AMSER Case of the Month August 2025

67-year-old woman with history of Sjögren's disease presenting with cough and dyspnea on exertion

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Patient Presentation

- HPI: A 67-year-old woman is referred to a pulmonologist for > 6 months of cough and dyspnea on exertion.
- PMH: Sjögren's disease (positive ANA, SSa and SSb) complicated by MALT lymphoma
- Social history: Quit smoking 25 years ago, 30 pack year smoking history
- PSH, medications, and family history non-contributory

What Imaging Should We Order?

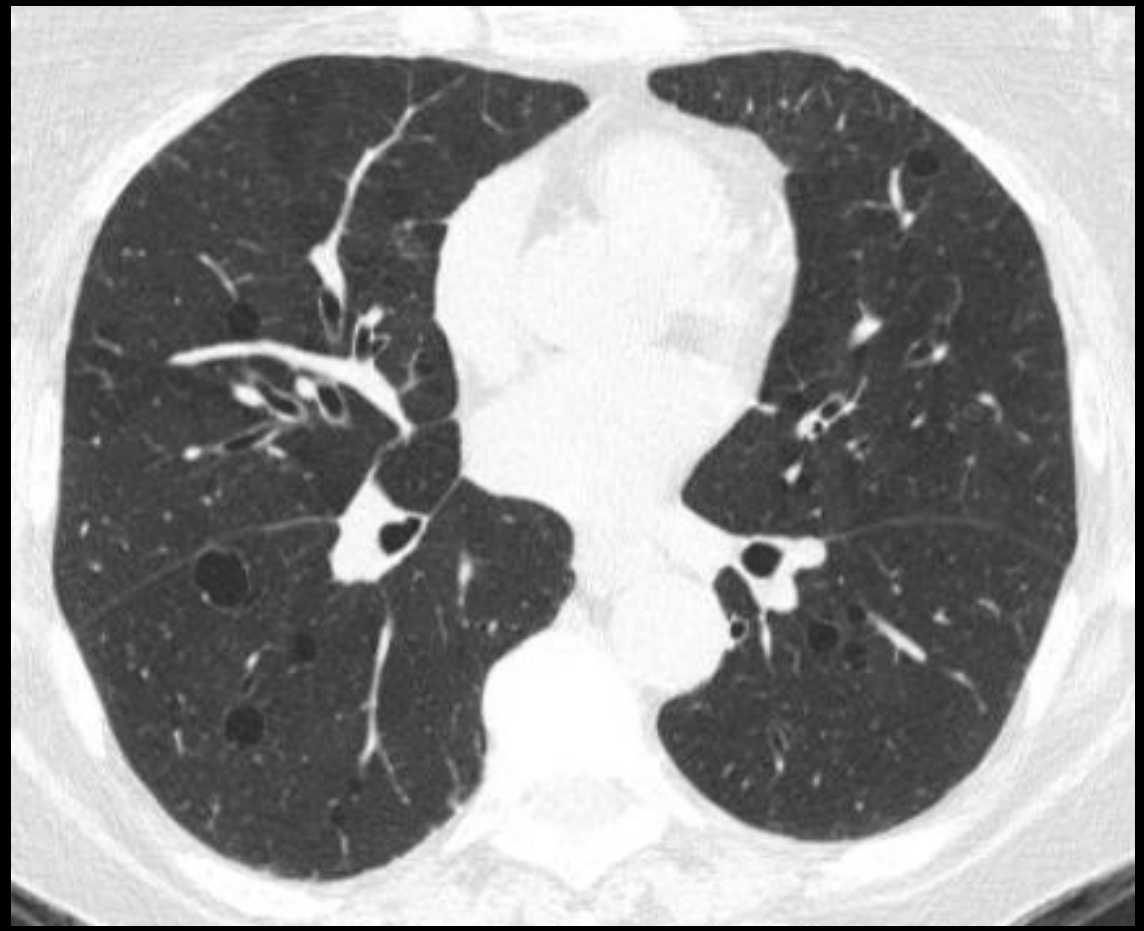
Select the applicable ACR Appropriateness Criteria

