RADY 401 Case Presentation

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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
**Variant 1:**

**Chronic dyspnea. Unclear etiology. Initial imaging.**

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<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tr>
<td>Radiography chest</td>
<td>Usually Appropriate</td>
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<tr>
<td>CT chest without IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
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<tr>
<td>CT chest with IV contrast</td>
<td>May Be Appropriate</td>
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<td>CT chest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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<td>MRI chest without and with IV contrast</td>
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<td>MRI chest without IV contrast</td>
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<td>US chest</td>
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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.

[2, 6, 7]
Treatment

• Smoking cessation +/- systemic glucocorticoid therapy
  • > 60% of patients show resolution or stabilization of disease with smoking cessation alone

• Refractory cases: chemotherapy (cladribine, cytarabine)
Imaging Discussion
Early disease: nodular predominance
Advanced disease: cystic predominance
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

[6, 7]
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
## Diagnosing PLCH

### Characteristic features of pulmonary Langerhans cell histiocytosis

| **Clinical** | - Typical age range 20 to 40 years  
| - Nearly all affected individuals are current or former cigarette smokers  
| - May be asymptomatic, with abnormal chest radiograph, or may report dyspnea, constitutional symptoms  
| - History of pneumothorax  
| - Diabetes insipidus (<10%)  
| - Bone lesions (<15%) |

| **Lung function tests** | - Normal or reduced lung volumes and reduced diffusing capacity  
| - 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)  
| - Upper and mid-lung zone distribution with sparing of costophrenic angles  
| - Can show scattered cystic lesions without nodules  
| - 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  
| - Langerhans-like cells express CD1a, langerin (CD207), and S100 |

| **Diagnosis based on clinical features and HRCT** | - Classic HRCT features  
| - BAL with <5% CD1a-positive cells, but without lymphocytosis  
| - Improvement with smoking cessation |

| **Diagnosis based on BAL or biopsy** | - Compatible clinical features and one of the following:  
| - BAL with ≥5% CD1a-positive cells  
| - Transbronchial or surgical lung biopsy demonstrating diagnostic histological features  
| - Compatible HRCT and extrapulmonary Langerhans cell histiocytosis confirmed by bone or skin biopsy |
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


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

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


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