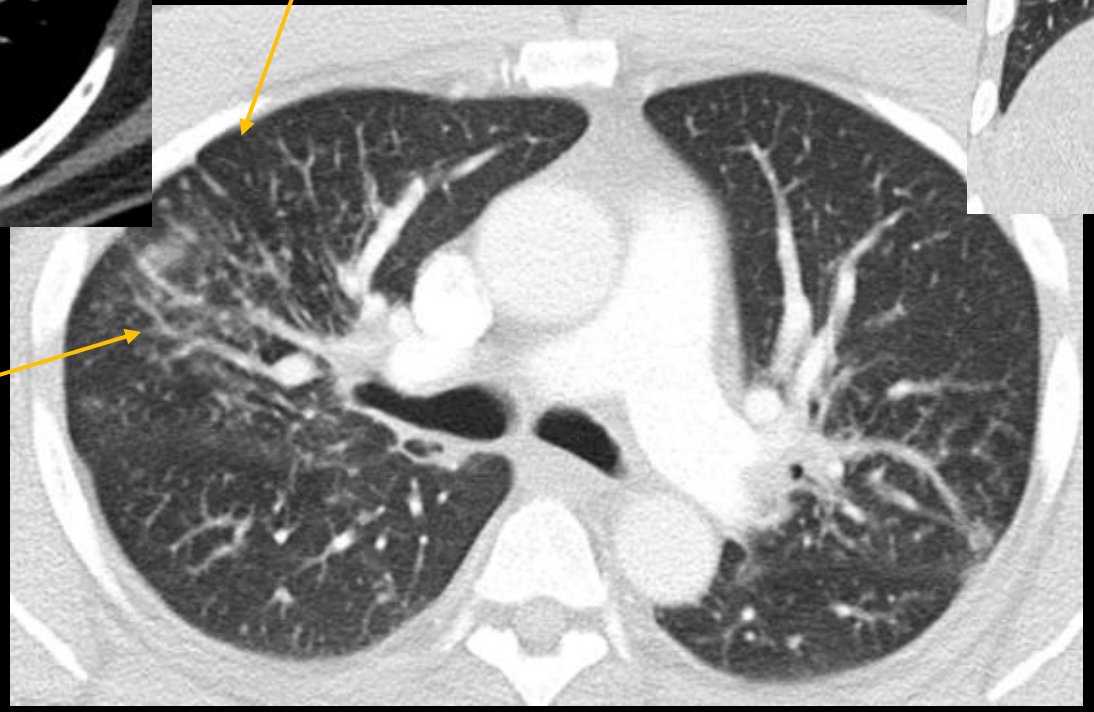
CTA Chest (Labeled)

Mild cardiomegaly



Ground-glass opacities in upper lobes





Perilymphatic pulmonary nodules

Pleural effusions

Rationale

- CTA Chest negative for PE
- Considering this, patient presentation likely due to non ischemic cardiomyopathy or congestive heart failure
- Evidence of pulmonary sarcoidosis also found on CTA Chest
- Due to suspicion for systemic disease, primary team proceeded with cardiac MRI….

Select the applicable ACR Appropriateness Criteria

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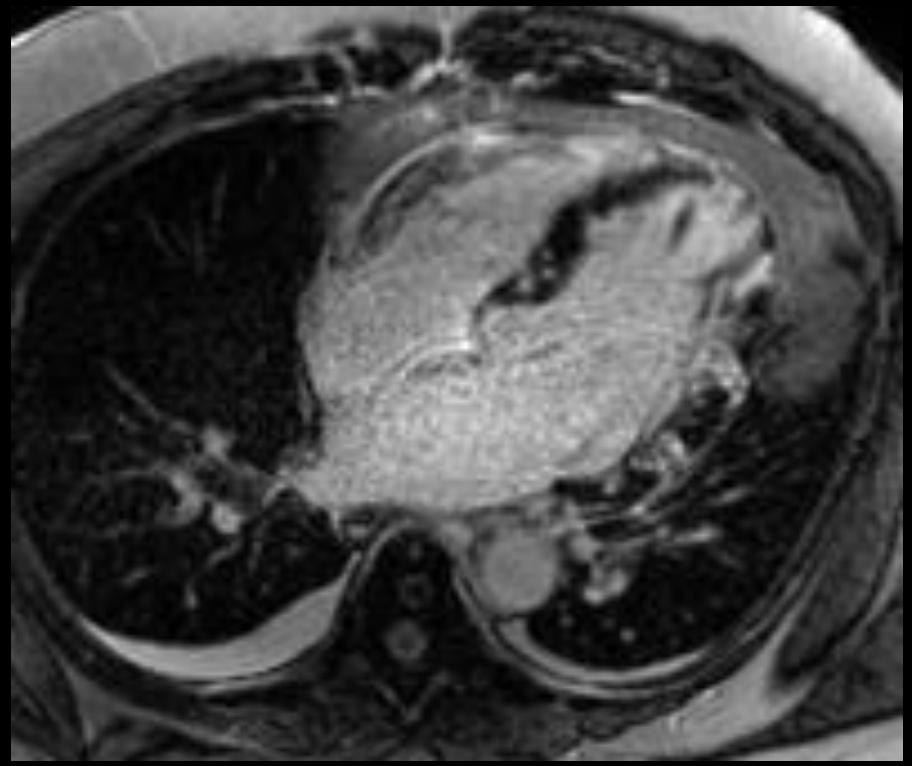
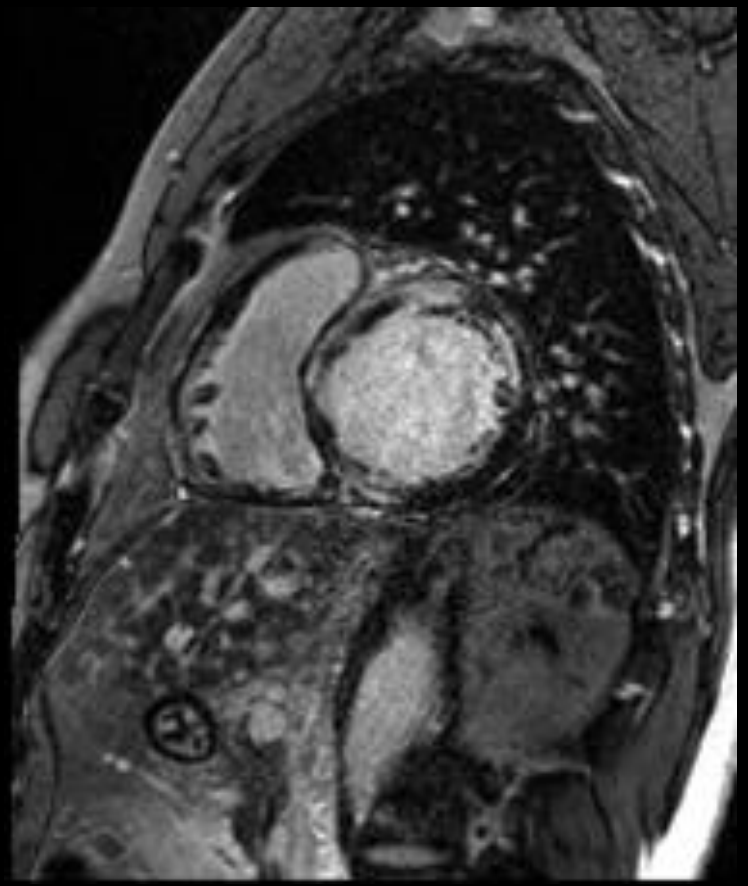
This imaging modality was ordered by the primary team