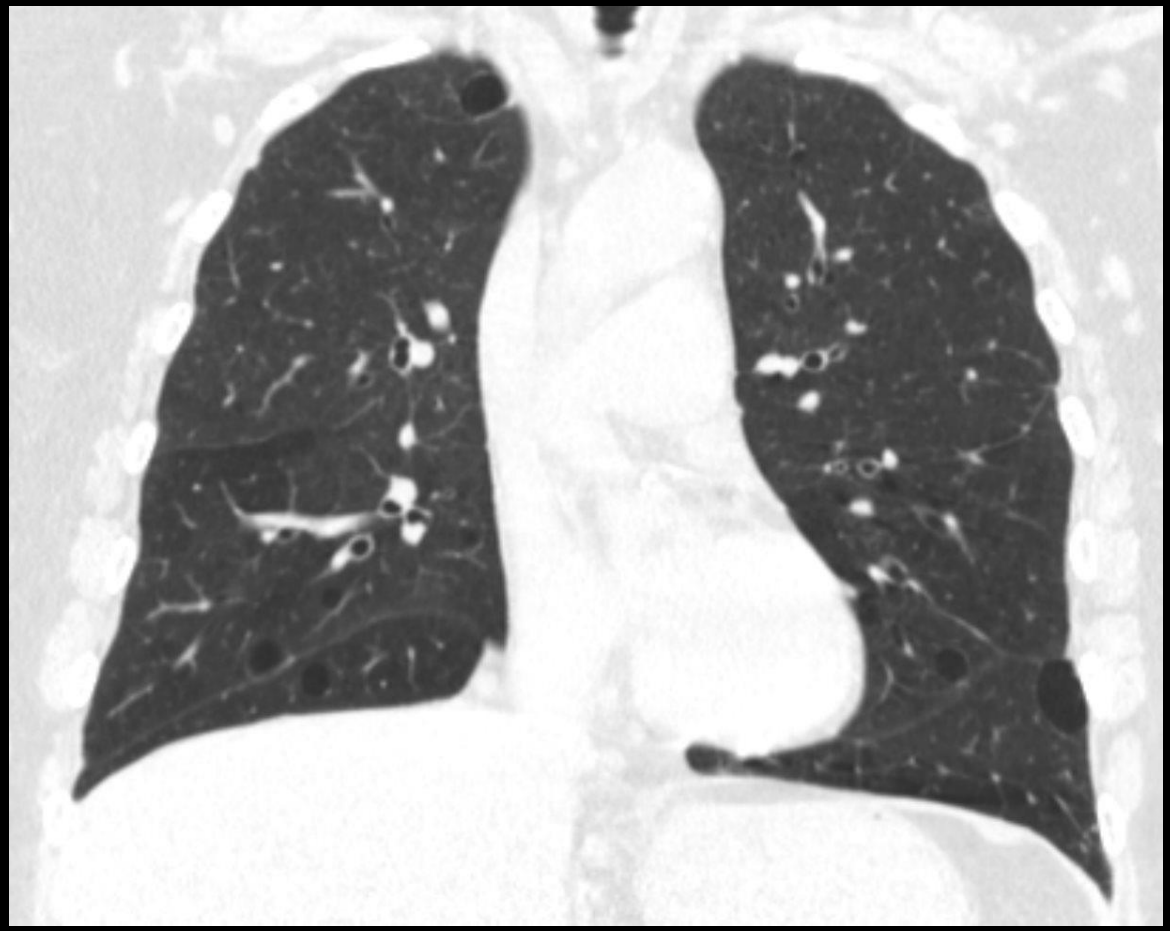
Suspected diffuse lung disease. Initial imaging.

