Cystic Lung Diseases: Spectrum of Radiologic Findings

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Learning objectives

- To be aware of the main etiologies of Cystic Lung Lesions.
- To become familiar with imaging findings Cystic Lung Diseases.
- To distinguish cysts from cavities and to categorize focal, multifocal and diffuse distribution.
- To learn about diagnostic pitfalls of Cystic Lung Diseases.

Background

We report a retrospective study that evaluates 67 patients with Cystic Lung Diseases: 35 males and 32 females with a mean age of 48.6 years (range, 4-72 years), seen during the four year-period from January 2010 to December 2013.

Patients were admitted with variant clinical features: cough, fever, expectoration, wheezing, and chest pain. All our patients underwent chest radiograph and CT scan.

Diagnosis of Cystic Lung Diseases were confirmed by imaging findings and if necessary with histopathological analysis.

The term cyst is used to mean a clearly defined air-containing space surrounded by a relatively thin (#4 mm) wall, which differentiates them from cavitary processes, which have thicker and irregular walls and will not be discussed in this pictorial essay.

Imaging findings OR Procedure details

The results that were obtained:

**Focal or multifocal cystic lesions**: blebs and bullae (n=16), pneumatoceles (n=14), several infectious processes, including hydatid disease (n=10), Pneumocystis jiroveci pneumonia (n=2) and coccidioidomycosis (n=1), congenital pulmonary airway malformation (n=4), lymphocytic interstitial pneumonia (n=3), cystic metastases (n=1).
Diffuse cystic lesions: honeycomb lung associated with advanced fibrosis (n=11), lymphangioleiomyomatosis (n=3), Langerhans cell histiocytosis (n=2).

**Focal or multifocal cystic lesions:**

**Bullae and blebs**

A bulla is generally defined as a sharply demarcated air-containing space of 1 cm or more in diameter within the lung that possesses a smooth wall that is less than 1 mm in thickness. Bullae usually result from coalescence of emphysematous spaces or from a Ball-valve type of airway obstruction (Fig.1).

A bleb is a localized collection of air in the immediate subpleural lung or within the pleura and is usually less than 1 cm in diameter. Blebs are usually located in the apex of the lung and have been attributed to congenital abnormalities, greater distending pressure at the apex of the lung, or obstructive airway diseases.

**Pneumatoceles**

Pneumatoceles are thin-walled, gas-filled spaces in the lung that are most frequently caused by pneumonia, trauma, or inhalation of hydrocarbon fluid. In the acute phase, pneumatoceles can be thick-walled. They tend to become thinner in the chronic phase (Fig.2).

**Infectious processes**

**hydatid disease**

The localization of hydatid cysts in humans is mostly hepatic (55 to 75 %), with the lungs being the second most frequent location in adults (10 to %).

Radiographically, the cysts are commonly seen as spherical, homogenous masses with smooth borders surrounded by normal lung tissue (Fig.3,4). An intact cysts is filled with clear fluid. Cysts may rupture spontaneously or due to trauma. As the cysts enlarges and erodes into the bronchioles, air enters the potential space between pericyst and endocyst and appears as a thin lucent crescent (crescent or meniscus sign)(Fig.5). When hydatid
cyst completely collapsed, the crumpled endocyst floats freely in the cyst fluid (water lily sign) (Fig.6).

In cases of completely empty cysts (Fig.7), a variety of diseases that produces or mimic air-containing cavities should be considered in the differential diagnosis.

Pneumocystis jiroveci pneumonia

Pneumocystis jiroveci pneumonia (PJP), formerly known as Pneumocystis carinii pneumonia, was classified as a protozoan but is now known to be a fungal infection. It is almost exclusively seen in immunosuppressed patients, especially in those with AIDS who have a CD4 count below 200 cells/mm³.

The symptoms can be insidious but may progress to respiratory failure and even death, often following a rapid, fulminant course if left untreated. Once a common opportunistic infection.

Although up to 90% of chest x-rays in patients with PCP are abnormal, appearances are often non-specific. Between 10 - 15% of patients with PCP have normal chest radiographs and close to 30% have non-specific or inconclusive findings.

The most common radiographic finding in patients with PJP consists of diffuse, bilateral symmetric reticulonodular ground-glass, or finely granular opacities occurring in a perihilar, or lower lung zone distribution (Fig.8). Less commonly, PJP will occur with upper lobe predominance. If untreated, these opacities may manifest as diffuse airspace consolidation.

Other radiographic findings include single or multiple nodules, miliary nodules, cavitation, hilar or mediastinal adenopathy, and pleural effusion.

On CT, PJP often appears as a pattern of bilateral, multifocal, mainly symmetric ground-glass opacities (Fig.8). More focal areas of consolidation are also common. PJP-related cysts tend to occur mostly after multiple infections, can have an apical predominance, and may lead to pneumothoraces, which portend a poor outcome. Other less frequent manifestations include a reticular or reticulonodular pattern and pulmonary nodules.

