AMSER Case of the Month

59-year-old male evaluation for lung transplant with history of COPD

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Patient Presentation

- HPI:
- 59-year-old male, ex-smoker (quit in 2000, 2 PPD) came in for evaluation for possible lung transplantation due to COPD (20 yr history) & bronchiectasis (diagnosed 04/2023).

- Uses supplemental O2 with activity & at night but not at rest. No hospitalizations for lung issues in 10+ yrs, never been intubated.

- Denies Right Heart Failure symptoms including syncope, leg edema.

Patient Presentation(cont.)

- He works full time as a steel mill inspector and would like to work in an office position soon.
- Had undergone multiple bronchoscopies revealing recurrent Pseudomonas. Managed with medications and vest therapy. Recently noted symptom improvement with inhaled tobramycin.
- Evaluated for cystic fibrosis 10 yrs ago with negative sweat chloride test. Has three children with no fertility issues.

Patient Presentation(cont)

- PMHx: Bronchiectasis/COPD, chronic Pseudomonas on BAL, OSA
- PSHx: Bronchoscopy
- Fam Hx: Father had lung disease with secretions (undiagnosed), and mother had lymphoma
- Medications: albuterol, azithromycin, tobramycin, aspirin, montelukast
- Vitals: BP: 146/83 Pulse: 98 Temp: 36.7C RR: 16 SpO2: 92% BMI: 27.45kg/m2

Pertinent Labs

- Spirometry:
  - -FVC: 3.15L (80%-mildly reduced)
  - -FEV1: 1.20L (severely reduced)
  - -FEV1/FVC: 0.38 (48%-severely reduced)

Lung Volume:

- -TLC: 7.64L (mild hyperinflation)
- -RV: 4.74L (severe air trapping)

- Diffusion Capacity:
  - -DLCO: 22.24ml/min/mmHg (mild reduction)
  - -VA: 4.84L (slightly reduced)
  - -DLCO/VA: 4.60 (normal)

Findings of moderate to severe COPD with significant air trapping and small airway disease. However, gas exchange remains relatively preserved.

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant 6: **Adult. Bronchiectasis. Assessment of complications or treatment response.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	☢
CT chest without IV contrast	Usually Appropriate	☢☢☢
CT chest with IV contrast	May Be Appropriate	☢☢☢
CTA chest with IV contrast	May Be Appropriate	☢☢☢
Radiography neck	Usually Not Appropriate	☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV contrast	Usually Not Appropriate	O
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT neck with IV contrast	Usually Not Appropriate	☢☢☢
CT neck without and with IV contrast	Usually Not Appropriate	☢☢☢
CT neck without IV contrast	Usually Not Appropriate	☢☢☢
CT neck and chest with IV contrast	Usually Not Appropriate	☢☢☢☢
CT neck and chest without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT neck and chest without IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢

This imaging modality was ordered initially.

