DPU
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(Deemed to be University)

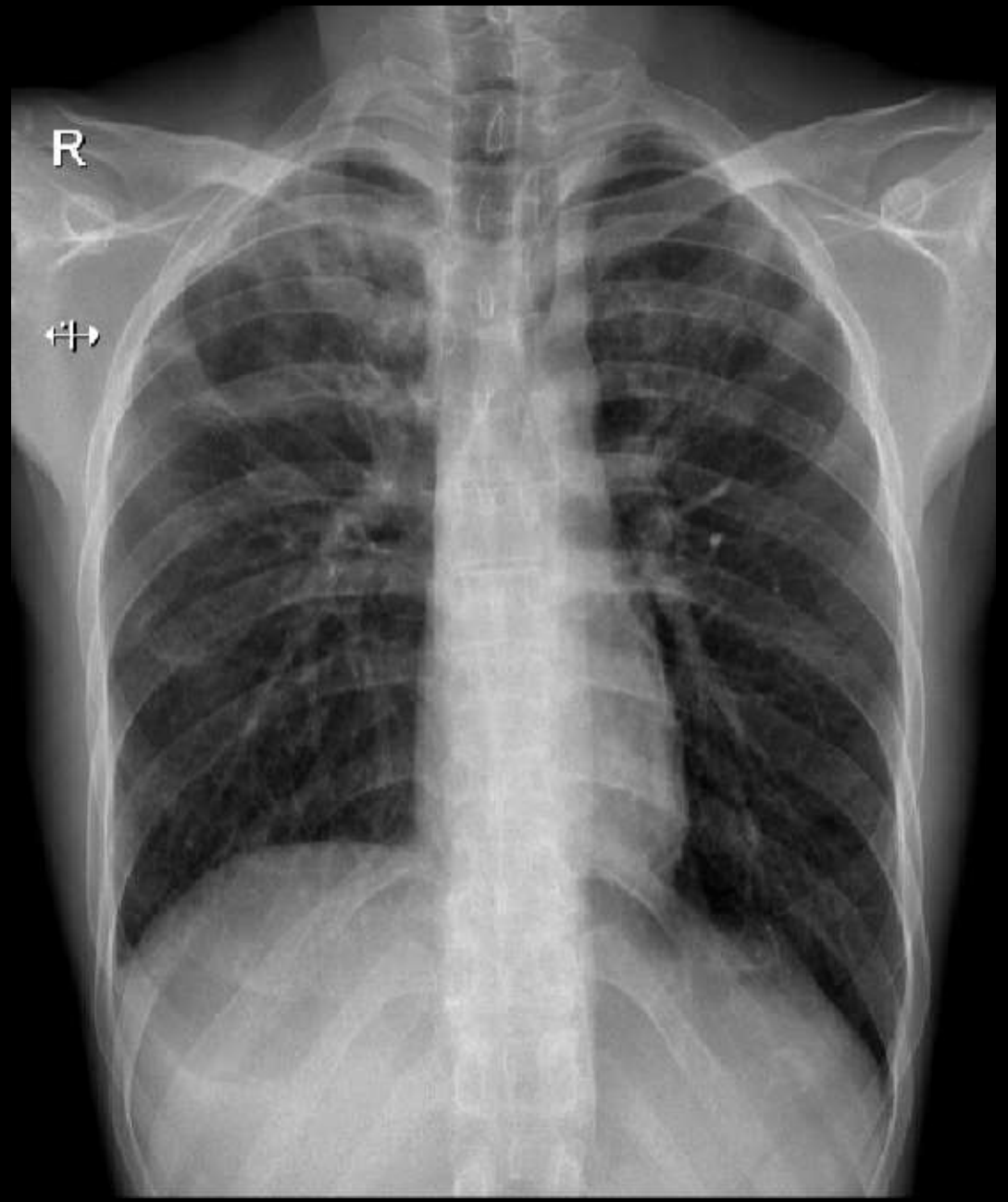
RADIOLOGY CLINICAL MEET 2025

RARE PRESENTATION OF INTERSTITIAL LUNG DISEASE DURING FOLLOW UP STUDY

Clinical profile:

27 years / Male

- Known case of scleroderma with interstitial lung disease since 5 years
- C/o chest pain and fever since 1 week
- H/o persistent cough and mild breathlessness
- H/o weight loss – approximately 6 kgs in 1 year
- No h/o hemoptysis / vomiting
- No h/o smoking / exposure to fumes
- HIV - negative status
- Patient had sclerodactyly
- Patient had regular follow ups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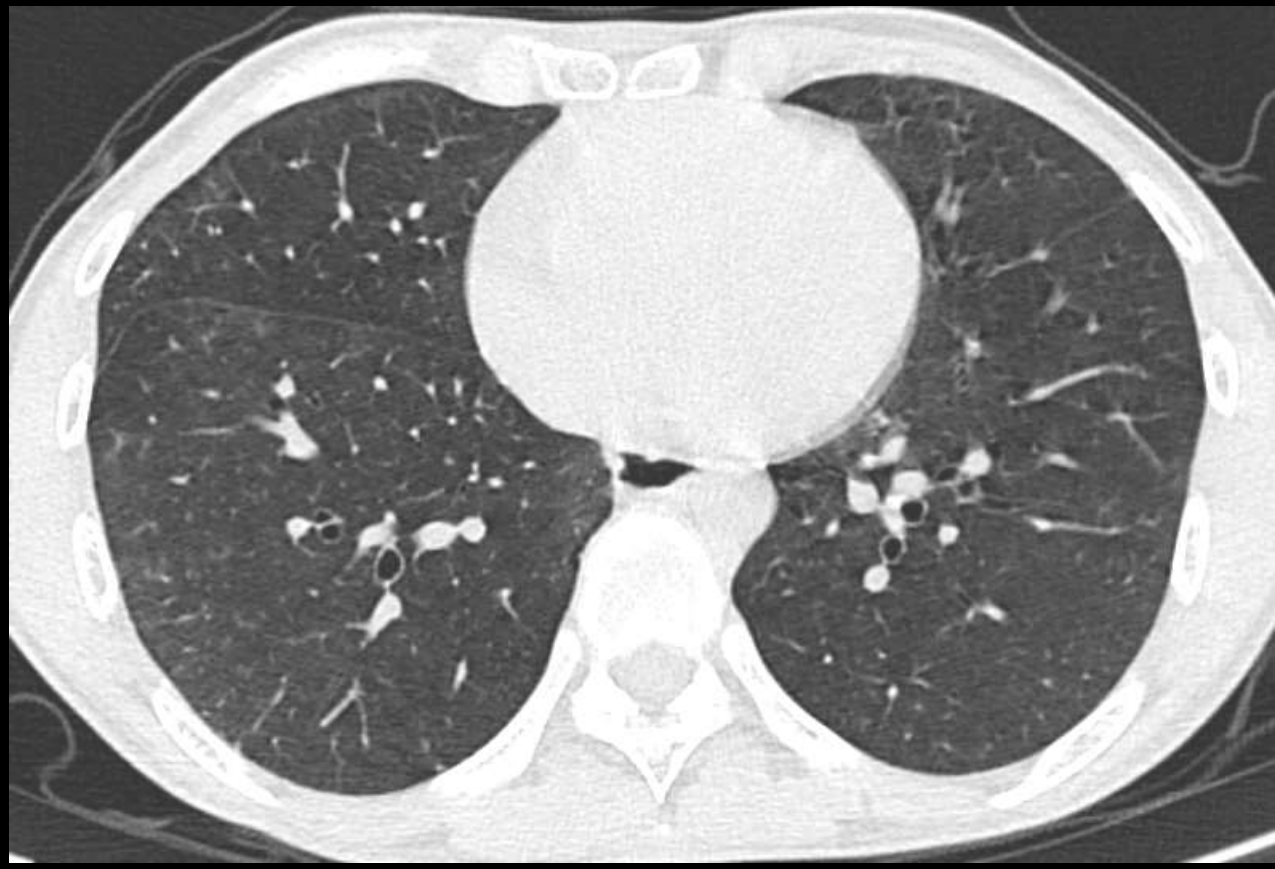
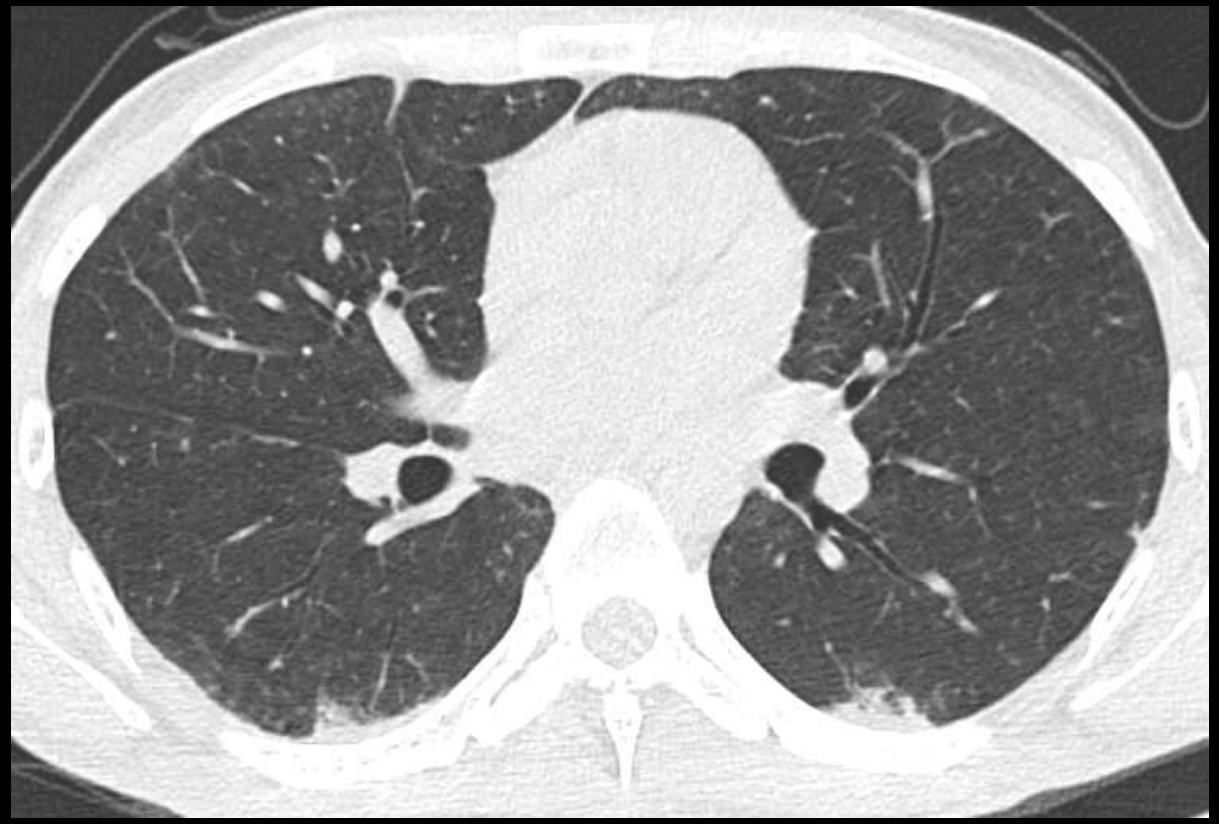
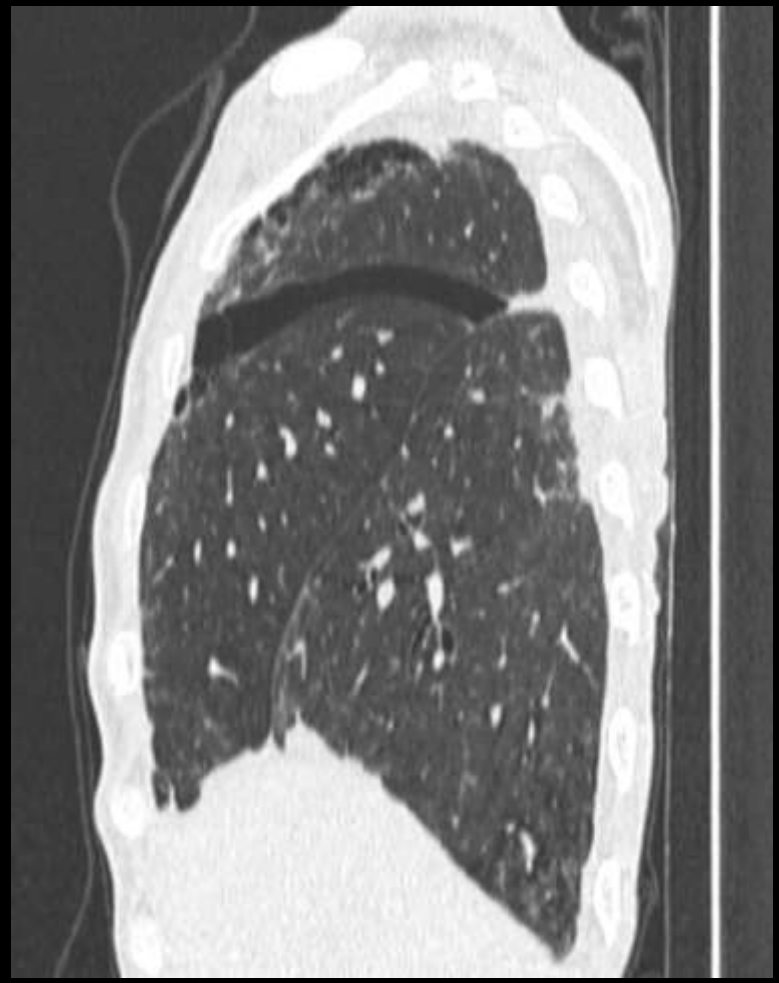
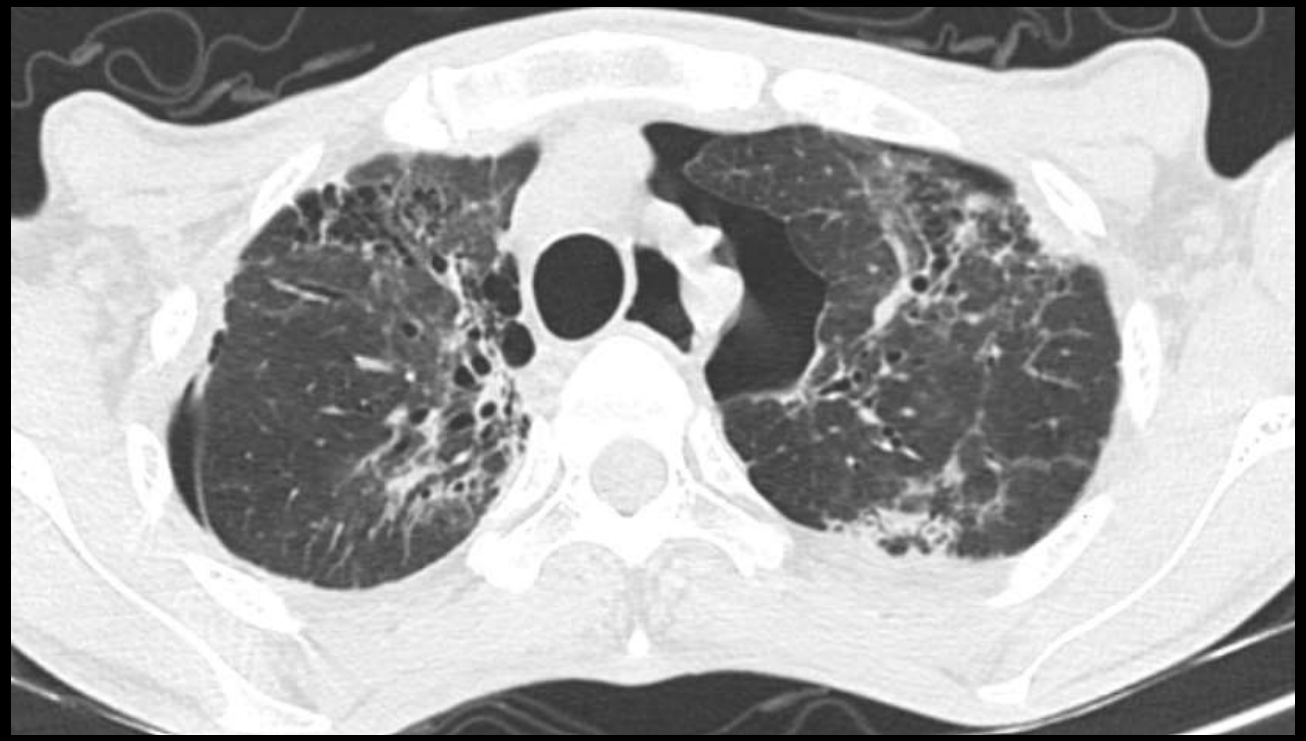
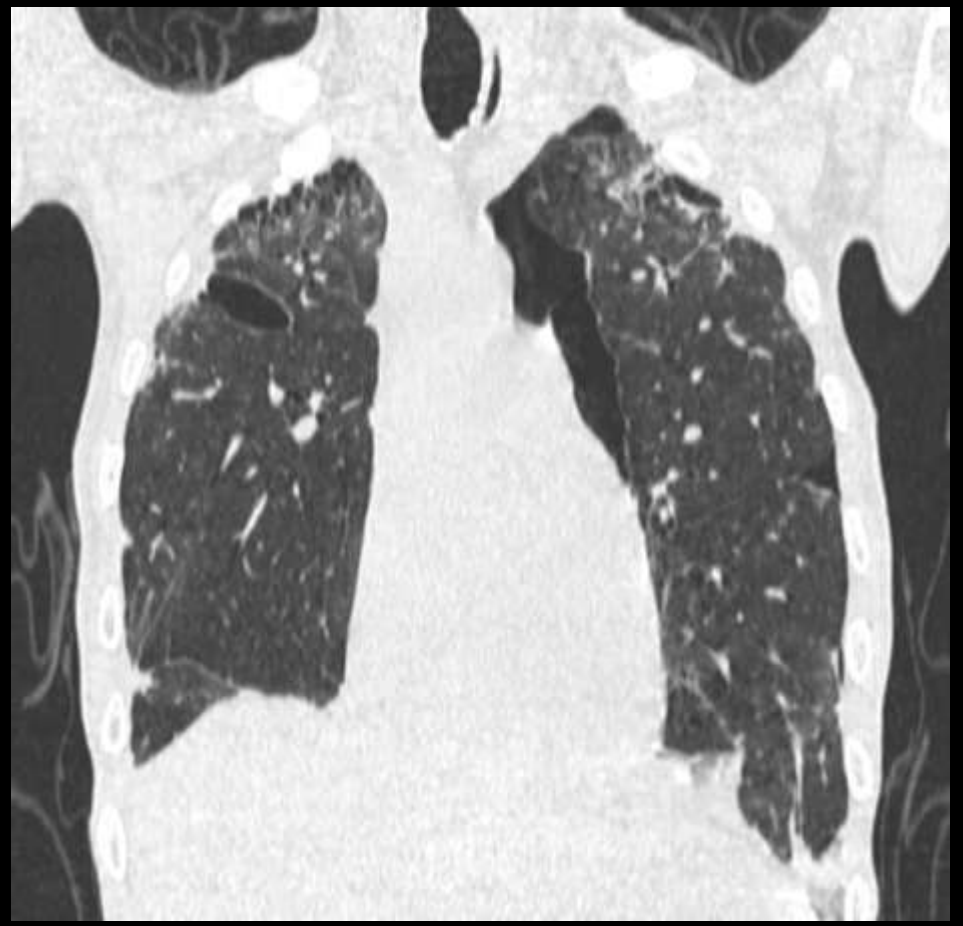


