Interstitial and Cystic Lung Disease:

An Unusual Case

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Agenda

• Our patient’s...
  – Clinical presentation
  – Radiologic work-up

• Review of pulmonary anatomy on CT

• Our patient’s differential diagnosis with discussion of radiologic findings in...
  – Smoking-related interstitial lung disease
  – Cystic lung disease

• Our patient’s follow-up and working diagnosis

• Take home pearls
Our patient: Clinical presentation

- 56 y/o woman with 2 months of low back pain with left lower extremity radiculopathy w/o any inciting events
  - Paresthesic radiation down lateral aspect of left lower extremity to the soles of her feet and several digits
  - Sxs exacerbated by bending, raising her leg and sitting
  - PT, pain meds and steroid injections have not helped
- PMH: Ménière’s disease
- Social hx: 10 pack-years
- ROS: no CP, SOB, N/V or incontinence
- PE:
  - temp 97.8, HR 115, BP 134/89, RR 18, SpO2 99%
  - MSK: Limited ROM lumbar spine, tenderness of paraspinal muscles and SI joints, (+) Patrick’s test
  - Neuro: CNs intact, 5/5 strength, no sensation deficits in S1 distribution, reflexes 2+, (+) SLR
What would you like from the menu of radiologic tests?

Choose your test, then let’s continue to review the ACR Appropriateness Criteria for this clinical picture
# ACR Appropriateness Criteria

## Clinical Condition:

### Low Back Pain

### Variant 4:

Acute, subacute, or chronic low back pain or radiculopathy. Surgery or intervention candidate with persistent or progressive symptoms during or following 6 weeks of conservative management.

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI lumbar spine without IV contrast</td>
<td>8</td>
<td>MRI is preferred. CT is useful if MRI is contraindicated or unavailable and/or for problem solving.</td>
<td>0</td>
</tr>
<tr>
<td>CT lumbar spine with IV contrast</td>
<td>5</td>
<td>MRI is preferred. CT is useful if MRI is contraindicated or unavailable and/or for problem solving.</td>
<td>📉booking 📉booking 📉booking 📉booking 📉booking</td>
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</tr>
<tr>
<td>MRI lumbar spine without and with IV contrast</td>
<td>5</td>
<td>This procedure is indicated if noncontrast MRI is nondiagnostic or indeterminate. Contrast is indicated if patient has history of prior lumbar surgery. See variant 5.</td>
<td>0</td>
</tr>
<tr>
<td>X-ray myelography and post myelography CT lumbar spine</td>
<td>5</td>
<td>MRI is preferred. This procedure can be indicated if MRI is contraindicated or nondiagnostic.</td>
<td>📉booking 📉booking 📉booking 📉booking 📉booking</td>
</tr>
</tbody>
</table>
Our patient: MRI lumbar spine

Please pause to evaluate the image and continue to view findings
Our patient: MRI lumbar spine

- Synovial cyst
- Facet arthropathy
- Disc bulge
Our patient

• Our patient was scheduled for an L5-S1 facetectomy and synovial cyst removal
• For the patient’s pre-operative evaluation, a chest radiograph was ordered

Continue to view the current ACR appropriateness criteria for a pre-operative chest radiograph
As there is no clinical concern for cardiopulmonary disease on history or physical exam and the patient and procedure in this case are not high-risk, today’s guidelines would not support a chest radiograph.
Our patient: Chest radiographs

Please pause to evaluate the image and continue to view findings.
Our patient: Chest radiographs

Cystic lesion
What would you like from the menu of radiologic tests to work-up this incidental finding?

