# RADY 401 Case Presentation

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UNC SCHOOL OF MEDICINE
Radiology

# HPI

- 32-year-old female with no significant past medical history
- Presents to her PCP with “several months” of a non-productive cough and mild (2/10) pleuritic chest pain
- 14 pack-year smoking history; denies other substance or alcohol use
  - Current every-day smoker

- Pertinent ROS
  - (-) sputum production, hemoptysis, weight loss, night sweats
  - (+) dyspnea on exertion, fatigue

# Workup

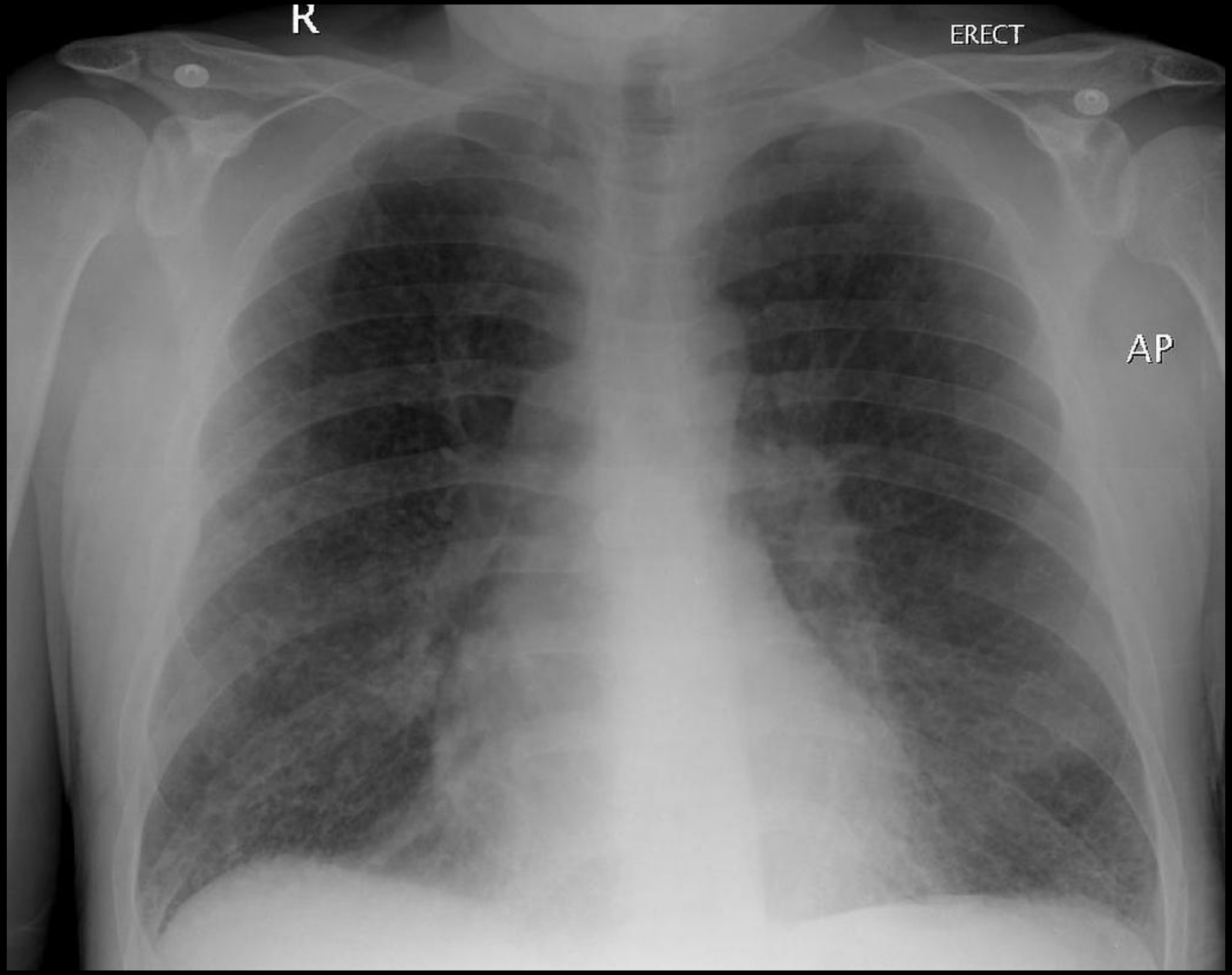
- CBC, CMP, UA within normal limits
- EKG unremarkable
- D-dimer negative

