AMSER Case of the Month January 2021

45-year-old female with chest pain

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Patient Presentation

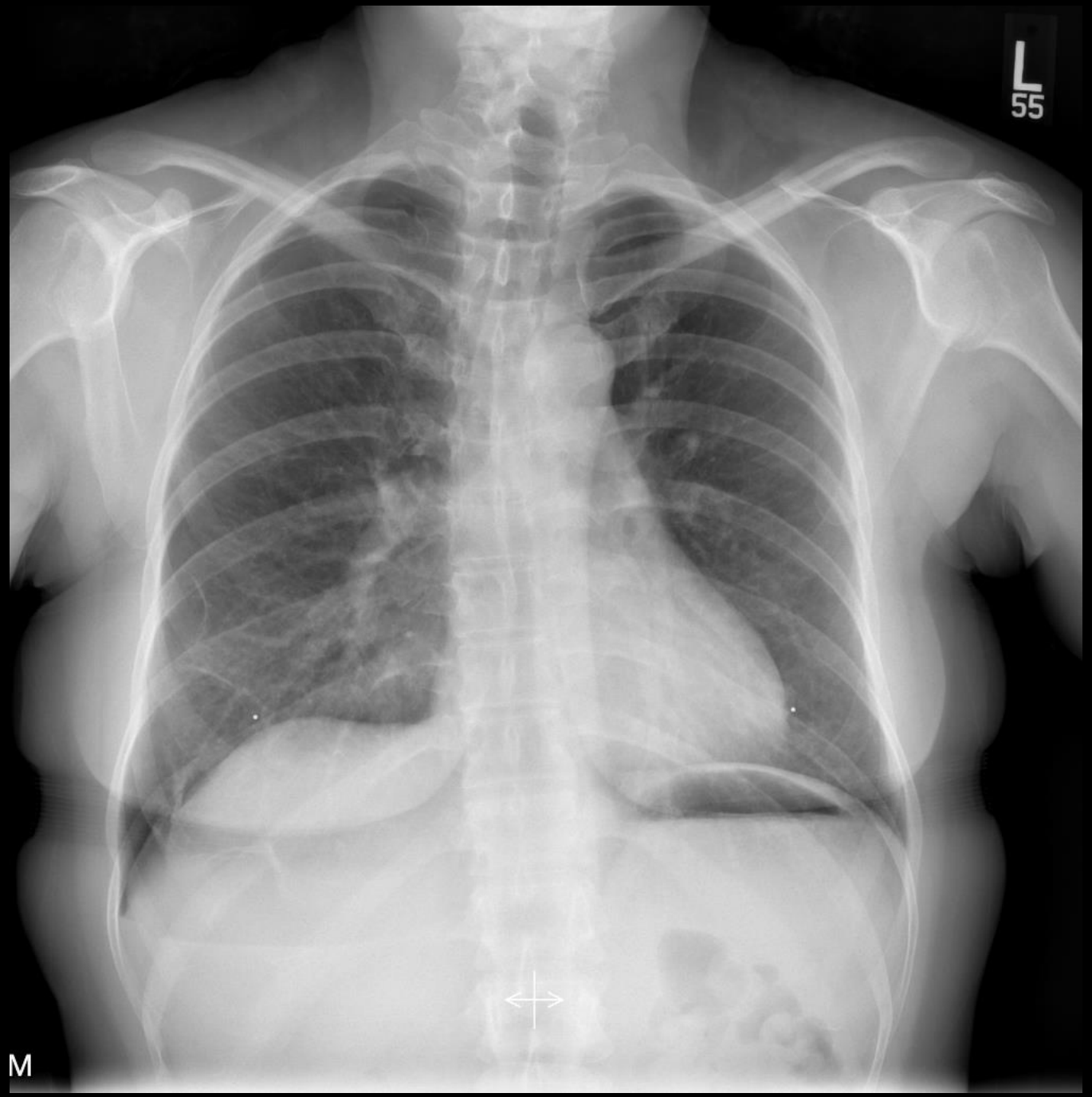
- HPI: 45-year-old female with history of “cysts” in lungs diagnosed at outside hospital, recurrent PTX status-post right mechanical pleurodesis presents to pulmonary clinic with right sided chest pain and SOB.
- SH: Non-smoker.
- FH: No history of COPD or ILD.
- ROS: Endorses pleuritic CP. Denies fevers, night sweats, HA, cough, palpitations, weight loss, fatigue.
- PE: Afebrile, BP 108/71, P 87, clear breath sounds, no respiratory distress.

