AMSER Case of the Month
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65-year-old man with worsening dyspnea on exertion

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Patient Presentation

• 65-year-old man with transthyretin amyloid cardiomyopathy was transferred to the cardiac care unit from an outside hospital with 3 weeks of worsening dyspnea on exertion and orthopnea

• Past medical history:
  • Transthyretin amyloid cardiomyopathy (2018)
  • Interstitial lung disease per outside hospital records

• Social history: denies tobacco/alcohol/drugs

• Family history: son and brother with cardiomyopathies
Pertinent Labs

- **PE:** Neck – JVP to ear; Cardiac – irregularly irregular rhythm, distant heart sounds; Lungs – wheezes on inspiration and expiration; Extremities – warm, 2+ pulses, 1+ edema
- **Vitals:** T 36.0 °C   HR 87 /min   BP 84/70 mmHg   RR 25/min   Oxygen Sat 99%
- **Labs:**
  - WBC 13.10
  - BNP 3,500 (baseline 700)
  - Troponin 0.2

• **Transthoracic Echo:**
  - LVEF: 10-15%
  - Dilated four chambers
  - Moderate to severe mitral regurgitation, posteriorly directed eccentric jet
  - Mild aortic, tricuspid and pulmonic regurgitation

What Imaging Should We Order?
### ACR Appropriateness Criteria for Dyspnea with Suspected Cardiac Origin

#### Variant 2:

**Dyspnea due to suspected nonischemic heart failure. Ischemia excluded.**

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<td>X-ray chest</td>
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<td>US echocardiography transthoracic resting</td>
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<td>MRI heart function and morphology without and with IV contrast</td>
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<td>US echocardiography transthoracic stress</td>
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<td>MRI heart with function and inotropic stress without and with IV contrast</td>
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*This imaging modality was ordered by the physician*
Findings (unlabeled)
Findings (labeled)

Bilateral bronchioloalveolar opacification

Pulmonary venous cephalization (Stag’s antler sign)

Moderate cardiomegaly

Trace pleural effusion
Findings (unlabeled)
Findings (labeled)

- Ground-glass opacity in right upper lobe
Final Diagnosis:

Unilateral pulmonary edema secondary to mitral regurgitation

Repeat CT following diuresis 2 weeks later
Differential Diagnoses:

- Infectious process
  - Organizing pneumonia
- Chronic interstitial diseases
  - UIP (usual interstitial pneumonia)
  - NSIP (nonspecific interstitial pneumonia)
  - Sarcoidosis
- Acute alveolar diseases
  - Pulmonary edema
  - Hypersensitivity pneumonitis
  - ARDS
Unilateral Pulmonary Edema (UPE)

- Cardiogenic pulmonary edema on chest radiograph:
  - Up to 20% of patients may have normal CXR
  - Mild pulmonary vascular redistribution
  - Bilateral perihilar alveolar edema
  - Cardiomegaly and bilateral interstitial markings in severe cases

- In 2% of cases, unilateral pulmonary edema is present and is caused by eccentric mitral regurgitation (MR)
  - Predilection for the right upper lobe
  - Related to lateralized direction of MR: posterior leaflet prolapse is more likely to be associated with right-sided UPE
  - Associated with delay in initiation of appropriate treatment
  - Independent increased risk of mortality
Pathophysiology of UPE

• The anatomy of the pulmonary veins in relation to the mitral valve may explain the predilection for the right upper lobe
  • Eccentric regurgitant jet (often posteriorly)

• Positioning of the patient

• Left-sided cardiac enlargement can physically impede blood flow in left pulmonary artery

• Differences in lymphatic draining capacity of the right and left lung

Image credit: Myrianthefs et al 2011
References:


