Cystic Lung Disease

: What Can Cause Holes in the Lung?

Hyun Jung Koo, Mi Young Kim, Jae Woo Song
Department of Radiology, Asan Medical Center, University of Ulsan College of Medicine
Financial Disclosures

- None
Pathogenesis

- Check valve air cyst or ball-valve effect
  - Pulmonary LCH, LAM, Lymphocytic interstitial pneumonia
- Drainage of necrotic lung parenchyma
  - Pneumatocele
- Vascular occlusion or ischemic necrosis
  - Septic embolism, metastasis
- Dilatation of bronchioles
  - Desquamative interstitial pneumonia, Light chain deposition disease
- Focal collection of air in interstitial tissue
**Cyst**

**Pathology**
Round circumscribed space surrounded by an epithelial or fibrous wall

**CT scans**
Round parenchymal lucency or low-attenuating area with a well-defined interface with normal lung
Usually thin-walled (2 mm) and occur without associated pulmonary emphysema
- Thin-walled airspaces: patients with LAM or LCH
- Thicker-walled honeycomb cysts: patients with end-stage fibrosis

Usually contain air but may contain fluid or solid material
**Bleb**

**Definition**

A small gas-containing space within the visceral pleura or in the subpleural lung, not larger than 1 cm in diameter

**CT scans**

A thin-walled cystic air space contiguous with the pleura
Bulla

**Pathology**
An airspace measuring more than 1 cm—usually several centimeters—in diameter, sharply demarcated by a thin wall that is no greater than 1 mm in thickness. Usually accompanied by emphysematous changes in the adjacent lung.

**CT scans**
A rounded focal lucency or area of decreased attenuation, 1 cm or more in diameter, bounded by a thin wall. Often associated with other signs of pulmonary emphysema.
Emphysema

Pathology

- Permanently enlarged airspaces distal to the terminal bronchiole with destruction of alveolar walls
- Classified in terms of the part of the acinus predominantly affected
  - proximal (centriacinar, more commonly termed centrilobular, emphysema)
  - distal distal (paraseptal emphysema)
  - whole acinus (panacinar or, less commonly, panlobular emphysema)

CT scans

- Focal areas or regions of low attenuation, usually without visible walls
Honeycombing

**Pathology**
- Destroyed and fibrotic lung tissue containing numerous cystic airspaces with thick fibrous walls.
- Late stage of various lung diseases, with complete loss of acinar architecture.
- Size from a few millimeters to several centimeters in diameter.
- Variable wall thickness, and lined by metaplastic bronchiolar epithelium.

**CT scans**
- Usually 3–10 mm but occasionally as large as 2.5 cm.
- Usually subpleural and characterized by well-defined walls.
- A CT feature of established pulmonary fibrosis.
- Often considered specific for pulmonary fibrosis and is an important criterion in the diagnosis of usual interstitial pneumonia.
Cavity

**Definition**

- Usually produced by the expulsion or drainage of a necrotic part of the lesion via the bronchial tree
- Sometimes contains a fluid level

**CT scans**

- A gas-filled space, seen as a lucency or low-attenuation area, within pulmonary consolidation, a mass, or a nodule
- Cavitating consolidation: the original consolidation may resolve and leave only a thin wall.
Diffuse cystic lung disease

- Pulmonary Langerhans cell histiocytosis (PLCH)
- Lymphocytic interstitial pneumonia (LIP)
- Desquamative interstitial pneumonia (DIP) and respiratory bronchiolitis–associated interstitial lung disease (RB-ILD)
- Birt-Hogg-Dube syndrome (BHD)
- Metastasis
Pulmonary Langerhans cell histiocytosis (PLCH)

17/M Chest pain, dyspnea, smoking (+)

- Reticulonodular pattern with upper and mid lung zone distribution
- Sparing of costophrenic angles
- Lung volumes preserved or enlarged
- Pneumothorax

Leader-cath inserted

Case 1
Pulmonary Langerhans cell histiocytosis (PLCH)

18/M Left chest pain (3 month earlier)

- Cysts seen approximately 80% and nodules seen 60~80% of patients
- End-stage disease may be difficult to differentiate from extensive bullous emphysema

Basic pathogenesis: Langerhans’ cell proliferation, centered around the membranous and the respiratory bronchiole

CD 1a : focal (+)
S-100 (+)
SMA (+)
Several thin-walled cystic lesions in both upper lobes.
Focal osteolytic lesion with soft tissue involving right scapula (yellow arrow).
→ Right scapula needle biopsy: Langerhans cell histiocytosis
Radiology report:
Multiple nodules with cavitary nodules in both lungs.
→ Possible metastasis from unknown primary site

Lung, core needle biopsy: Langerhans cell histiocytosis
Lymphangioleiomyomatosis (LAM)

30/F Left chest pain

- Bilateral thin-walled cysts scattered throughout the lungs with no zonal predominance.
- More uniform in size, round, and diffuse in distribution of cysts.
- Diffuse throughout the lung including costophrenic angles.
Lymphangioleiomyomatosis (LAM)
Lung: Lymphangioleiomyomatosis (LAM)
Lymphangioleiomyomatosis (LAM) and tuberous sclerosis (TSC)

26/F, Chest pain, recurrent pneumothorax (x 5)

✓ Basic pathogenesis: idiosyncratic smooth muscle cell proliferation then cyst formation
Lymphocytic interstitial pneumonia (LIP)

- GGO and areas of consolidation
- Peribronchovascular thickening
- Interlobular septal thickening
- Subpleural nodules
- Lymphadenopathy

- Benign lymphoproliferative disorder
- Characterized by pulmonary interstitial infiltration of lymphocytes with varying admixtures of plasma cells, and lymphoid follicles.
Lymphocytic interstitial pneumonia (LIP)

2016-3-30
On steroid therapy

- Thin-walled cysts with a basal predominance (1–30 mm)
- Typically less numerous than in LAM or LCH.