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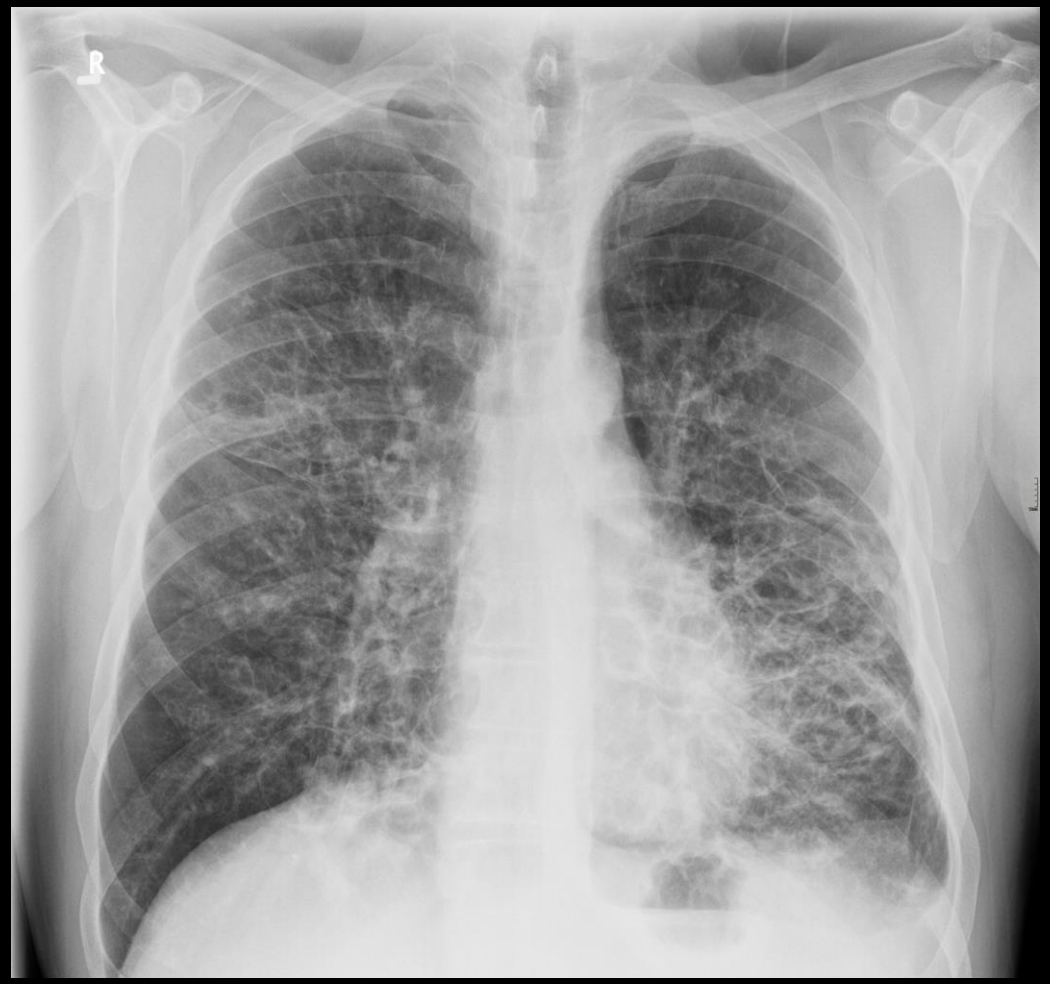
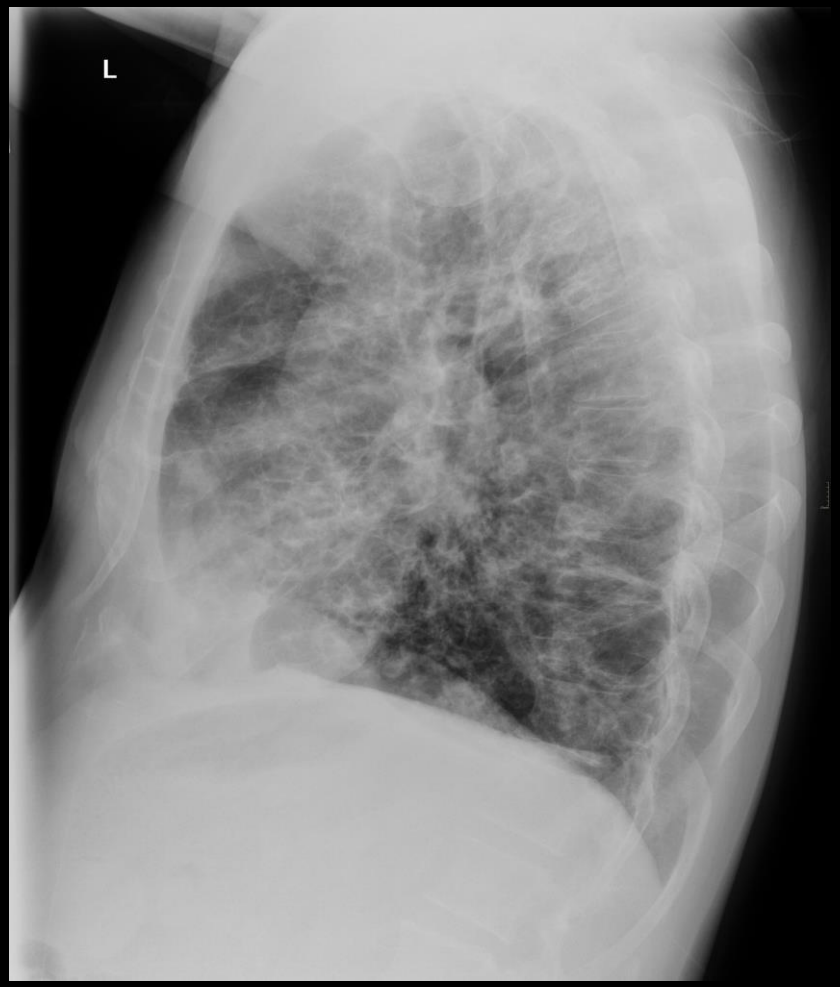
Findings: (unlabeled)