Select the applicable ACR Appropriateness Criteria

Variant 4: **Chronic dyspnea. Suspected interstitial lung disease. Initial imaging.**

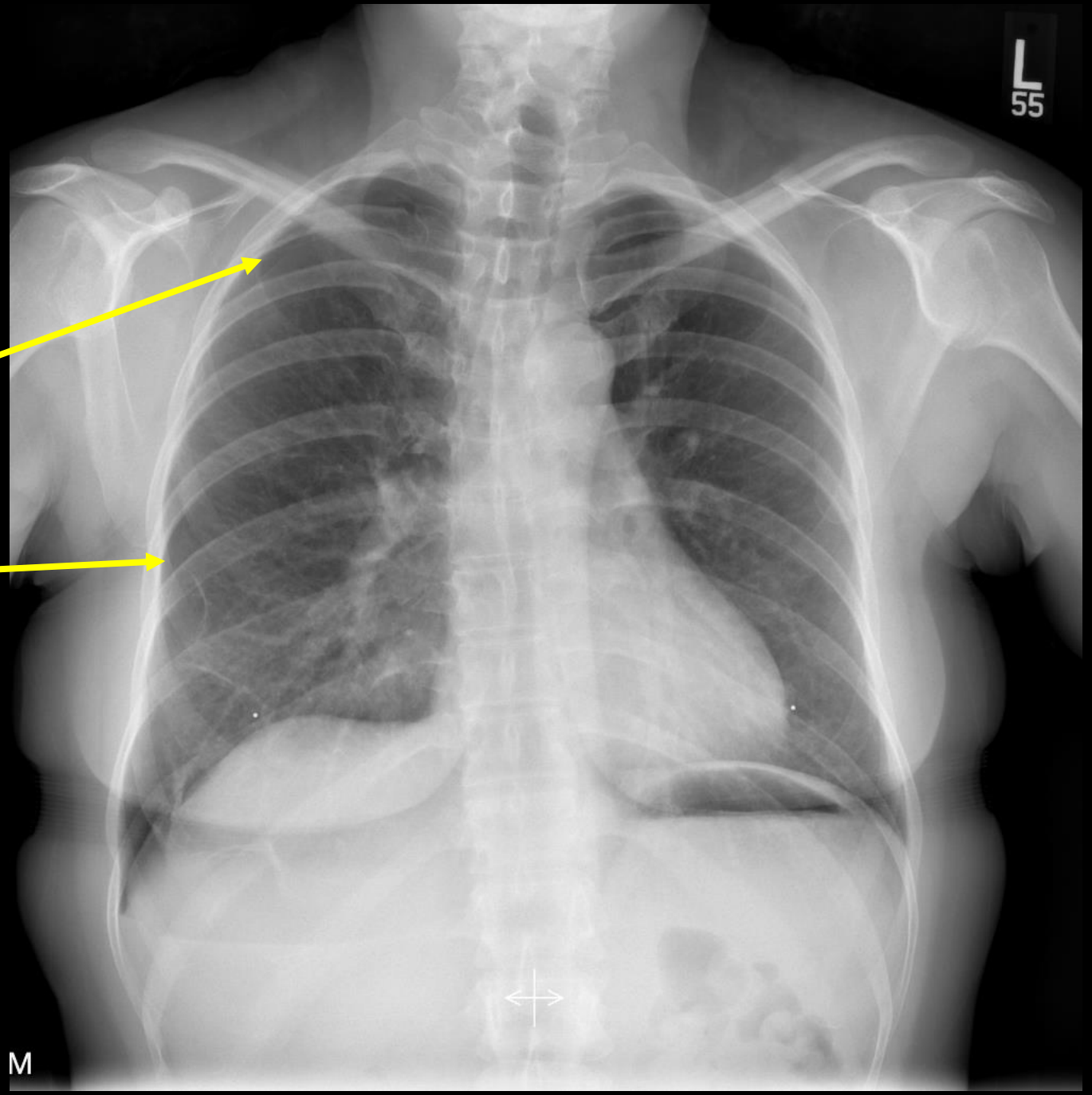
Procedure	Appropriateness Category	Relative Radiation Level
CT chest without IV contrast	Usually Appropriate	☢☢☢
Radiography chest	Usually Appropriate	☢
CT chest with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
MRI chest without and with IV contrast	Usually Not Appropriate	O
MRI chest without IV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