- Imaging ordered: CXR → CT chest

# Workup

**Variant 1:** **Chronic dyspnea. Unclear etiology. Initial imaging.**

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

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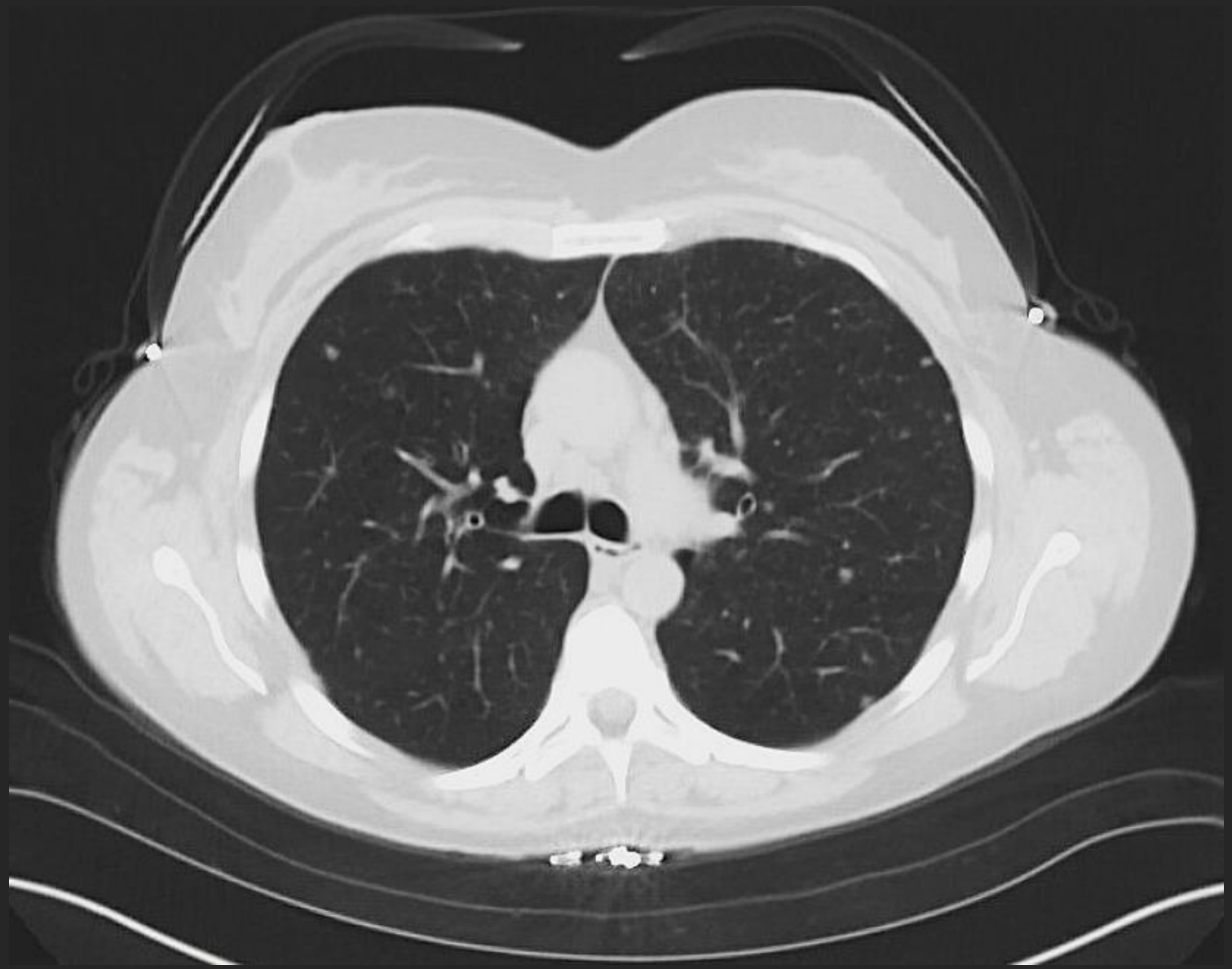
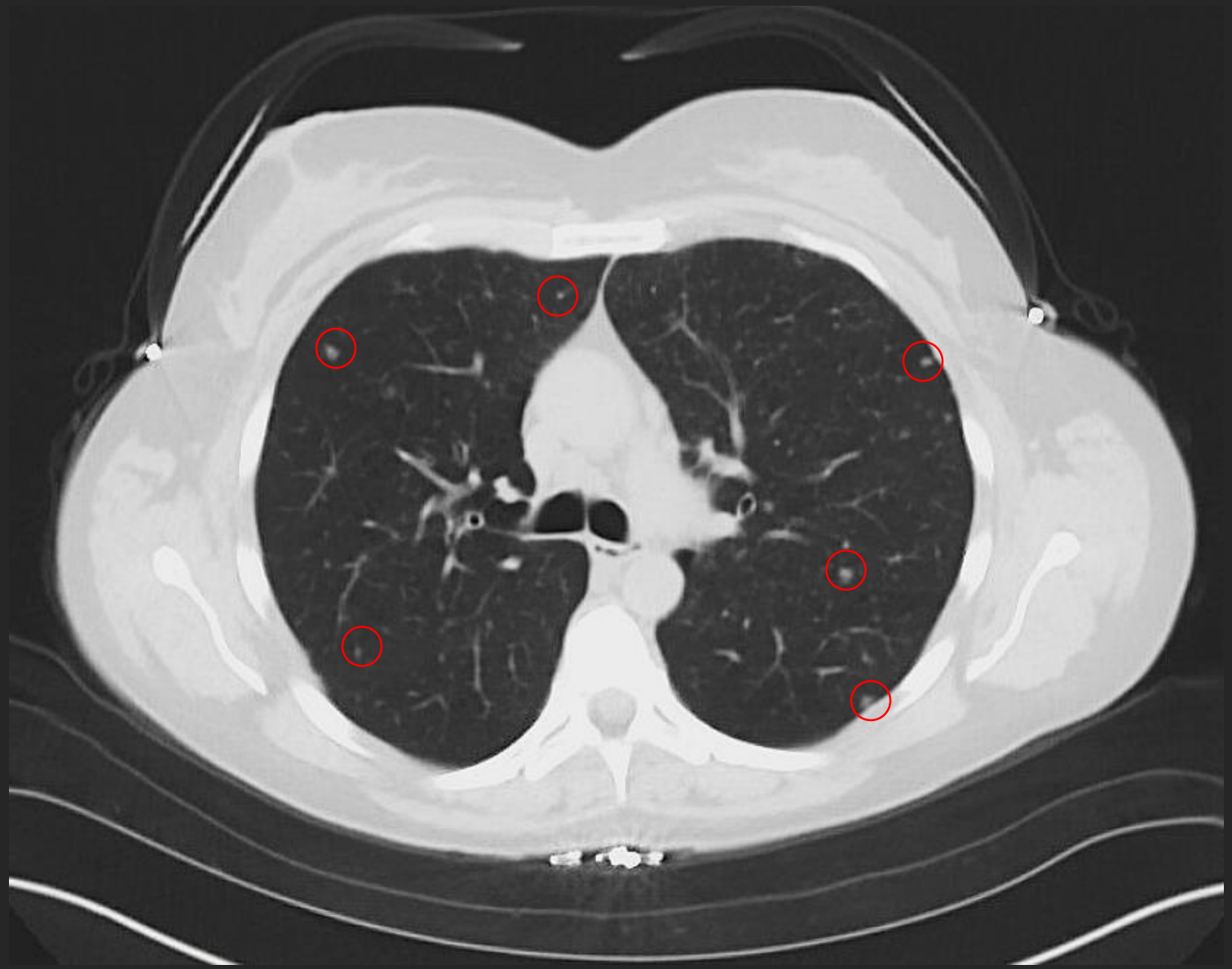
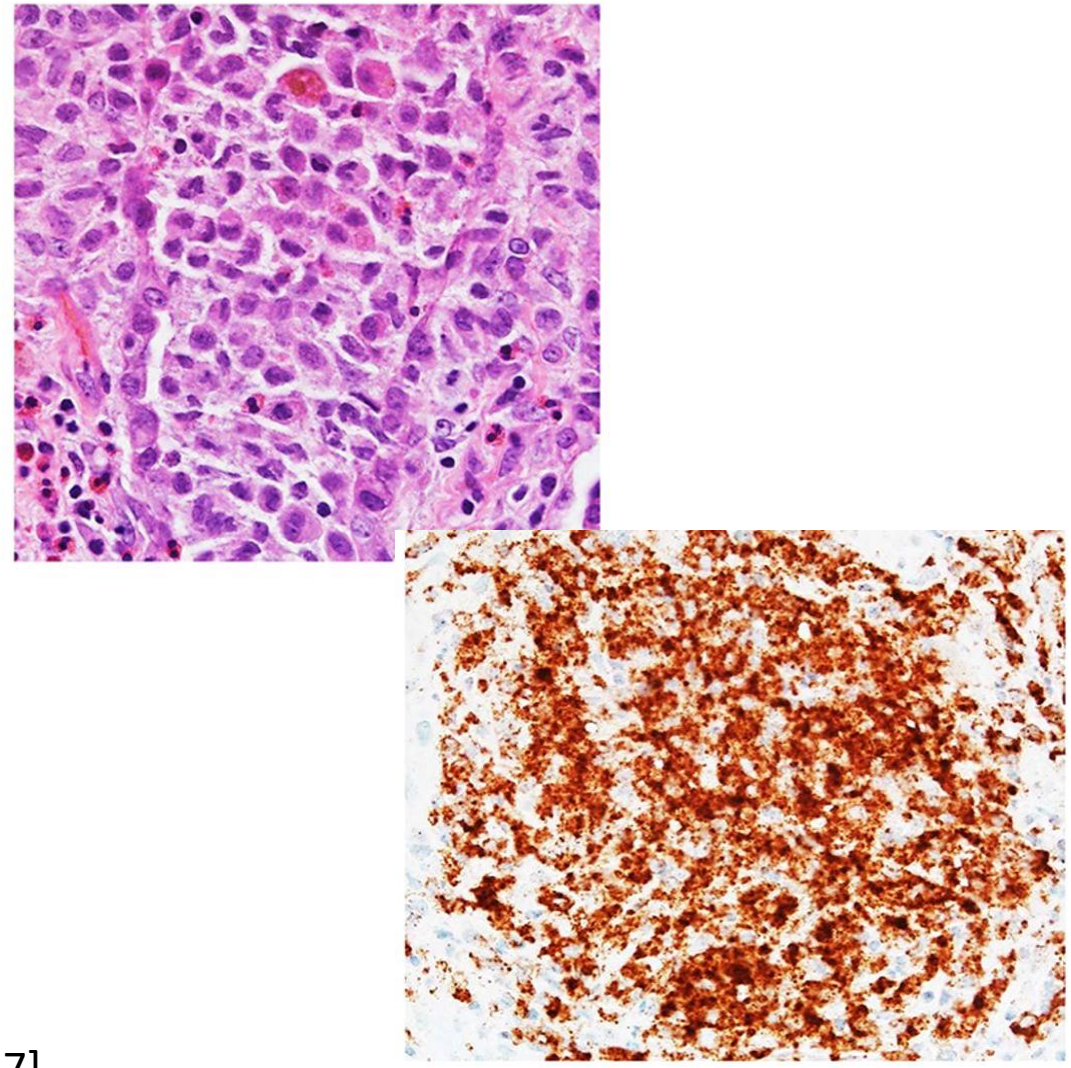




# Pulmonary Langerhans cell histiocytosis (PLCH)



- Epidemiology: 20-40 yo, Caucasian
- Presentation: dyspnea, dry cough, constitutional symptoms
  - 25% asymptomatic
- Pathophysiology: Langerhans cells proliferate in bronchial epithelium and form granulomas
  - Evolution from nodules to cysts
- Associations: History of smoking in 95% of cases. Can be associated with AML, ALL.

