

AMSER Case of the Month

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Shortness of Breath in a 50-Year-Old Male with
Congestive Heart Failure

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Patient Presentation

- **HPI:** A 50-year-old male presented for an outpatient transesophageal echocardiogram and was noted to have a respiratory rate of 30 with increased work of breathing. He denied any dyspnea, chest pain, syncope, palpitations, edema, or exertional symptoms.
- **PMH:** Congestive Heart Failure (EF 55%), Pulmonary Hypertension, Hypertension, Renal Cell Carcinoma s/p right partial nephrectomy (stable), Thalassemia s/p splenectomy
- **PSH:** Partial right nephrectomy, left ureteroscopy with left stent insertion, right heart catheterization, splenectomy
- **Vitals:** BP - 104/68, HR - 88, RR - 28
- **Labs:** Hb - 13.1, WBC - 17.7, Troponin - 9, BNP - 196

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant: 1 Dyspnea due to suspected valvular heart disease. Ischemia excluded. Initial imaging.

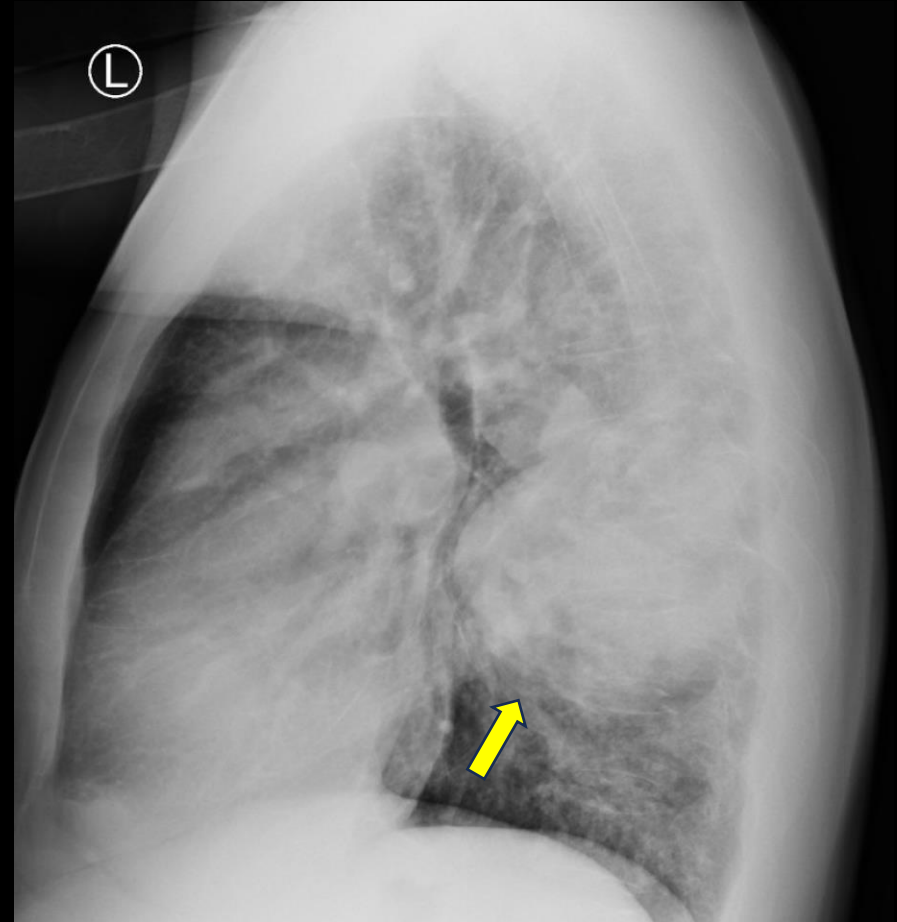
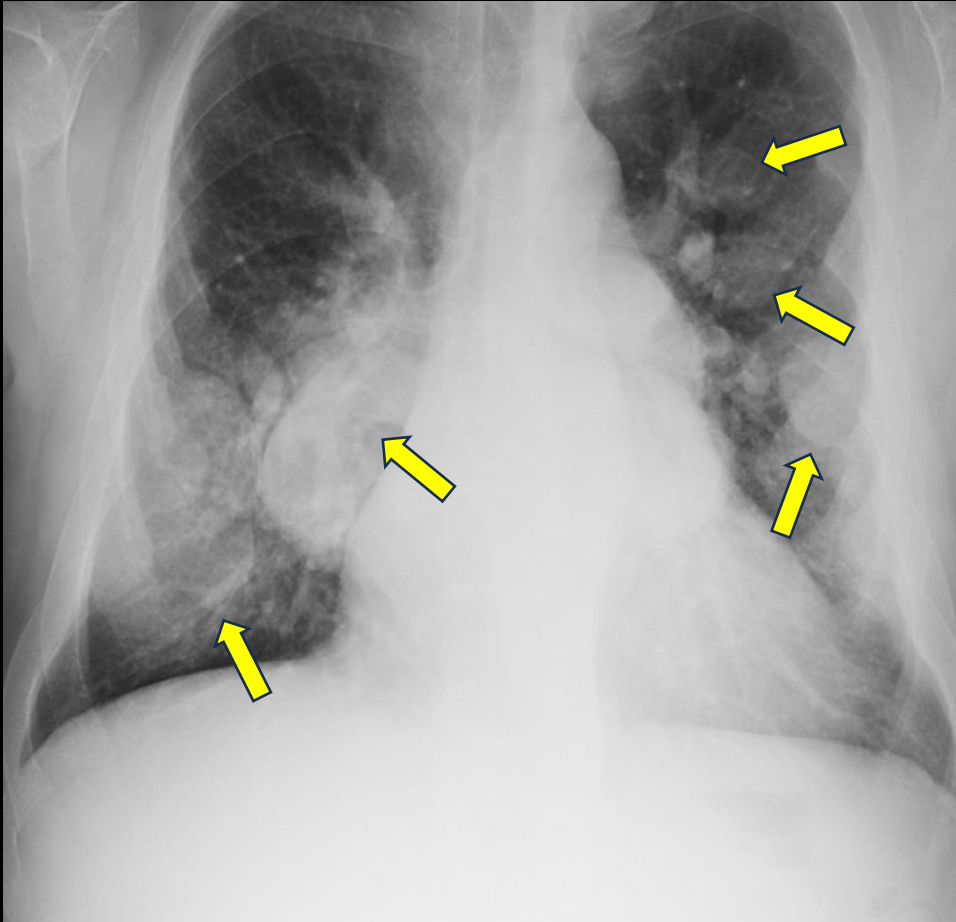
Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	○
Radiography chest	Usually Appropriate	☢
US echocardiography transesophageal	May Be Appropriate	○
US echocardiography transthoracic stress	May Be Appropriate	○
MRI heart function and morphology without and with IV contrast	May Be Appropriate	○
MRI heart function and morphology without IV contrast	May Be Appropriate	○
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	○
MRI heart function with stress without IV contrast	Usually Not Appropriate	○
CT coronary calcium	Usually Not Appropriate	☢☢☢
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
Rb-82 PET/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢

This imaging modality was ordered by the ER physician

Findings (unlabeled)



Findings (labeled)



Bilateral chest wall and posterior mediastinal masses.

What Imaging Should We Order?