Sjogren’s syndrome-Lymphocytic interstitial pneumonia
Lymphocytic interstitial pneumonia (LIP)

Sjogren’s syndrome-Lymphocytic interstitial pneumonia, Amyloidosis

Case 9
Desquamative interstitial pneumonia (DIP)

48/M, 30 PY

- A peripheral subpleural and basal predominance of ground-glass opacity
- Small cystic spaces may develop within the areas of ground-glass opacity
- Coexistent emphysema
Desquamative interstitial pneumonia (DIP)

52/M

- These small cysts are shown to represent bronchiolectasis and dilated alveolar ducts
- About a third of patients
Respiratory Bronchiolitis (RB)-ILD

50/M, Current smoker

RB-ILD with emphysema? Advanced pulmonary LCH?

Lung, wedge resection: Smoking associated respiratory bronchiolitis
Birt-Hogg-Dube Syndrome

23/F, Dyspnea, familial history of pneumothorax

Lung wedge resection:
- Subpleural bullae.
- Single parenchymal cysts with focal smooth muscle proliferation.

Discrete thin-walled cysts, mostly in the lower lobes
Vary in size and shape
Large lung cysts are frequently multiseptated.
Total extent of lung involvement being less than 30%

✓ Susceptibility to kidney tumor (RCC, oncocytoma), renal and pulmonary cysts, and noncancerous tumors of hair follicles, called fibrofolliculomas
Metastasis

67/M, Dry cough

- RUL wedge resection: adenocarcinoma, moderately differentiated, micropapillary predominant type, EGFR/ALK mutation (-)
Metastasis

67/M, Dry cough

- VASCULAR TUMOR, FAVOR ANGIOSARCOMA.

- Sarcoma including angiosarcoma, osteosarcoma, epithelioid hemangioendothelioma
- Squamous cell carcinoma, head and neck cancer in men and cervix cancer in women
- Adenocarcinoma including colorectal cancer
Focal or Multifocal cystic lung disease

- Pneumatocele
- Fungal infection including invasive aspergillosis, mucormycosis
- Tuberculosis
- Pneumocystis jiroveci pneumonia (PJP)
- Paragonimiasis
After pneumonia, thin-walled air cyst lesion is appeared in the right upper lobe.

Sequelae of previous septic embolism with pneumatocele caused by MRSA
Invasive pulmonary aspergillosis

30/F AML BMT GVHD Bronchiolitis obliterans

Introduction          Terminology                 Diffuse CLD             Multifocal CLD             Quiz          Summary

Tx: Voriconazole

2016-3-28

2016-7-11
Pulmonary tuberculosis

19/M

2007-7-9

Tx: AntiTBc med.

2007-9-20

Case 19
Pneumocystis jiroveci pneumonia (PJP)

35/M

Paragonimiasis

63/M

- 5-15 mm filled with either fluid or gas, within the consolidation
- About 60% of patients
- Subpleural linear opacities communicating with a cyst, suggestive of worm migration tracks

Case 20/21
Pneumocystis jiroveci pneumonia

47/M

Pulmonary LCH

17/M
Birt-Hogg-Dube Syndrome

57/M

Paragonimiasis

63/M
<table>
<thead>
<tr>
<th>Disease</th>
<th>Clinical feature</th>
<th>HRCT feature</th>
<th>Association</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary LCH</td>
<td></td>
<td>Smoker</td>
<td>Young adults</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Pneumothorax</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Early centrilobular nodules</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Sparing of CPA</td>
</tr>
<tr>
<td>Pulmonary HT</td>
<td></td>
<td></td>
<td>LAM and tuberous sclerosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Sporadic</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- TS complex</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Pneumothorax</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Chylous effusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Perimenopausal women</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Seizures, skin lesions</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Throughout the lungs</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>No zonal predominance</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Tiny nodules, MMPH</td>
</tr>
<tr>
<td>Angiomyolipoma</td>
<td></td>
<td>AD inherited</td>
<td>AD inherited</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LIP</td>
<td>Follicular Bronchiolitis &amp; LIP</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Amyloidosis</td>
<td>Light chain deposition Ds.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Light chain deposition Ds.</td>
<td>Sjögren disease, cardiovascular Ds., AIDS</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Renal failure, Sjögren disease</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Peri-vascular cysts, smaller nodules</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cysts and larger nodules</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lymphoproliferative disorder, lymphoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Multiple myeloma, Macroglobulinemia</td>
<td></td>
</tr>
<tr>
<td>DIP and RB–ILD</td>
<td></td>
<td>Smoker, male</td>
<td>Subpleural and basal predominance of ground-glass opacity</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Coexistent emphysema</td>
</tr>
<tr>
<td>Birt-Hogg-Dube syndrome</td>
<td>Skin lesions</td>
<td>Fibrofolliculomas</td>
<td>Basilar subpleural lentiform cysts</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Renal oncocytoma, RCC, AD inherited</td>
</tr>
<tr>
<td>Cystic metastasis</td>
<td>History of malignancy</td>
<td>CA-125 (+)</td>
<td>Peripheral nodules</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>GGO, by hemorrhage</td>
</tr>
<tr>
<td>Septic embolism</td>
<td></td>
<td></td>
<td>Pulmonary or extra-pulmonary infection</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**References**


Thank you for your attention.

Contact: mimowdr@gmail.com