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# Treatment

- Smoking cessation +/- systemic glucocorticoid therapy
  - > 60% of patients show resolution or stabilization of disease with smoking cessation alone
- Refractory cases: chemotherapy (cladribine, cytarabine)

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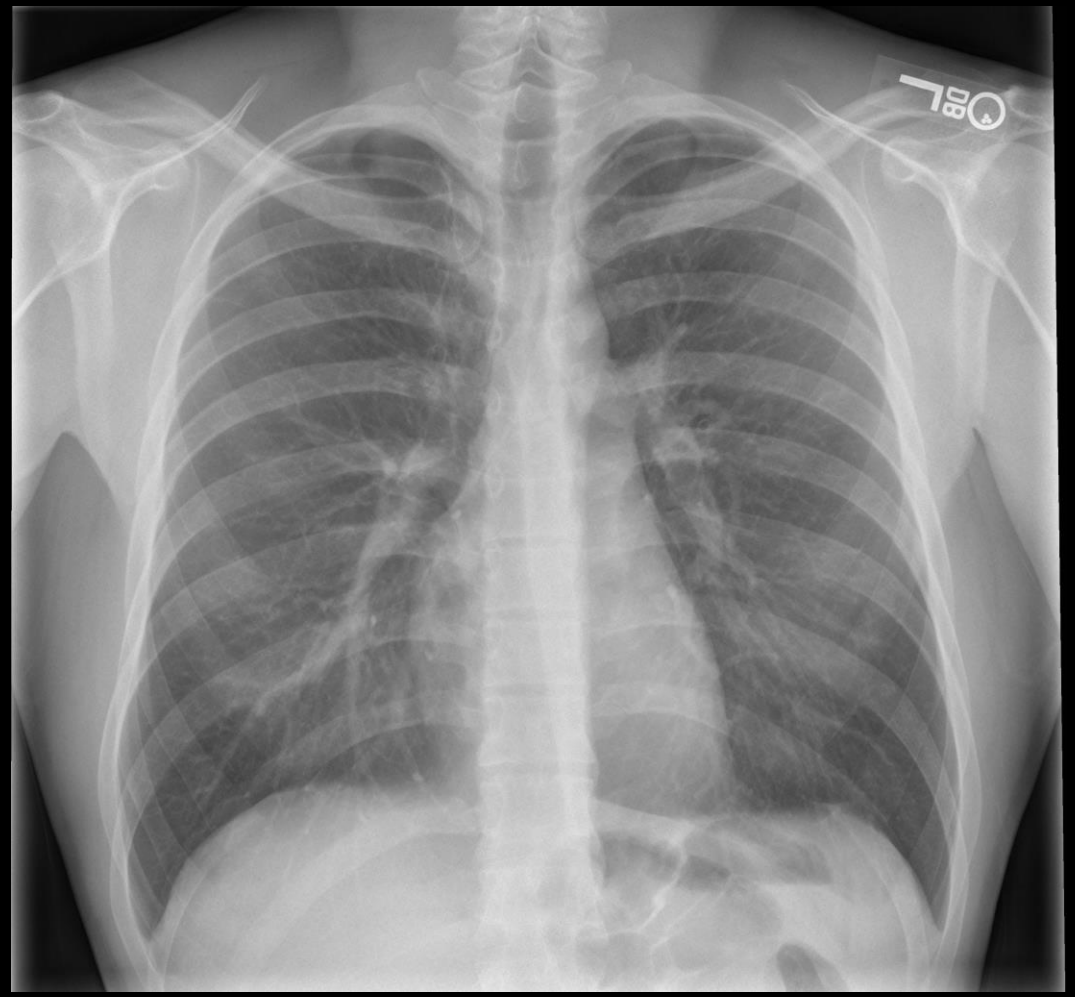
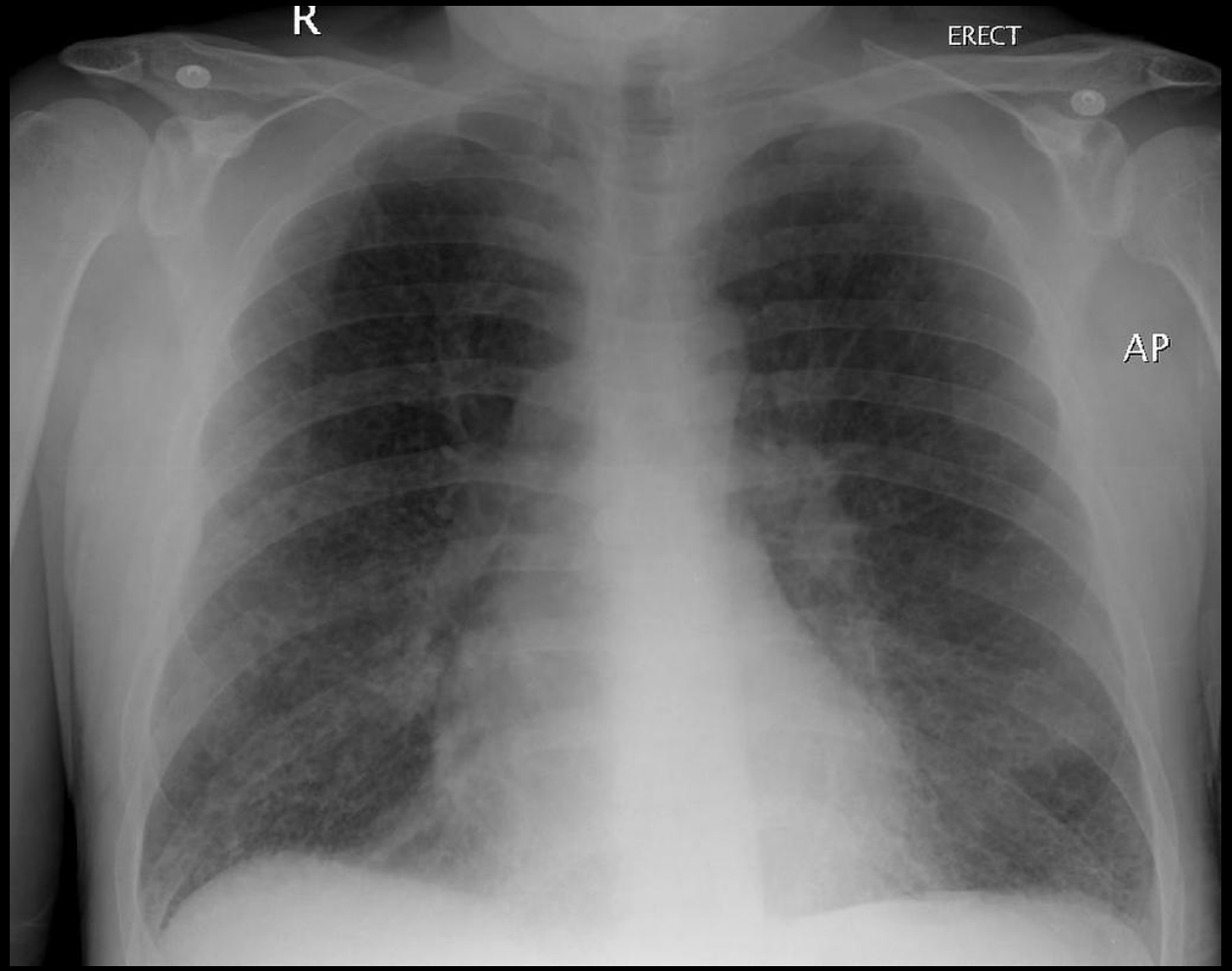
# Imaging Discussion

