Learning objectives

To review the definition of pulmonary cysts

To differentiate from other air filled pulmonary lesions, like an emphysematous bulla, cystic bronchiectasis, honeycombing, and a cavitating malignancy

To describe the differential diagnosis of cystic lung disease

To review the imaging findings on high resolution computed tomography (HRCT) that may help in the differential diagnosis

Background

A cyst appears as a round parenchymal lucency or low attenuating area with a well-defined interface with normal lung. Cystic lung disease can occur in a number pathological condition. Awareness of these conditions and their distinguishing imaging features, such as size, wall thickness, distribution as well as of the associated pulmonary and extra-pulmonary disease patterns may lead to the correct diagnosis.

Findings and procedure details

The first step in evaluating air filled pulmonary lesions on CT is to identify the lesions type. We present definitions of the possible lesions based on the Fleischner Society Glossary of 2008 [1].

Pulmonary cyst and their mimics

- Pulmonary cyst (Fig. 1 on page 5)

A cyst is any round circumscribed space that is surrounded by an epithelial or fibrous wall of variable thickness. On CT, cysts appear as lucent or low attenuation parenchymal lesions with well-defined borders, distributed through normal lung parenchyma.

Mimics of pulmonary cyst

- Bulla (Fig. 2 on page 6)
Bulla is airspace that result from the destruction of pulmonary parenchyma and 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. On CT, a bulla appears as a rounded focal lucency or area of decreased attenuation, 1 cm or more in diameter, bounded by a thin wall.

- Bronchiectasis (Fig. 3 on page 7)

Bronchiectasis is irreversible localized or diffuse bronchial dilatation, usually resulting from chronic infection, proximal airway obstruction, or congenital bronchial abnormality.

-Honeycombing (Fig. 4 on page 8)

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. On CT, the appearance is of clustered cystic air spaces, typically of comparable diameters on the order of 3-10 mm.

Cystic lung disease

Cystic lung disease can occur in a number of pathological conditions. Important clues to the differential diagnosis are:

- Shape of cyst
- Distribution of cysts
- Presence of ancillary findings

1. Pulmonary Langerhans cell hysti cytosis (PLCH) (Fig. 5 on page 9)

- Clinical manifestations

Most cases of PLCH occur in young adults between 20 and 40 years of age. Etiology is unknown, but cigarette smoking plays a primary role.

When patients present with symptoms, the most common symptoms include nonproductive cough, dyspnea, chest pain, fatigue and fever.

- Imaging features [2, 3]
  
  • Cysts may be round but are often irregular or bizarre shapes.
  • Upper lung zones are predominantly involved. Relative sparing of lung bases
• There may be few or innumerable nodules, usually with irregular margins and in a centrilobular or peribronchial distribution.

- Associated features
  • Skeletal involvement: lytic or expansile
  • Lymphadenopathy

2. Pulmonary lymphangioleiomyomatosis (LAM) (Fig. 6 on page 10)

- Clinical manifestations

The most common presentations in patients with sporadic LAM are those of progressive dyspnea and spontaneous pneumothorax.

LAM has rarely been reported in men, almost always in association with definite or probable tuberous sclerosis complex (TSC).

- Imaging features [2]
  • Diffuse thin-walled cysts surrounded by normal lung without regional sparing
  • Cysts are typically round or ovoid
  • Patchy ground-glass opacities

- Associated features
  • Pneumothorax
  • Chylothorax

3. Birt-Hogg-Dubé syndrome (BHD) (Fig. 7 on page 11)

- Clinical manifestations

BHD syndrome is an inherited syndrome in which affected individuals are at risk for the development of bilateral, multifocal kidney cancer, as well as various pulmonary and dermatologic lesions.

Approximately 80 percent of patients with BHD have multiple pulmonary cysts. Spontaneous pneumothorax may be seen in up to one-fourth of patients.

- Imaging features [4]
  • Multiple round or irregular shaped cysts of various sizes with lower medial lung zone predominance

- Associated features
4. Lymphocytic interstitial pneumonia (LIP) (Fig. 8 on page 12)

- Clinical manifestations

LIP is one entity within a spectrum of lymphoproliferative disorders that can involve the lung. Most cases of LIP are associated with other conditions, although idiopathic cases still rarely occur. Sjögren syndrome is associated with one-fourth of reported cases of LIP. Fewer than 5 percent of LIP patients are asymptomatic at presentation.

- Imaging features [5]
  - Bilateral ground-glass opacities
  - Bronchovascular bundle thickening
  - Interlobular septal thickening
  - Poorly defined centrilobular nodules (perivascular and subpleural distribution)
  - Multiple thin-walled cysts

5. Neurofibromatosis type 1 (NF-1) (Fig. 9)

- Clinical manifestations

NF1 is an autosomal dominant genetic disorder with an incidence of approximately 1 in 2600 to 1 in 3000 individuals. The typical order of appearance of clinical manifestations is café-au-lait macules, axillary and/or inguinal freckling, iris hamartomas, and neurofibromas. Interstitial lung disease and cystic lung disease have been reported.

- Imaging features [6]
  - Cystic and bullous lesion with upper lung zone predominance
  - Fibrosis with lower lung zone predominance

- Associated features
  - Neurofibromas
  - Scoliosis

Summary of distinguishing features of cystic lung disease (Fig. 9 on page 13, Table 1 on page 14)

Images for this section:
Fig. 1: Pulmonary cyst. CT shows multiple air filled well-circumscribed lesions with thin walls.
Fig. 2: Bullae. CT shows focal lucency of decreased attenuation bounded by a thin wall.
**Fig. 3:** Bronchiectasis associated with agammaglobulinemia. CT shows air-filled space that branches and connects with the airway.
Fig. 4: Honeycombing. CT shows clustered subpleural air spaces with relatively thick walls.
Fig. 5: Pulmonary Langerhans cell hystiocyteisis. CT shows multiple thin wall cysts. Multiple cysts, some with bizarre shape, predominantly affect the upper lung zone and spare the bases.
Fig. 6: Pulmonary lymphangioleiomyomatosis. CT shows multiple thin wall cysts in all lung zones.
Fig. 7: Birt-Hogg-Dubé syndrome. CT reveals lower lung-predominant thin-walled cysts that vary in size and shape.
**Fig. 8:** Lymphocytic interstitial pneumonia associated with Sjögren syndrome. CT shows multiple thin-walled cysts and multiple nodules.
**Fig. 9:** Distribution and shape of pulmonary cysts.
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Conclusion

Awareness of spectrum of HRCT findings of cystic lung disease may help reach a correct diagnosis.

Personal information

References