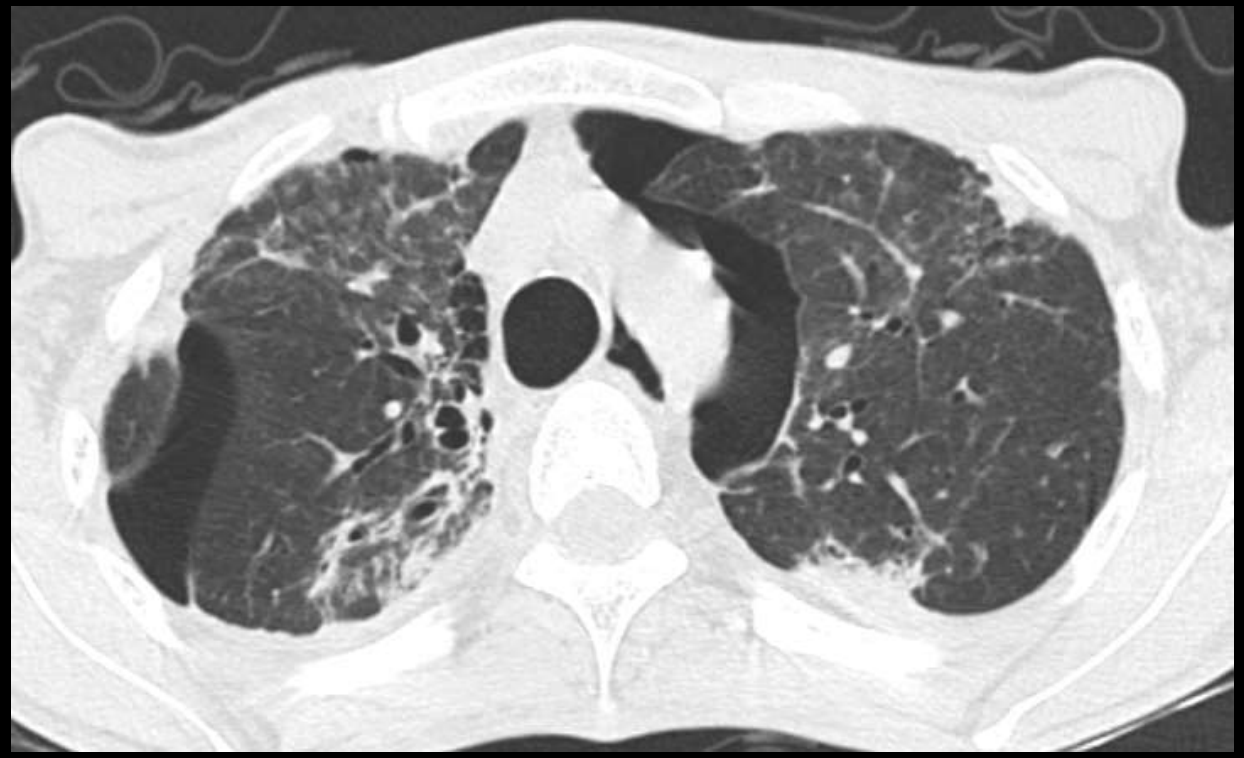


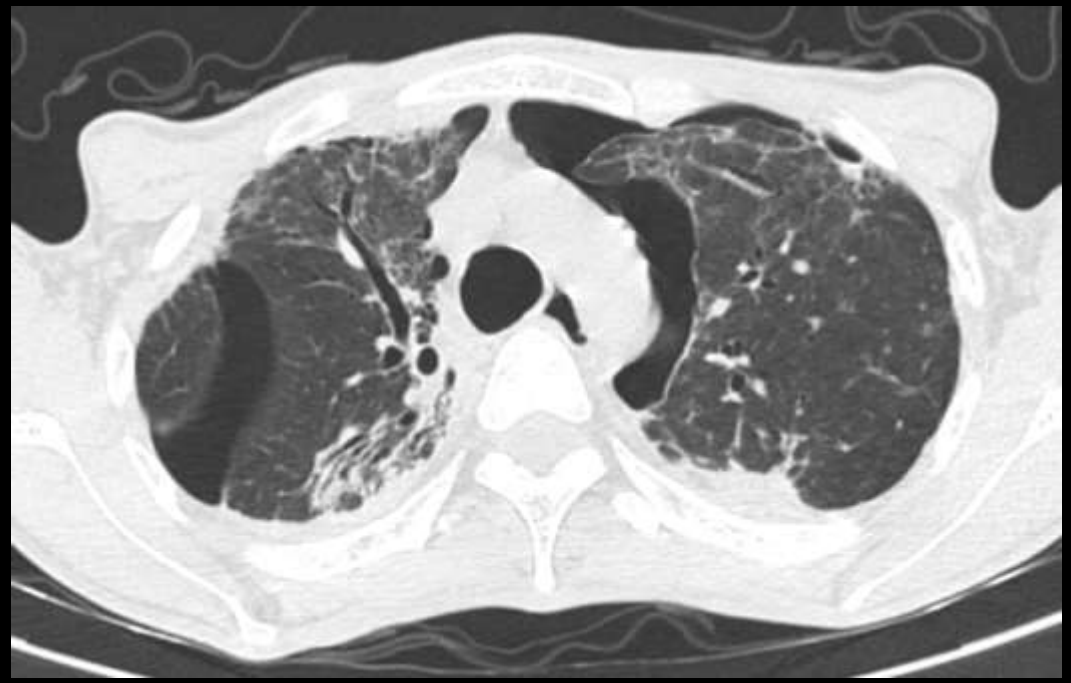
These finding were suggestive of **early changes of interstitial lung disease**