# Early disease: nodular predominance

# Normal



# Patient

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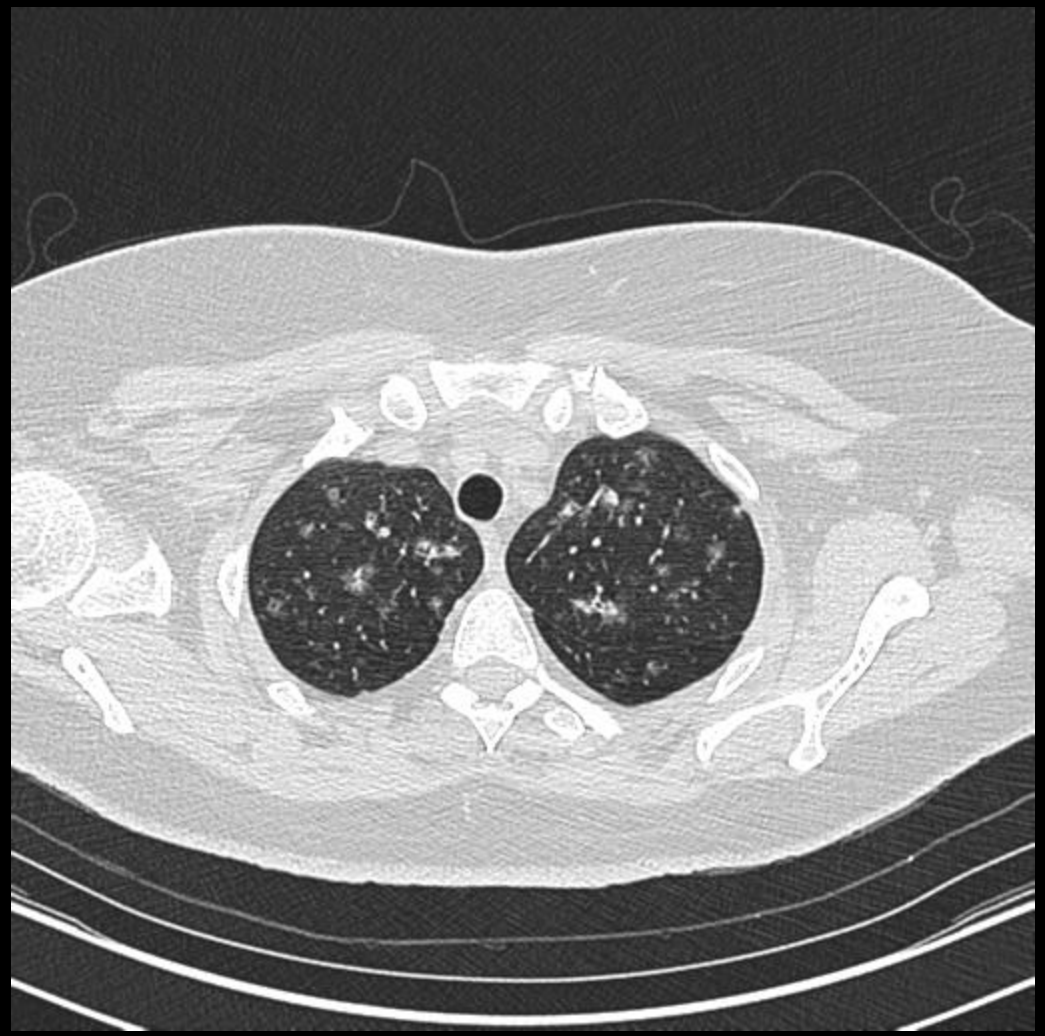

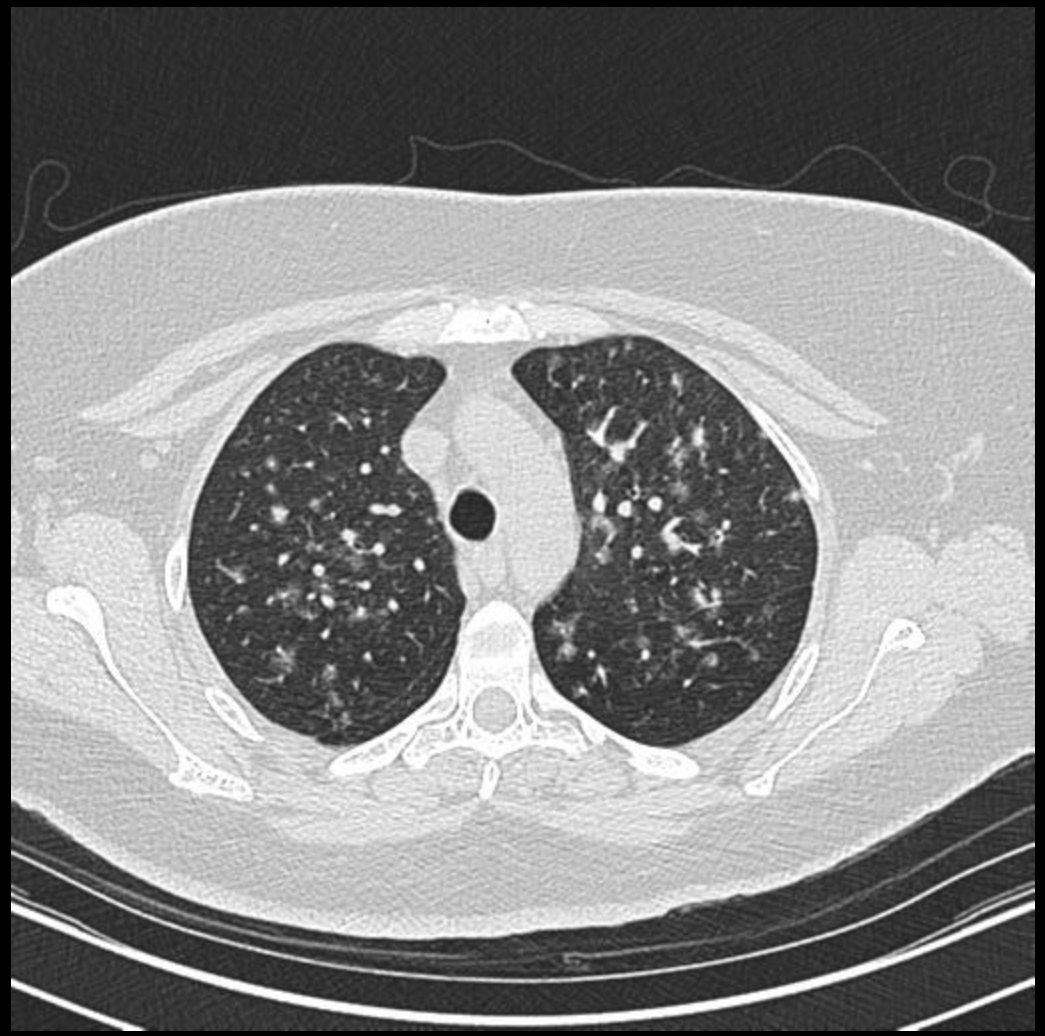

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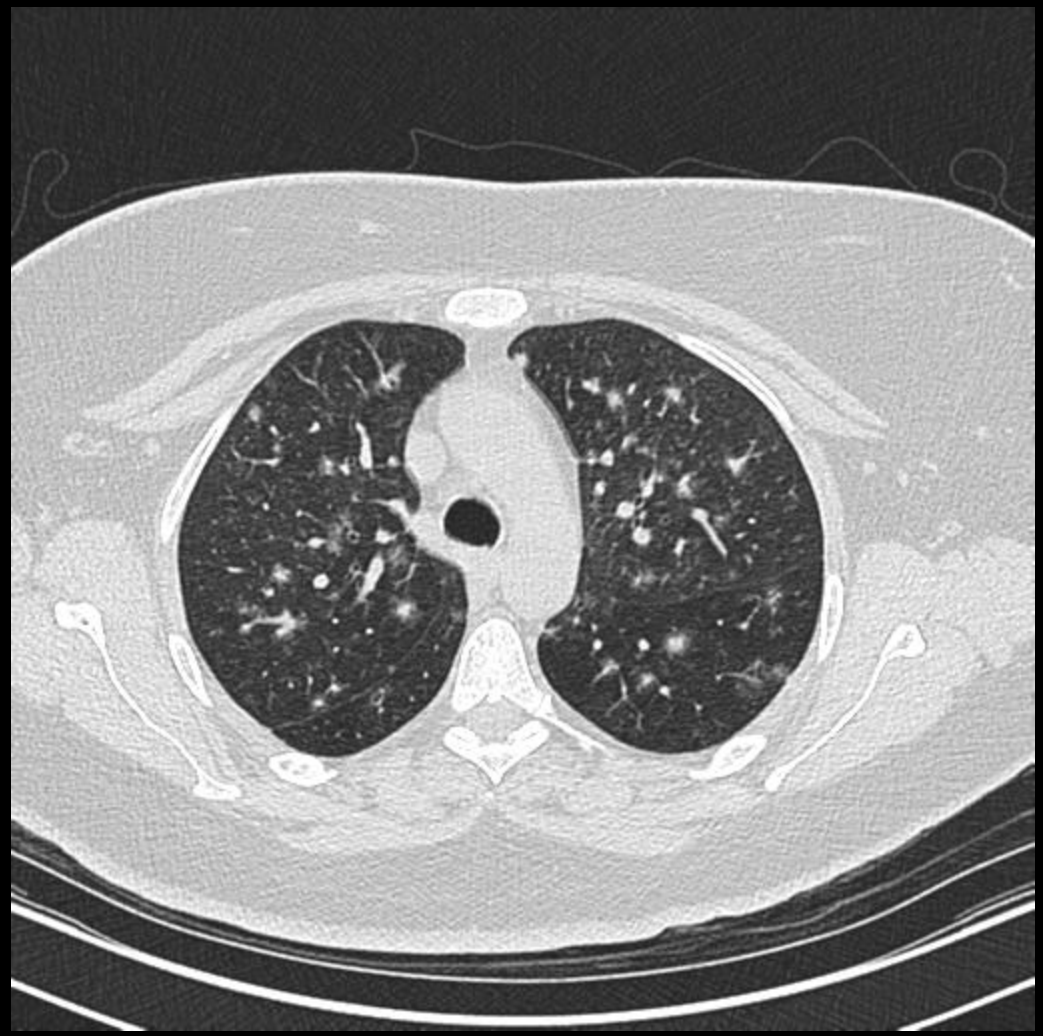