CXR (unlabeled)

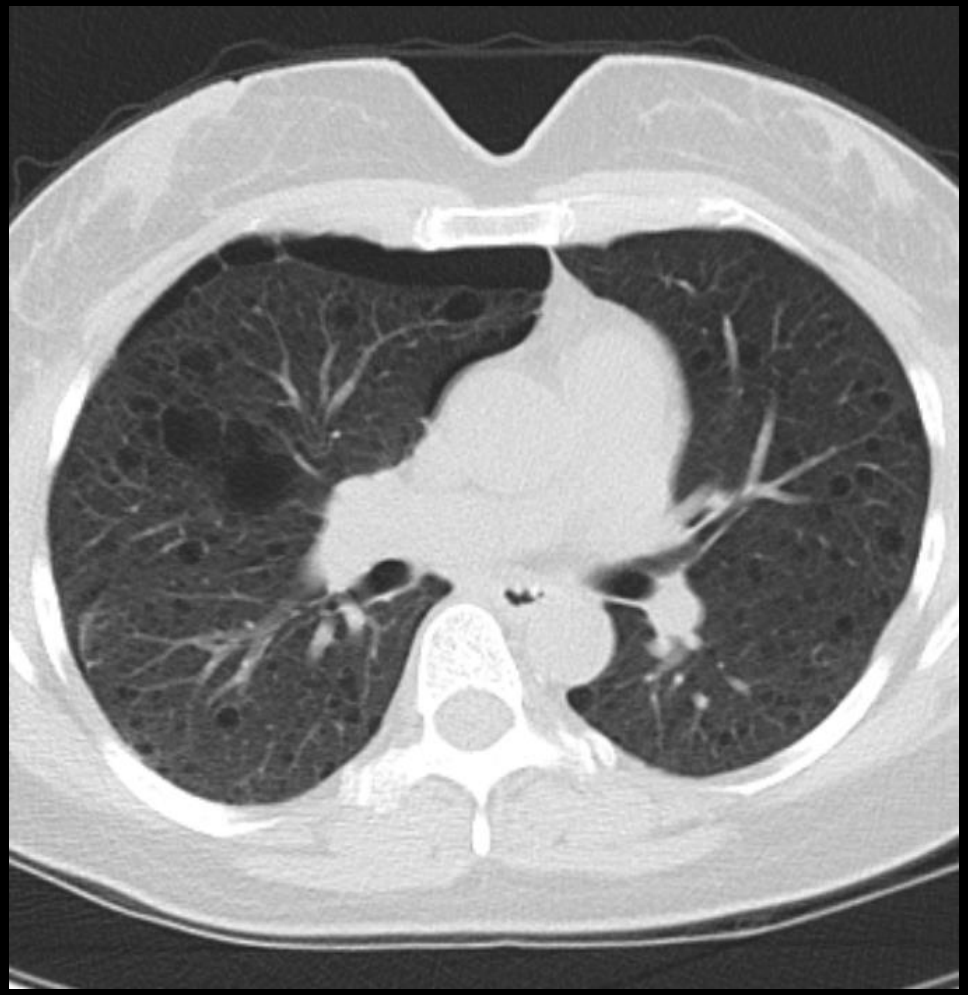


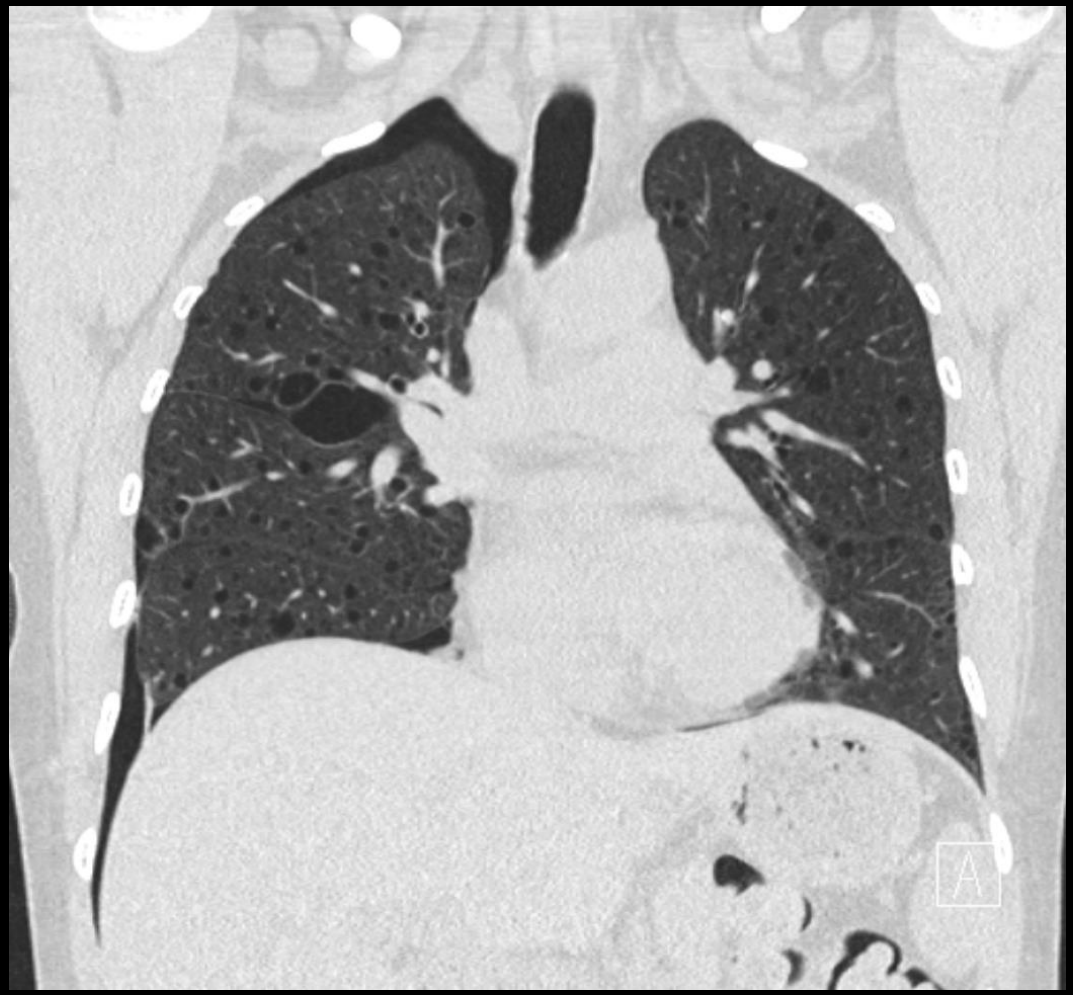
CXR (labeled)