Pulmonary coccidioidomycosis infection
Cavitation is a major characteristic of chronic pulmonary coccidioidomycosis, and the
wall of the focal lesion is often thin (Fig.9). A thin-walled cyst also can represent residue from a resolved primary infection.

**Congenital Pulmonary Airway Malformation**

Congenital pulmonary airway malformation, formerly known as congenital cystic adenomatoid malformation, is a rare multicystic, intralobar mass of disorganized lung tissue, most often seen in the lower lobes.

It is usually diagnosed in children but can be left unrecognized until adulthood.

It can be categorized into 3 subtypes on the basis of its appearance.

**Type 1** is most frequent and consists of multiple cysts 2 cm or greater in diameter. On CT, the disease appears as a large, air-filled multicystic lesion (Fig.10).

**Type 2** is defined as multiple cysts smaller than 2 cm.

In **type 3**, the least frequent type, there are numerous microscopic cysts that often have a more solid appearance on imaging.

Complications of congenital pulmonary airway malformation include infection and malignant transformation.

Treatment of congenital cystic lesions usually consists of simple resection of the involved tissue or lobectomy.

**lymphocytic interstitial pneumonia**

Lymphocytic interstitial pneumonia (LIP) is a benign lymphoproliferative disorder characterised by lymphocyte predominant infiltration of the lungs.

It is classified as a subtype of interstitial lung disease.

It also falls under the umbrella of non lymphomatous pulmonary lymphoid disorders.

LIP is typically associated with collagen-vascular diseases, especially Sjögren syndrome, and AIDS. Other less frequently reported disorders associated with LIP include
autoimmune thyroid disease, Castleman disease, systemic lupus erythematosus, myasthenia gravis, pernicious anemia, and chronic active hepatitis.

LIP may produce a fine linear or reticulonodular pattern on chest radiographs.

On CT, the lung cysts are usually less numerous than in LCH or LAM. During the acute phase, diffuse or multifocal ground-glass opacities are seen. However, these opacities may regress so that cysts are the only residual finding in more chronic cases (Fig.11). Less frequently, LIP presents with a reticular or reticulonodular pattern, lung nodules, or consolidation. Lymph node enlargement can also occur.

The usual treatment is steroid therapy, after which ground-glass opacities can resolve. The cysts, however, persist even after therapy.

**Cystic metastases**

Thin-walled cystic metastases develop primarily in sarcoma (Fig.12), squamous cell cancer, transitional cell carcinoma of the bladder (Fig.13), and melanoma. Less frequently, this pattern may be seen with lymphoma.

The appropriate history of primary malignancy is critical to suggest this diagnosis.

As with other metastatic lesions, cystic metastases tend to have different sizes and a basilar predominance.

A potential complication of cystic metastases is pneumothorax, which may require placement of a chest tube.

**Diffuse cystic lesions**

**honeycomb lung associated with advanced fibrosis:**

Honeycombing is one of the manifestations of fibrotic lung disease. The most common cause of honeycombing is usual interstitial pneumonia (UIP), but this appearance can also be seen in conditions such as nonspecific interstitial pneumonia, sarcoidosis, chronic hypersensitivity pneumonitis, asbestosis, and postradiation fibrosis.
CT demonstrates clustered cystic air spaces (between 0.3-1.0 cm in diameter), which are usually subpleural and basal in distribution. The walls of the cysts are well-defined and often thick (1-3 mm). It is usually accompanied by other signs of fibrosis, such as traction bronchiectasis or bronchiolectasis, reticulation, and ground-glass opacities (Fig.14,15).

**Lymphangioleiomyomatosis**

Lymphangioleiomyomatosis (LAM) is a rare hamartomatosis characterized by smooth cell hyperplasia along the terminal bronchioles, lymphatic vessels and blood vessels. (LAM) primarily affects women of childbearing age. However, it also can present after menopause in women undergoing estrogen hormonal treatment.

On chest radiographs, there are increased lung volumes and often a suspicion of linear or ringlike lucencies.

Characteristic CT features of lymphangioleiomyomatosis are diffuse thin-walled cysts surrounded by normal lung without regional sparing (Figs. 1-3). Cysts are usually 2-5 mm but can be as large as 25-30 mm. Cysts are typically round or ovoid, but they may become polygonal with severe parenchymal involvement (Fig.16). Small centrilobular nodules corresponding to hyperplastic muscle or pneumocyte hyperplasia have been reported.

Focal ground-glass opacities may be due to smooth-muscle proliferation, hemosiderosis, or pulmonary hemorrhage. Lymphatic obstruction may cause septal thickening.

Lung transplantation is the definitive treatment of this progressive disease, but recurrence can occur in the transplanted lung.

**Langerhans cell histiocytosis**

Pulmonary LCH is a smoking-related lung disease, with 80-100% of cases seen in patients who smoke or have a history of smoking and occurs most frequently in young adults.

LCH is characterized pathologically by the formation of granulomas containing Langerhans cells.
Clinical findings include cough, dyspnea, and fatigue. Pneumothorax can also occur, either at the time of presentation or later during the course of the disease.