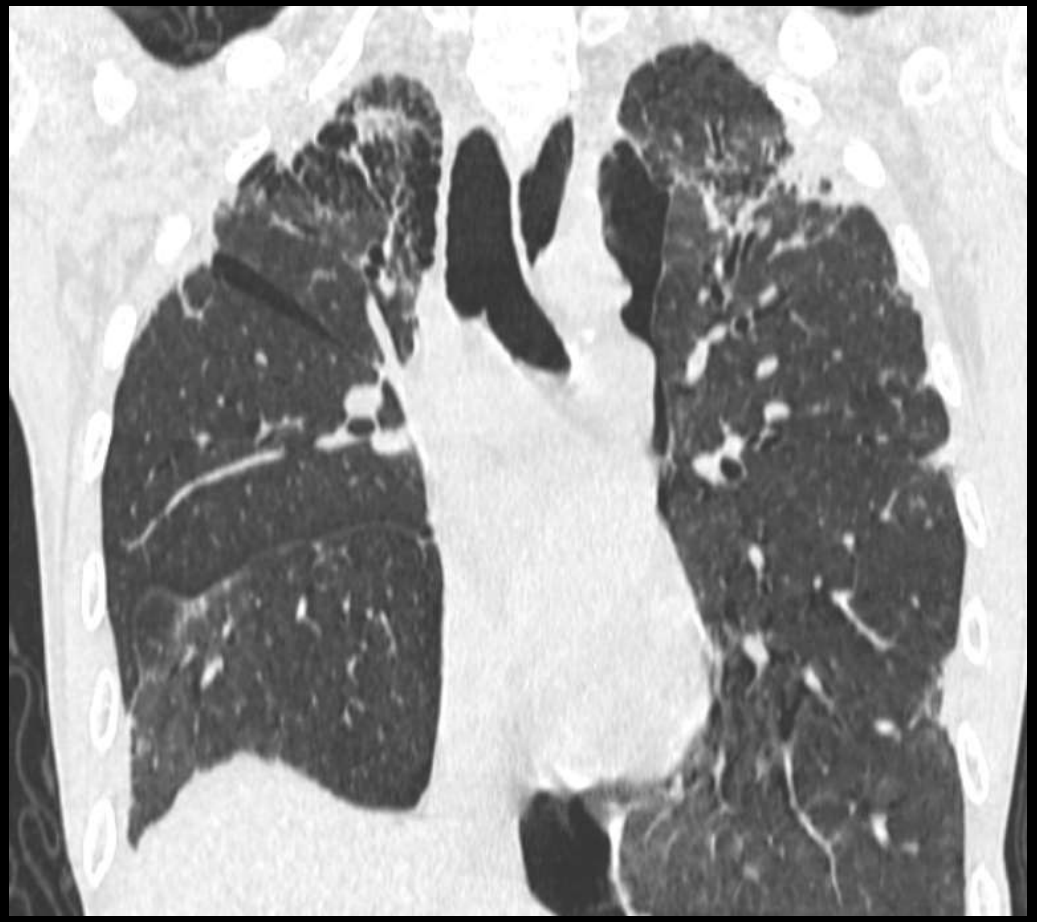


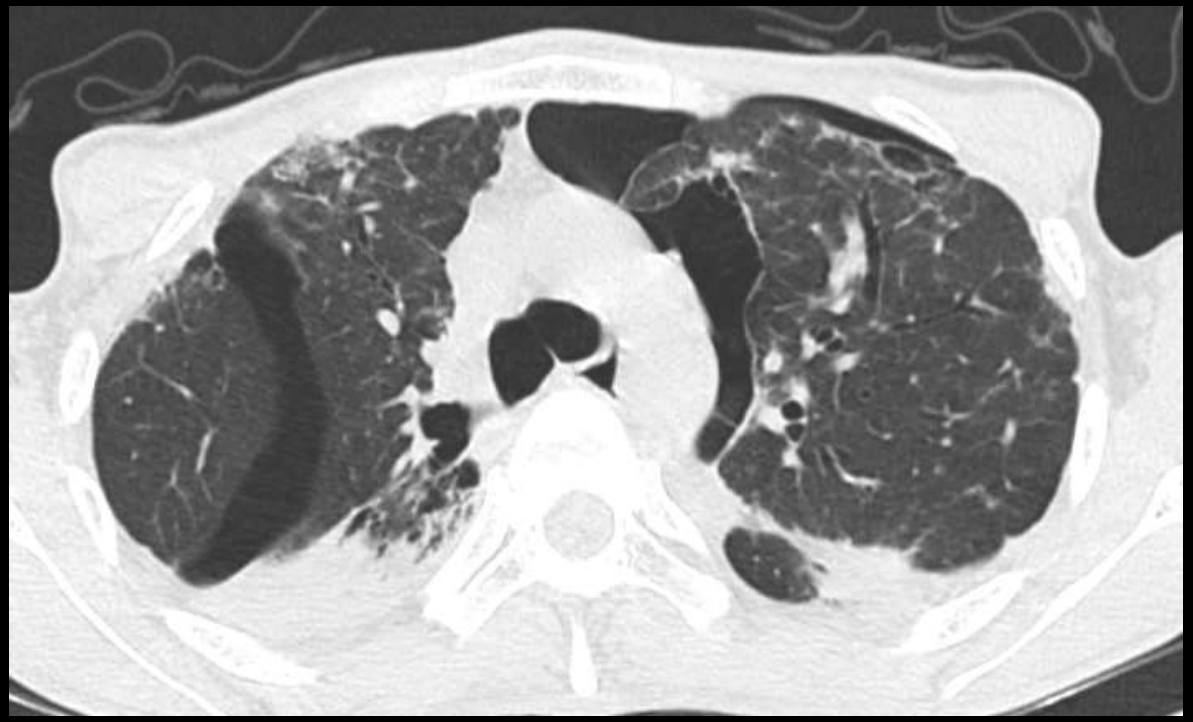


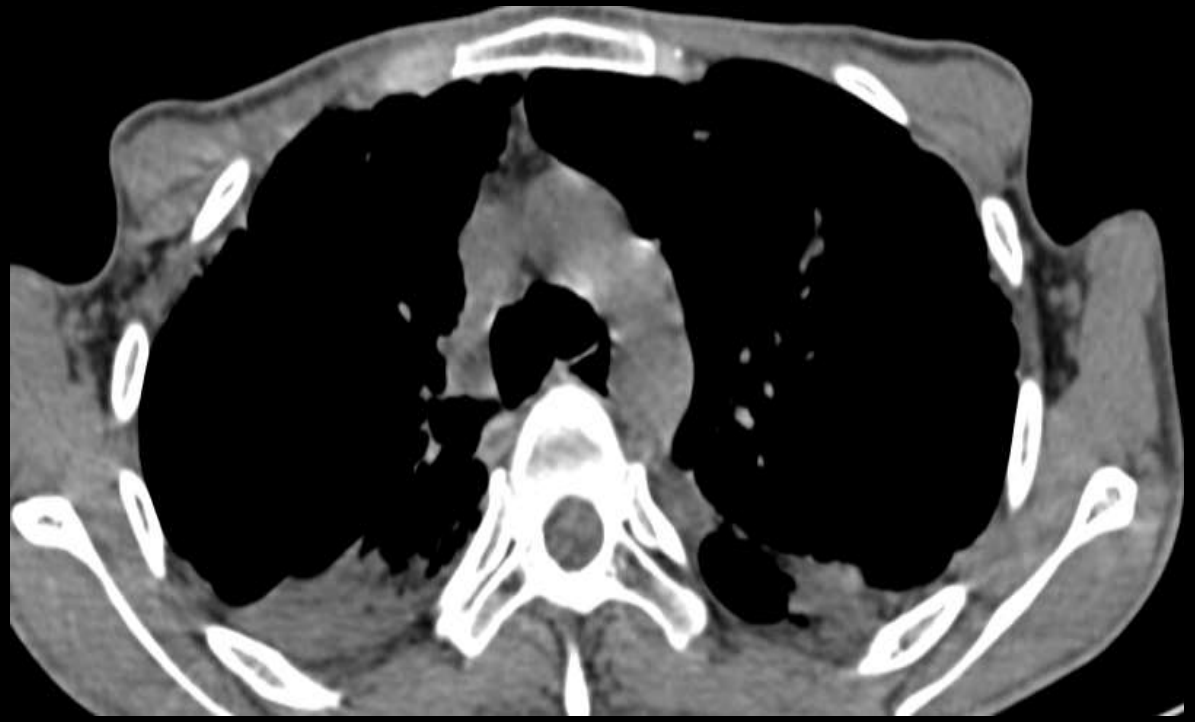