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

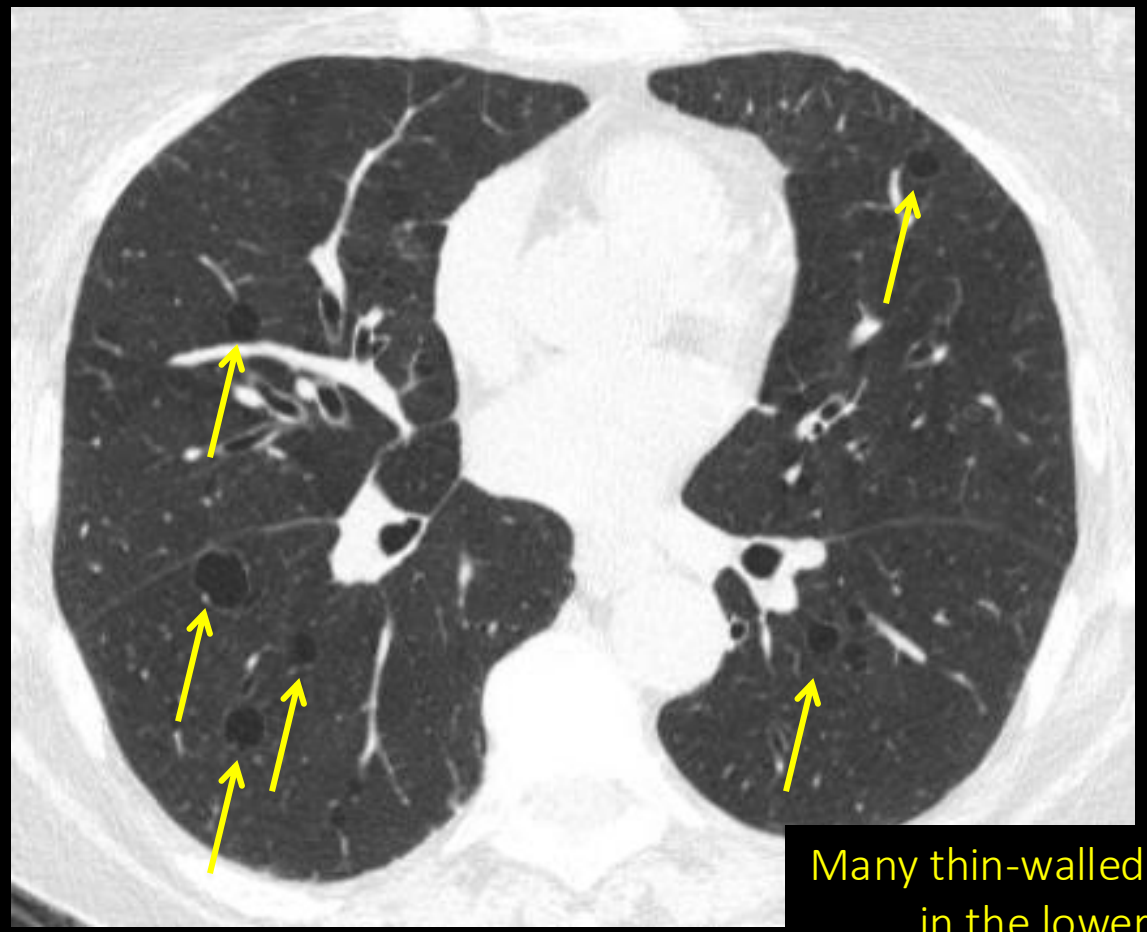
This imaging modality was ordered by the ER physician