Complex pneumothorax on the right with both apical and basilar components

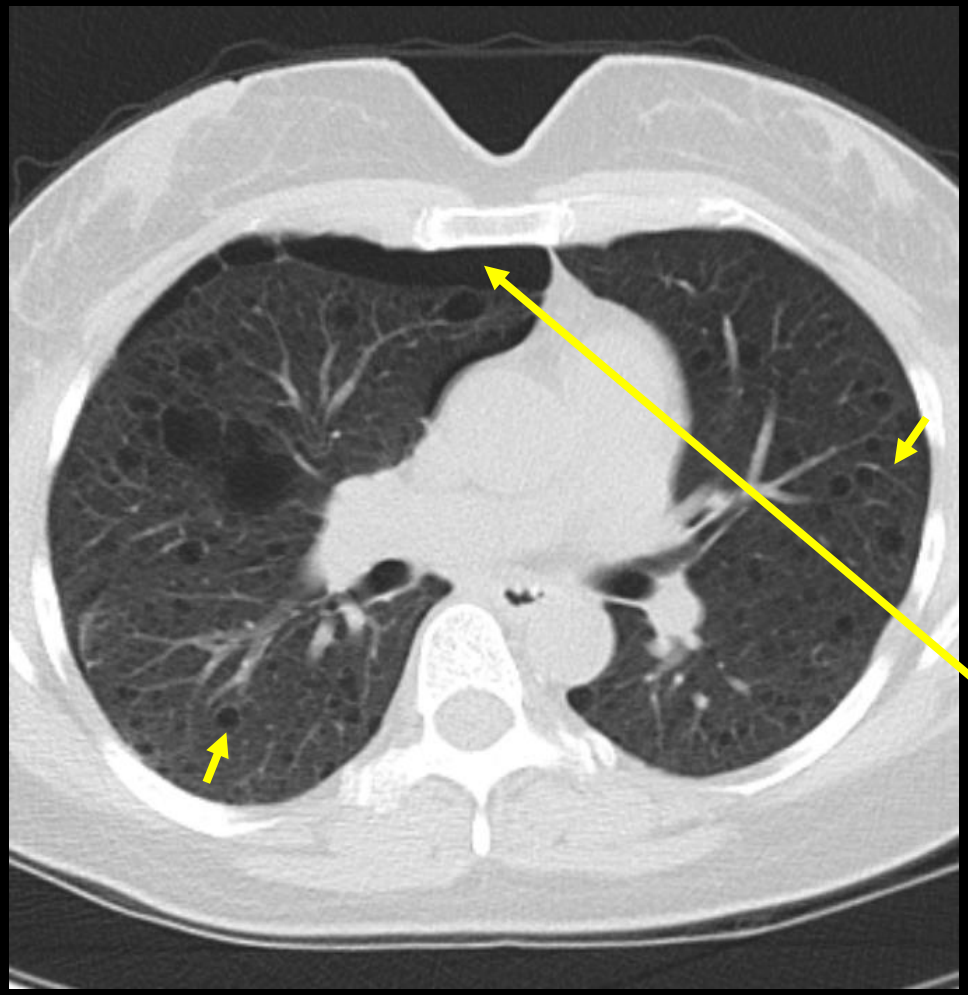


