Cystic Lung Diseases

Melissa Price
Gillian Lieberman, MD
Advanced Radiology Clerkship
Beth Israel Deaconess Medical Center
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How do we define a cyst of the lung?

Fleischner Society: Glossary of Terms for Thoracic Imaging[1]

Figure 21

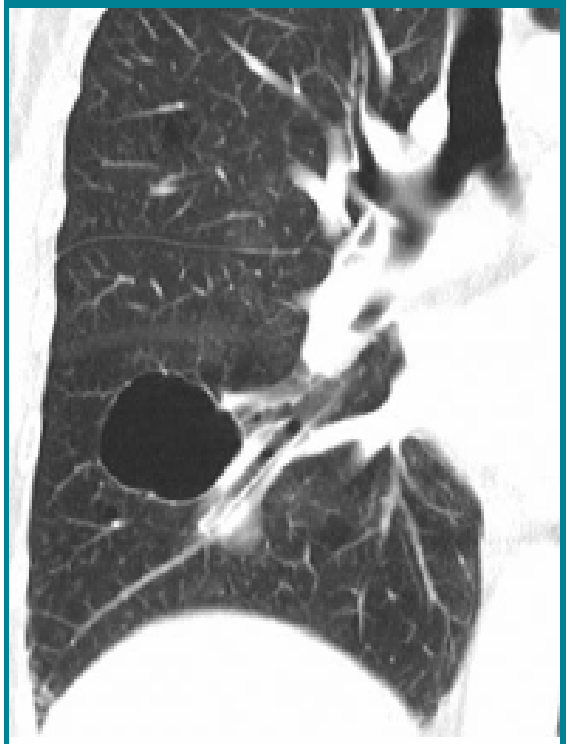


Figure 21: Coronal CT scan shows a cyst.

cyst

Pathology.—A cyst is any round circumscribed space that is surrounded by an epithelial or fibrous wall of variable thickness (51).

Radiographs and CT scans.—A cyst appears as a round parenchymal lucency or low-attenuating area with a well-defined interface with normal lung. Cysts have variable wall thickness but are usually thin-walled (<2 mm) and occur without associated pulmonary emphysema (Fig 21). Cysts in the lung usually contain air but occasionally contain fluid or solid material. The term is often used to describe enlarged thin-walled airspaces in patients with lymphangioleiomyomatosis (52) or Langerhans cell histiocytosis (53); thicker-walled honeycomb cysts are seen in patients with end-stage fibrosis (54).

Hansell DM, Bankier AA, MacMahon H, et al. Fleischner Society: Glossary of terms for thoracic imaging. Radiology 2008;246(3):697-722. Accessed via Pubmed.com

Differential Diagnosis of Cystic Lung Disease

- Non-infectious:
  - Pulmonary Langerhans' cell histiocytosis (PLCH)
  - Lymphangioleiomyomatosis (LAM)
  - Tuberous sclerosis (TS)
  - Lymphoid interstitial pneumonia (LIP)
  - Idiopathic pulmonary fibrosis (IPF)
- Infectious:
  - *Pneumocystis carinii* pneumonia (PCP)
  - Tuberculosis
  - Staphylococcus
- Mimics:
  - Cystic bronchiectasis
  - Centrilobular emphysema
  - Panlobular emphysema

Hartman TE. CT of cystic diseases of the lung. Radiologic Clinics of North America. 2001;39(6):1231-43.

Cystic Lung Disease & Mimics

Disease	Findings	Distribution	Assoc. Findings
IPF	Honeycomb cysts	Subpleural, basilar predominance	Irregular lines of attenuation, ground-glass
PLCH	Thin-walled cysts	Random, spares bases	Nodules
LAM	Thin-walled cysts	Random, diffuse	Chylous effusion
TS	Thin-walled cysts	Random, diffuse	Angiomyolipomas of kidneys and liver
LIP	Thin-walled cysts	Basilar predominance	Ground-glass attenuation
Cystic Bronchiectasis	Cystic structures contiguous with bronchial tree	Diffuse or focal	Signet ring sign: each cystic space has an attendant vessel
Centrilobular Emphysema	Cystic airspaces without discernible wall	Upper lobe predominant	Vessels present within cystic airspace
Panlobular Emphysema	Cystic airspaces	Subpleural	One cystic space immediately adjacent to the pleura

Adapted from: Hartman TE. CT of cystic diseases of the lung. Radiologic Clinics of North America. 2001;39(6):1231-43.

Cystic Lung Disease: IPF

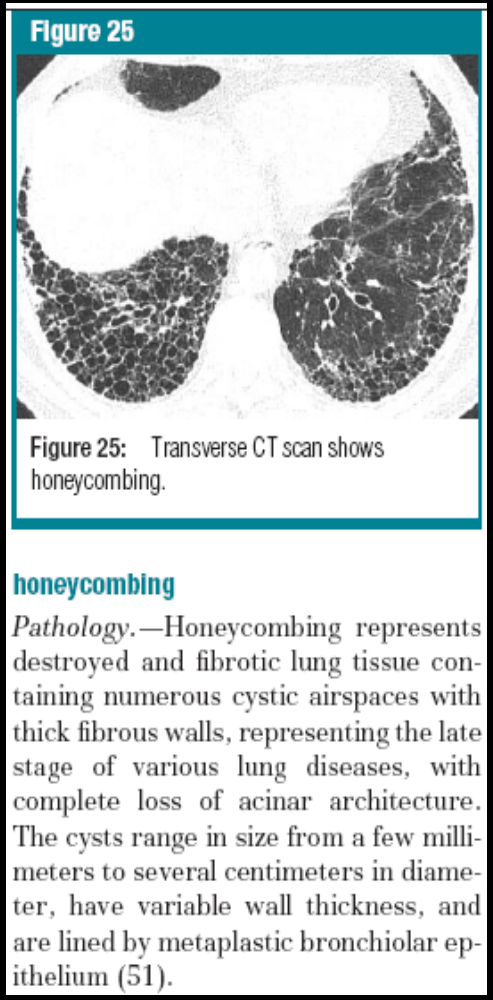


Figure 25

Figure 25: Transverse CT scan shows honeycombing.

