Lung diseases presenting with multiple cysts: a diagnostic approach

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Learning objectives

- To review the definition of a lung cyst and differentiate it from other lesions capable of simulating a cyst, such as emphysema, cystic bronchiectasis, honeycomb cyst and lung cavity.

- To present and illustrate the radiological differential diagnosis of multiple cystic lung diseases (CLDs) and to highlight imaging characteristics.

- To review other conditions that may accompany cystic lung diseases.
Background

A lung cyst is an air-filled structure defined by an outer wall (usually ≤ 2 mm) and radiologically seen as a round low attenuation area. Although, lung cysts can be detected incidentally in high resolution computed tomography (HRCT) images in healthy individuals, may be indicating some systemic diseases when they are multiple. At first, lung cysts should be distinguished primarily from pathologies, such as emphysema, bronchiectasis, cystic metastases or cavity, that can mimic lung cyst in imaging. Afterwards, the presence of multiple lung cysts may be associated with some of the types of infrequent lung diseases, so it is important to know the differential diagnosis of cystic lung diseases (CLDs) and accompanying imaging findings. Integration with clinical information is often important to achieve diagnosis in CLD. Important clues in differential diagnosis are: patient age and gender, distribution, shape and size of the cysts, the presence of any ancillary computed tomography (CT) findings, smoking and family history, environmental and occupational exposure, immune status and the presence of accompanying other diseases. Patients with CLD usually have nonspecific clinical findings and spontaneous pneumothorax can be a sentinel event leading to the diagnosis. The knowledge of HRCT findings in CLDs is of great importance for diagnosis. This pictorial essay will describe the clinical and imaging findings of CLD’s and differential diagnosis.
Imaging findings OR procedure details

Parenchymal lucencies that may mimic lung cysts

**Emphysema**

Emphysema appear as polygonal low-attenuation area, and a central dot can be seen which represents the pulmonary artery within the secondary pulmonary lobule (Fig. 1). Emphysema has no walls. However, interlobular septa surrounding the emphysema areas of the lung may be misinterpreted as walls and it may be difficult to differentiate some forms of atypical emphysema from lung cysts (Fig. 2). In contrast, cystic lung disease in advanced stages can mimics emphysema.

**Honeycombing**

Honeycombing represents the advanced stage of lung fibrosis that lead to enlarged airspaces with thick fibrous walls. On HRCT, honeycombing appears as small, clustered lucencies, with well-defined contours and tend to be subpleural, stacked together in contiguous rows and sharing common walls (Fig. 3). Honeycombing usually associated with traction bronchiectasis and bronchiolectasis, architectural distortion and subpleural reticulation. The presence of these findings helps to distinguish honeycombing from cystic lung disease.

**Bronchiectasis**

Cystic bronchiectasis may mimic lung cysts and it can be differentiated from CLD based on their continuity with an airway (Fig. 4). Also tram-track sign and signet ring sign on HRCT are ancillary findings for discrimination (Fig. 5).

**Cavity**

Lung cavities are typically thick-walled (usually $>4$ mm), air-filled spaces seen as a low-attenuation area within an area of lung consolidation, a nodule or a mass (Fig. 6). A cavity is usually produced by the drainage of a necrotic part of the lesion via the bronchus, and it may contain a fluid level or fungus ball.

**Pneumatocele**

Pneumatocele is a thin-walled, air-filled space in the lung. They most commonly arise in the setting of acute pneumonia, blunt chest trauma, barotrauma or aspiration of hydrocarbon fluid. *Pneumocystis jirovecii* pneumonia in immunosuppressed patients may cause pneumatoceles in the ground-glass opacity areas (Fig. 7 and 8). In appropriate clinical conditions, the absence of pneumatocele walls allows differentiation from lung cysts.
Neoplastic processes

Rarely, primary lung carcinoma or metastatic lung disease can be present as multiple cystic lung lesions. On CT, these lesions appear as solid nodules, thin and thick-walled cystic lesions. As with other metastatic lesions, these lesions tend to have different sizes and a basilar predominance (Fig. 9).

Cystic lung diseases

Lymphangioleiomyomatosis (LAM)

Lymphangioleiomyomatosis (LAM) is a rare multi-system disorder that almost exclusively affects women. LAM can be encountered either sporadically or in association with the tuberous sclerosis complex (TSC). Most patients with LAM have nonspecific symptoms, such as dyspnea, cough, and recurrent pneumothorax. Pneumothorax may occur in 60-80% of patients during the course of disease. On HRCT, thin-walled, well circumscribed and round shaped cysts are seen and typically profuse (>10), display limited variability in size and shape, and evenly distributed throughout both lungs. Usually, the lung parenchyma outside the cysts appears normal, but patchy ground-glass opacity areas or areas of air trapping can be seen on HRCT. Upper-lobe predominant, benign, peripheral lung nodules are rarely seen in patients with TSC associated LAM (Fig. 10). Chylous pericardial or pleural effusion, hepatic or renal angiomyolipoma thoracic duct enlargement, menigioma and abdominal or thoracic lymphangioleiomyoma can be seen.