CT (unlabeled)

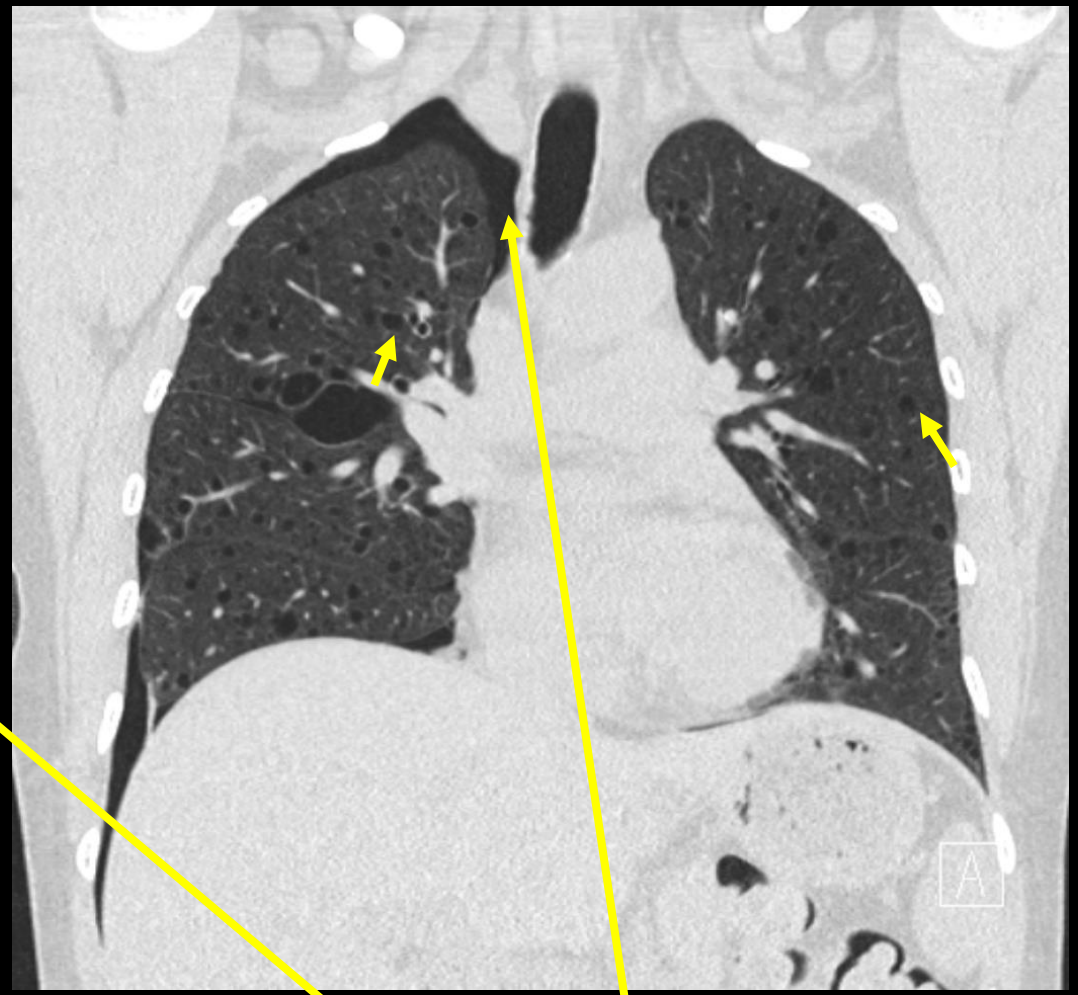




CT (labeled)