honeycombing

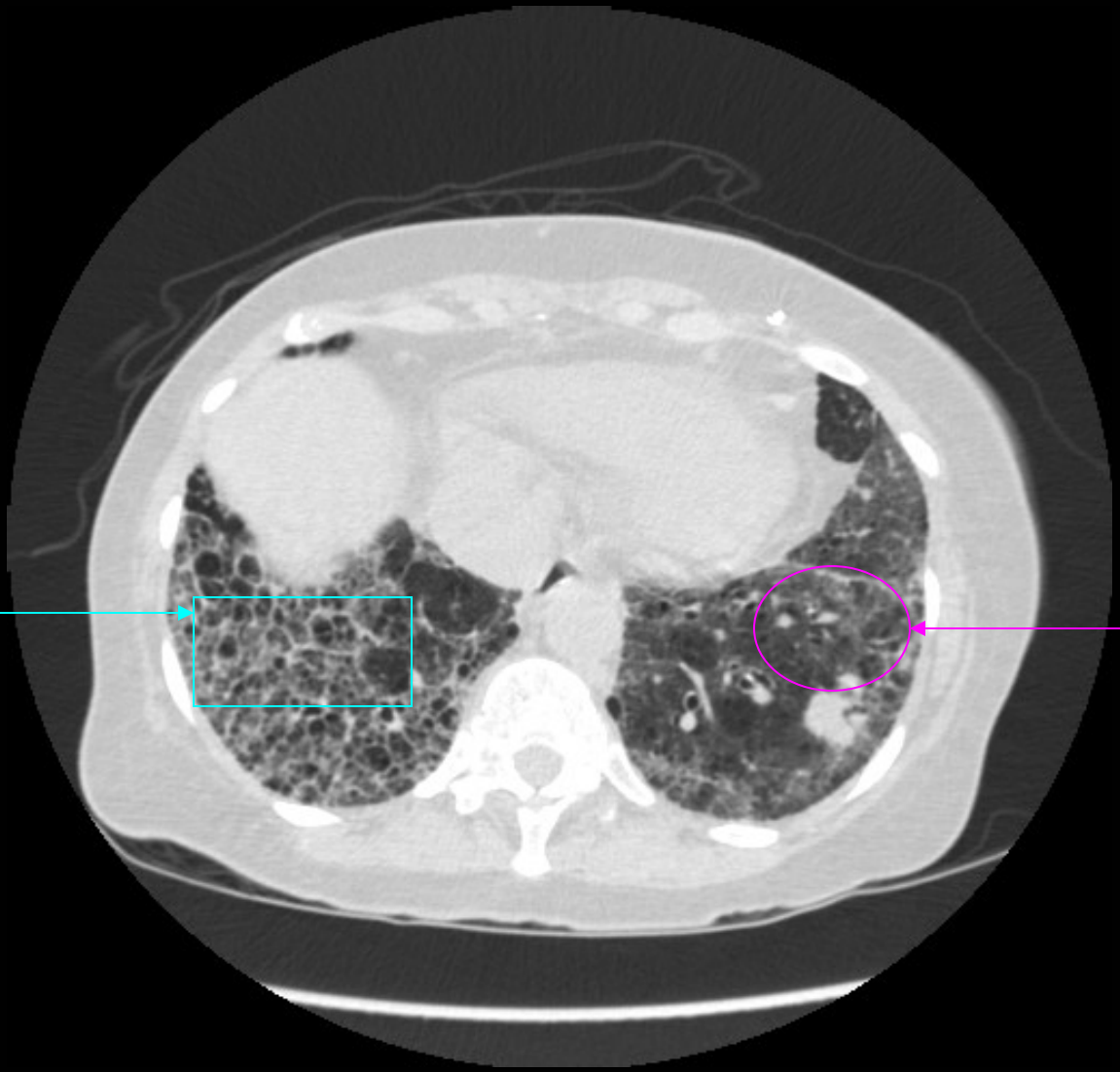
Pathology.—Honeycombing represents destroyed and fibrotic lung tissue containing numerous cystic airspaces with thick fibrous walls, representing the late stage of various lung diseases, with complete loss of acinar architecture. The cysts range in size from a few millimeters to several centimeters in diameter, have variable wall thickness, and are lined by metaplastic bronchiolar epithelium (51).