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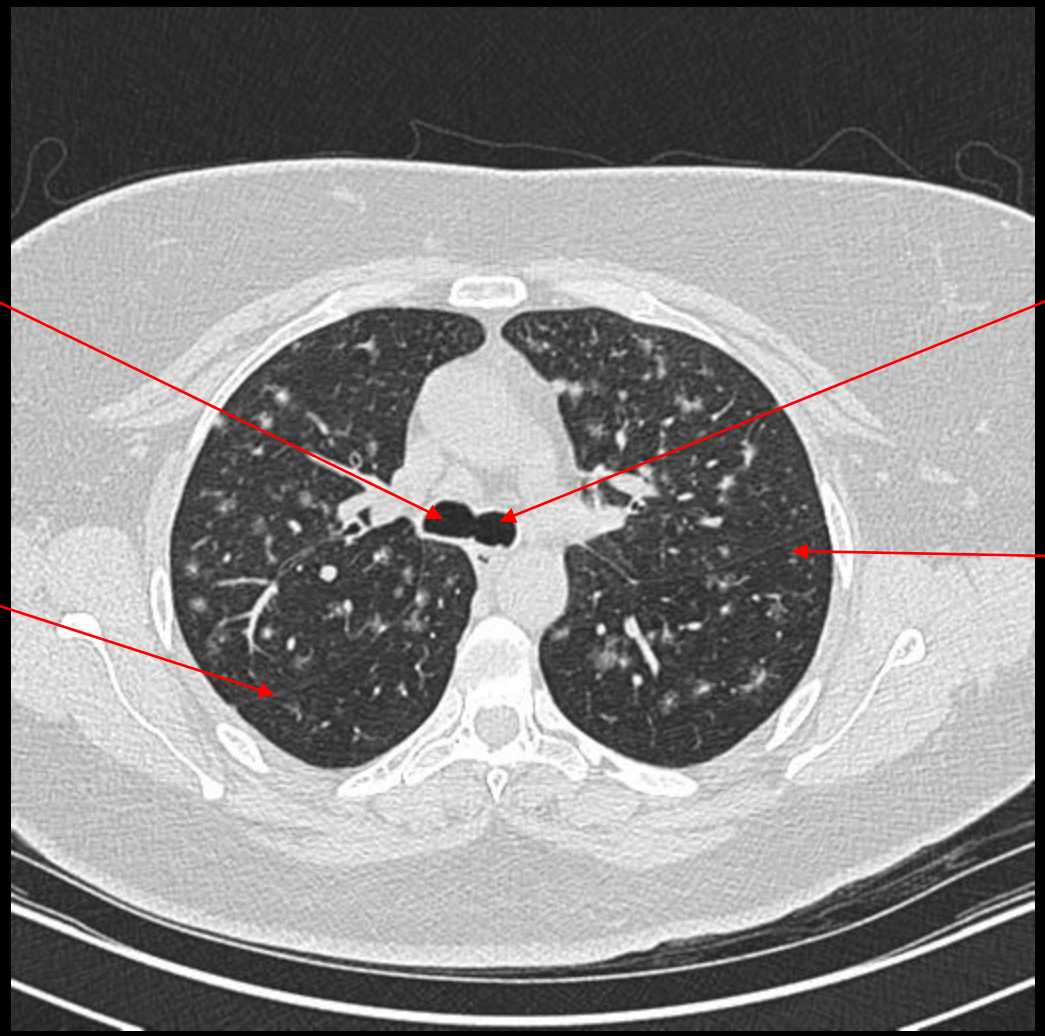
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L mainstem bronchus
Major fissure
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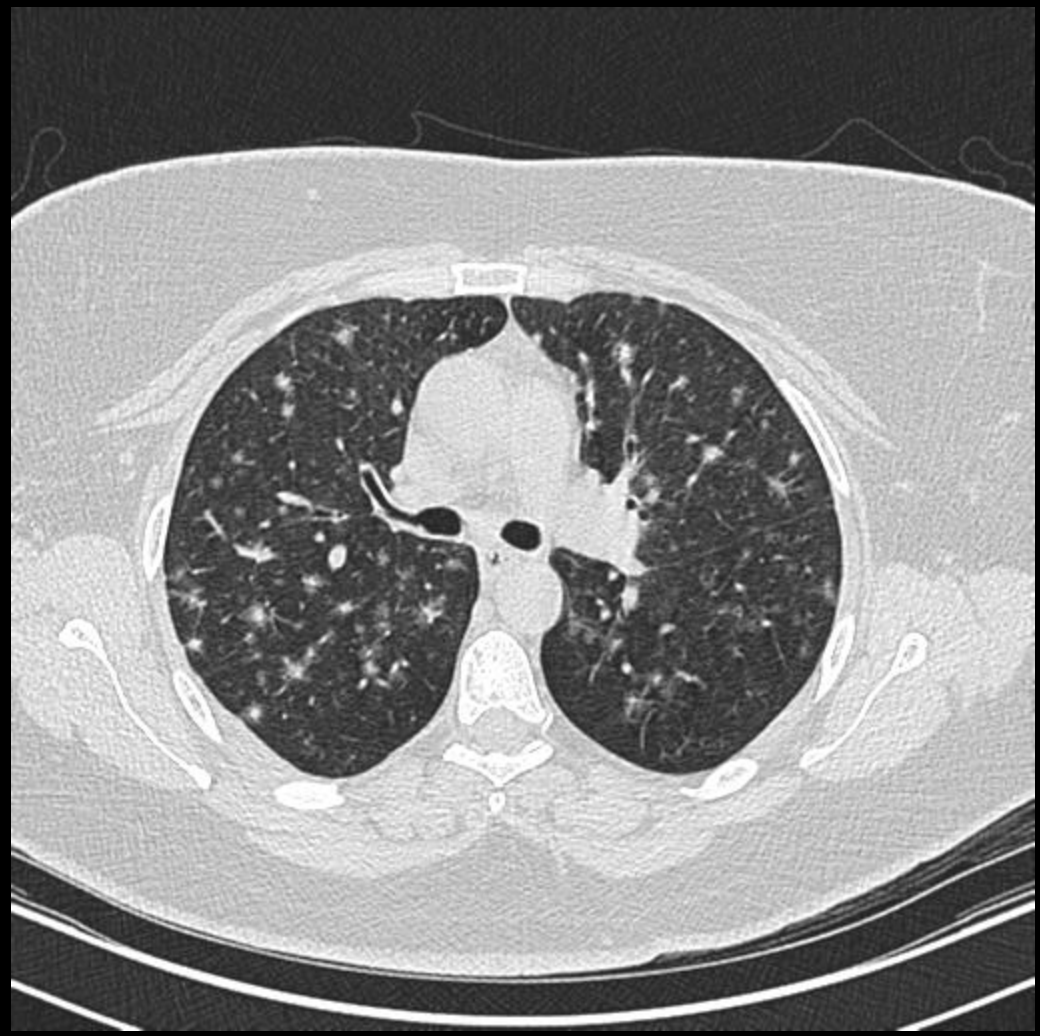

