AMSER Case of the Month
July 2026

46-year-old female w/ progressively worsening cough & intermittent pneumonia

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HENRY FORD HEALTH

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Patient Presentation

- HPI: 46-year-old female with PMH of asthma, severe microcytic anemia 2/2 uterine fibroids, and NSTEMI (10/2024), who presented to ED on 12/14/25 for pneumonia (PNA)
  - Past 12 months:
    - H/O chronic progressively worsening cough post-COVID in Nov 2024
    - Treated for reactive airways disease & intermittent PNA with persistent symptoms
- SH: Never smoked; office worker
- Prior Imaging:
  - CXR 8/25 & 10/25 with B/L reticular infiltrates (R>L)
  - CT 10/25 B/L diffuse ground glass with predominantly right sided interlobular septal thickening & L>R AVM
- Current course – presented to HFH following hospitalization for PNA
  - Vitals stable, SpO_2 100% on O_2 no resp distress

Pertinent Labs

- Inflammatory markers:
  - LDH= 283 IU/L (normal<250)
  - CRP= 1.6 mg/dL (normal<0.5)
  - ESR= 126 mm/Hr (normal<20)
- Immunologic markers:
  - GM-CSF< 15pg/ml (normal <15)
    - Antibody test performed at outside hospital
  - IgG= 1,551 mg/dL (normal: 700-1600)-(Jan 2026)
- Hematologic labs:
  - Hemoglobin: 8.8-9.8 g/dL
  - Hct: 28.5-31.0%
  - Platelets and WBCs unremarkable
- Rest of the labs WNL

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Home Occupational Lung Diseases

Variants

1. **Occupational exposure, suspected interstitial lung disease. Initial imaging.**

Documents

Documents

Narrative

Evidence Table

Lit Search

Appendix

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Occupational exposure, interstitial lung disease suspected, initial imaging	3164918	Radiography chest	<0.1 mSv ☢	<0.03 mSv [ped] ☢	Usually appropriate
		CT chest without IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually appropriate
		MRI chest without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		MRI chest without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		CT chest with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		CT chest without and with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		FDG-PET/CT skull base to mid-thigh	10-30 mSv ☢☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate

This imaging modality was ordered by the ER physician

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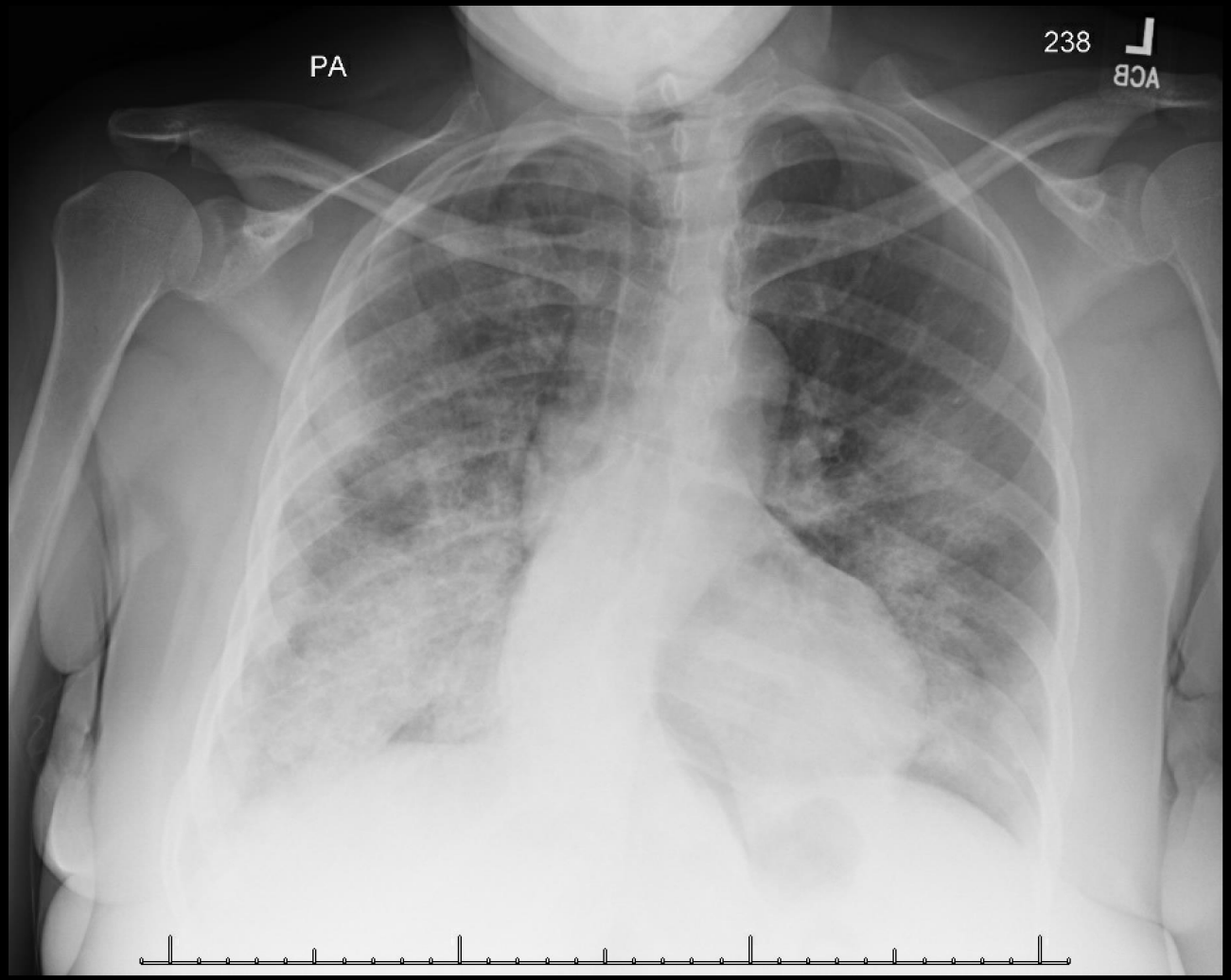
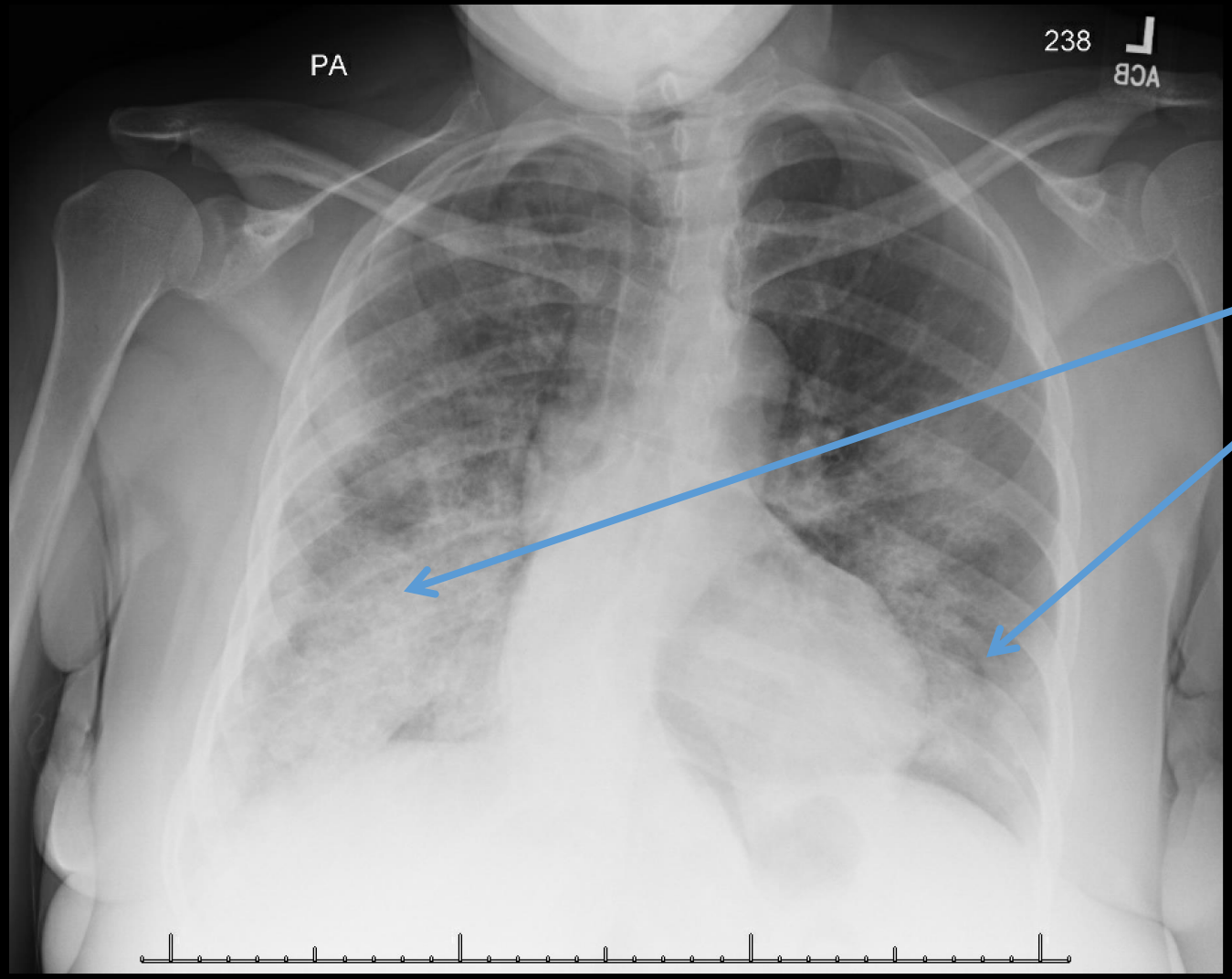
Findings (unlabeled)- CXR