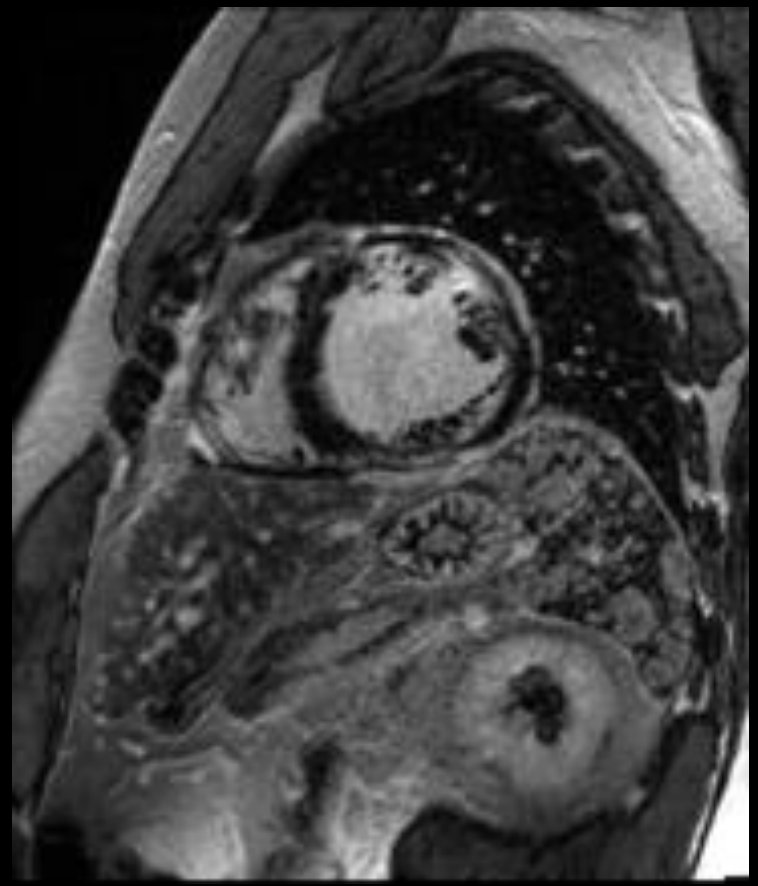
Cardiac MRI (Unlabeled)