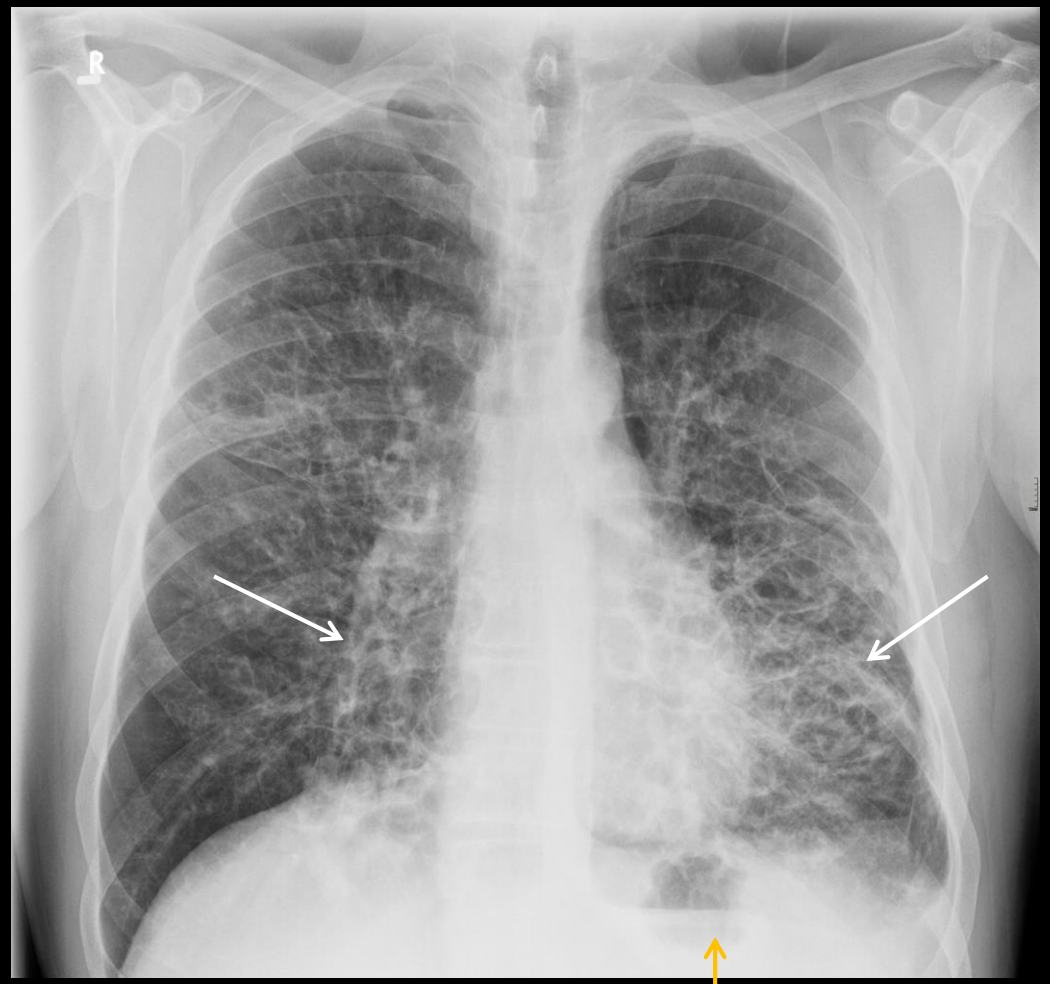




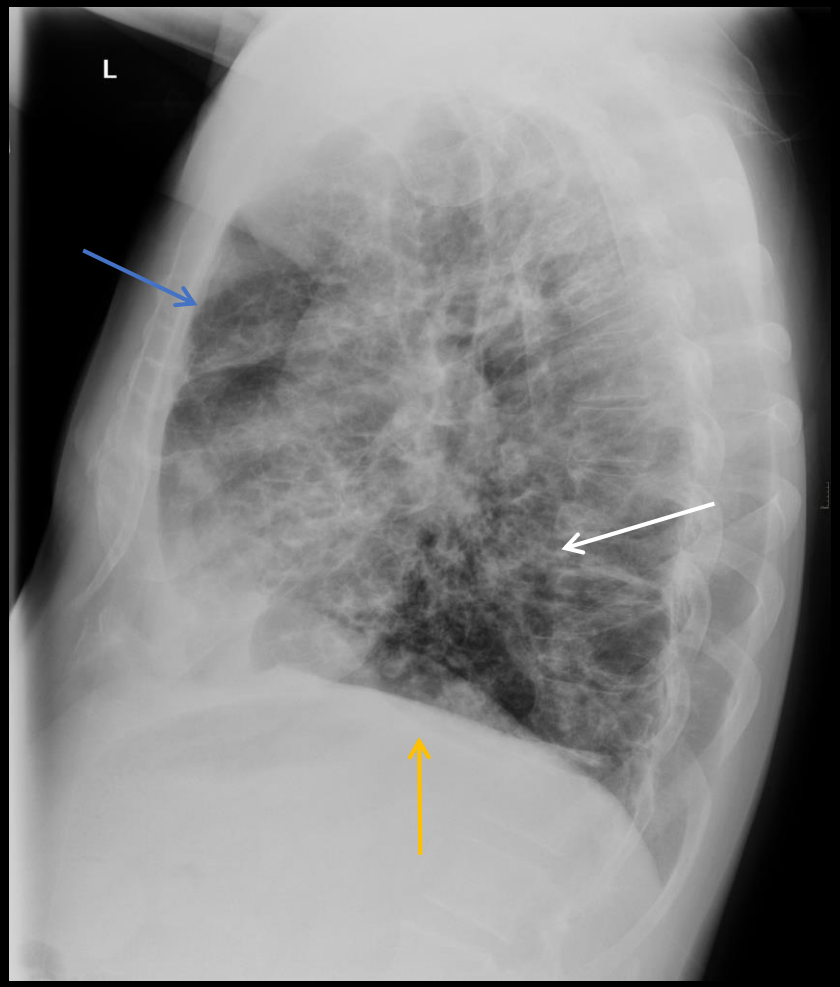
Chest X Ray: A) PA view B) Lateral View

Findings: (labeled)

A



B



Chest X Ray: A) PA view B) Lateral View: Hyperinflated lungs with flattened left hemidiaphragm (yellow arrows). Increased retrosternal clear space in figure B (blue arrow). Reticular pattern