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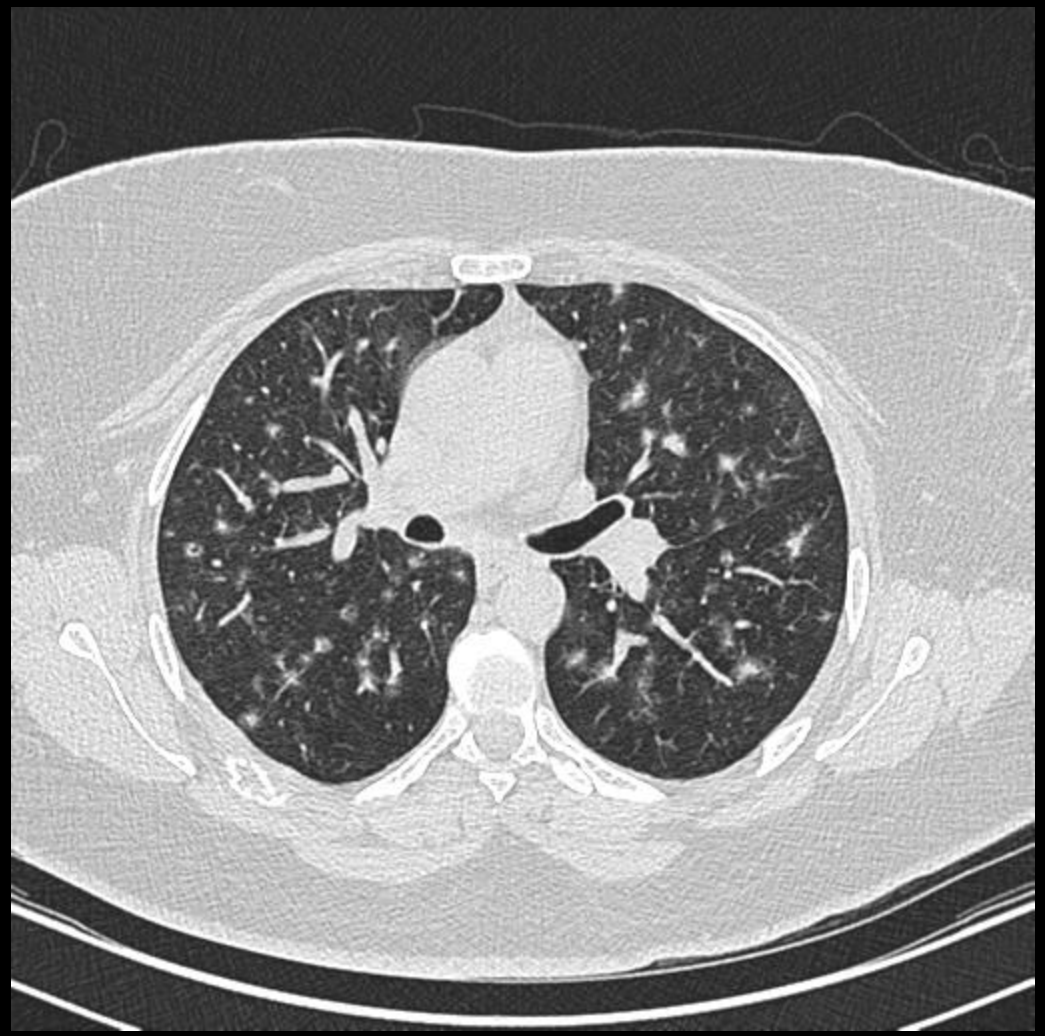

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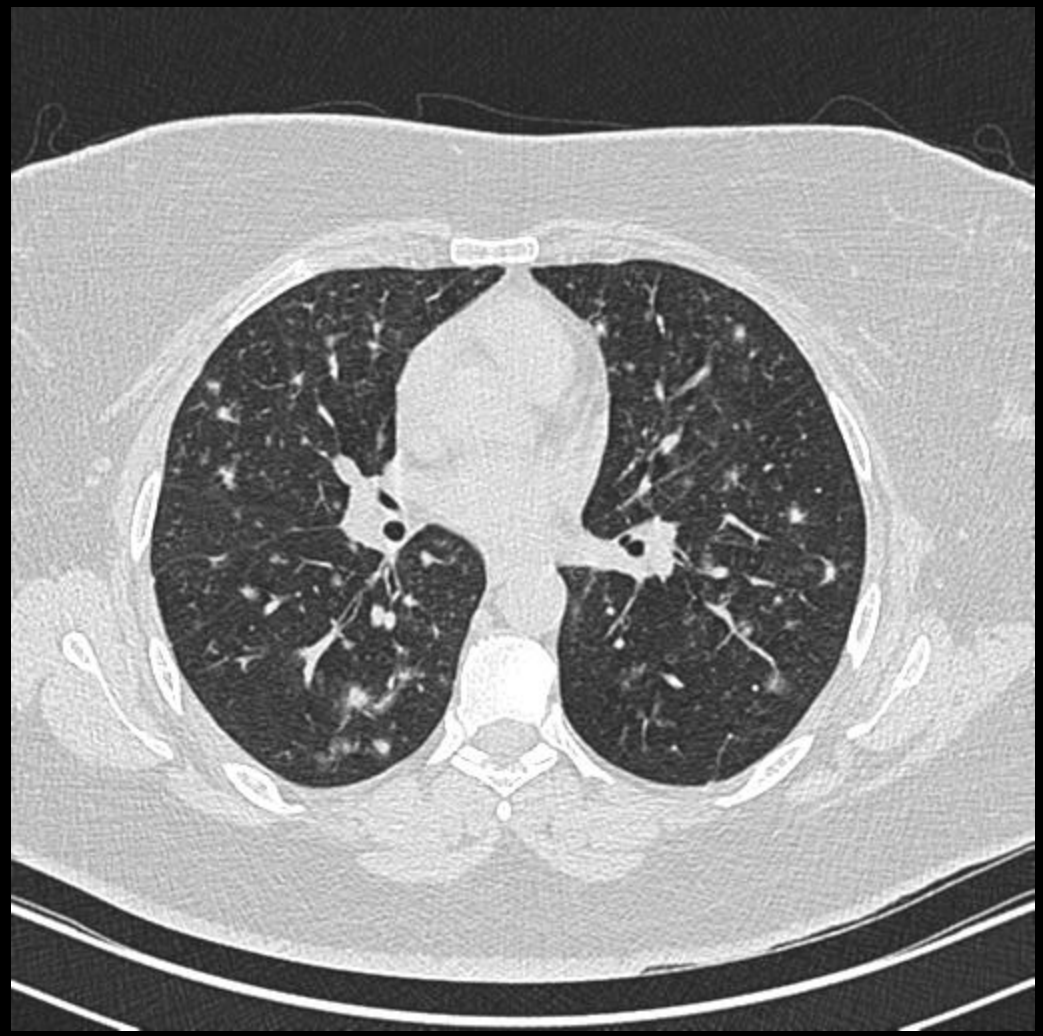