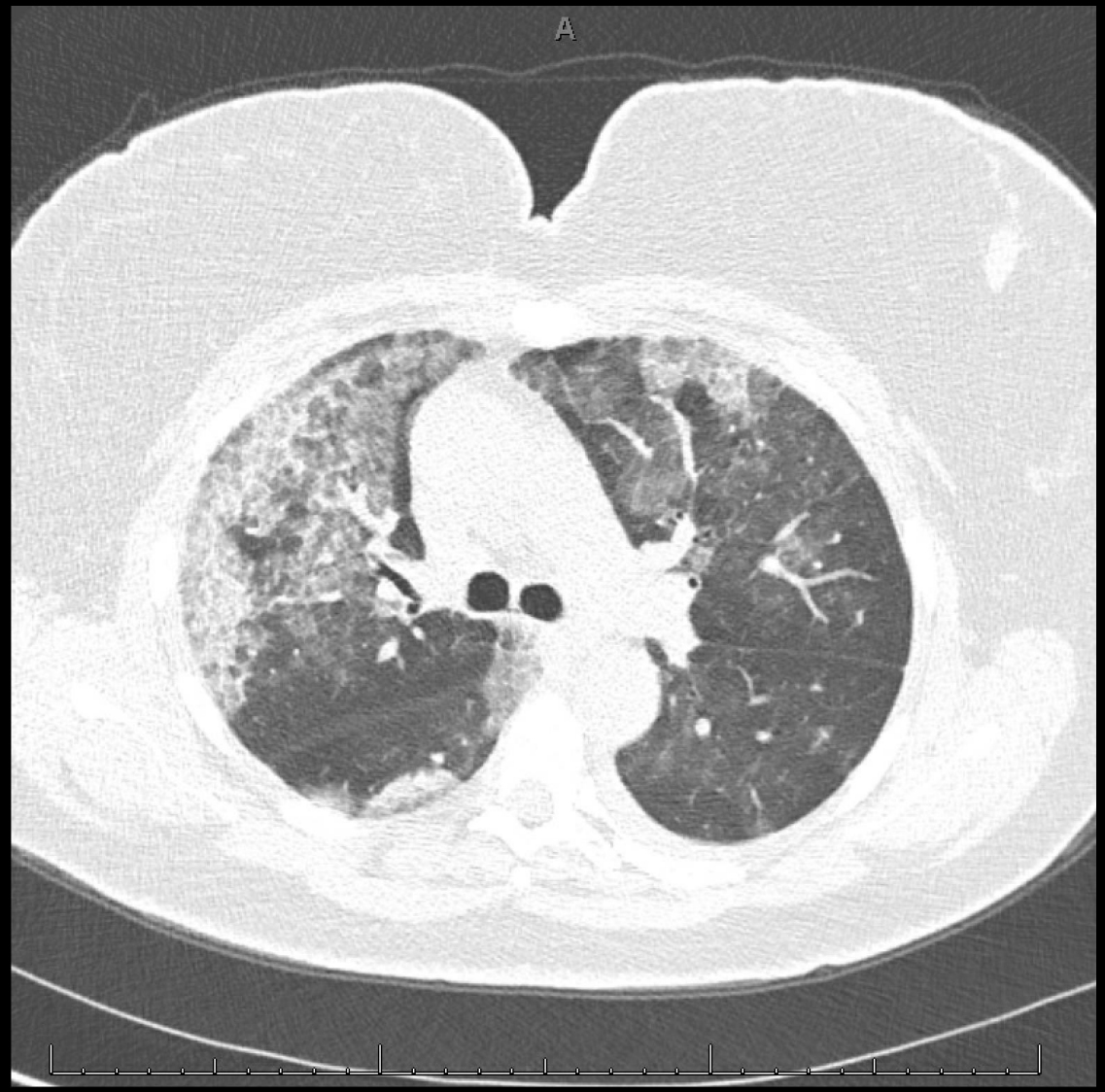


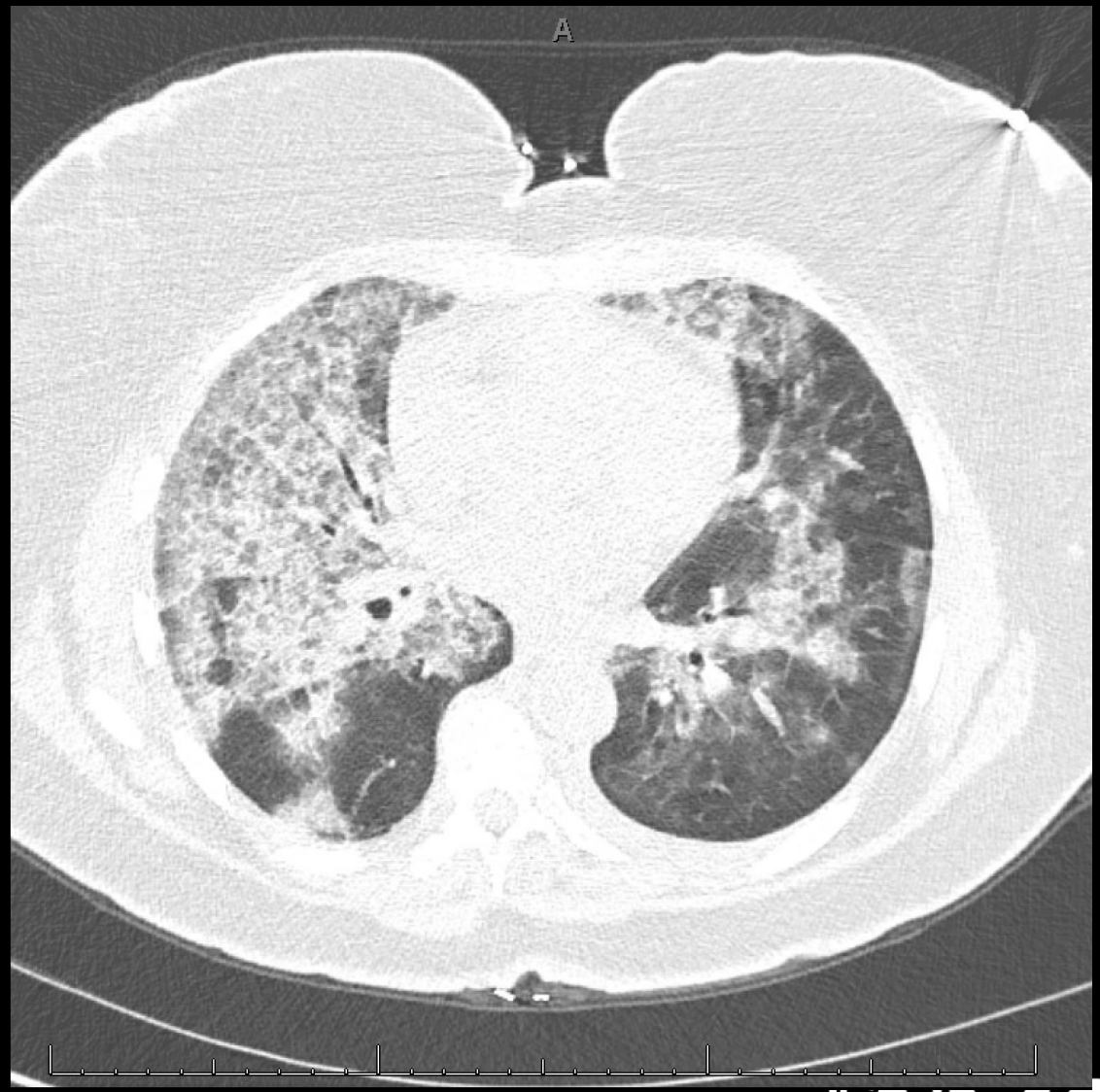
Findings (labeled)- CXR



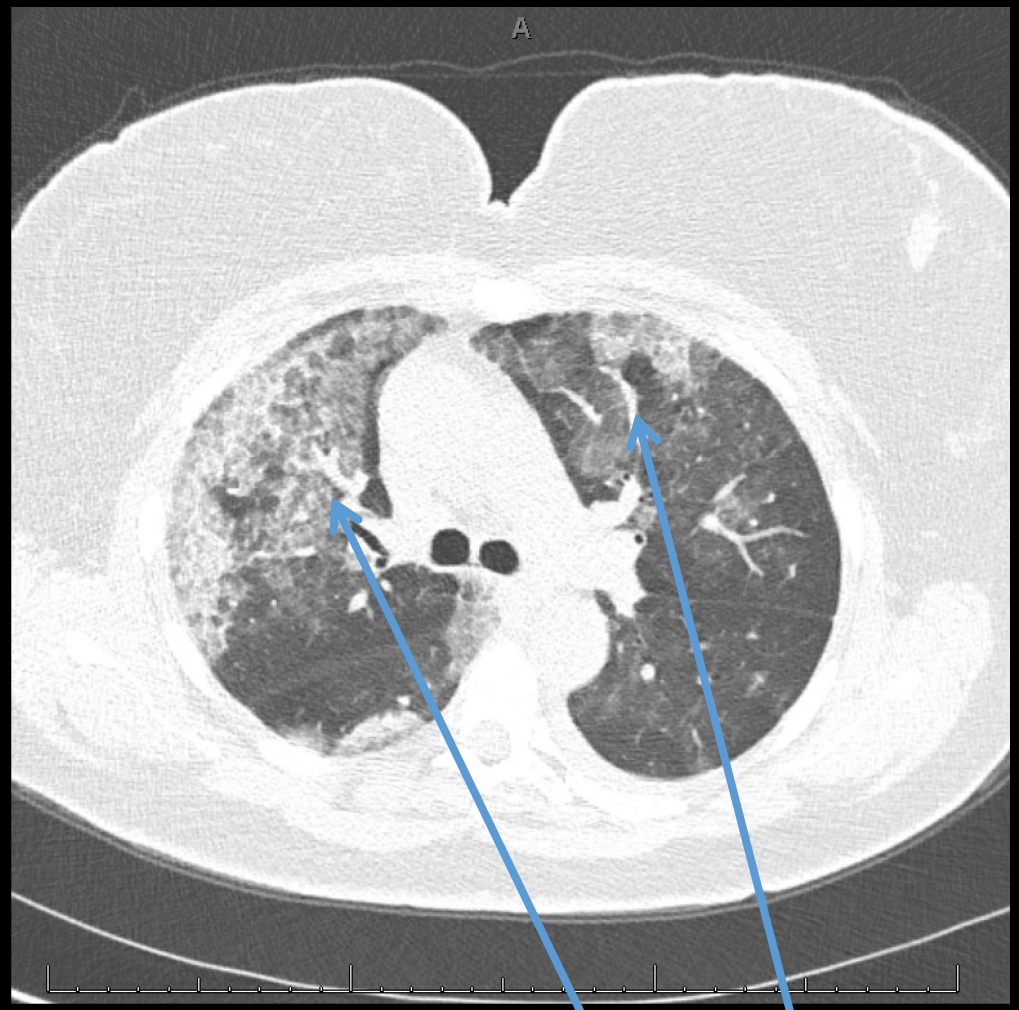