- Honeycomb cysts are a feature of idiopathic pulmonary fibrosis (IPF) and typically have a subpleural location
- Walls of the cysts are clearly defined and thickened – a sign of fibrosis
- On high-resolution CT (HRCT) there is an interstitial pattern: irregular lines of attenuation are present with a subpleural, lower lobe predominance
- Honeycomb cysts in IPF can cause architectural distortion of the lung parenchyma and are often located adjacent to regions of traction bronchiectasis

Hansell DM, Bankier AA, MacMahon H, et al. Fleischner Society: Glossary of terms for thoracic imaging. Radiology. 2008;246(3):697-722. Accessed via Pubmed.com

Patient DB: Honeycombing on Chest CT

Patient History: 72 year-old female with known idiopathic pulmonary fibrosis



Honeycombing in the right lower lobe: thickened walls indicate fibrosis

Ground-glass opacification

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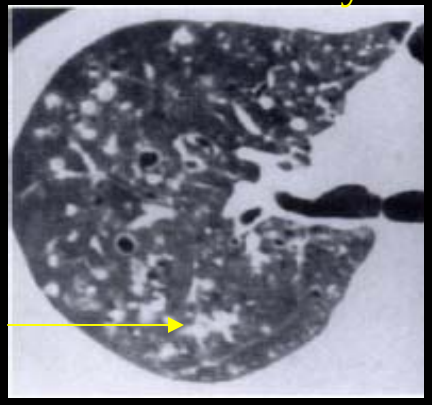
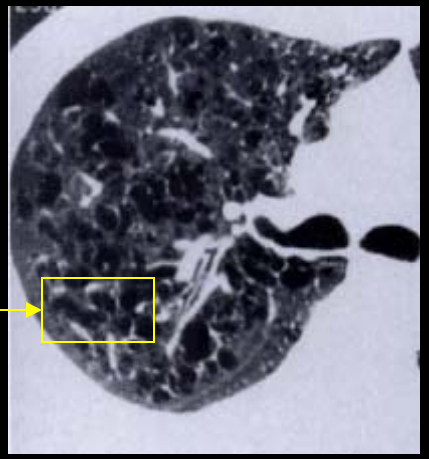
Cystic Lung Disease: Pulmonary Langerhans' Cell Histiocytosis

Initial CT scan of 46 y/o male:



CT scan 20 months later:

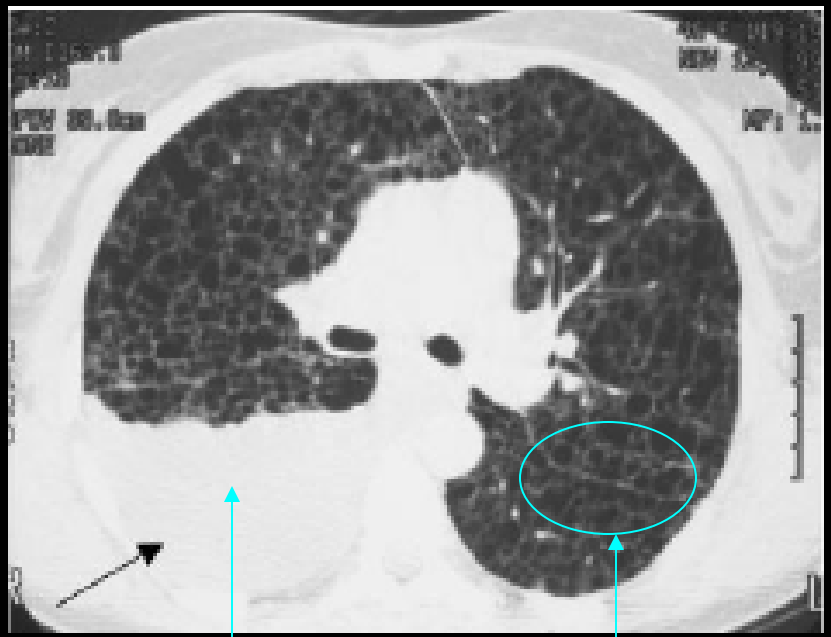


- PLCH is typically a disease of young adults which predominately affects the lungs and bones
- Very strong association with smoking
- Pulmonary disease in PLCH is characterized by peribronchiolar 1-10 mm nodules in the early stages
- In later stages of PLCH, the major pulmonary finding is cysts (present in 80% of patients) and there may be no nodules present
- Lung bases are relatively spared at all disease stages
- PLCH can be differentiated from IPF on the basis of lung volumes: IPF has decreased lung volumes and PLCH tends to have slightly increased lung volumes

Brauner MW, Grenier P, Tijani K. Pulmonary Langerhans' cell histiocytosis: Evolution of lesions on CT scan. Radiology 1997;204:497-502. Accessed via Pubmed.com

Cystic Lung Disease: Lymphangioleiomyomatosis (LAM)

- LAM is a rare disease affecting females *exclusively* which results in vascular proliferation of smooth muscle in the lung
- Presentation is typically between 30-35 years of age
- HRCT findings: multiple cysts with thin-walls which are adjacent to normal lung parenchyma
- There is no regional predominance of the cysts in the lung – diffuse distribution of cysts helps to differentiate from PLCH
- Cysts in LAM are typically less than 2 cm in size
- Cysts can rupture and results in pneumothoraces (30-40%)
- Proliferation of smooth muscle in pulmonary lymphatics can result in obstruction and chylothoraces
- Identical pulmonary findings are seen in 1% of patients with tuberous sclerosis



McCormack FX. Lymphangioleiomyomatosis: A clinical update. Chest 2008;133:507-516. Accessed via Pubmed.com

Cystic Lung Disease: Lymphoid Interstitial Pneumonia (LIP)