As with other smoking-related diseases, such as emphysema, respiratory bronchiolitis, and desquamative interstitial pneumonia, LCH typically has upper lobe predominance with relative sparing of both bases.

Radiographically, LCH appears as multiple nodules that range from 1 to 10 mm. The presence of cysts usually occurs only during the later stages of the disease.

On CT, bizarre-shaped cysts are associated with noncavitary nodules that progressively cavitate to become thickwalled cavitary nodules and then thin-walled cysts (Fig. 17).

The CT findings of LCH are often characteristic and reflect the temporal heterogeneity of the disease.

Treatment consists of smoking cessation, which can result in improvement of the abnormalities.

**Pitfalls**

Emphysematous changes sometimes can be mistaken for multifocal lung cysts. In contrast to true cysts, the cystlike lucencies caused by the destruction of lung parenchyma in emphysema do not have walls (Fig. 18). Nevertheless, emphysema can be accompanied by multiple cysts, such as bullae, or in association with LCH.

Cystic bronchiectasis can also be mistaken for cysts at CT if thin-section images are not reviewed. Multiplanar reformations can be helpful to distinguish cystic bronchiectasis, in which there is continuity of a bronchus with thin-walled lucencies and true cysts, in which such communication does not exist (Fig. 19).

**Images for this section:**
**Fig. 1:** Multiple upper lobe predominant bullae (arrow) in emphysematous patient. Note a right pleural effusion.

**Fig. 2:** Thoracic trauma in 4 year-old-boy Chest radiograph (A) and axial CT scan (B) show a pneumotocele with relatively thin walled in left lower lobe(arrow). Note a left pleural effusion
Fig. 3: Unruptured pulmonary cystic echinococcosis in a 40 year-old-man. Posteroanterior chest radiograph showing well-defined, rounded opacities, involving both lungs (Fig.3)
Fig. 4: The same case as Fig 3. Axial CT scan show a well defined thin walled fluid attenuation lesions in both lungs(arrow).

Fig. 5: Ruptured hydatid cyst in a 45 year-old -man. Posteroanterior chest radiograph showing rounded opacities with a thin lucent crescent (arrowhead), involving the right lung.
Fig. 6: Hydatid cyst in 35 year-old -man. The CT scan shows the water lily sign created by collapsed and crumpled endocysts floating freely in the most dependent part of the cyst.

Fig. 7: Ruptured hydatid cyst in a 45 year-old -woman. CT scan shows an "empty cyst" after complete evacuation parasitic membranes (arrowhead).
**Fig. 8:** Pneumocystis jiroveci pneumonia. A, Chest radiograph shows bilateral, symmetric, mostly perihilar and bibasilar ground-glass opacity. B, Transverse CT image confirms bilateral, widespread ground-glass opacity and shows scattered lung cysts (arrow).
Fig. 9: Posteroanterior chest radiograph shows a cystic lesion in the left mid lung due to coccidioidomycosis infection (arrow).
Fig. 10: Congenital pulmonary airway malformation type 1 in 4 year-old-boy. (A) chest radiograph shows thin walled cysts with multiple internal septa expand left lung and displace diaphragm and mediastinum (B) CT scan shows replacement of the left lung by cystic lesions of varying sizes.
Fig. 11: Lymphocytic interstitial pneumonia in patient with Sjögren syndrome. High-resolution CT image shows thinwalled cysts of various sizes, some of which are subpleural.
Fig. 12: Cystic metastases in woman with soft-tissue sarcoma of the right thigh. Axial and coronal CT scan show thin-walled cystic lesion in the right upper (white arrow) lobe with bilateral lung nodules (red arrow).

Fig. 13: Cystic metastasis in man with metastatic bladder cancer. CT scan shows a small thin-walled cyst in the left lower lobe (white arrow).
Fig. 14: pulmonary fibrosis with prominent honeycombing a 55 year-old man.
**Fig. 15:** The honeycombing in patient with non-specific interstitial pneumonia. Axial CT scan shows honeycomb cysts with a distinct predominance in the peripheral and subpleural regions.
Fig. 16: Axial MDCT image, obtained in patient with progressive dyspnea and pulmonary hypertension shows the round, and thin-walled cysts typical of Lymphangioleiomyomatosis.
**Fig. 17:** Histiocytosis X in a 45 year-old-man. Axial CT scan shows a combination of cysts (red arrows) and nodules (white arrow). The cysts are round and well defined.
Fig. 18: Emphysema. Transverse CT image shows upper lobe predominant lucencies (arrows) with no visible wall, caused by destruction of lung parenchyma.
**Fig. 19:** Cystic bronchiectasis in a 35 year-old-man. Axial CT image shows cystlike lesions (white arrow) contiguous with dilated bronchi (red arrow).
Conclusion

Cystic lung lesions can be caused by a diverse array of pathologic processes.

High-resolution CT findings including morphology, location, distribution of the lung cysts, correlated with the tempo of the disease process and the clinical context provide the basis for management of Cystic Lung Diseases in order to avoid unnecessary procedure or delayed treatment.

References


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