Pulmonary Langerhans Cell Histiocytosis (PLCH)

Pulmonary Langerhans Cell Histiocytosis (PLCH) is a smoking-related cystic lung disease, and most of cases have a history of smoking. PLCH has no sex-based predominance. Most patients with PLCH have nonspecific symptoms, such as dyspnea, dry cough, fever, weight-loss and spontaneous pneumothorax. Peribronchiolar and peribronchial Langerhans and inflammatory cells infiltration results in stellate bronchiolocentric nodules. The nodules may subsequently cavitate and form thick- and thin-walled cysts thought to represent enlarged airway lumina as a result of bronchiole wall inflammation and fibrosis. The cysts are irregular and bizarrely shaped, have prominent walls, and tend to involve the upper lobes with relative sparing of the costophrenic angles, anterior tips of the middle lobe and lingula. Usually, both nodules and cysts are seen on HRCT (Fig. 11). In late phase of PLCH, secondary pulmonary hypertension may occur and may cause death.

Birt-Hogg-Dubé syndrome (BHD)

Birt-Hogg-Dubé syndrome (BHD), also known as folliculin gene-associated syndrome, is an autosomal-dominant, multisystemic disorder, with mostly affects the lungs, skin and kidneys. Rarely, patients with BHD may have only multiple pulmonary cysts, with no
other systemic feature. In this context, the detection of lung cysts may be useful for early diagnosis of BHD, and chest HRCT may play an important role. Affected patients may be asymptomatic, but the most common presentation is skin lesions (fibrofolliculomas and trichodiscomas). Kidney findings can range from simple cortical cysts to multifocal renal cell carcinomas. The most common presentation of lung involvement with BHD is recurrent and spontaneous pneumothorax and patients may have a family history of pneumothorax. On HRCT, bilateral, thin-walled, irregular, oval or lentiform shaped cysts shows a lower-medial lung zone and subpleural predominance, and some cysts may contain internal septa or encircling adjacent bronchovascular bundle (Fig. 12). The remaining parenchyma is usually normal.

**Lymphocytic interstitial pneumonitis (LIP)**

Lymphoid or lymphocytic interstitial pneumonia (LIP) is a rare form of interstitial pneumonia and characterized by infiltration of the pulmonary interstitial and alveolar spaces by lymphocytes, plasma cells, and histiocytes. LIP usually seen in association with various underlying disorders, such as Sjögren's syndrome, acquired immunodeficiency syndrome, Castleman disease, rheumatoid arthritis, and systemic lupus erythematosus. Most patients with LIP have nonspecific symptoms, such as dyspnea, cough, fever and weight-loss. Rarely, recurrent pneumothorax can be seen in patients with LIP. HRCT usually demonstrates a combination of ground-glass opacities, interlobular septal thickening, consolidations and scattered lung cysts. Cysts are usually seen within areas of ground-glass opacities and they vary in shape (Fig. 13). The diagnosis of LIP should be considered in a patient with a few scattered lung cysts and an underlying disorder. In a minority of patients with LIP, lymphoma may develop.

**Pneumocystis jiroveci pneumonia (PJP)**

Pneumocystis jiroveci pneumonia (PJP) is a rare fungal infection, is seen almost exclusively in immunosuppressed patients. Most patients with PJP have nonspecific symptoms, such as dyspnea, nonproductive cough, fever and spontaneous pneumothorax. However, if left untreated, it can progress to respiratory failure and death. On HRCT, bilateral, multifocal and mainly symmetric ground-glass opacities and consolidations are seen, which are distributed in the central and upper portions of the lungs. Small cysts within the ground-glass opacities can be seen (Fig. 7 and Fig. 8).

**Amyloidosis**

Amyloidosis refers to systemic disease resulting from extracellular deposition of amyloid (also known as insoluble fibrillar protein) in tissue. This disease usually shows systemic involvement (80-90%), but rarely occurs with localized disease, such as pulmonary amyloidosis. Amyloid deposition in the chest can be manifest as multiple lung nodules, tracheobronchial disease, pleural disease, and mediastinal or hilar lymphadenopathy. Multiple lung nodules are usually associated with light chain form of amyloidosis. Amyloidosis of the lung can also present with multiple and scattered lung cysts. On HRCT,
multiple, thin-walled and peripherally located cysts are seen. Multiple lung nodules (may be calcified, usually in association with Sjögren's syndrome and light chain amyloidosis) and interlobular septal thickening may also be present (Fig. 14). Isolated pulmonary amyloidosis also has a good prognosis.

**Light chain deposition disease**

Light-chain deposition disease (LCDD) is a multisystem disease that characterized by the systemic accumulation of immunoglobulin light chains. Lung involvement is extremely rare and has been described in only a few cases. On HRCT, multiple, thin-walled lung cysts and nodules are usually seen. Pulmonary LCDD should be considered in any patient with a history of hematological malignancy (especially multiple myeloma) or lymphoma presenting with multiple lung cysts and nodules.