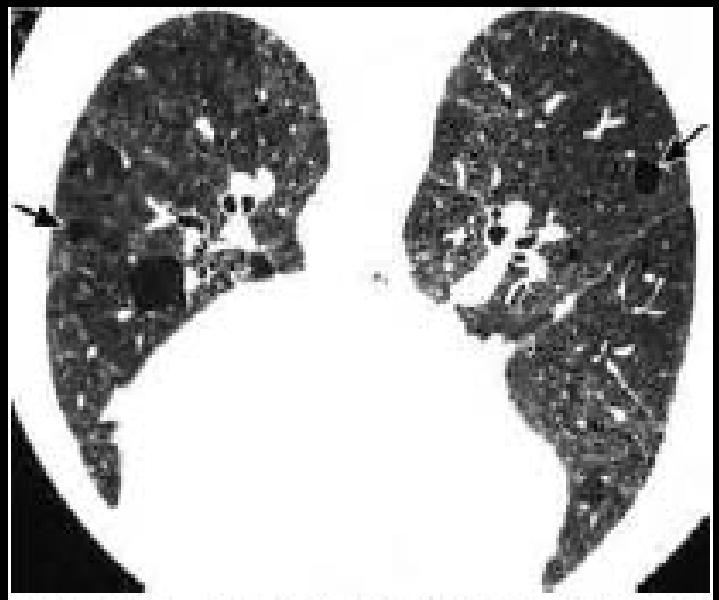


Figure 27. LIP caused by Sjögren syndrome in a 62-year-old woman. Transverse thin-section CT image obtained with patient prone shows diffuse ground-glass opacification and multiple lung cysts (arrows).

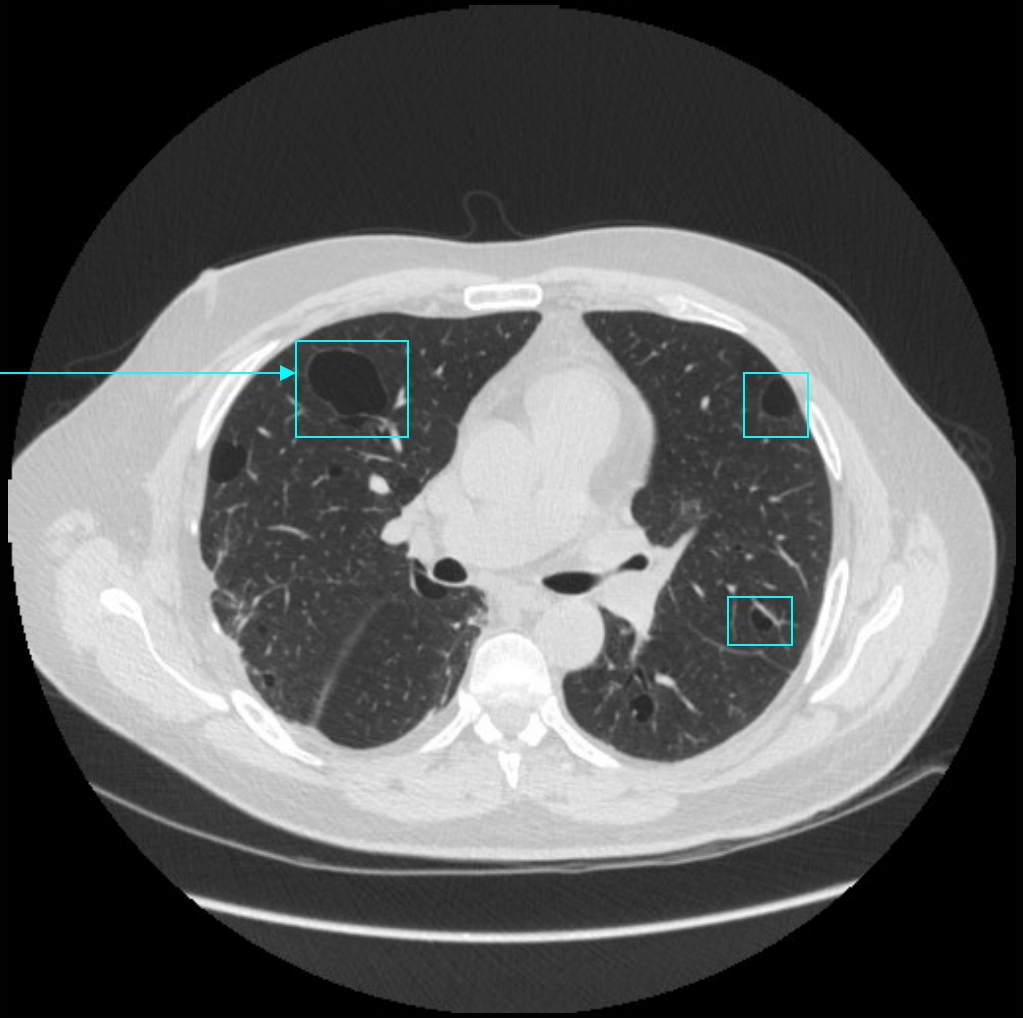
- LIP is caused by lymphocytic proliferation along the alveolar septum of the lung
- It can progress to low-grade lymphoma, however this is rare
- LIP is associated with immune function disturbances – most commonly AIDS as well as connective tissues disorders, especially Sjögren's syndrome
- Ground-glass attenuation is the most common finding on HRCT in LIP
- Cysts can also be present in LIP and typically have a perivascular distribution
- The cystic form of LIP is more common among patients with Sjögren's syndrome

Lynch DA, Travis WD, Müller NL, Idiopathic interstitial pneumonias: CT features. Radiology 2005;236:10-21. Accessed via Pubmed.com