opacities indicating possible scarring, bronchiectasis, ILD (white arrows).

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FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢

This imaging modality was ordered next.

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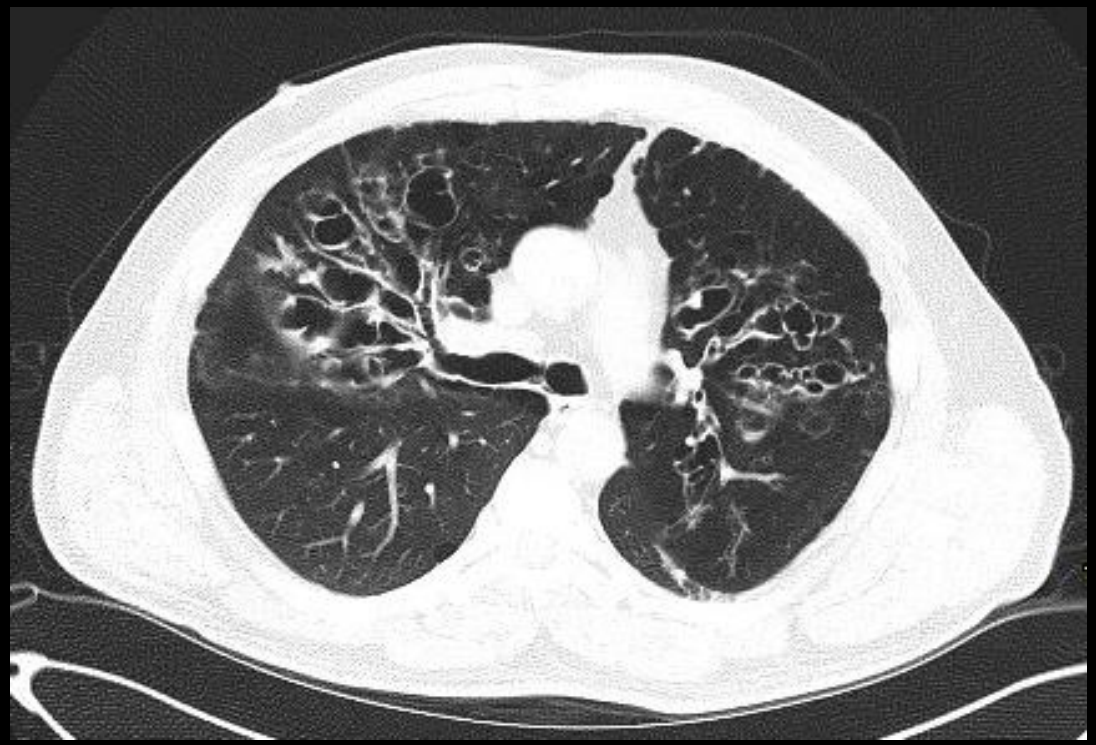
Findings: (unlabeled)

