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**

**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).
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

## Cystic Lung Disease & Mimics

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Cystic Lung Disease: IPF

- 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

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
Cystic Lung Disease:
Pulmonary Langerhans’ Cell Histiocytosis

- 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

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

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.

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
Companion Patient A: LIP on Chest CT

Cyst with thin wall (<3mm)

Cylindrical bronchiectasis; bronchi extend to the periphery of the lung and do not taper
Mimics: Cystic Bronchiectasis

- 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

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

Major bronchi have a normal appearance and configuration

Bronchi can be seen extending to the lung periphery - bronchi should not be visible within 1 cm of the pleura in normal lung
Companion Patient B: Cystic Bronchiectasis on Chest CT

- Signet ring sign of bronchiectasis
- Dilated airways larger in size than adjacent pulmonary arteries
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

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
Companion Patient C: *Pneumocystis carinii* Pneumonia on Chest CT

- Thin-walled cyst
- Relatively normal appearing lung
- Atelectasis
References

- PACS, Beth Israel Deaconess Medical Center.
The End!