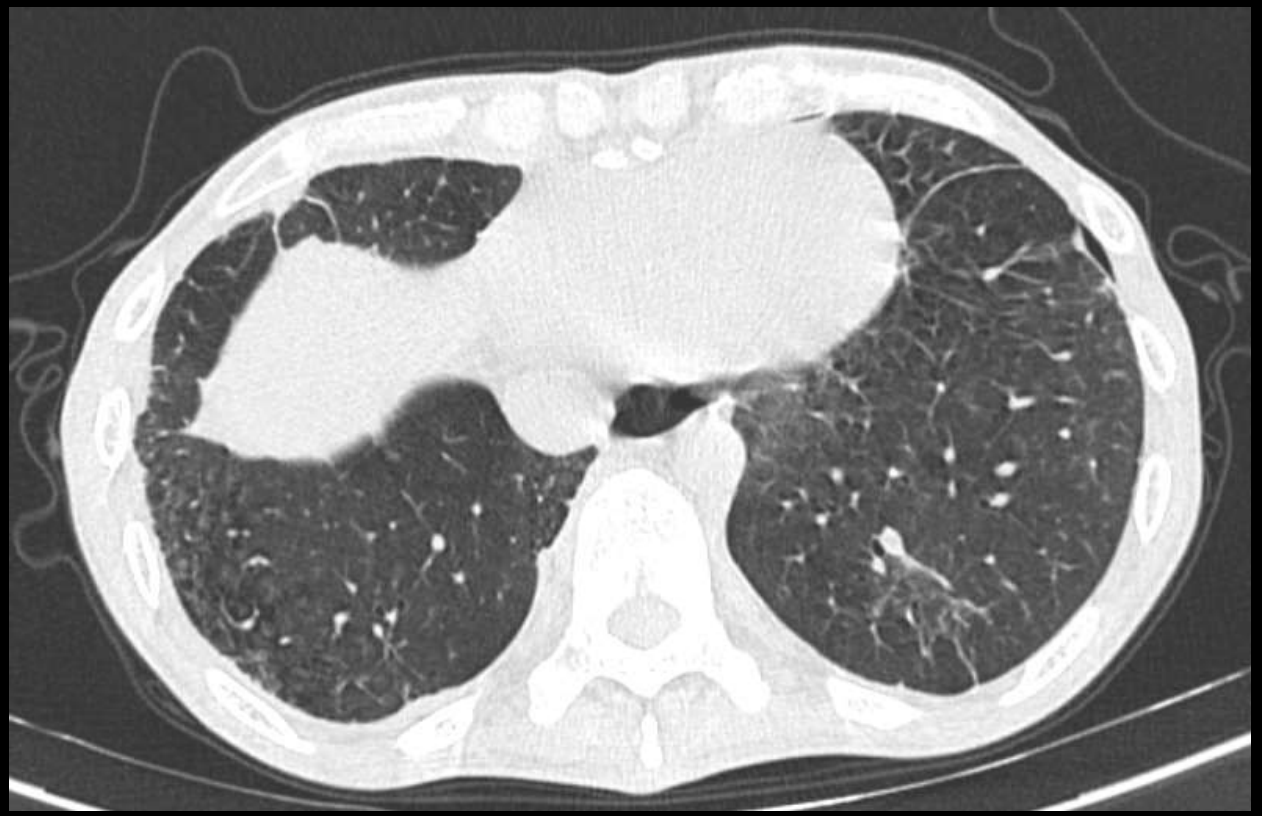


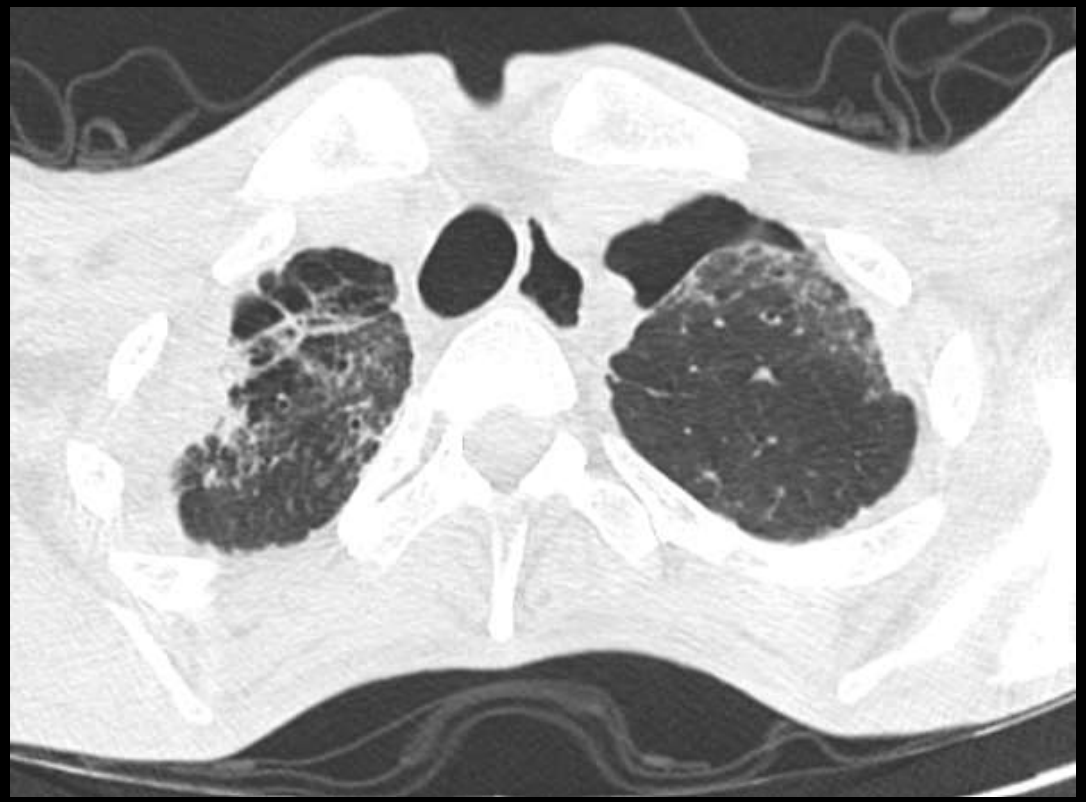