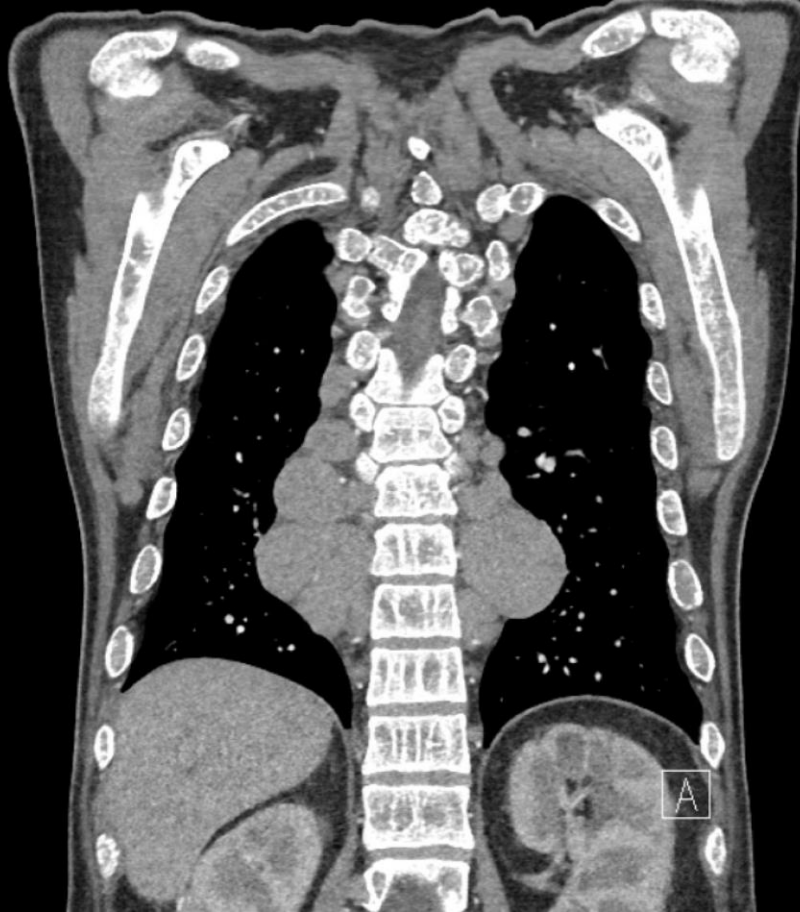
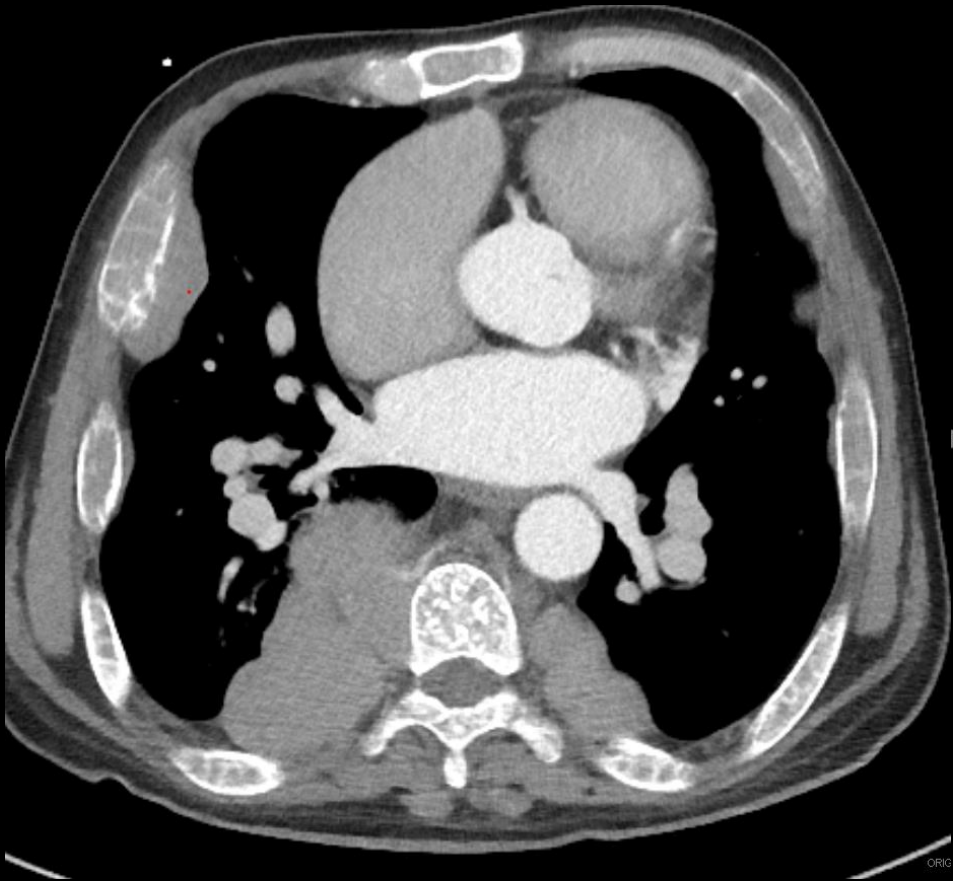
ACR Appropriateness Criteria