Diffuse B/L airspace suggestive of multifocal pneumonia or edema R>L.
Note: relative apical sparing.

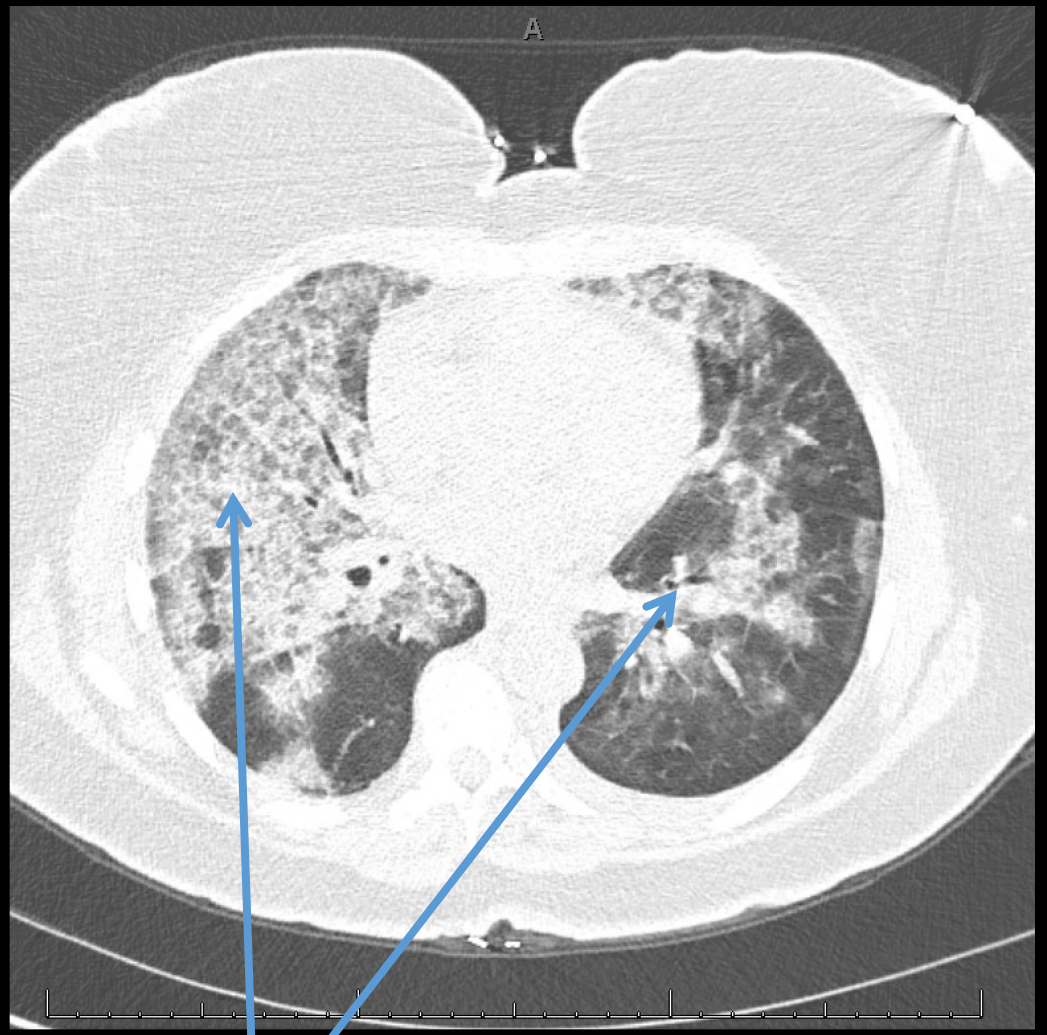
Findings (unlabeled)- CT Chest w/o IV contrast