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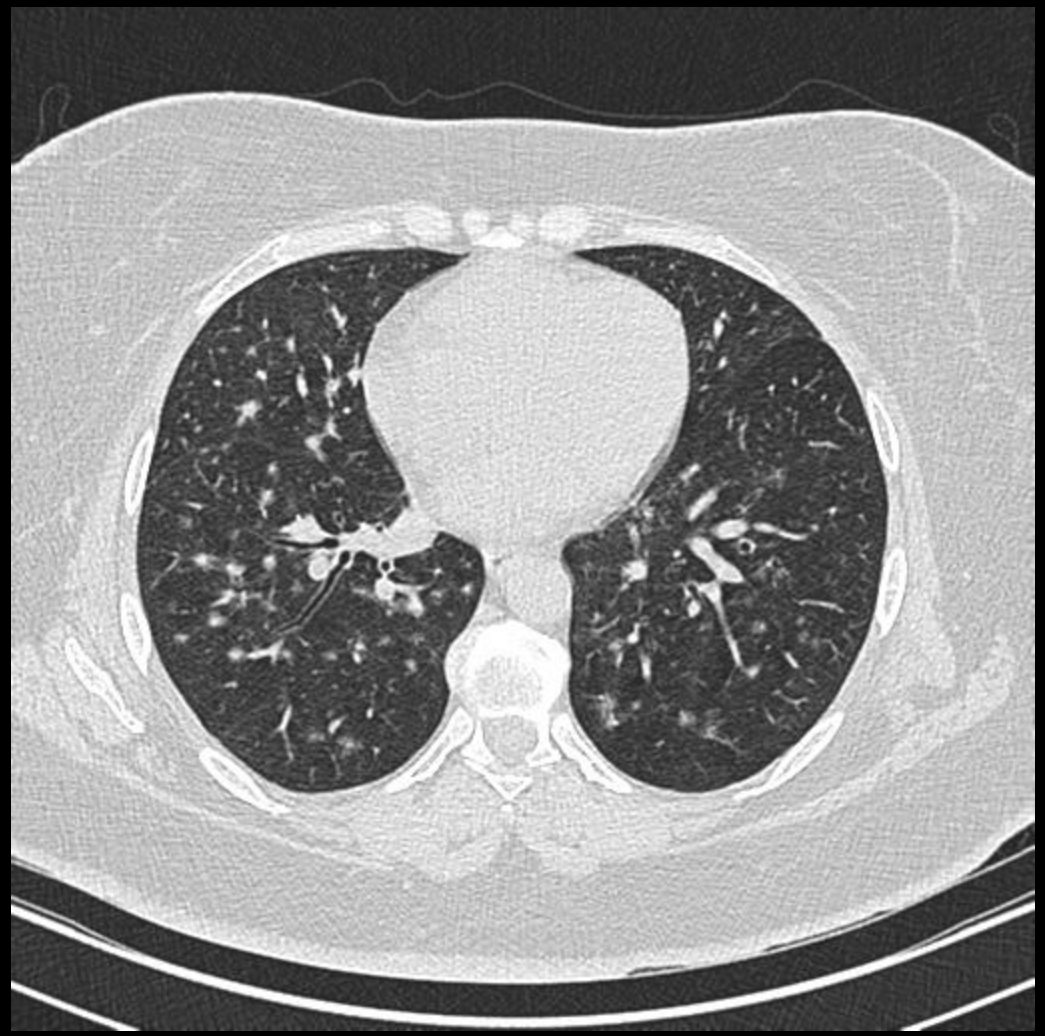
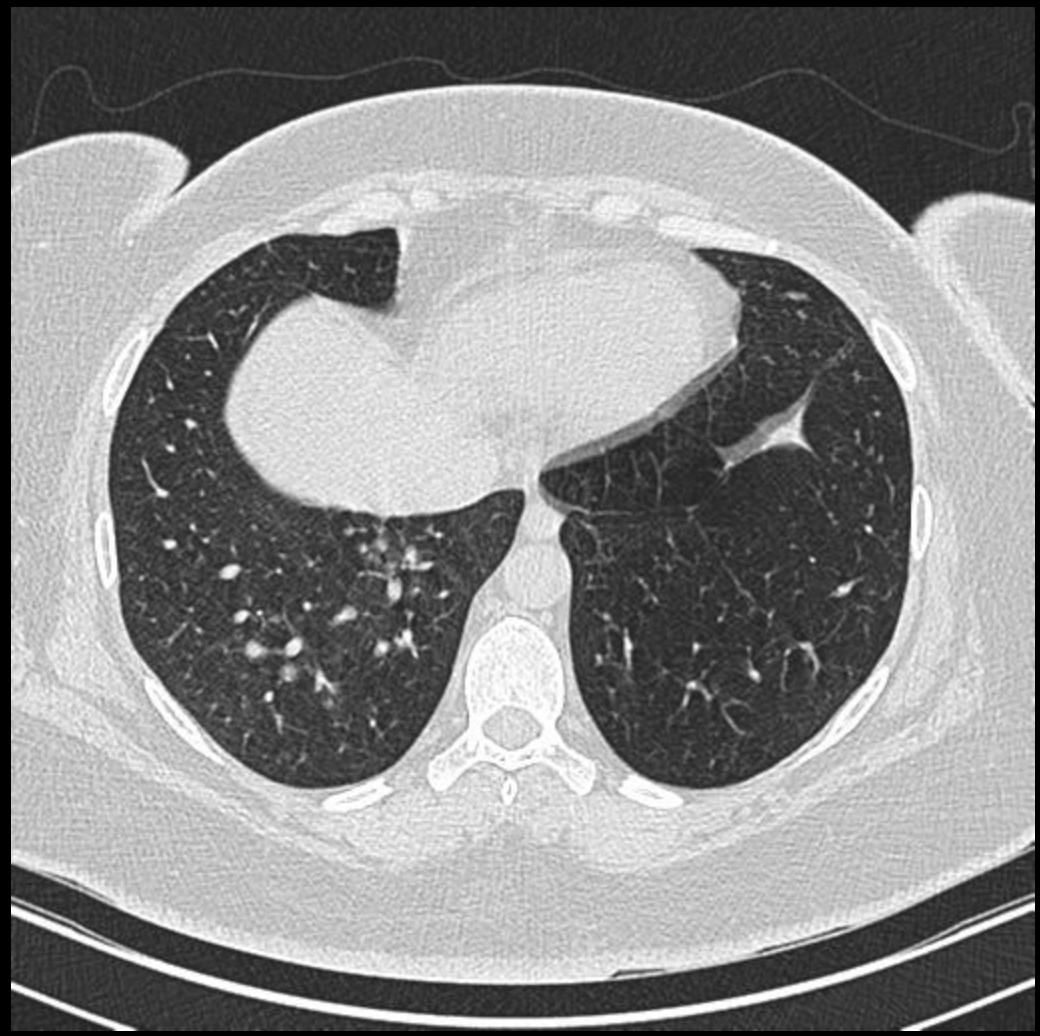
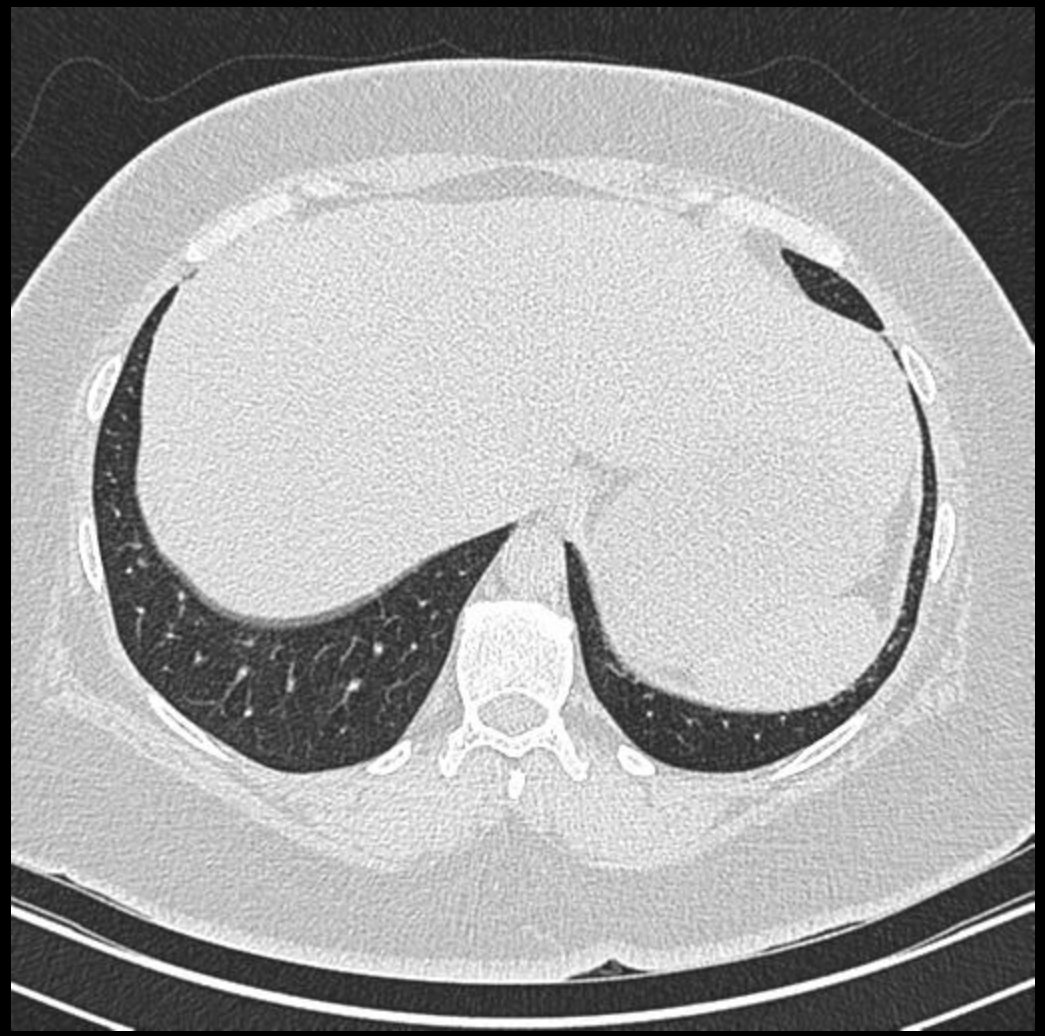
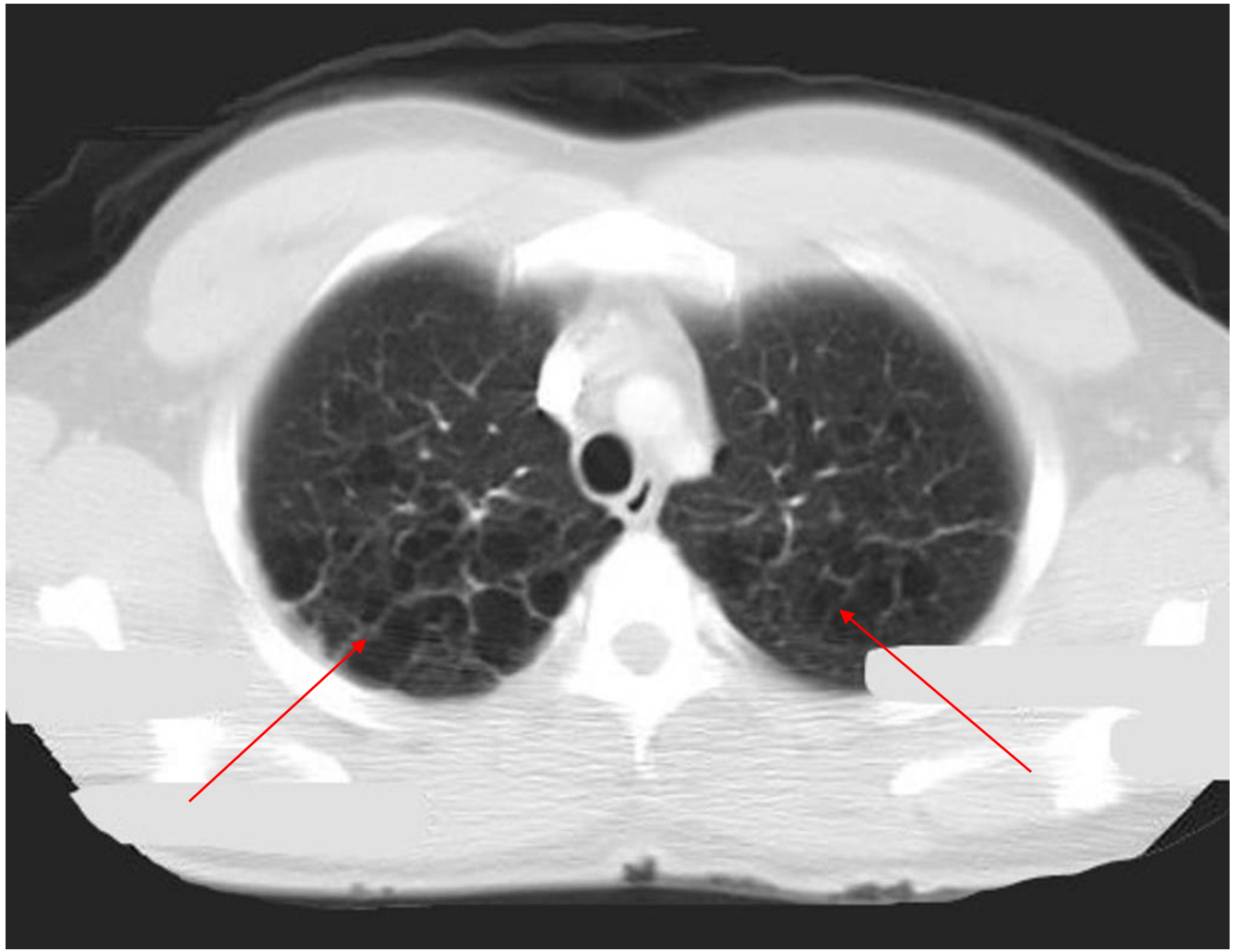