Companion Patient A: LIP on Chest CT

Patient history: 61 year-old male with Sjögren's syndrome

Multiple thin-walled cysts with a perivascular distribution



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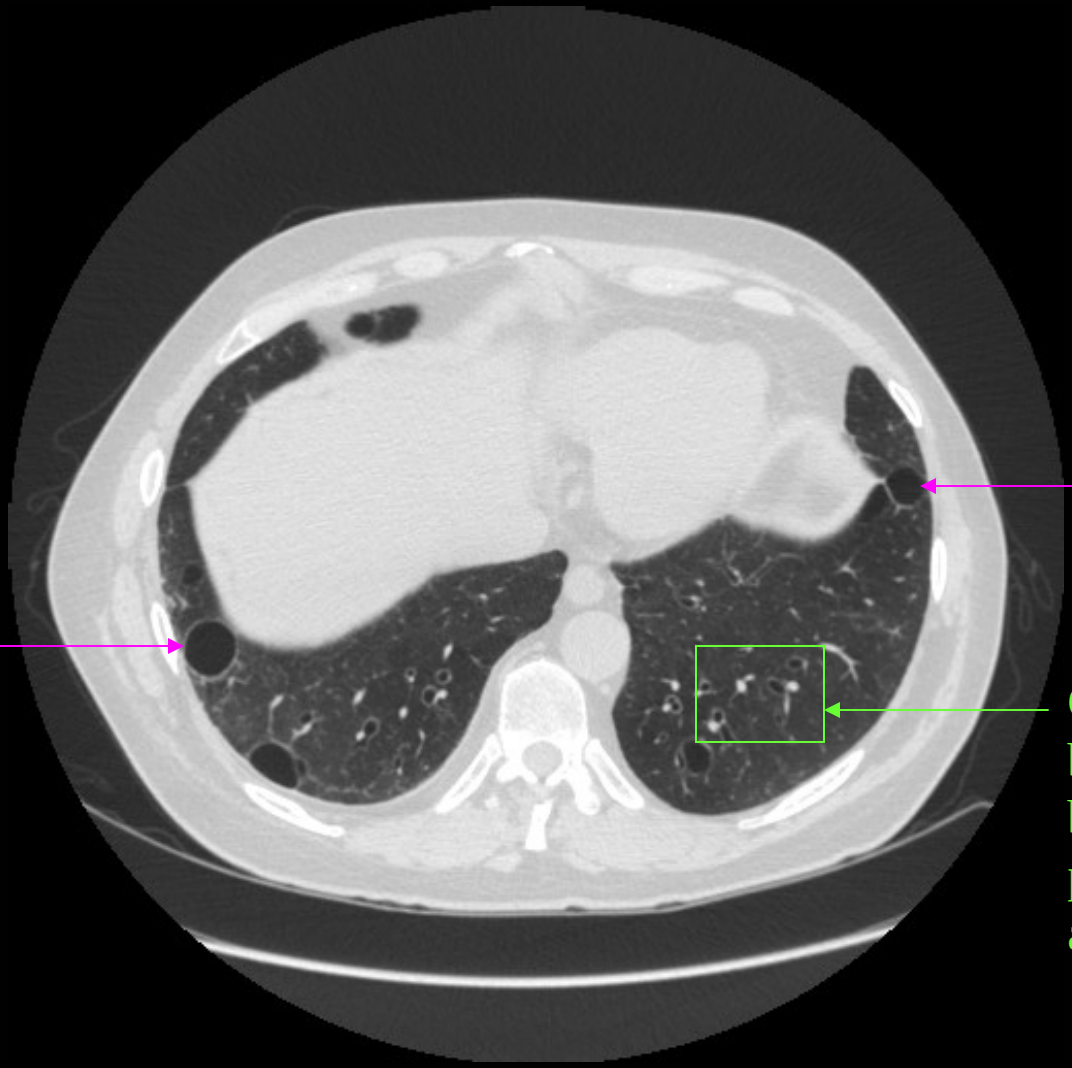
Companion Patient A: LIP on Chest CT



Cyst with thin wall (<3mm)

Cyst with thin wall (<3mm)

Cylindrical bronchiectasis; bronchi extend to the periphery of the lung and do not taper

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Mimics: Cystic Bronchiectasis

bronchiectasis

Pathology.—Bronchiectasis is irreversible localized or diffuse bronchial dilatation, usually resulting from chronic infection, proximal airway obstruction, or congenital bronchial abnormality (26). (See also *traction bronchiectasis.*)