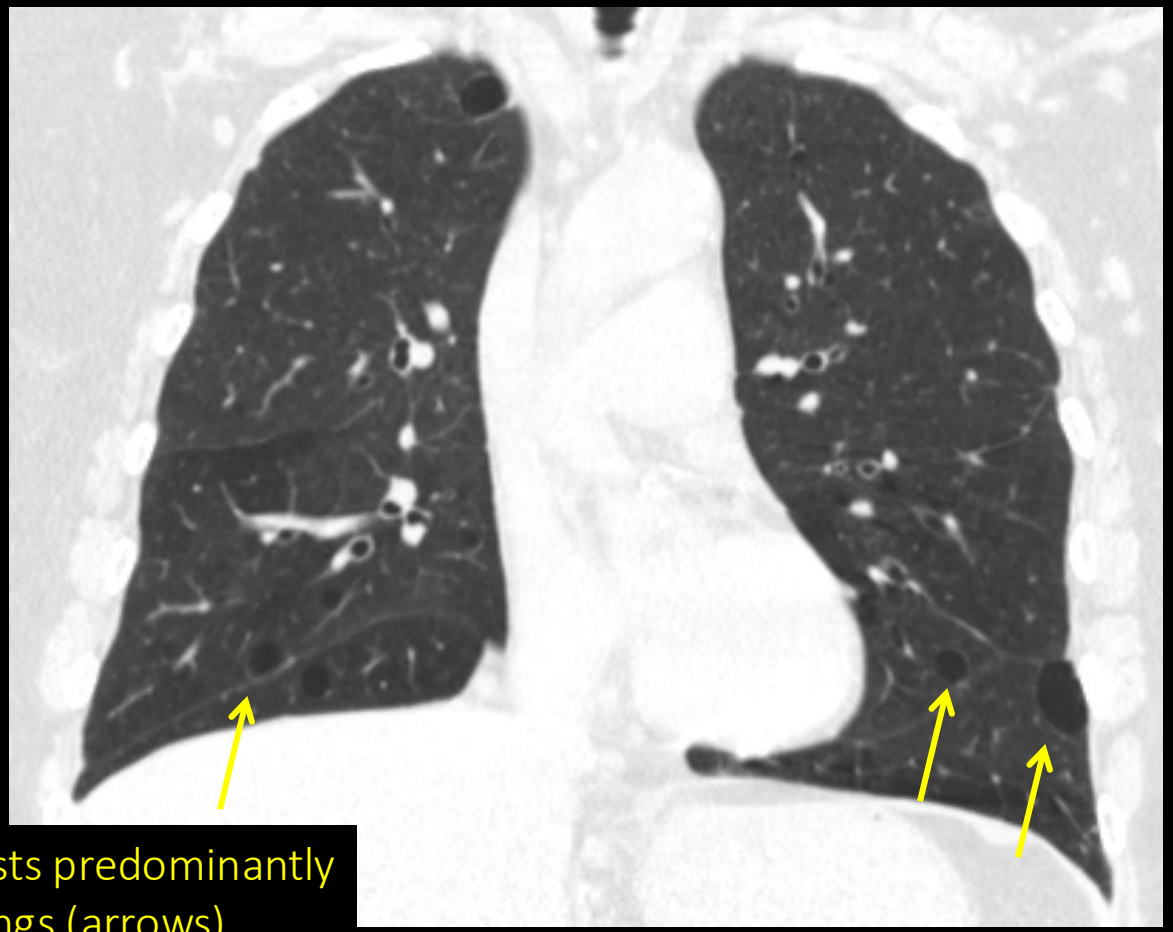
Findings (unlabeled)





Findings (labeled)





Many thin-walled cysts predominantly in the lower lungs (arrows)

Final Dx:

Lymphoid interstitial pneumonia (LIP)

Case Discussion

Typical imaging findings [1]

- Thin-walled cysts; lower lung predominance
- May also have centrilobular nodules, ground glass opacities, enlarged lymph nodes, and/or septal thickening

Etiology

- Classically thought to be due to interstitial lymphocytic infiltrates pathologically [2]
- Recent studies show discordance between radiologic and pathologic diagnosed LIP[3]
    - Controversy regarding this diagnosis includes whether an idiopathic form exists and the pathophysiologic mechanisms at play

Case Discussion

Associations [4]

- Autoimmune connective tissue disease (especially Sjögren's disease)
- Infections (HIV and EBV)
- Most patients have dysproteinemia (often polyclonal gamma globulins)

Treatment [4]

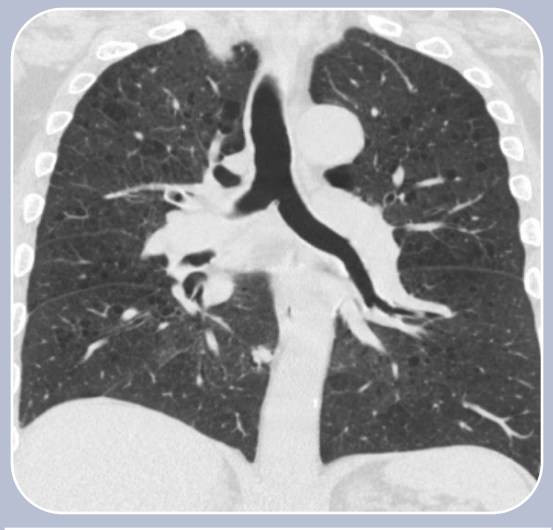
- Treated with corticosteroids, management of underlying conditions

Prognosis and complications [4]

- Variable- some patients spontaneously improve, some develop pulmonary fibrosis
- Five year mortality is estimated to be 30-50% (although studies are limited by small sample size and advanced age of participants)
- Roughly 5% of patients acquire B-cell lymphoma

Case Discussion

Differential diagnosis for multiple cysts on CT [1]