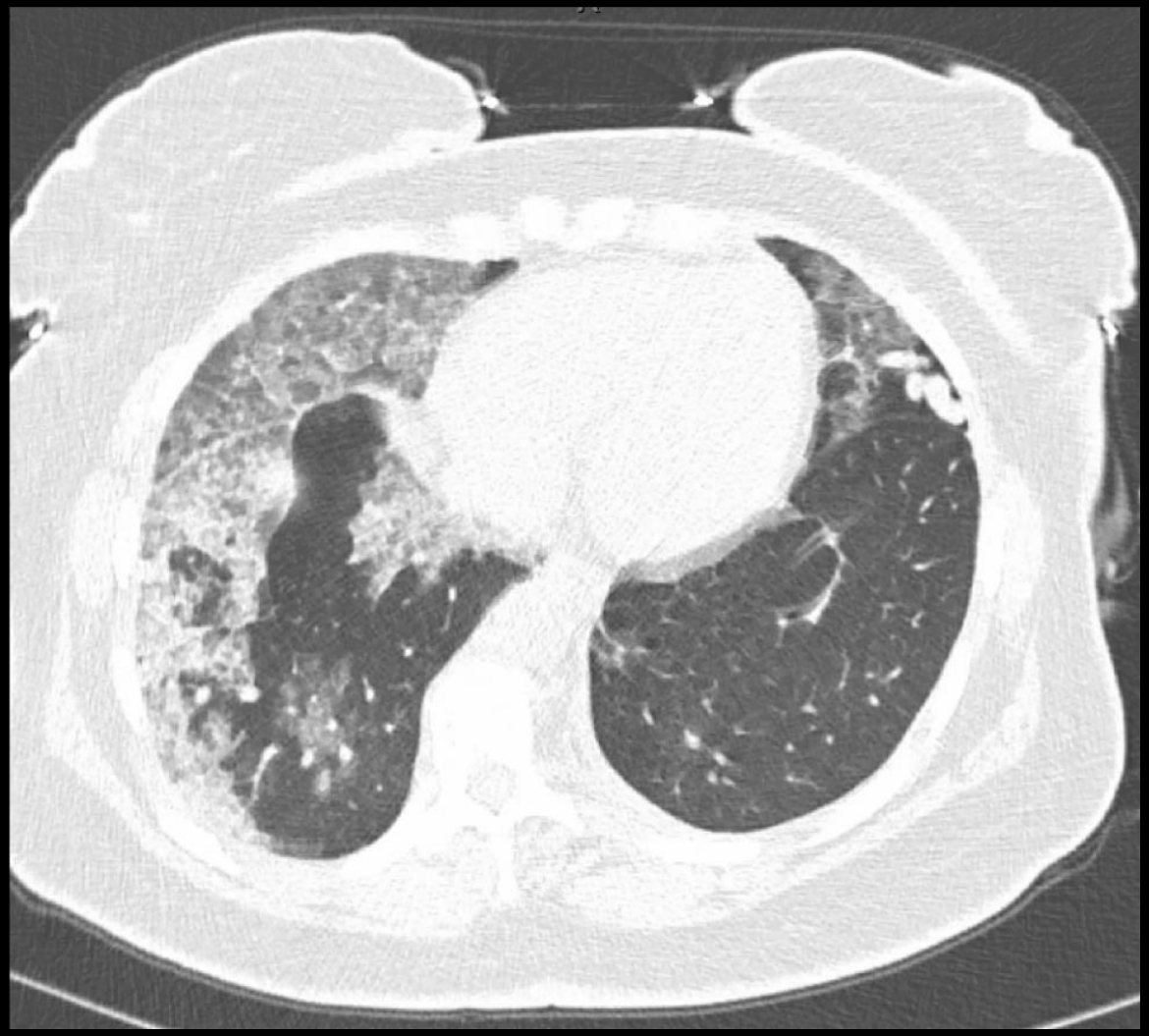
Findings (labeled)- CT Chest w/o IV contrast