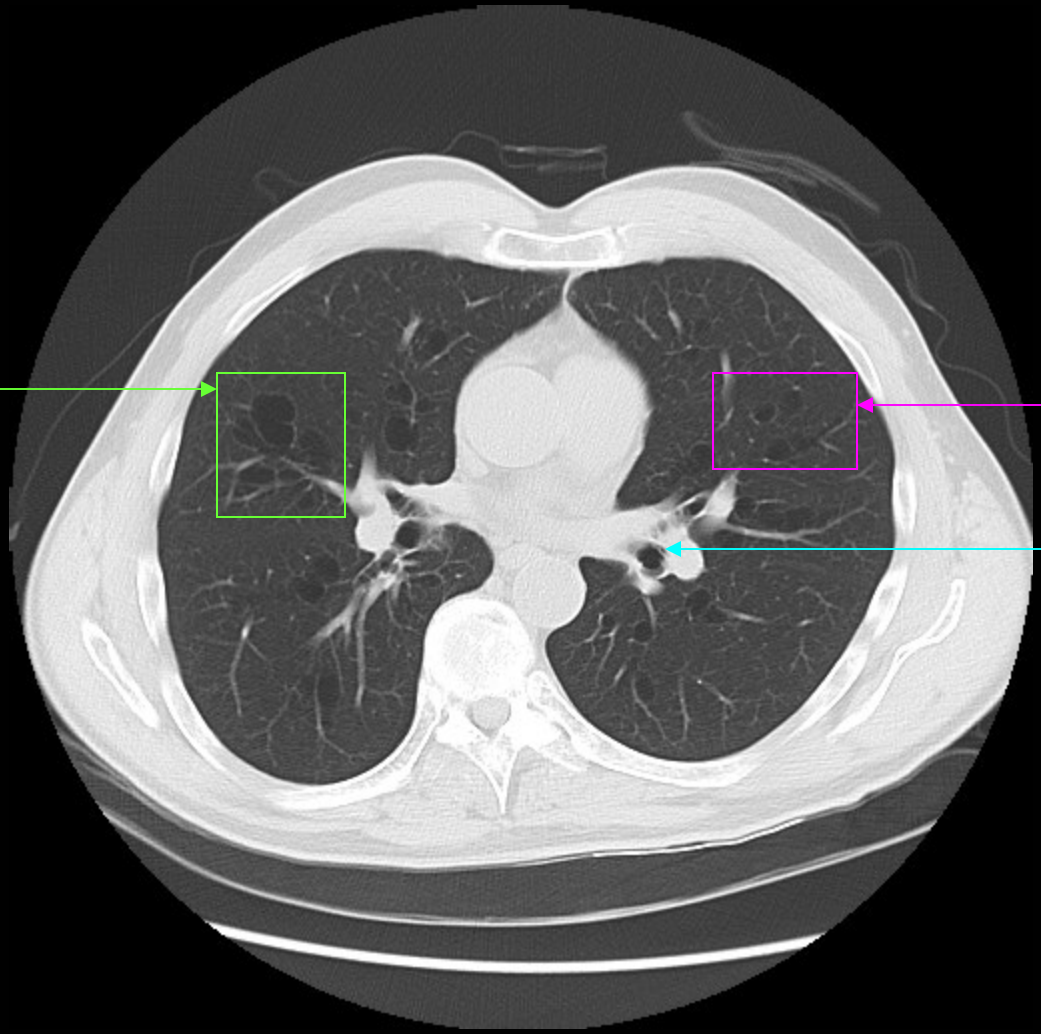
Radiographs and CT scans.—Morphologic criteria on thin-section CT scans include bronchial dilatation with respect to the accompanying pulmonary artery (signet ring sign), lack of tapering of bronchi, and identification of bronchi within 1 cm of the pleural surface (27) (Fig 11). Bronchiectasis may be classified as cylindric, varicose, or cystic, depending on the appearance of the affected bronchi. It is often accompanied by bronchial wall thickening, mucoid impaction, and small-airways abnormalities (27–29). (See also *signet ring sign.*)

- Cystic bronchiectasis can be differentiated from true cystic lung disease by the continuous relationship of the cystic structure to bronchial tree
- The signet ring sign is characteristic of bronchiectasis, with the “stone” of the ring representing the pulmonary artery
- The signet ring sign is present on transverse sectional imaging
- Diseases that produce cystic bronchiectasis include tracheobronchomegaly, Williams-Campbell syndrome, and cystic fibrosis

Hansell DM, Bankier AA, MacMahon H, et al. Fleischner Society: Glossary of terms for thoracic imaging. Radiology. 2008;246(3):697-722. Accessed via Pubmed.com

Companion Patient B: Cystic Bronchiectasis on Chest CT

Patient history: 34 year-old male with Williams-Campbell syndrome



Saccular bronchiectasis is present in fourth and higher-order bronchi

Bronchi can be seen extending to the lung periphery - bronchi should not be visible within 1 cm of the pleura in normal lung