A



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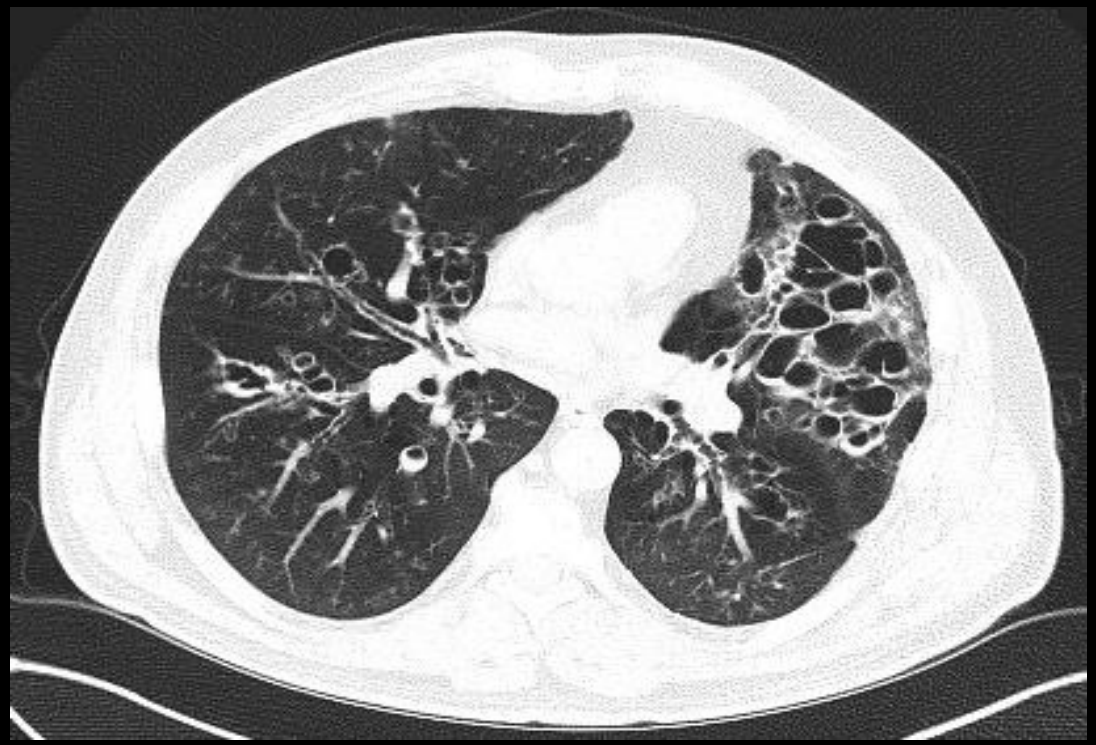
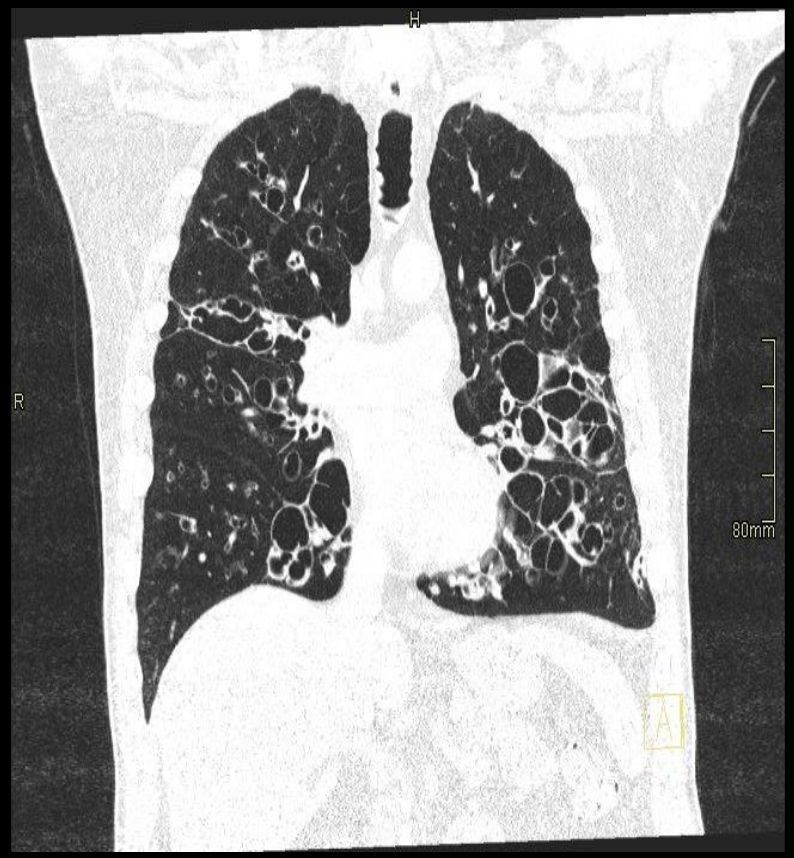
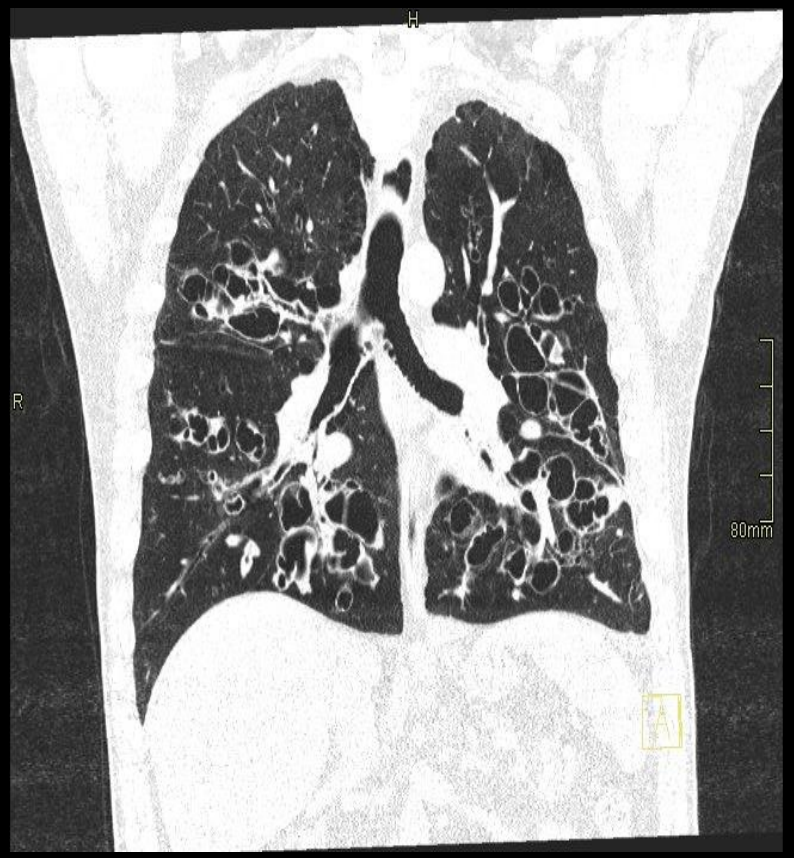