**Desquamative interstitial pneumonia (DIP)**

Desquamative interstitial pneumonia (DIP) is a smoking-related interstitial lung disease. It is considered one of the rarest of idiopathic interstitial pneumonias. CT usually shows bilateral symmetric, basal and peripheral ground-glass opacities and small cysts within the ground-glass opacities in the lower lobes (Fig. 15).

**Neurofibromatosis type 1 (NF-1)**

Neurofibromatosis type 1 is an autosomal dominant neurocutaneous disorder and can involve lung parenchyma. On CT mediastinal and cutaneous neurofibromas, meningoceles and small, round shaped lung cysts can be seen (Fig. 16).
**Fig. 1:** CT scan through the apices obtained in a 47-year-old male with emphysema shows polygonal low-attenuation area, and central dots can be seen which represents the pulmonary artery within the secondary pulmonary lobule (arrows).

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Fig. 2: CT scan through the apices obtained in a 55-year-old male with bullous emphysema shows interlobular septa surrounding the emphysema areas mimicking walls. Emphysema has no walls.

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Fig. 3: Systemic sclerosis in a 62-year-old female patient. CT scan shows luminal dilatation of distal esophagus (red arrow), honeycomb cysts with multiple layering (blue arrow) and traction bronchiectasis (yellow arrows). CT findings consistent with fibrotic non-specific interstitial pneumonia pattern.

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Fig. 4: Williams-Campbell syndrome in a 30-year-old female patient. (A) CT scan demonstrate the presence of thin walled cystic bronchiectasis diffusely distributed throughout the lungs (arrows). (B) Sagittal reconstruction with minimun intensity projection shows cystic bronchiectasis (arrow).

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Fig. 5: Cystic fibrosis in a 17-year-old female patient. (A, B) CT scan demonstrate the presence of thic-walled cystic and tubular bronchiectasis and Tram-track sign (arrows).

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Fig. 6: Granulomatosis with polyangiitis in a 61-year-old male patient. (A) CT scan shows a cavitated mass in the right upper lung lob (arrow). (B) 6-months, later, CT scan in the same patient shows resolution of the cavitated mass with residual scarring and thin-walled cavity that mimicks lung cyst (arrow).

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Fig. 7: Pneumocystis jiroveci pneumonia in a 51-year-old male patient with hematological malignancy. CT scan shows patchy ground-glass opacities and small lung cysts.

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Fig. 8: Pneumocystis jiroveci pneumonia in a 51-year-old male patient with hematological malignancy. Sagittal CT image with minimum intensity projection (MinIP) shows patchy ground-glass opacities and small cysts.

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Fig. 9: CT images (A, B) in a patient with squamous cell lung carcinoma shows cystic metastatic nodules in both lower lobes (red arrows), irregular interlobular septal and bronchovascular interstitial thickening compatible with lymphangitic carcinomatosis (yellow arrows).

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**Fig. 10:** Thirty-six-year-old female patient with tuberous sclerosis. Chest CT images show multiple well-defined lung cysts are seen in both lungs (arrows). There is also an angiomyolipoma in the left kidney (dashed arrow).

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**Fig. 11:** Nineteen-year-old male patient with Langerhans cell histiocytosis. On HRCT images there are multiple bizarrely shaped lung cyst and nodules. In addition, the pituitary MRI also shows a thickened infundibulum (dashed arrows).

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Fig. 12: Chest CT images in a 42-year-old female patient with Birt-Hogg-Dubé syndrome shows multiple, thin-walled, lentiform shaped cysts with a lower-medial lung zone predominance (arrows).

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Fig. 13: Sixty-nine-year-old female patient with Sjogren's syndrome and lymphoid interstitial pneumonia. Chest CT images show multiple, scattered thin-walled cysts. There is also a lung mass in the right hilar region. The mass was diagnosed as primary pulmonary lymphoma.

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Fig. 14: Chest CT images in a 72-year-old male patient with Sjögren syndrome and amyloidosis shows multiple irregular lung nodules (arrows) and bizarre shaped lung cysts with a lower lung zone predominance.

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**Fig. 15:** Chest CT images in a 66-year-old male patient with heavy smoking history shows ground-glass opacities in the lower lobes and small cysts within the ground-glass opacities.

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**Fig. 16:** Chest CT images (A, B) in a 26-year-old male patient with NF-1 shows multiple cutaneous neurofibromas (dashed arrows) and a small lung cyst. CT with coronal reconstruction shows small lung cysts (arrows).

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Conclusion

The differential diagnosis of multiple cystic lung disease is extensive and associated with different pathological processes. Although LAM and PLCH are the most frequent causes of multiple cystic lung disease on HRCT, this educational exhibit summarizes the radiological approach to multiple cystic lung diseases.
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