Variant: 2 Indeterminate mediastinal mass on radiography. Next imaging study.

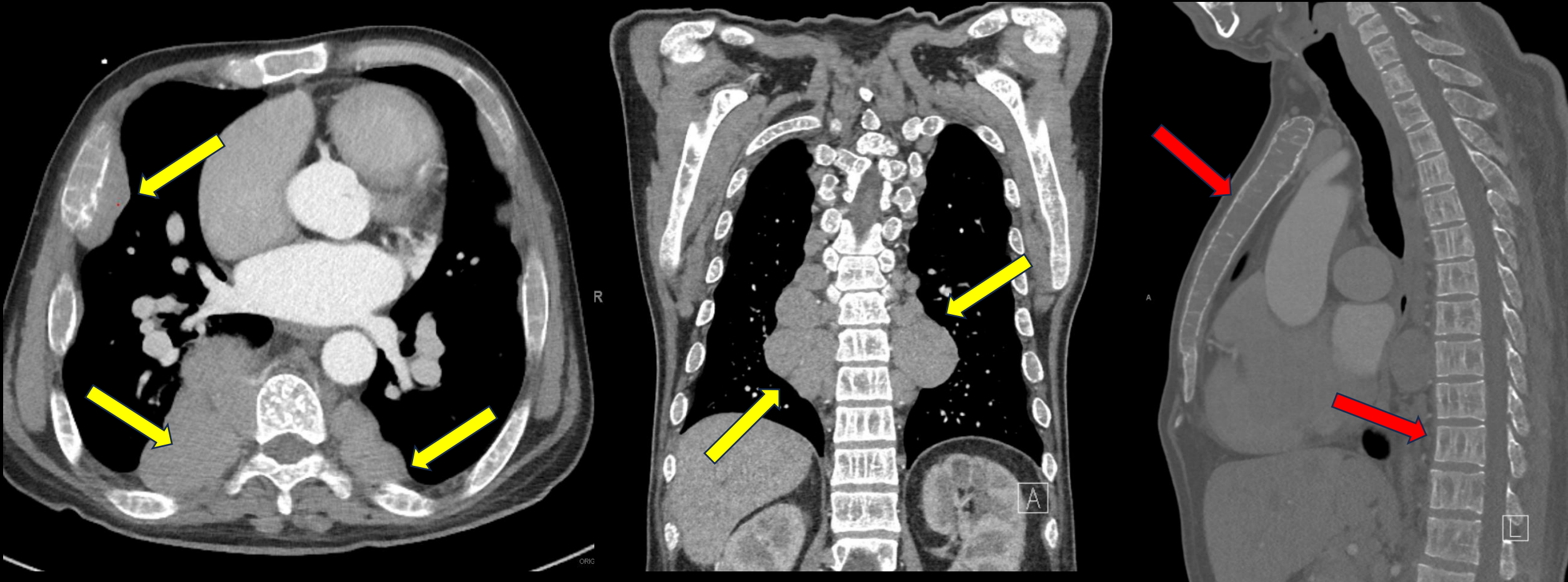
Procedure	Appropriateness Category	Relative Radiation Level
MRI chest without and with IV contrast	Usually Appropriate	○
MRI chest without IV contrast	Usually Appropriate	○
CT chest with IV contrast	Usually Appropriate	☢☢☢
CT chest without IV contrast	Usually Appropriate	☢☢☢
US chest	Usually Not Appropriate	○
Image-guided transthoracic needle biopsy	Usually Not Appropriate	Varies
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢

This imaging modality was ordered by the physician

Findings (unlabeled)



Findings (labeled)



Diffuse abnormalities of the thoracic skeleton with expansion of the bones and increased trabeculation. Multiple periosteal soft tissue masses adjacent to the ribs and thoracic vertebral bodies measuring up to 11.6cm.

Final Dx

Extramedullary Hematopoiesis due to Chronic
Thalassemia

Case Discussion

Extramedullary hematopoiesis (EMH) is compensatory blood cell production outside the bone marrow. In thalassemia, chronic anemia and ineffective erythropoiesis drive marrow expansion and EMH.²

- **Epidemiology⁵**

- EMH is most often seen in non-transfusion-dependent thalassemia or undertreated transfusion-dependent disease. Common sites include the liver, spleen, and paraspinal regions.

- **Pathophysiology^{1,2}**

- Reduced globin chain synthesis causes chronic microcytic hemolytic anemia.
- Ineffective erythropoiesis leads to erythroid hyperplasia and marrow expansion.
- Persistent erythropoietin stimulation promotes hematopoiesis outside marrow.
- Paraspinal EMH may arise from marrow extrusion through vertebral trabeculae or embryologic hematopoietic remnants.

Case Discussion

- **Radiographic features**^{3,4}
 - Location: Classically thoracic paraspinal, may be bilateral
 - X-ray: Well-demarcated paraspinal masses, widened ribs, coarse trabeculation, thickened calvarium
 - CT: Smooth paravertebral soft-tissue masses, usually without bony destruction
 - MRI: Modality of choice, especially for spinal canal compression
 - Chronic lesions may show fatty change, fibrosis, or iron deposition causing variable signal

Case Discussion

- **Management³**

- Observation if asymptomatic; transfusion and hydroxyurea to suppress EMH; radiotherapy or surgical decompression if symptomatic or cord compression is present.

- **Prognosis^{1,3}**

- Good with early recognition, but delayed diagnosis can cause irreversible neurologic injury. Follow-up MRI and hematology management are recommended.

References

- 1) Haidar R, Mhaidli H, Taher AT. Paraspinal extramedullary hematopoiesis in patients with thalassemia intermedia. Eur Spine J. 2010 Jun;19(6):871-8. doi: 10.1007/s00586-010-1357-2. Epub 2010 Mar 5. PMID: 20204423; PMCID: PMC2899982.
- 2) Kalchiem-Dekel O, Greenbaum U. Extramedullary Hematopoiesis in β -Thalassemia. Mayo Clin Proc. 2015 Nov;90(11):1591-2. doi: 10.1016/j.mayocp.2015.07.031. PMID: 26541258.
- 3) Radswiki T, Weerakkody Y, Silverstone L, et al. Extramedullary hematopoiesis. Reference article, Radiopaedia.org (Accessed on 30 Jun 2026) <https://doi.org/10.53347/rID-12220>
- 4) Roberts AS, Shetty AS, Mellnick VM, Pickhardt PJ, Bhalla S, Menias CO. Extramedullary haematopoiesis: radiological imaging features. Clin Radiol. 2016 Sep;71(9):807-14. doi: 10.1016/j.crad.2016.05.014. Epub 2016 Jul 1. PMID: 27377325.
- 5) Taher AT, Musallam KM, Cappellini MD. Guidelines for the Management of Non-Transfusion-Dependent β -Thalassaemia [Internet]. 3rd edition. Nicosia (Cyprus): Thalassaemia International Federation; 2023. Chapter 9, EXTRAMEDULLARY HAEMATOPHOIESIS. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK599494>