Chest CT without Contrast-Axial View

Findings (unlabeled)

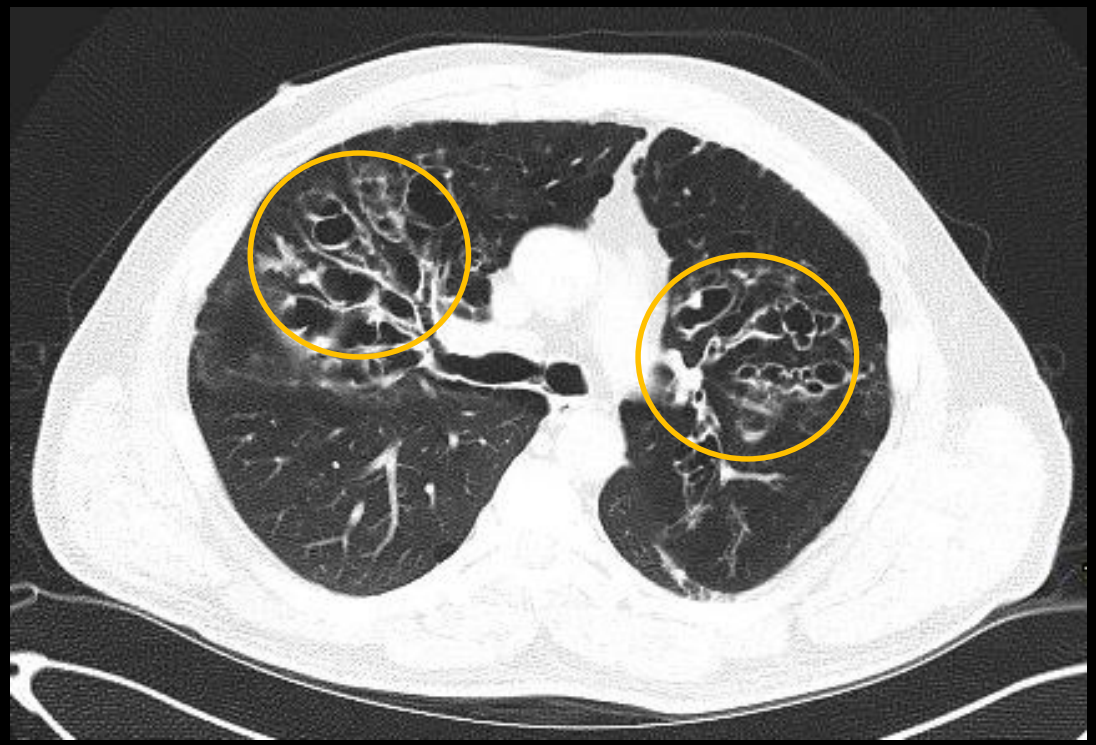




Chest CT without Contrast-Coronal View

Findings: (labeled)