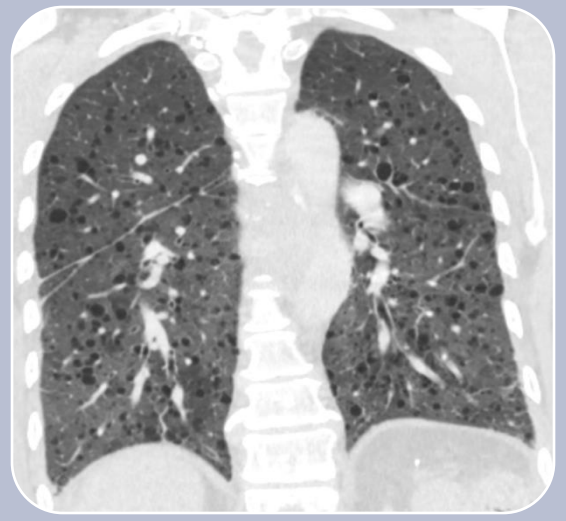


Pulmonary Langerhans cell histiocytosis

Upper lung predominant

Bizarre shapes with ± thick walls ± nodules

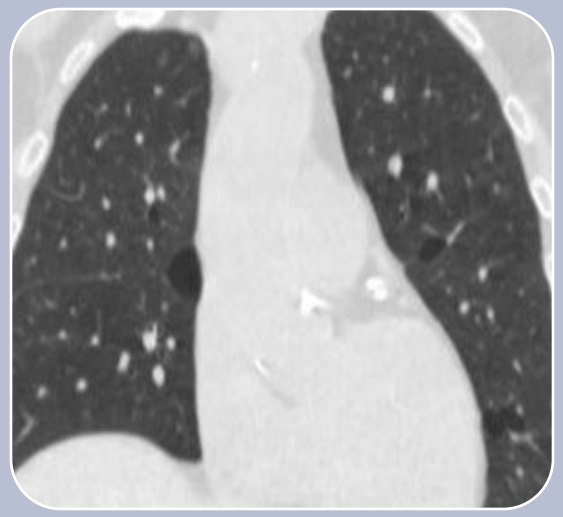
Smoking related



Lymphangioleio-myomatosis

Diffuse

Association with tuberous sclerosis or can see form with renal AMLs in women of childbearing age

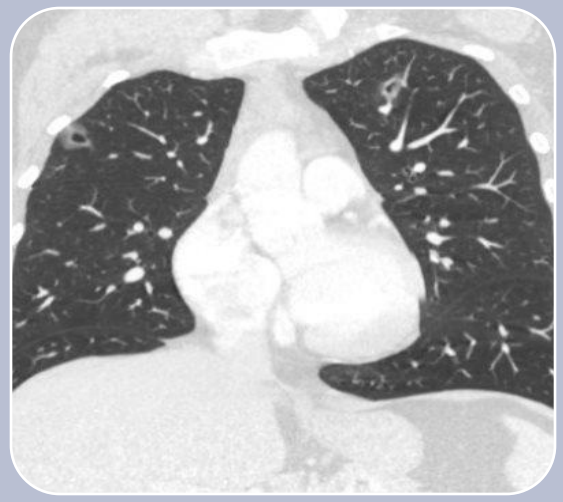


Birt-Hogg-Dubé

Lower lung predominant

Cysts may be elongated

Other manifestations include cutaneous fibrofolliculomas and renal tumors



Metastasis

Variable distribution

Various tumors can lead to cystic metastatic disease

References:

1. Raoof S, Bondalapati P, Vydyula R, et al. Cystic Lung Diseases: Algorithmic Approach. Chest. 2016;150(4):945-965. doi:10.1016/j.chest.2016.04.026
2. Panchabhai TS, Farver C, Highland KB. Lymphocytic interstitial pneumonia. Clinics in Chest Medicine. 2016;37(3):463-474. doi:10.1016/j.ccm.2016.04.009
3. Fraune C, Churg A, Yi ES, et al. Lymphoid interstitial pneumonia (Lip) revisited: a critical reappraisal of the histologic spectrum of “radiologic” and “pathologic” lip in the context of diffuse benign lymphoid proliferations of the lung. American Journal of Surgical Pathology. 2023;47(3):281-295. doi:10.1097/PAS.0000000000002014
4. Swigris JJ, Berry GJ, Raffin TA, Kuschner WG. Lymphoid interstitial pneumonia. Chest. 2002;122(6):2150-2164. doi:10.1378/chest.122.6.2150