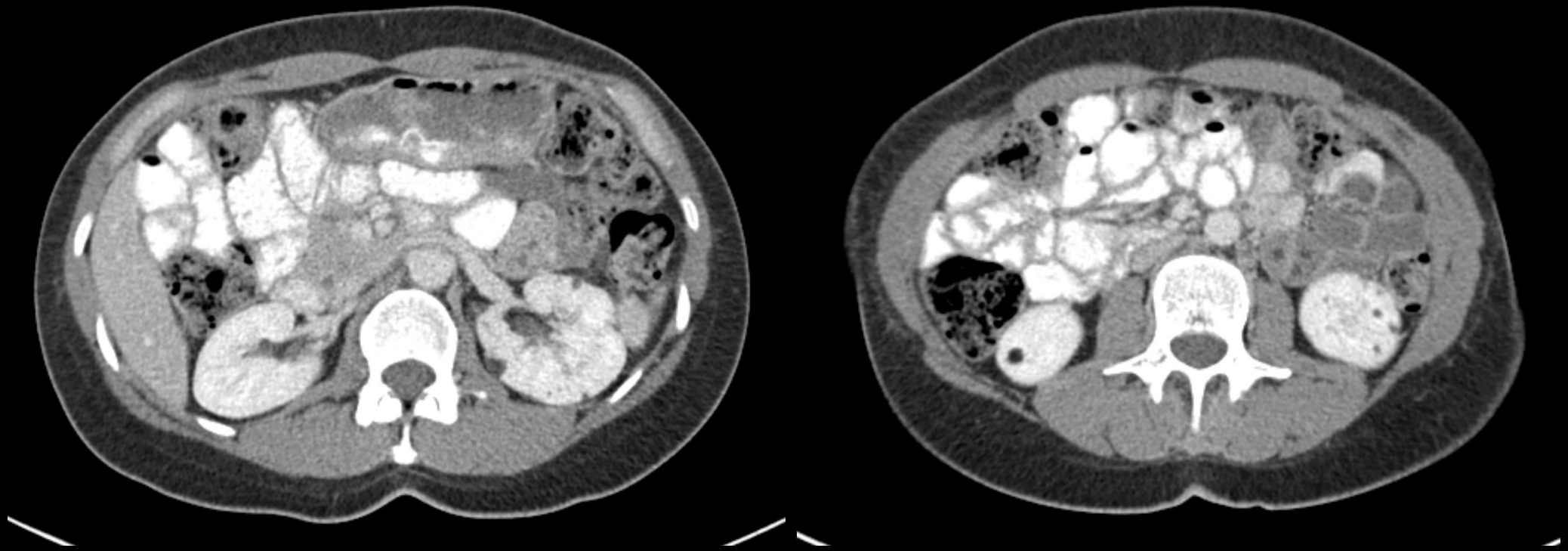


Small cystic lucencies seen throughout both lungs

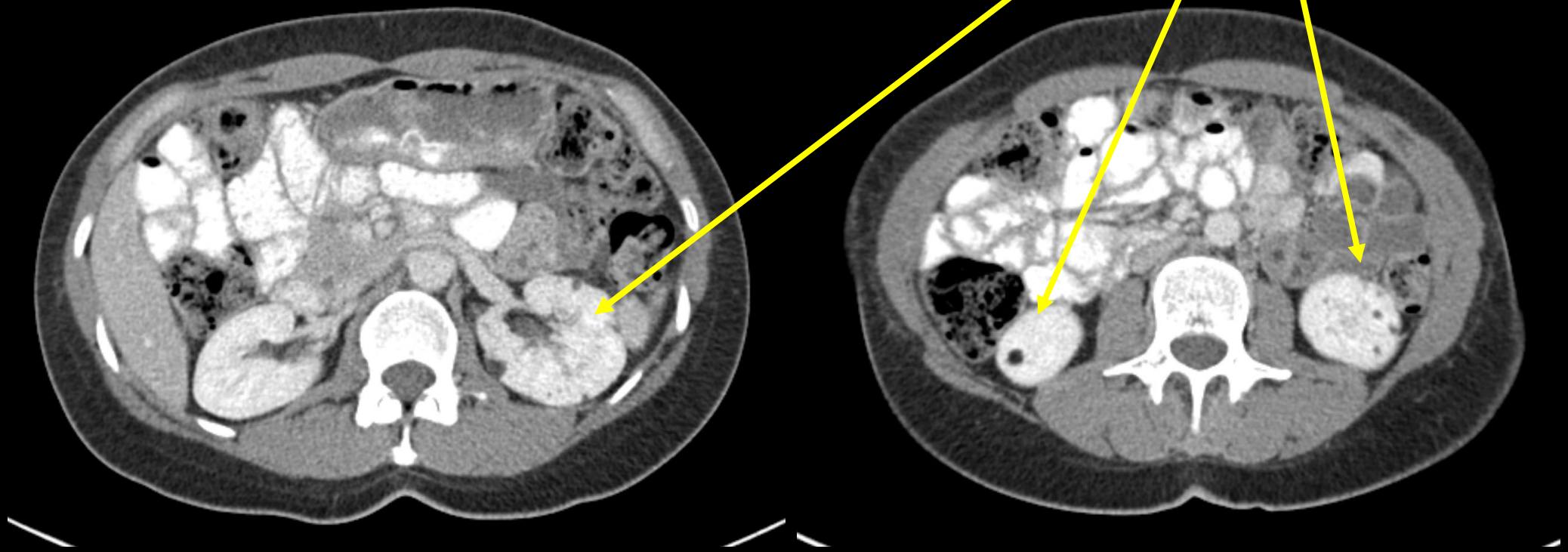


Small loculated right pneumothorax

Additional CT imaging (unlabeled)



Additional CT imaging (labeled)