A



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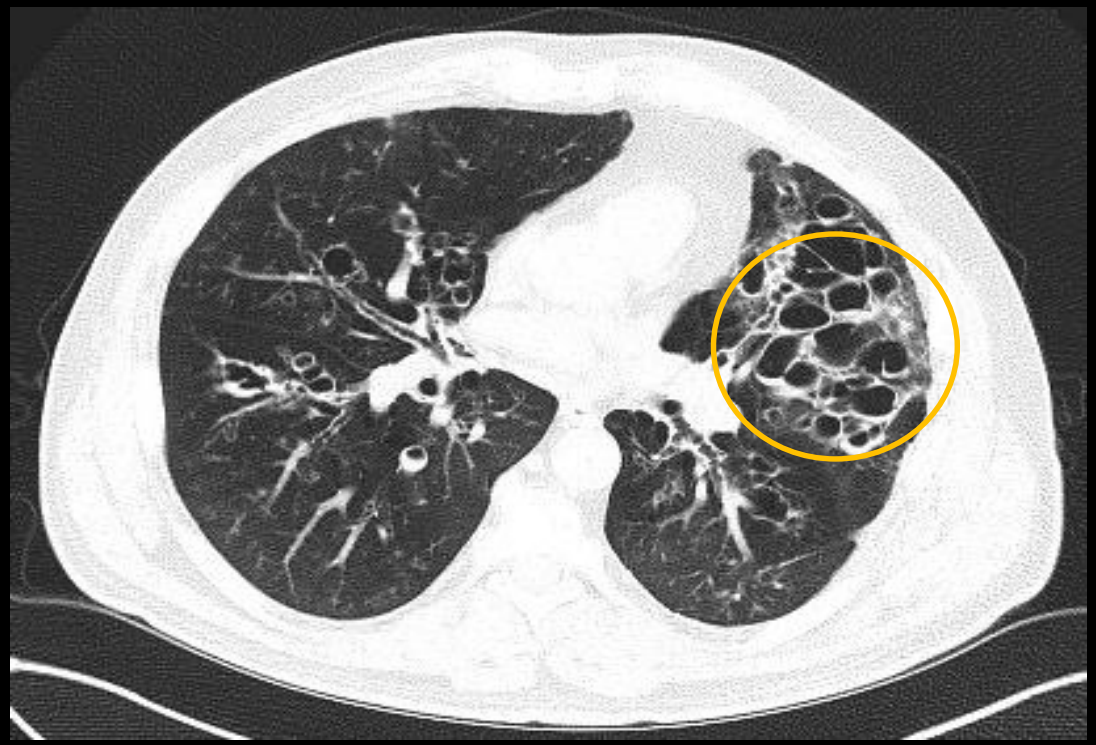
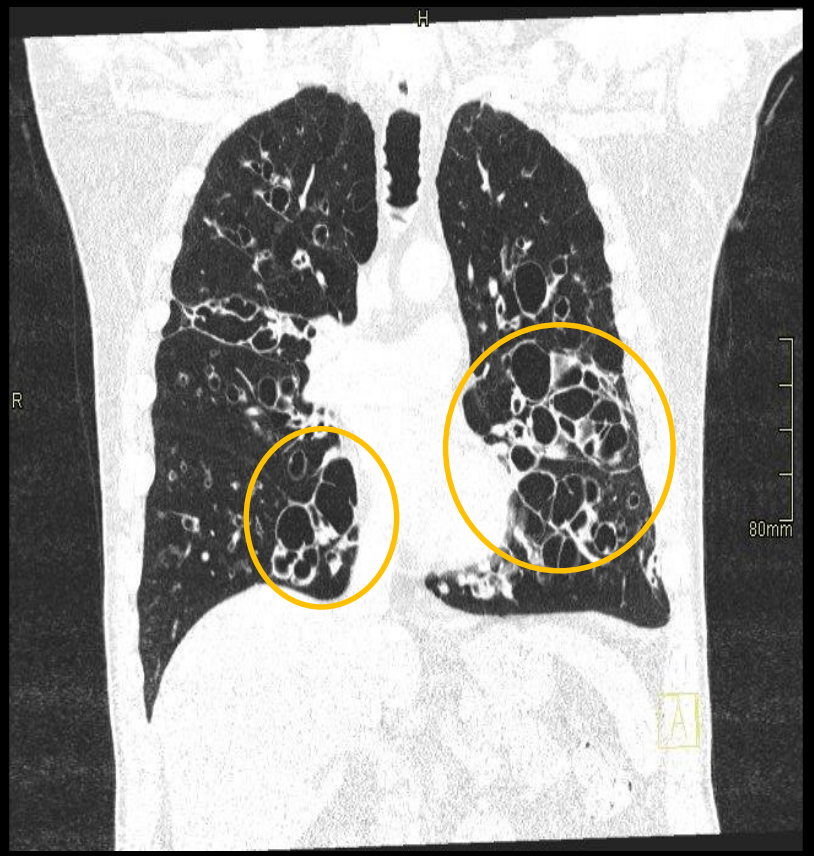


Chest CT without Contrast-Axial View: Mosaic attenuation pattern along with cylindrical bronchiectasis of subsegmental bronchi (encircled)

Findings: (labeled)