Major bronchi have a normal appearance and configuration

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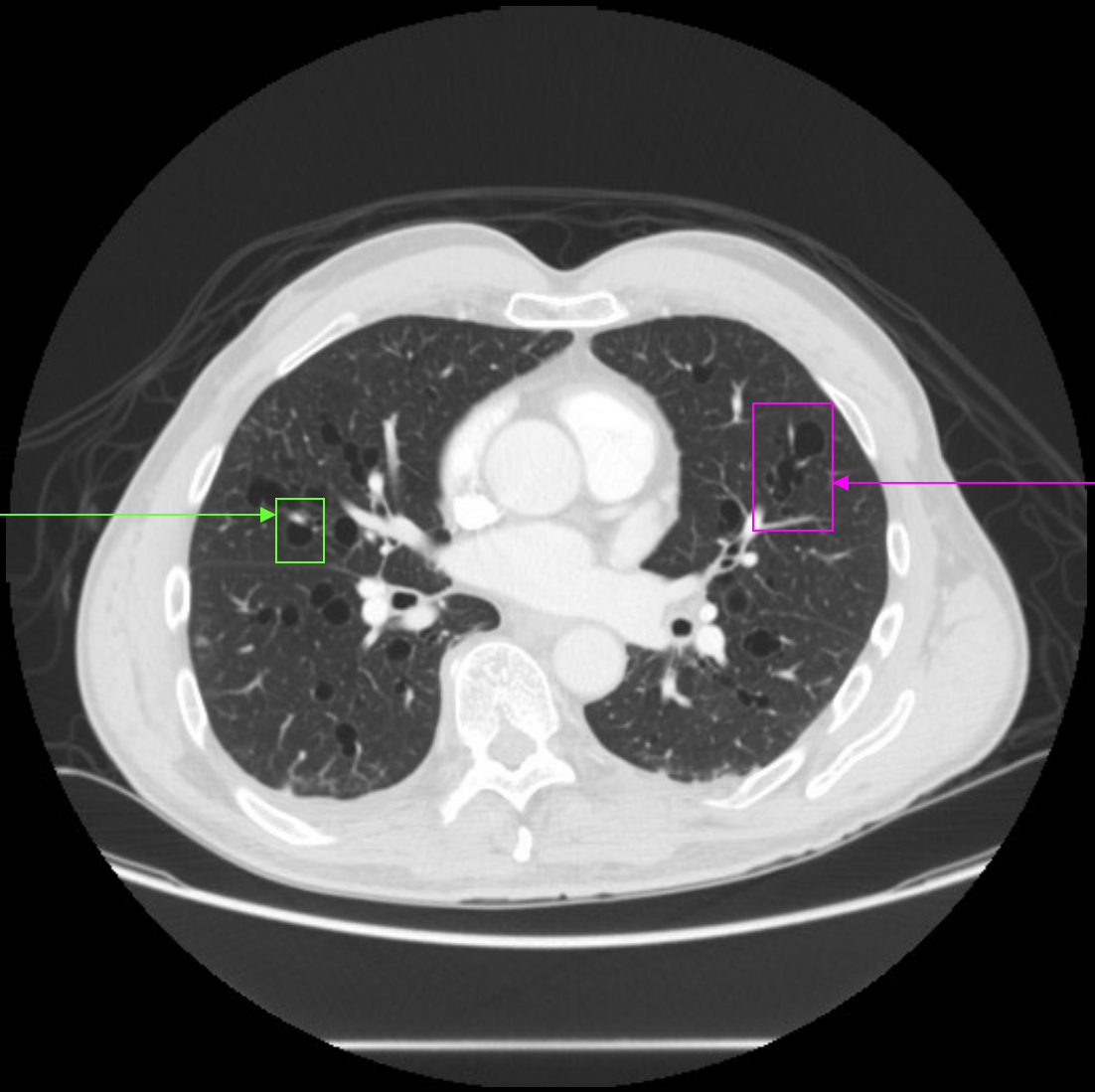
Companion Patient B:
Cystic Bronchiectasis on Chest CT



Signet ring sign of bronchiectasis

Dilated airways larger in size than adjacent pulmonary arteries

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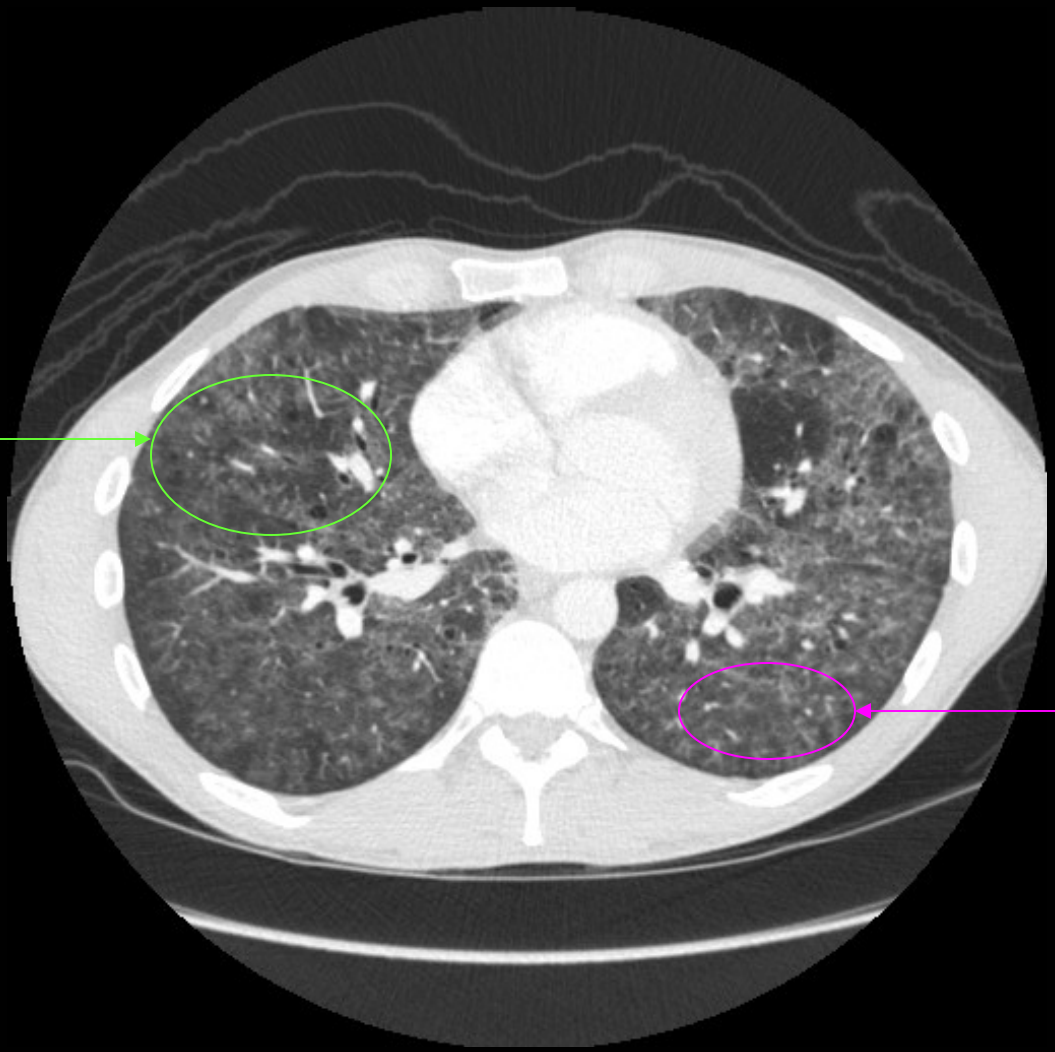
Infectious Etiologies: *Pneumocystis carinii* Pneumonia