# Advanced disease: cystic predominance



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# Summary of PLCH radiographic features

- Reticular and nodular opacities
- Cysts or honeycombing
- Preservation of lung volume
- Costophrenic angle sparing
- No hilar or mediastinal lymphadenopathy

# Differential diagnosis for reticulonodular pattern on chest imaging

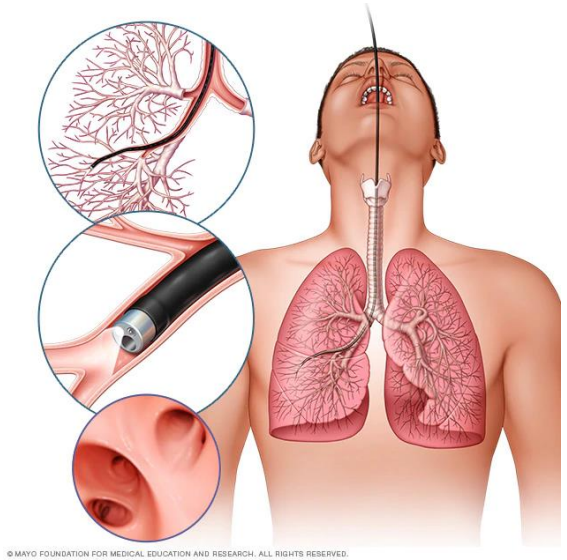
- Hypersensitivity pneumonitis
- Idiopathic interstitial pneumonias

# Differential diagnosis for cystic pattern on chest imaging

- Pulmonary lymphangioleiomyomatosis (LAM)
- Lymphoid interstitial pneumonia (LIP)
- Sarcoidosis

# Diagnosing PLCH

- Clinical findings alone: low sensitivity and specificity
  - Add HRCT: increases sensitivity, but not specificity
  - Add BAL: increases specificity, but not sensitivity

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# Other Imaging Modalities

- Gallium-67 scans are generally negative
- V/Q scans are generally negative
  - Can show non-specific findings: non-homogeneous uptake, non-segmental perfusion defects, air trapping

[6]

# Diagnosing PLCH

**Characteristic features of pulmonary Langerhans cell histiocytosis**

| | |
|---|---|
| **Clinical** | ▪ Typical age range 20 to 40 years<br>▪ Nearly all affected individuals are current or former cigarette smokers*<br>▪ May be asymptomatic, with abnormal chest radiograph, or may report dyspnea, constitutional symptoms<br>▪ History of pneumothorax<br>▪ Diabetes insipidus (<10%)<br>▪ Bone lesions (<15%) |
| **Lung function tests** | ▪ Normal or reduced lung volumes and reduced diffusing capacity<br>▪ Airflow limitation and hyperinflation less common, typically in patients with more advanced, cystic disease |
| **HRCT** | ▪ Mix of nodules (2 to 10 mm) and thick-walled cysts (early stages) or bizarrely shaped cysts varying in size and shape (advanced stages)<br>▪ Upper and mid-lung zone distribution with sparing of costophrenic angles<br>▪ Can show scattered cystic lesions without nodules<br>▪ Can show peribronchial nodules without cysts |
| **Bronchoalveolar lavage** | ▪ BAL with ≥5% CD1a-positive cells is considered diagnostic, but frequently a lower percentage of CD1a cells is noted |
| **Histopathology** | ▪ Peribronchial inflammatory lesions containing an admixture of Langerhans cells, eosinophils, lymphocytes, and neutrophils<br>▪ Langerhans-like cells express CD1a, langerin (CD207), and S100 |
| **Diagnosis based on clinical features and HRCT** | ▪ Classic HRCT features<br>▪ BAL with <5% CD1a-positive cells, but without lymphocytosis<br>▪ Improvement with smoking cessation |
| **Diagnosis based on BAL or biopsy** | ▪ Compatible clinical features and one of the following:<br>• BAL with ≥5% CD1a-positive cells<br>• Transbronchial or surgical lung biopsy demonstrating diagnostic histological features<br>• Compatible HRCT and extrapulmonary Langerhans cell histiocytosis confirmed by bone or skin biopsy |

[7]

# Take Home Points

- PLCH is an uncommon disease in young adults that presents with dyspnea, dry cough, and constitutional sx.
- Radiographically, it is characterized by a reticulonodular pattern that over time becomes more cystic.
- Often associated with smoking, cessation is typically the only treatment necessary. Corticosteroids can be used.

# References


[1] Case courtesy of Dr. Michael Sargent, Radiopaedia.org, rID: 6088

[2] Gupta, N., Vassallo, R., Wikenheiser-Brokamp, K. A., & McCormack, F. X. (2015). Diffuse cystic lung disease. Part I. American journal of respiratory and critical care medicine, 191(12), 1354-1366.

[3] https://radiologypics.com/

[4] Case courtesy of Prof Oliver Hennessy, Radiopaedia.org, rID: 33062

[5] Case courtesy of Dr. Frank Gaillard, Radiopaedia.org, rID: 9507

[6] Tazi, A. (2006). Adult pulmonary Langerhans' cell histiocytosis. European Respiratory Journal, 27(6), 1272-1285.

[7] UpToDate: Pulmonary Langerhans cell histiocytosis, King et al.