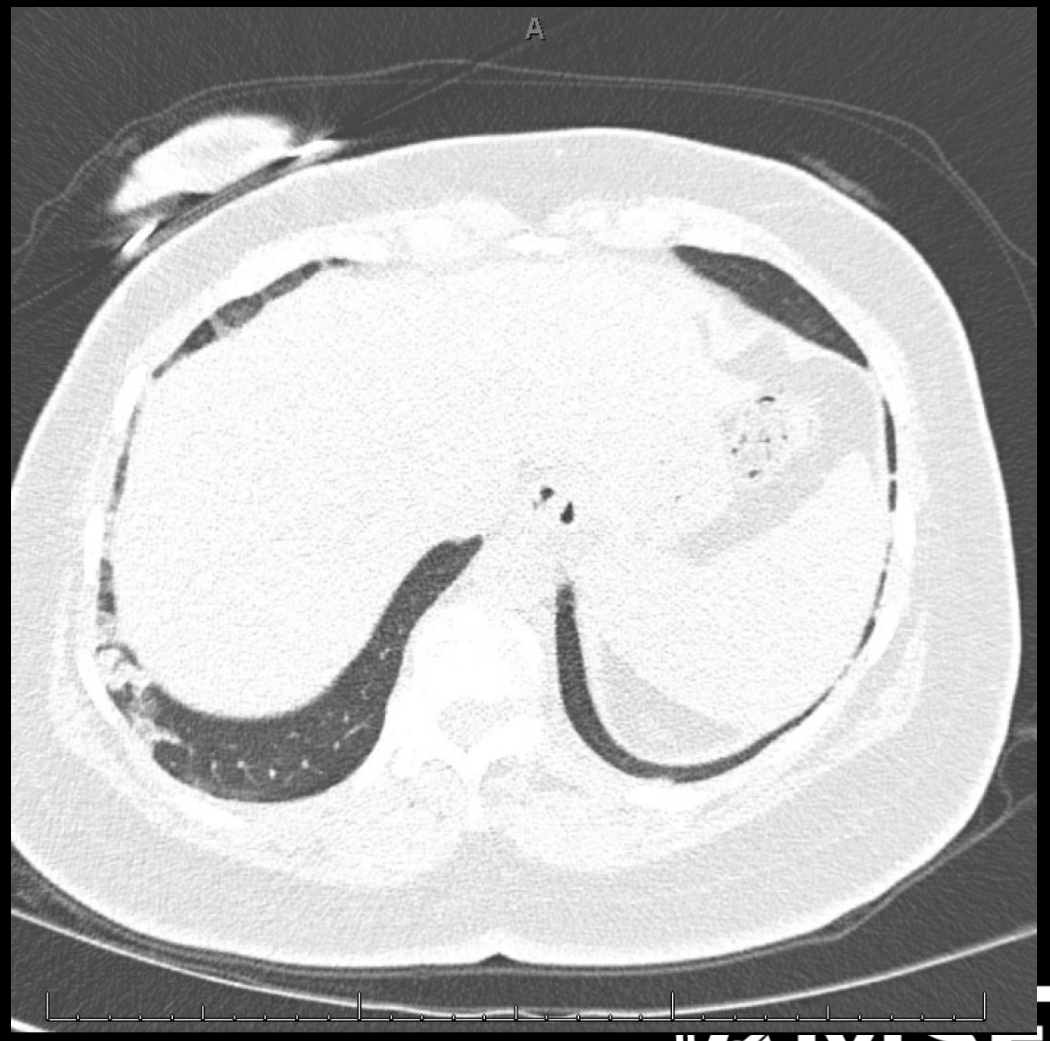




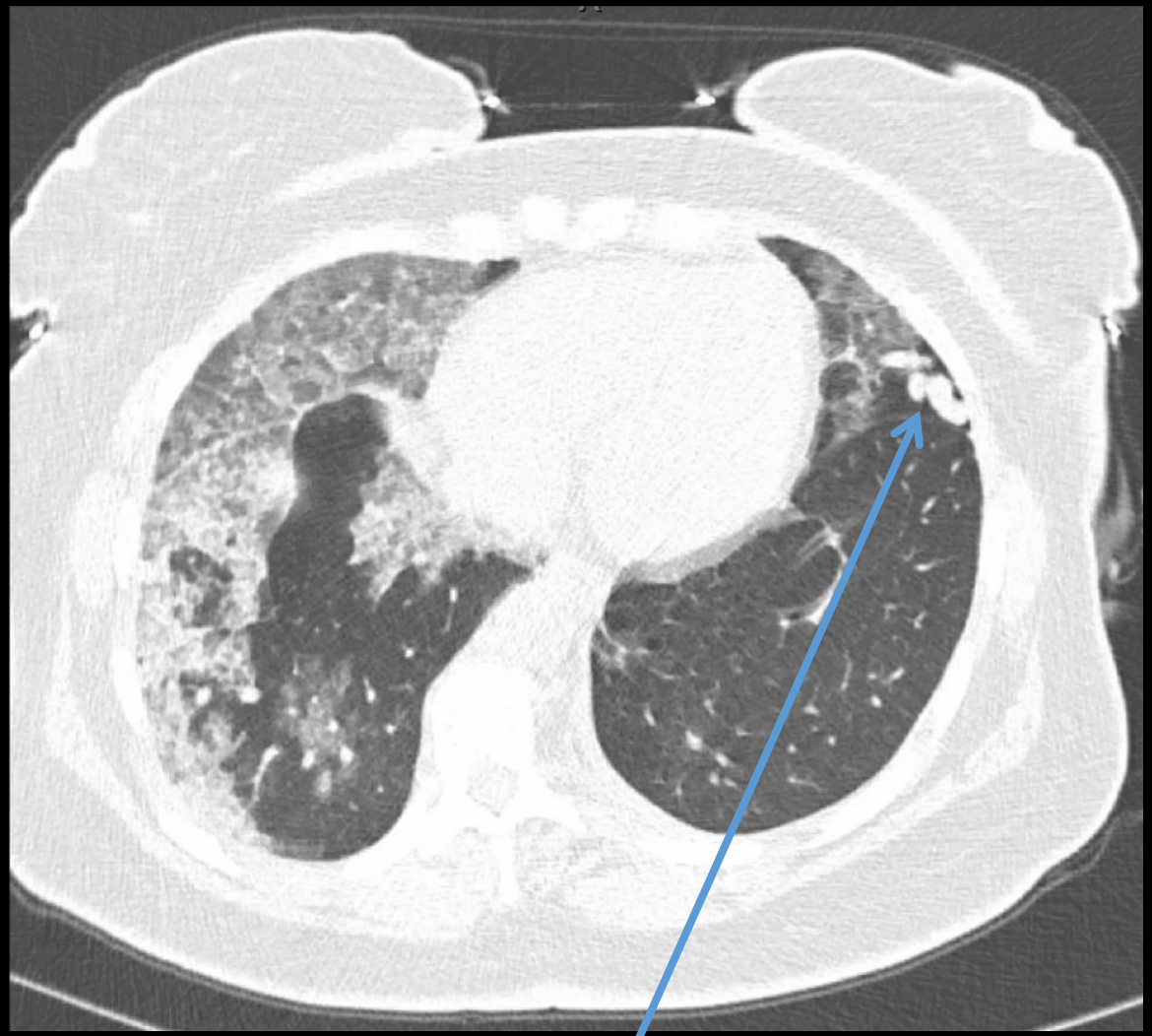
Multiple ground glass opacities with anterior & intralobular septal thickening

Findings (unlabeled)- CT Chest w/o IV contrast (continued)