DDX (based on imaging)

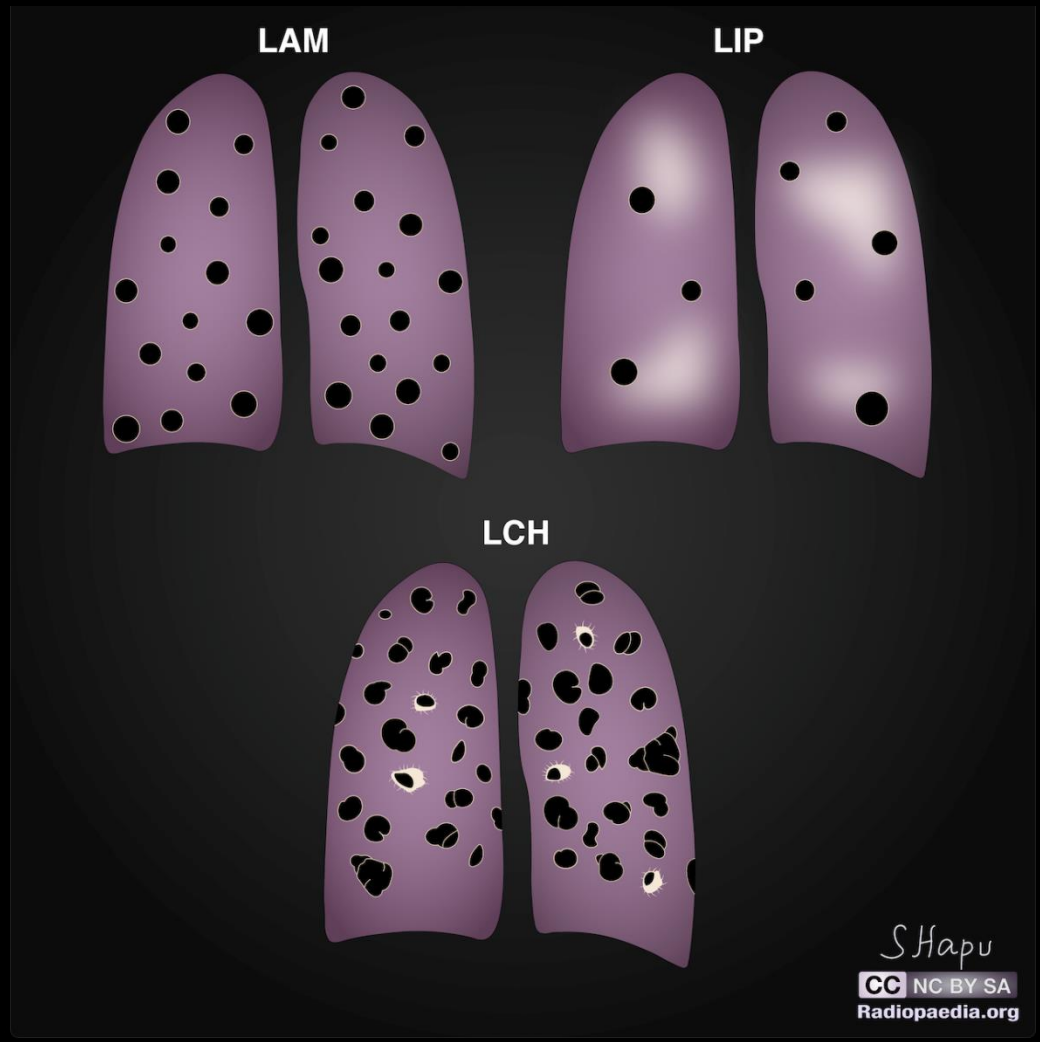
- 3 most common cystic lung diseases in adults:
  - **Lymphangioleiomyomatosis (LAM)**
  - **Pulmonary Langerhans cell histiocytosis (LCH)**
  - **Lymphoid interstitial pneumonia (LIP)**