A



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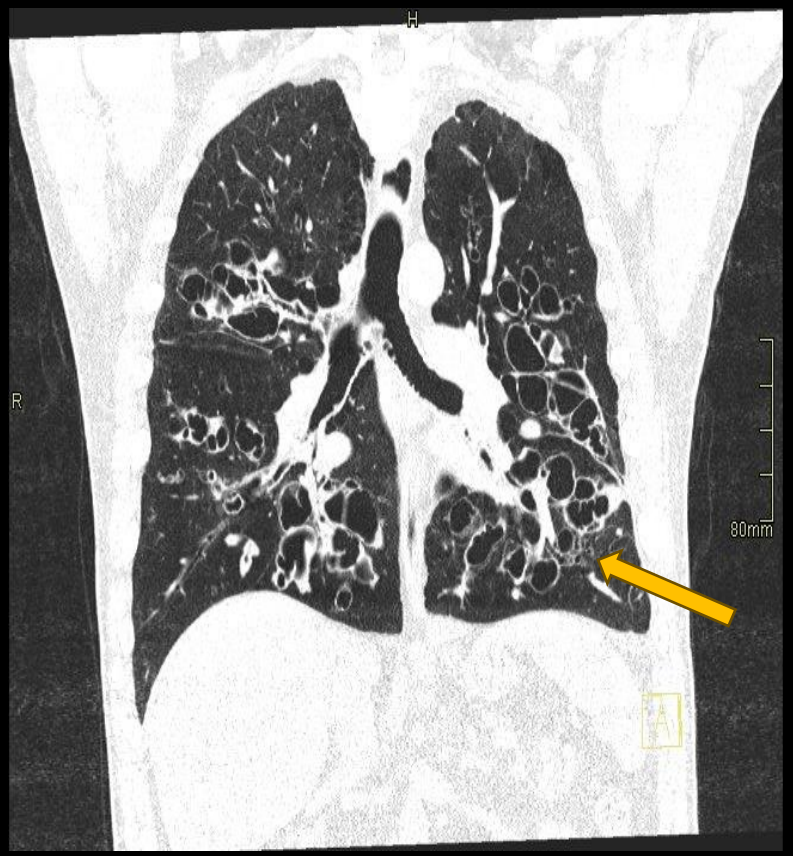

Chest CT without Contrast-Coronal View: Extensive bronchiectasis, with basilar predominance (encircled in fig A). Bronchial and bronchiolar wall thickening (arrow in fig B).

Differential Diagnosis:

- Cystic Fibrosis → Usually involves upper lobe bronchiectasis & had –ve sweat chloride test
- ABPA → Involves upper lobe bronchiectasis & was –ve for Aspergillosis on BAL
- Primary Ciliary Dyskinesia → No issues with fertility, has 3 kids
- Mounier-Kuhn Syndrome → Involves dilation of trachea and main stem bronchi
- Williams-Campbell Syndrome → Most likely as it involves bronchiectasis of 4th order bronchi predominantly in basilar zone of the lungs

Final Dx:

Williams-Campbell Syndrome

Case Discussion

- **Definition:** Williams-Campbell Syndrome is a rare pulmonary disorder characterized by absence of cartilage in subsegmental bronchial walls, leading to collapse of those bronchi and air trapping distal to the collapsed bronchi.

- **Etiology:** Exact cause unclear but may be associated with
  1. Post infection 2. Environmental toxins/irritants exposure
  3. Autoimmune/inflammatory changes leading to small airway fibrotic changes
  4. Genetic predisposition

Case Discussion (cont.)

- **Clinical Features:** **D**yspnea, chronic productive cough, wheezing, pulmonary HTN, recurrent pneumonia, barrel chest deformity, Harrison sulcus

- **Radiographic findings:**
  1. CT: Bronchiectasis involving 4^{th}-6^{th} order bronchi, mosaic attenuation pattern due to air trapping, bronchial wall thickening
  2. Chest X Ray: Hyperinflation and reticular pattern opacities

Case Discussion (cont.)

- **Prognosis:** Depends upon severity and extent of fibrosis. Early diagnosis and treatment can slow disease progression and improve quality of life but in advanced cases, respiratory failure occurs requiring Long-term Oxygen Therapy/Lung Transplantation.

- **Management:** No definitive cure, but supportive treatment includes bronchodilators, corticosteroids, oxygen therapy, pulmonary rehabilitation, antibiotics, and lung transplantation.

References:

1.Iqbal S, Weerakkody Y. Williams-Campbell syndrome. *Radiopaedia.org*. Published online May 20, 2010. doi:10.53347/rid-9743

2.Johnson L, et al. William Campbell syndrome: Clinical features and management strategies. *Respir Med.* 2021;12(1):78-85.

3.National Pulmonary Association. *Guidelines for the diagnosis and management of rare pulmonary disorders.* Published 2019.

4.Aldave AN, Saliski DW. The clinical manifestations, diagnosis and management of williams-campbell syndrome. *North American Journal of Medical Sciences*. 2014;6(9):429.