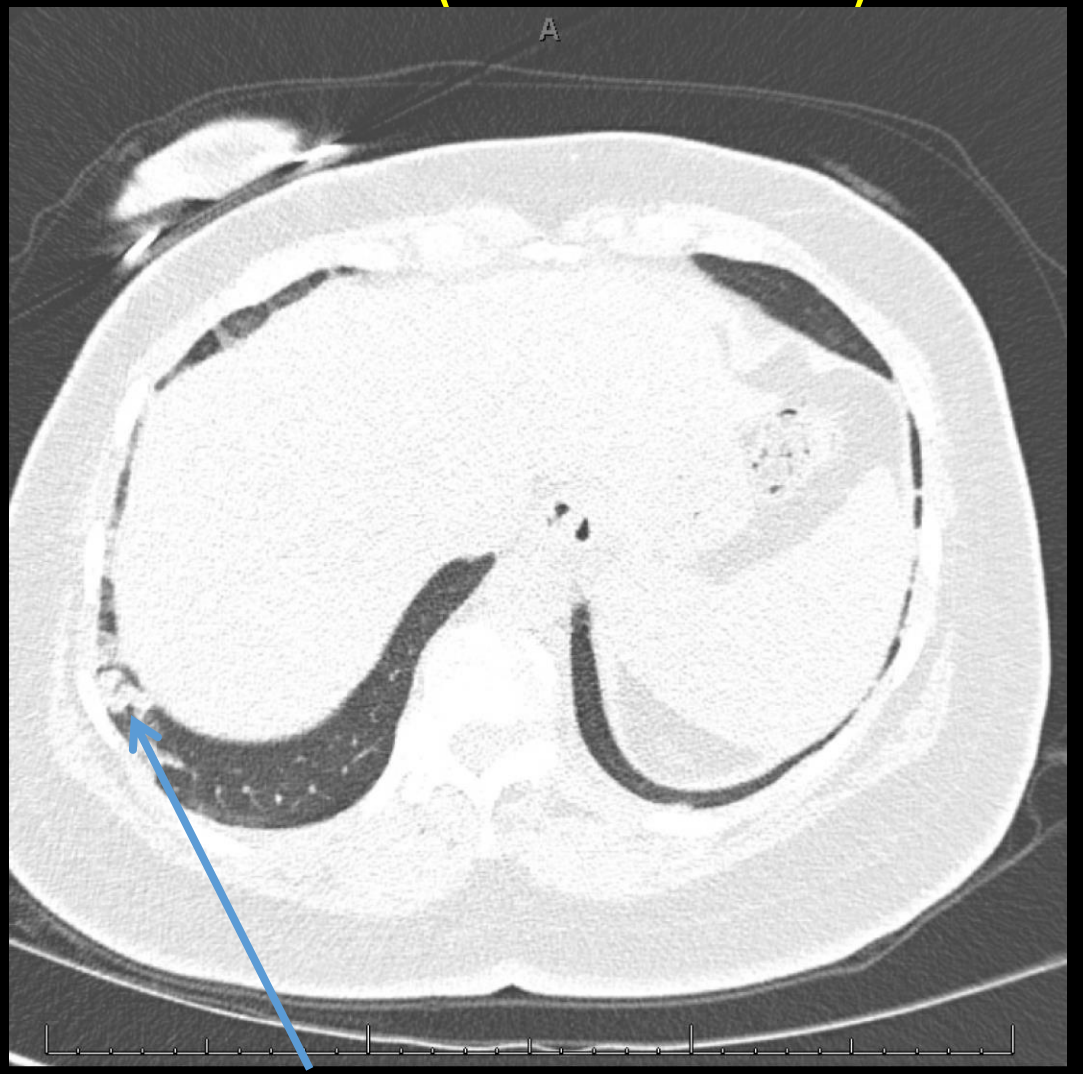




Findings (labeled)- CT Chest w/o IV contrast (continued)



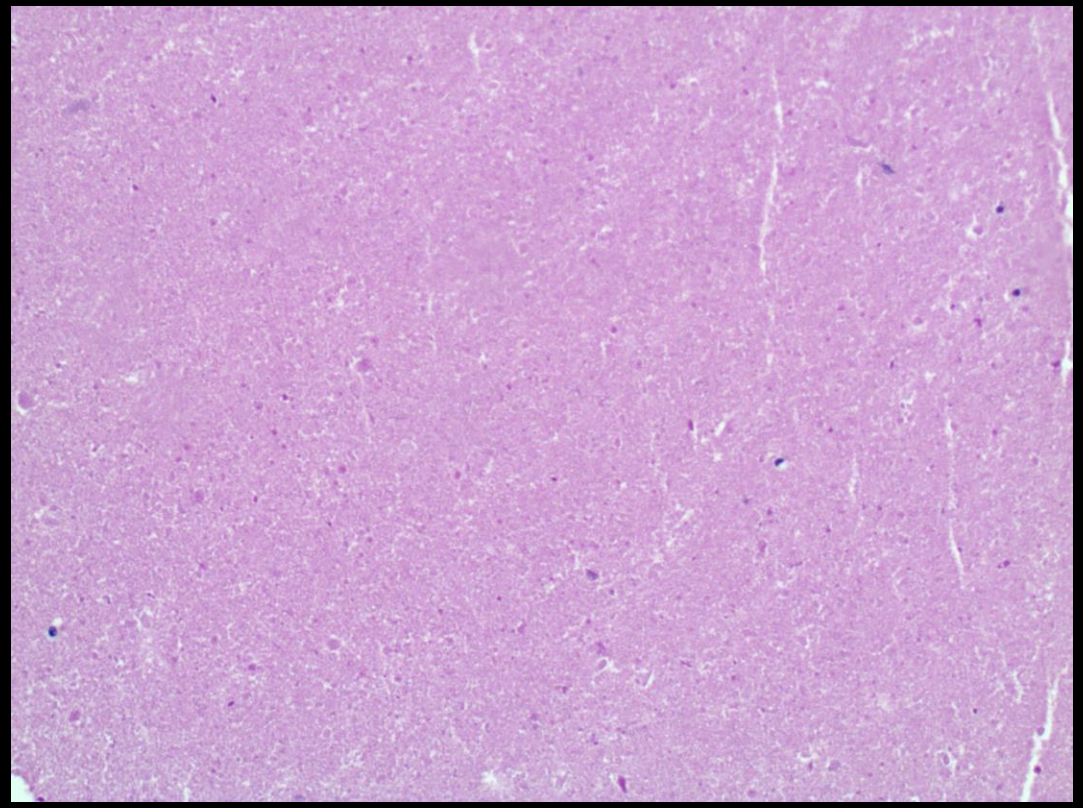
Pulmonary arteriovenous malformation, Feeding artery 5mm in diameter
-Location: Inferior Lingula