Choose your test, then let’s continue to review the ACR Appropriateness Criteria for this clinical picture.
ACR Appropriateness Criteria

**Clinical Condition:** Radiographically Detected Solitary Pulmonary Nodule

**Variant 1:** Solid nodule ≥1 cm, low clinical suspicion for cancer.

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<td>8</td>
<td>To detect occult calcifications, fat, bronchus sign, etc.</td>
<td>🌐🌐🌐</td>
</tr>
<tr>
<td>FDG-PET/CT whole body</td>
<td>8</td>
<td>If nodule is indeterminate on HRCT.</td>
<td>🌐🌐🌐🌐</td>
</tr>
<tr>
<td>Transthoracic needle biopsy</td>
<td>8</td>
<td>If nodule shows contrast enhancement or PET scan is positive.</td>
<td>Varies</td>
</tr>
<tr>
<td>CT chest with IV contrast</td>
<td>6</td>
<td>Probably not indicated if PET is performed.</td>
<td>🌐🌐🌐</td>
</tr>
<tr>
<td>CT chest without and with IV contrast</td>
<td>6</td>
<td>Can look at washout.</td>
<td>🌐🌐🌐</td>
</tr>
<tr>
<td>Watchful waiting with CT follow-up</td>
<td>4</td>
<td>Reasonable at short interval.</td>
<td>Varies</td>
</tr>
<tr>
<td>MRI chest without IV contrast</td>
<td>2</td>
<td>Limited data.</td>
<td>O</td>
</tr>
<tr>
<td>MRI chest without and with IV contrast</td>
<td>2</td>
<td>Limited data.</td>
<td>O</td>
</tr>
</tbody>
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_Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate_
Our patient: Chest CT without contrast

Please pause to evaluate the image and continue to view some of the findings
Our patient: Chest CT without contrast

Irregular nodules

Large cysts

Ground-glass opacities
Let’s pause to review some of the pulmonary anatomy and their radiologic correlates demonstrated by our patient’s CT.
Anatomy Review: Secondary Pulmonary Lobule

Nodule Distribution (in relation to secondary lobule)

Centrilobular distribution
Periphery spared

Terminal bronchiole
Centrilobular artery
Acinus

Interlobular septum
- Lymphatics
- Pulmonary veins

Ground-glass opacification

Radiologyassistant.nl
Let’s now discuss the differential diagnoses most consistent with our patient’s radiologic presentation
Differential diagnosis

- Smoking-related interstitial lung disease
  - Respiratory bronchiolitis-associated interstitial lung disease (RB-ILD)
  - Desquamative interstitial pneumonia (DIP)
- Cystic lung disease
  - Pulmonary Langerhans cell histiocytosis (PLCH)
  - Lymphangioleiomyomatosis (LAM)
  - Lymphocytic interstitial pneumonia (LIP)
RB-ILD

• **Respiratory bronchiolitis-interstitial lung disease**
  • **Clinical features**
    • 30+ pack-years
    • 30-40 y/o
    • Progressive SOB, chronic cough, inspiratory crackles (~50%)
  • **Diagnostic findings**
    • **HRCT**
      • Centrilobular nodularity
      • Ground-glass opacity
      • Bronchial wall thickening
      • Upper lobe predominance
    • **PFT:** nl or mixed obstructive
    • **Bx:** Pigmented macrophages

Image: A.Prof Frank Gaillard, Radiopaedia.org, rID: 6535
Mavridou & Laws, 2004
DIP

- Desquamative interstitial pneumonia
  - End-stage RB-ILD
  - Clinical features
    - 40-50 y/o
    - M:F 2:1
    - Dyspnea, dry cough, inspiratory crackles (60%), digital clubbing (50%)

- Diagnostic findings
  - HRCT
    - Ground-glass opacity
    - Reticular opacities
    - Subpleural and basal predominance
  - PFT: abnl (esp. DLCO < 50%)
  - Bx: More diffuse pigmented macrophages

Attili et al., 2008
• Pulmonary Langerhans cell histiocytosis
  • Clinical features
    • Heavy smokers
    • 20-40 y/o
    • Dry cough (60%), dyspnea (60%)
    • 25% asx
  • Diagnostic findings
    • HRCT
      • Cysts (most common)
      • Middle and upper zone irregular nodules
      • Lesion sequence: nodules → cysts
    • PFT: reduced DLCO
    • Bx: stellate nodules, CD1a IHC

