Cystic and Nodular Lung Disease

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KEYWORDS

• Cystic lung disease • Nodular lung disease • High-resolution computed tomography

KEY POINTS

- A lung cyst is a lucent structure with a thin perceptible wall. When multiple lung cysts are present, the spectrum of cystic lung disease should be considered.
- Recognizing zonal predominance and cyst morphology is helpful in reaching an accurate diagnosis of cystic lung disease. Abnormalities within the intervening lung parenchyma and outside the thorax may also be helpful.
- Entities such as emphysema, honeycombing, cavities, and bronchiectasis can mimic true cystic lung disease.
- Nodular lung disease is best characterized by the relationship to the secondary pulmonary lobule (SPL), with the 3 main patterns being centrilobular, perilymphatic, and random.
- A multimodality approach including clinical history, physical examination, and imaging is often essential to narrow down the differential diagnosis of nodular lung disease.

INTRODUCTION

Diffuse cystic and nodular lung diseases often have characteristic imaging findings, which may allow the radiologist to be the first to suggest the diagnosis. Diffuse cystic lung diseases are rare entities. The most common causes of cystic lung disease are lymphangioleiomyomatosis (LAM) and Langerhans cell histiocytosis (LCH); differentiation of LAM from LCH on computed tomography (CT) of the chest is readily made in most clinical cases. Other less common cystic lung diseases include Birt-Hogg-Dube syndrome, lymphocytic interstitial pneumonitis, and light chain deposition disease. Although emphysema, honeycombing, cavities, and bronchiectasis mimic cystic lung disease, careful inspection of CT images usually allows one to differentiate true cystic lung disease from these entities. Diffuse nodular lung disease are categorized into 3 main categories based on nodule relationship to the SPL: centrilobular, perilymphatic, and random. Each of these categories carries a unique differential diagnosis. In diffuse nodular lung disease, a specific diagnosis can be achieved through a combination of history, physical examination, and imaging findings.

CYSTIC LUNG DISEASE

The Fleischner Society defines a cyst as a round parenchymal lucency or low-attenuating area with a well-defined interface with normal lung.[1] The presence of multiple cysts throughout the lung signals the presence of cystic lung disease, which carries a short differential diagnosis. A specific diagnosis can be made based on the imaging appearance of lung cysts and their axial and zonal distribution.

Lymphangioleiomyomatosis

LAM is a rare multiorgan disorder, which occurs almost exclusively in women of childbearing

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age.[2,3] Pathologically, LAM is characterized by smooth muscle proliferation along the pulmonary interstitium affecting the lymphatics, vessels, airways, alveolar septa, and pleura.[4] LAM can occur sporadically or in association with tuberous sclerosis complex (TSC-LAM), although TSC-LAM is 5 to 10 times more common than sporadic LAM. LAM occurs in 30% of women with TSC.[2] Clinically, patients present with progressing dyspnea on exertion, recurrent pleural effusions (chylothorax), and spontaneous pneumothoraces.[5] In a series of 32 patients with LAM, exertional dyspnea (47%) and spontaneous pneumothorax (53%) were the most common findings at presentation, whereas 81% of patients within this cohort exhibited spontaneous pneumothorax at some point during the course of the disease.[6]

Appearance on chest radiography varies with disease severity. The radiograph may show normal results or increased reticular markings related to superimposition of cyst walls. As the disease progresses-, the cysts become more apparent and may have a lower lung zone predominance. Lung volumes are normal to increased. Pleural effusion or spontaneous pneumothorax can occur and is usually readily apparent on imaging.[3,6–8]

Results on CT are almost always abnormal, and CT can demonstrate parenchymal abnormality even when the radiograph shows normal results.[9] High-resolution computed tomography (HRCT) is superior to traditional CT in defining the parenchymal abnormality in LAM, with the characteristic features being diffuse, thin-walled cysts with normal intervening parenchyma (**Fig. 1**). The cysts tend to be round and uniform in shape measuring 3 to 5 mm, although cysts measuring up to 25 to 30 mm have been reported.[3,4,9] Occasionally, the cysts may appear polygonal in shape or fused.[10,11] Cysts enlarge and become more numerous as the disease progresses.[9–11]

The prevalence of LAM in patients with TSC varies, although age seems to be a critical risk factor for the development of cystic lung disease, as Cudzilo and colleagues[12] reported a cystic lung disease prevalence of 81% in subjects with TSC older than 40 years. Multifocal micronodular pneumocyte hyperplasia (MMPH) is an additional feature seen in patients with TSC. MMPH is a hamartomatous process of the lungs, which manifests at imaging with diffusely scattered, randomly distributed nodules measuring 1 to 8 mm in diameter.[13,14]

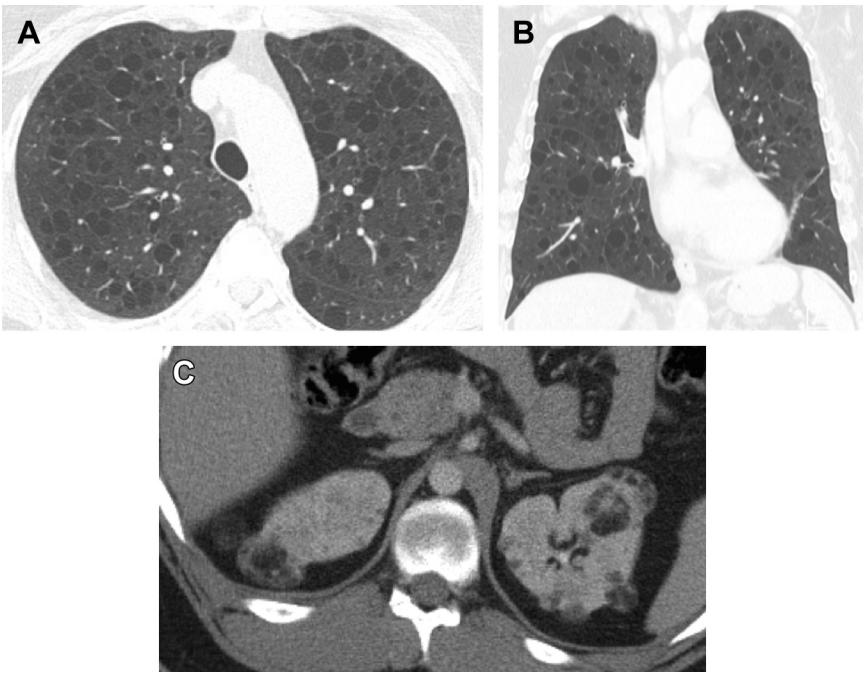

Fig. 1. Lymphangioleiomyomatosis. (*A*) Axial CT shows round, uniform cysts with normal intervening lung parenchyma. (*B*) Coronal reformation showing diffuse distribution of cysts without zonal predominance. (*C*) Axial CT of the abdomen showing multiple fat-attenuation lesions in both kidneys, consistent with angiomyolipomas in a patient with tuberous sclerosis. Grossly, one can infer that the lesions are fatty by comparing the internal density with that of adjacent abdominal fat.