- Additional rare causes include amyloidosis, Birt-Hogg-Dubé syndrome (BHD), bronchopulmonary dysplasia, Erdheim-Chester disease, Fire-eater's lung disease, fungi (eg, coccidiomycosis, Pneumocystis jirovecii), hypersensitivity pneumonitis, Hyperimmunoglobulin-E syndrome, light chain deposition disease, paragonimiasis, primary and metastatic tumors, recurrent respiratory papillomatosis, smoking-related small airways injury, staph PNA

DDX (based on imaging)

Adult cystic lung disease differential highlights

- **Lymphangioleiomyomatosis (LAM)**
  - Variably sized thin-walled cysts surrounded by normal lung parenchyma throughout entire lung
- **Pulmonary Langerhans cell histiocytosis (LCH)**
  - Mid to upper lobe distribution with preservation of the costophrenic angles
  - Cysts in LCH tend to be more irregular in shape
  - Children and young adults who heavily smoke
- **Lymphoid interstitial pneumonia (LIP)**
  - Smooth cysts with ground-glass attenuation and nodules, often associated with autoimmune disease



Case courtesy of Dr Sachintha Hapugoda, Radiopaedia.org, rID: 69938

Final Dx:

Lymphangioleiomyomatosis (LAM)

Lymphangioleiomyomatosis (LAM)

- Epidemiology
  - Almost exclusively affects young women; the estimated incidence is 1:400,000
  - Can occur either sporadically or in association with tuberous sclerosis
- Definitive diagnosis per 2016 American Thoracic Guidelines if patient:
  - Has compatible clinical history: young to middle-aged female, presenting with worsening dyspnea and/or pneumothorax/chylothorax in the absence of features suggestive of other cystic lung diseases
  - Has a characteristic HRCT of the chest
  - Has one or more of following features:
    - Tuberous sclerosis, renal angiomyolipoma, elevated VEGF-D > 800 pg/ml, thoracic or abdominal chylous effusion, lymphatic malformations, demonstration of LAM cells or LAM cell clusters on cytological examination of effusions or lymph nodes, histopathological confirmation of LAM by lung biopsy or biopsy of retroperitoneal or pelvic masses

Lymphangioleiomyomatosis (LAM)

Image Findings

- Chest x-ray : Chylothorax, chylous pleural effusion, hyperinflation, diffuse bilateral reticulonodular densities, recurrent PTX
- High resolution CT: Variably sized thin-walled cysts surrounded by normal lung parenchyma throughout entire lung, interlobular septal thickening, may see dilated thoracic duct and/or hemorrhages as areas of increase attenuation
- Additional potential imaging findings:
  - Abdominal CT: May see renal angiomyolipomas (most common abdominal finding), splenic infarcts, chylous ascites, uterine fibroids, abdominal lymphadenopathy
  - Head and neck CT: May see cystic hygroma, massive osteolysis with little or no periosteal reaction

Lymphangioleiomyomatosis (LAM)

Management:

- Sirolimus has been shown to slow progression of disease
- Supportive therapy (e.g. O2, bronchodilators)
- Management recurrent PTX (e.g. mechanical or chemical pleurodesis)

Prognosis

- Tends to be progressive with most of the disease severity due to pulmonary disease
- Often requires lung transplant

References

- American College of Radiology. ACR Appropriateness Criteria®. Available at https://acsearch.acr.org/list. Accessed September 22, 2020.
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