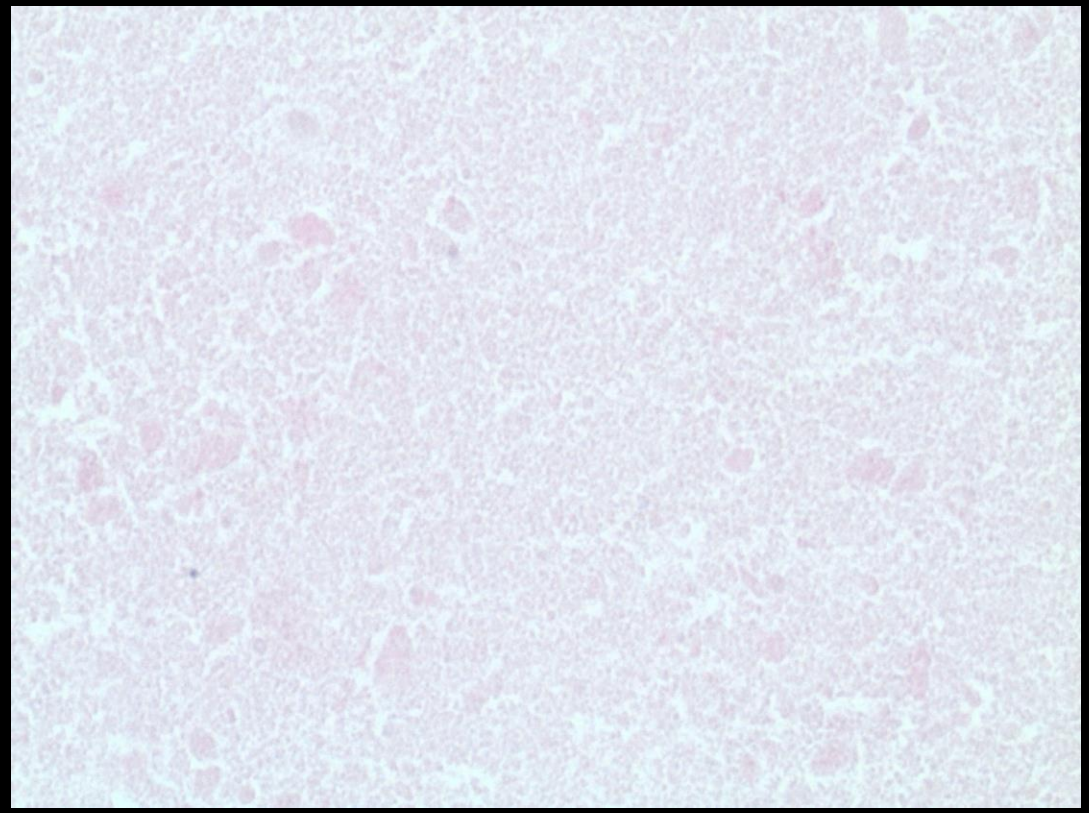
Pulmonary arteriovenous malformation, Feeding artery 4mm in diameter
-Location: Lateral basilar RLL

Pathology

- Bronchoalveolar Lavage findings:
  - Negative for malignant cells
  - PAS stains positive for proteinaceous material
    - Suggestive of pulmonary alveolar proteinosis (PAP)
- PAS Stain:



H&E stain:



Final Dx:

Pulmonary Alveolar Proteinosis (PAP) with Pulmonary AVM

Case Discussion

- PAP characterized by progressive accumulation of alveolar surfactant & dysfunction of alveolar macrophages.[1]
  - Result:
    - Progressive dyspnea of insidious onset
    - Hypoxemic respiratory failure
    - Secondary infections
    - Pulmonary fibrosis
- Epidemiology
  - Prevalence: rare (7 per million individuals in US/Japan)[1]
    - Underestimated due to underdiagnosis
  - Risk factors:
    - Age: most common in young-middle aged adults, 20-50yrs[2]
    - Smoking
    - Male:female ratio= 2:1

Case Discussion

- Classifications of PAP
  - **Primary PAP:** Disruption of granulocyte–macrophage colony-stimulating factor (GM-CSF) signaling[3], subdivided into:
    - **Autoimmune PAP:** Caused by GM-CSF autoantibodies (accounts for >90% of cases)
    - **Hereditary PAP:** Due to mutations in GM-CSF receptor genes (CSF2RA or CSF2RB)
  - **Secondary PAP:** Results from diseases, exposures, or drugs that reduce the number/function of alveolar macrophages[1]
    - Examples: hematological disorders, malignancies, immune deficiencies, toxic inhalation
  - **Congenital PAP:** Caused by mutations affecting surfactant production[1]
    - SFTPB & SFTPC (genes encoding pulmonary surfactant-associated protein B)
    - Other genes: ABCA3, NKX2-1

Case Discussion

- Diagnostic Workup
  - Radiographic Features:
    - CXR: Bilateral symmetric airspace opacities, usually in mid and lower lung field zones[4]
      - Opacities can demonstrate "bat-wing" pattern with relative sparing of apices and costophrenic angles (represents accumulation of proteinaceous material in alveoli)[4]
      - CXR has lower sensitivity and specificity (requires further imaging)
    - CT: Ground glass opacities w/ interlobular septal thickening= crazy paving pattern[4]
  - Labs: usually normal and/or nonspecific (elevated inflammatory markers-LDH)
  - Bronchoalveolar Lavage:
    - Standard of care for symptomatic primary PAP[5]; physically removes excess surfactant
    - +BAL or serum anti-GM-CSF antibodies
- Management
  - **GM-CSF therapy:** Inhaled GM-CSF is promising for autoimmune PAP[1,3,4,5]; subcutaneous GM-CSF also available (but does not directly target lungs)
    - Inhalational GM-CSF therapy leads to greater deposition of GM-CSF in alveoli (higher response rates and improvements in A-a gradient and PaO2 compared to subcutaneous GM-CSF)[3]
  - **Other therapies:** Plasmapheresis, rituximab (B-cell depletion)[1,3,4,5], and statins[1,3,4,5] (targeting cholesterol homeostasis) are under investigation
  - **Lung transplantation:** Considered for end-stage or refractory cases

References:

1. Trapnell, B.C., Nakata, K., Bonella, F. *et al.* Pulmonary alveolar proteinosis. *Nat Rev Dis Primers* **5**, 16 (2019). https://doi.org/10.1038/s41572-019-0066-3
2. Weerakkody Y, Sharma R, Silverstone L, et al. Pulmonary alveolar proteinosis. Reference article, Radiopaedia.org (Accessed on 20 May 2026) https://doi.org/10.53347/rID-8206
3. Wołoszczak J, Wrześniewska M, Hrapkowicz A, Janowska K, Szydziak J, Gomułka K. A Comprehensive Outlook on Pulmonary Alveolar Proteinosis-A Review. Int J Mol Sci. 2024 Jun 28;25(13):7092. doi: 10.3390/ijms25137092. PMID: 39000201; PMCID: PMC11241585.
4. Morton C, DeBiasi E. Pulmonary Alveolar Proteinosis. Clin Chest Med. 2025 Jun;46(2):373-382. doi: 10.1016/j.ccm.2025.02.014. Epub 2025 Mar 26. PMID: 40484510.
5. Bonella F, Borie R. Pulmonary Alveolar Proteinosis. Clin Chest Med. 2025 Dec;46(4):633-647. doi: 10.1016/j.ccm.2025.07.005. Epub 2025 Aug 25. PMID: 41110926.