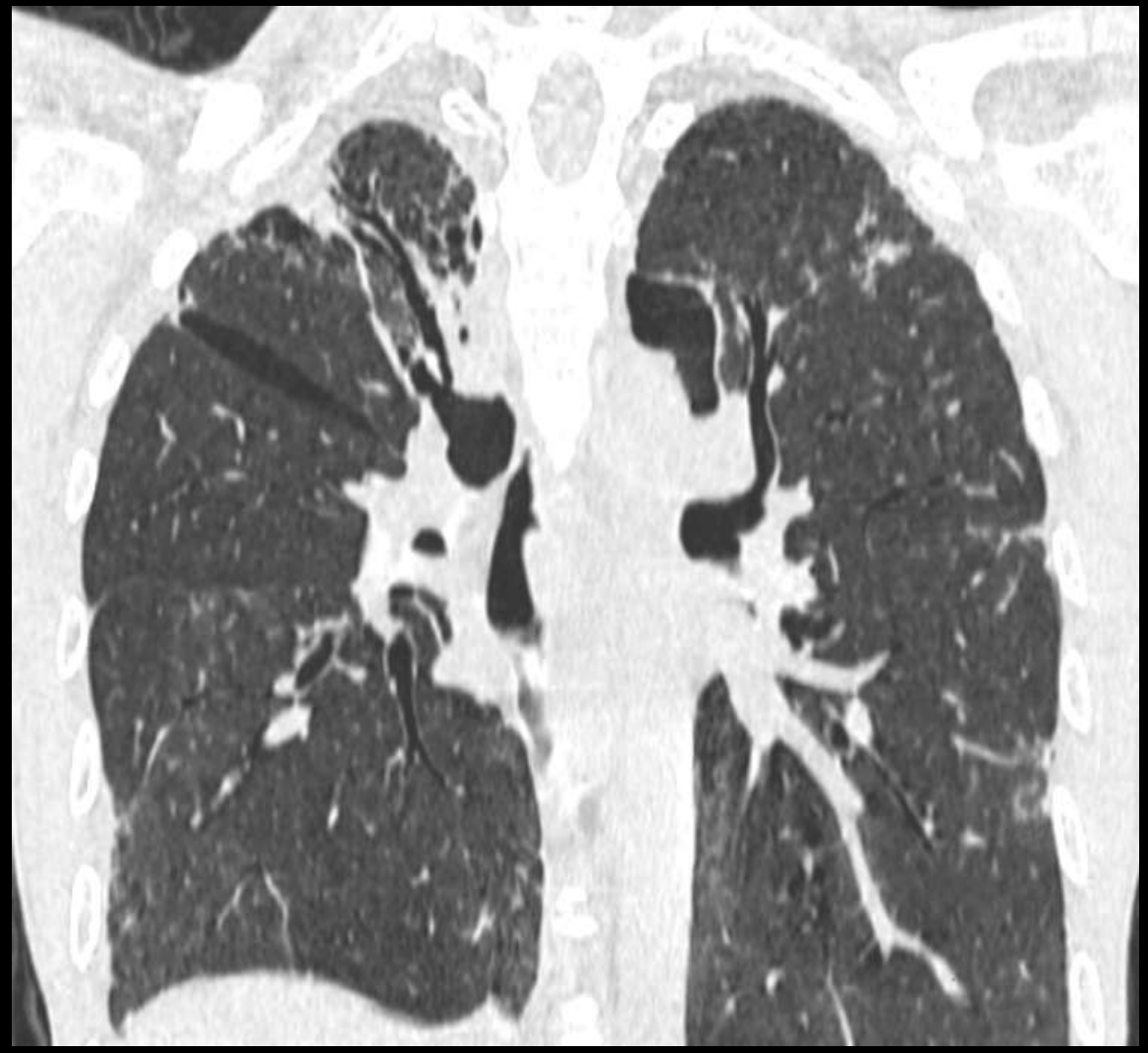


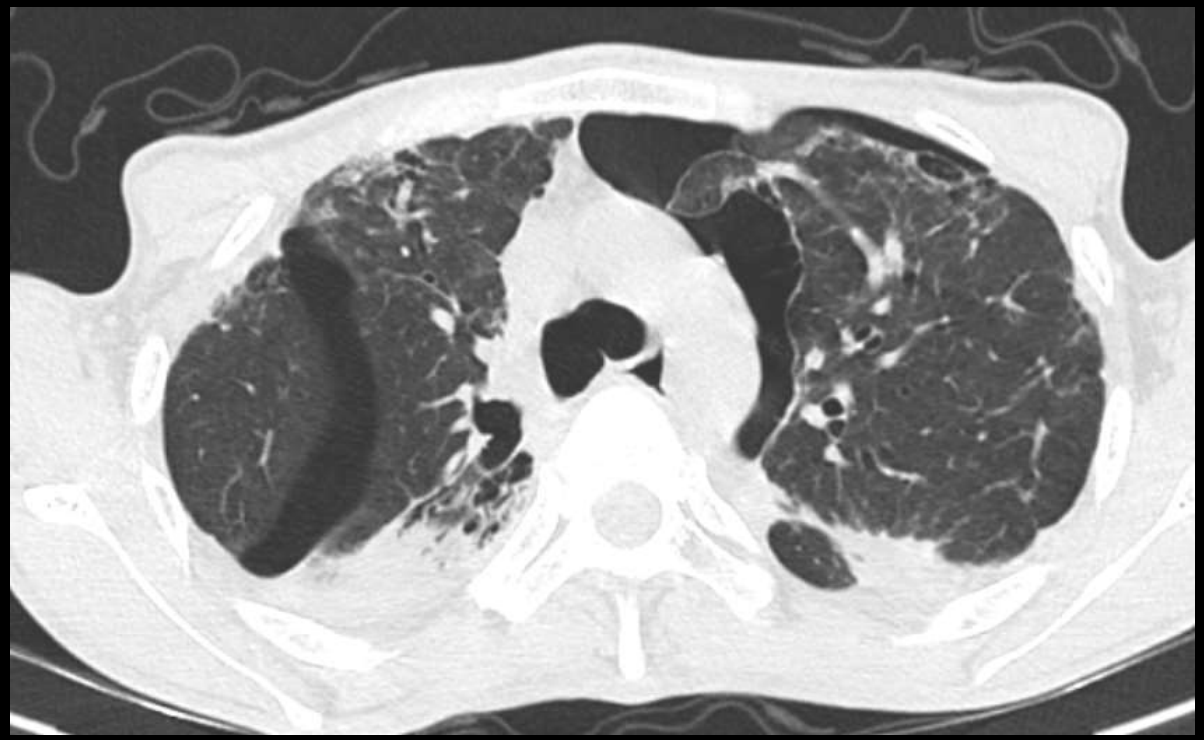












Based on the imaging features:

- Findings suggestive of **Interstitial lung disease**
- **Bilateral pneumothorax**
- Multiple areas of subsegmental consolidation in subpleural location - likely due to **superimposed infection**

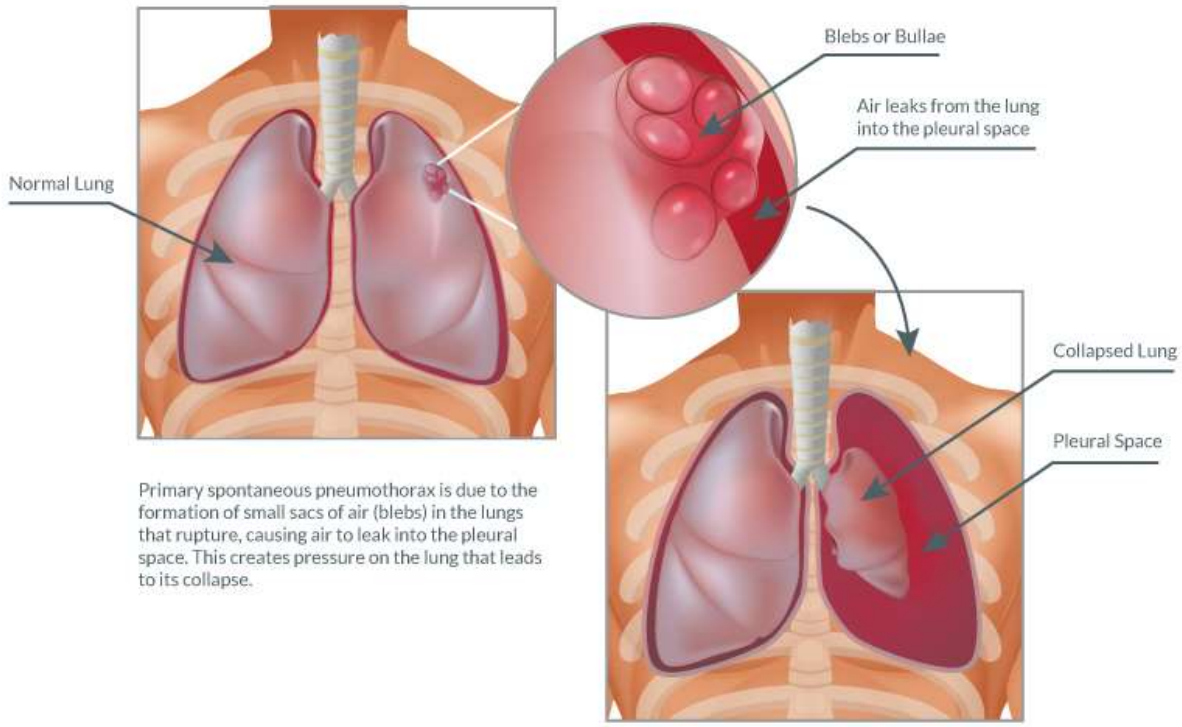
Bilateral pneumothorax in interstitial lung disease due to connective tissue disorder

Introduction

- **Pneumothorax in patients with Interstitial Lung Disease (ILD) due to connective tissue disorders (CTDs) is a rare but serious complication.**
- Spontaneous pneumothorax in ILD is relatively uncommon, and it has been documented in few case reports involving connective tissue disorders such as **systemic sclerosis (scleroderma)**, **rheumatoid arthritis (RA)** and **dermatomyositis**.
- These can lead to progressive lung damage, including fibrosis, cyst formation and bullae which predispose patients to pneumothorax.

Pathophysiology of Pneumothorax in CTD-Related ILD:

- In patients with **connective tissue disorders**, lung fibrosis often progresses with the development of **cystic changes** or **bullae** in the lung parenchyma. These changes increase the likelihood of **spontaneous pneumothorax** because the cysts or bullae may rupture, leading to accumulation of air in the pleural space.



- Persistent inflammation increases the **pleural porosity** which accelerates air leakage.
- **Corticosteroids** may lead to refractory pneumothorax by suppressing immune response and delaying tissue repair.
- **Immunosuppressive therapies** (commonly used to treat CTDs) increase the risk of infections or lung injury, which can further complicate the development of pneumothorax.

Predictive Factors for Pneumothorax in CTD-ILD

1. **Lower BMI:**
   - linked to **bleb formation** which can cause pneumothorax.

2. **Reticular Abnormalities on HRCT:**
   - Indicate lung damage.

3. **Methylprednisolone Pulse Therapy:**
   - Associated with higher disease activity
   - May impair healing by suppressing inflammation and vascular permeability.

Clinical Implications:

- **Spontaneous pneumothorax** is a life-threatening emergency and often requires **chest tube insertion** to re-expand the lung and relieve respiratory distress.
- In some cases, **surgical interventions** such as **pleurodesis** or **bullectomy** may be needed to prevent recurrence.
- For patients with **connective tissue disorders**, early detection and monitoring of lung function are crucial. **High-resolution CT (HRCT)** can help identify **subpleural cysts** or **bullae.**
- Prognosis depends on the underlying disease, the extent of lung involvement, and the response to treatment.

Journal Name	No. of Patients	Connective Tissue Disorder	No. of patients with Pneumothorax	Unilateral or Bilateral Pneumothorax
Chest	1	Systemic sclerosis (scleroderma)	1	Unilateral
Journal of Rheumatology	2	Systemic sclerosis (scleroderma)	2	Unilateral
The Journal of Rheumatology	1	Rheumatoid Arthritis (RA)	1	Unilateral
Annals of the Rheumatic Diseases	1	Rheumatoid Arthritis (RA)	1	**Bilateral**
European Respiratory Journal	1	Dermatomyositis	1	Unilateral
Lung	1	Mixed Connective Tissue Disease (MCTD)	1	Unilateral

Summary:

- Bilateral pneumothorax is a rare complication of ILD due to connective tissue disorders.
- **Systemic sclerosis**, **rheumatoid arthritis**, and **dermatomyositis** are among the connective tissue disorders where pneumothorax has been observed, most likely due to the presence of **lung cysts** or **bullae** that are prone to rupture.
- Management typically involves chest tube drainage, and in some cases, surgical intervention to prevent recurrence.

Thank you