Delayed contrast-enhanced
Horizontal Long axis

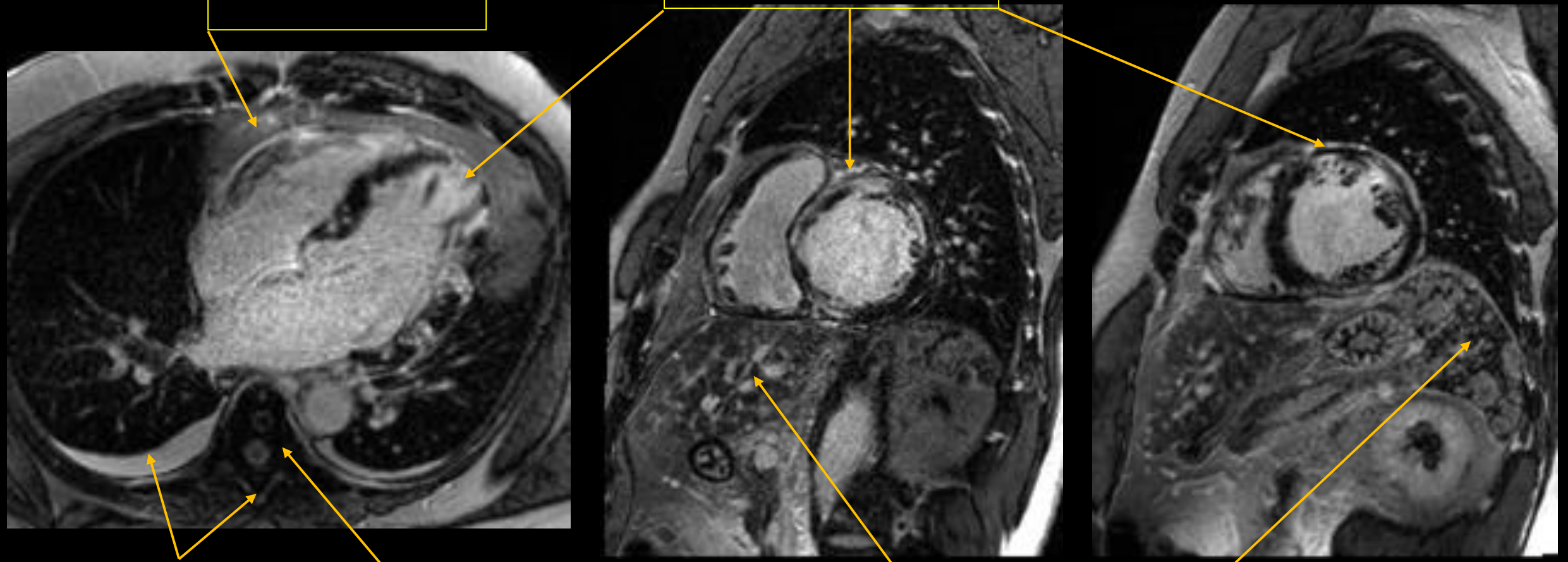


Delayed contrast-enhanced
Short axis view



Delayed contrast-enhanced
Short axis view

Cardiac MRI (Labeled)



Final Dx:

Cardiac Sarcoidosis

Cardiac Sarcoidosis

Etiology:

Exposure to antigens (bacteria, environmental agents, etc.) → Immune response leading to non-caseating granuloma formation by helper T cells in cardiac myocytes

Clinical Presentation:

- Congestive heart failure symptoms (shortness of breath, fatigue, edema)
- Arrhythmias (complete heart block, ventricular tachycardia), chest pain

Differential Diagnosis:

- Ischemic Cardiomyopathy – Hx of longstanding CAD and possible MI, usually dilated cardiomyopathy picture (HFrEF)
- Constrictive Pericarditis – Hx of pericarditis or other risk factors (surgery, radiation, autoimmune), pericardial effusion or signs of tamponade, pericardial calcifications

Cardiac Sarcoidosis - Diagnosis

Serum Biomarkers: ACE – no longer considered helpful diagnostically, may be used to monitor progression

ECG: Atrioventricular block, tachycardia, repolarization abnormalities

Echocardiography: LV systolic/diastolic dysfunction, pericardial effusion, valvular abnormalities

Nuclear Medicine PET/CT: May be useful in assessing disease activity

MRI: Imaging modality of choice – segmental myocardial motion abnormalities, increased myocardial T2 signal intensity, late gadolinium enhancement highly sensitive

Histology: Cardiac biopsy is also an option! Would show non-caseating granulomas in myocytes, “Invasive” and low sensitivity due to sampling error, not usually done

Cardiac Sarcoidosis – Treatment

- Corticosteroids – slows progression of inflammation and fibrosis, doses of 60-80 mg of prednisone are generally prescribed
- Immunotherapy – methotrexate or azathioprine commonly prescribed for patients who cannot tolerate steroids or do not respond to steroids
- Pacemakers/Defibrillators – reserved for patients with impairing degrees of heart block and tachyarrhythmias, respectively
- Heart Transplant – an option for younger patients with severe disease, although rarely done

Cardiac Sarcoidosis – Prognosis

Prognosis

- Heart involvement is life-threatening- requires prompt treatment with steroids and immunosuppressive therapy

- Overall mortality not well defined- depends largely on the impairment of left ventricular function (symptomatic patient survival mostly limited to two years, another study records 5-year mortality of 40-60%)

References:

1. Doughan, A., & Williams, B. (2006, February). Cardiac sarcoidosis. Retrieved July 28, 2020, from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1860791/
2. Dubrey, S., Bell, A., & Mittal, T. (2007, October). Sarcoid heart disease. Retrieved July 28, 2020, from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2600123/
3. Kusano, K., & Satomi, K. (2016, February 01). Diagnosis and treatment of cardiac sarcoidosis. Retrieved July 28, 2020, from https://heart.bmj.com/content/102/3/184.full
4. Okada, D., Bravo, P., Vita, T., Agarwal, V., Osborne, M., Taqueti, V., . . . Blankstein, R. (2018, August). Isolated cardiac sarcoidosis: A focused review of an under-recognized entity. Retrieved July 28, 2020, from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5540795/
5. Hulten, E., Aslam, S., Osborne, M., Abbasi, S., Bittencourt, M., & Blankstein, R. (2016, February). Cardiac sarcoidosis-state of the art review. Retrieved July 28, 2020, from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4731586/