Abbott et al. 2004
LAM

- Lymphangioleiomyomatosis
  - Clinical features
    - Women of childbearing age
    - Dyspnea (59%), dry cough (39%)
    - Crackles
    - Extrapulmonary manifestations
  - Diagnostic findings
    - HRCT
      - Diffuse bilateral thin-walled cysts (does not spare bases)
      - Intervening parenchyma anodular
    - PFT: obstructive, reduced DLCO
    - Bx: LAM cells (HMB-45 IHC)

Abbott et al. 2005
LIP

- Lymphocytic interstitial pneumonia
  - Clinical features
    - Mostly women, 50s
    - Strong autoimmune disease association (esp. Sjögren’s)
    - Dyspnea (61%), cough (71%)
  - Diagnostic findings
    - HRCT
      - Ground-glass attenuation
      - Centrilobular nodules
      - Interstitial thickening
      - Thin-walled perivascular cysts
    - PFT: reduced lung volumes with preserved flow
    - Bx: alveolar septal infiltration with polyclonal lymphocytes, plasma cell and histiocytes

Image: Dr Sinéad Culleton, Radiopaedia.org, rID: 40514
Ferguson & Berkowitz, 2012
Let’s return to our patient’s clinical course...

• Spirometry, lung volumes and DLCO normal
• Asked to avoid smoking and second-hand smoke
• No clinical complaints
• Monitored with CTs 1-2x/year
Our patient: Chest CT without contrast

Please pause to evaluate the image and continue to view findings
Our patient: Chest CT without contrast

2010

Cyst → Nodule

2016
Our patient: Chest CT without contrast

Let’s follow one of our patient’s lesions which first presented as a nodule
Our patient’s follow-up

- From 2010-2016...
  - PFTs remained normal
  - Systemic inflammatory/autoimmune markers all negative
  - CTs 1-2x/year
    - Most nodules stable
    - Some nodules grew but always low-suspicion for malignancy
    - Several minimally invasive biopsies were non-diagnostic
  - 2016, VATS right lower lobe wedge resection ruled out malignancy
Our patient’s working diagnosis and prognosis

• Nodule $\rightarrow$ cystic lesion sequence suggests pulmonary Langerhans cell histiocytosis (PLCH)

• As long as patient continues to be asymptomatic, no intervention or further imaging will be performed from 2017 onwards

• Instead, patient will undergo spirometry and DLCO every 6 months and chest imaging only if symptomatic
Take Home Pearls

• The utility of pre-operative chest radiographs in guiding treatment is controversial
  • Research suggests that they should only be used before high-risk procedures and/or in high-risk patients
  • Incidental findings may lead to adverse outcomes, especially for asymptomatic patients

• Smoking-related interstitial lung disease
  • **Desquamative interstitial pneumonia (DIP)** represents end-stage **Respiratory-bronchiolitis interstitial lung disease (RB-ILD)**
    • Both: centrilobular nodularity, ground-glass opacification
    • RB-ILD: upper lobe predominance
    • DIP: subpleural/basal predominance, worse PFTs, more diffuse histological changes
• Cystic lung diseases have unique etiologies, effects on pulmonary function, and findings on biopsy and HRCT; however they all typically present with dyspnea and cough
  • **Pulmonary Langerhans cell histiocytosis (PLCH)**
    • Heavy smokers
    • Middle/upper zone cysts and nodules that may become cystic
  • **Lymphangioleiomyomatosis (LAM)**
    • Women of child-bearing age
    • Diffuse thin-walled cysts with anodular intervening parenchyma
  • **Lymphocytic interstitial pneumonia (LIP)**
    • Underlying systemic disease
    • Centrilobular nodules, perivascular cysts, interstitial thickening
References


Acknowledgments

• Out patient
• Fourie Bezuidenhout, MD
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• Gillian Lieberman, MD