- PCP is the most common diffuse lung disease in AIDS
- On HRCT: ground –glass opacities (attenuation of the lung parenchyma is increased but pulmonary vasculature remains visible)
- A mosaic pattern of the ground glass attenuation may be present: reflects diseased secondary pulmonary lobules adjacent to normal ones
- Ground-glass attenuation can be more predominant in perihilar regions
- The opacification may be more prevalent in the upper lobes of the lung, particularly if the patient has been given prophylaxis with pentamidine
- Cysts are commonly interspersed among the ground-glass opacification
- Other CT findings can include increased interstitial markings and septal thickening

Primack SL, Müller NL. High-resolution computed tomography in acute diffuse lung disease in the immunocompromised patient. Radiol Clin North Amer 1994 July;32:731-44.

Companion Patient C:
Pneumocystis carinii Pneumonia on Chest CT

Patient history: 32 year-old male with recent HIV diagnosis and 3 week history of dyspnea and cough



Mosaic pattern: normal lung adjacent to diseased lung parenchyma

Diffuse ground-glass opacities without obscuration of pulmonary vasculature

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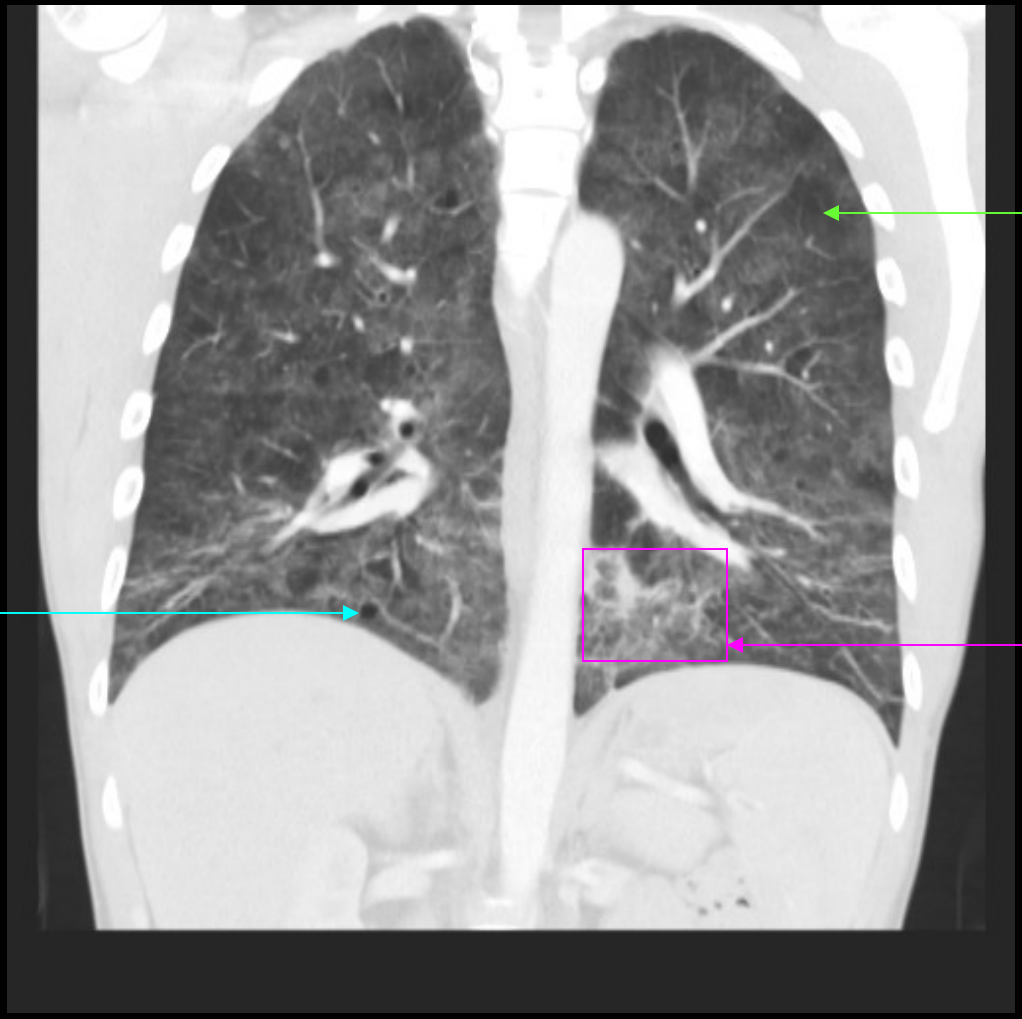
Companion Patient C:
Pneumocystis carinii Pneumonia on Chest CT



PACS, BIDMC

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- Brauner MW, Grenier P, Tijani K. Pulmonary Langerhans' cell histiocytosis: Evolution of lesions on CT scan. Radiology 1997;204:497-502.
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- Webb WR. High-resolution computed tomography of obstructive lung disease. Rad Clin North Am. 1994 July;32(4):745-57.

The End!