Systematic approach to cystic diseases of the lung

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Authors: J. Coleman, S. Abbara, K. Jordan, M. Landay, K. Batra; Dallas/US
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Learning objectives

The purpose of this exhibit is to detail a simple and effective framework for analyzing and characterizing cystic diseases of the lung based on clinical and radiological features, as well as outlining the role of biopsy in definitive assessment of these entities.
Background

Cystic diseases of the lung are almost uniformly uncommon or rare, however, when identified, careful consideration is warranted since many of the responsible etiologies have far-reaching clinical implications, from complications, particularly if left untreated. Potential causes of cystic lung disease include congenital, genetic, infectious, inflammatory and neoplastic etiologies which can be effectively characterized based on pulmonary and extrapulmonary radiological features, often in concert with clinical and pathological findings. (Table with list of Causes) (Table with decision tree for diagnosis)

Essential in the initial assessment of a cystic lung lesion is differentiation from the often similar, but altogether different cavitary lesion. By definition, a pulmonary cyst is a parenchymal lucency with a wall of less than 2-3mm which is most often air-filled, but rarely can be fluid containing. In contrast, a cavitary pulmonary lesion is an air-filled structure developing within a region of consolidation with a wall-thickness of greater than 4mm. Similarly, pulmonary blebs and bullae, though they also feature a thin wall, typically imperceptible, can be differentiated from true parenchymal cysts by their subpleural location and wall thickness of less than 1mm. The diagnostic considerations for each of these lesions are vastly different which makes differentiating these types of lesions of the utmost importance. (Figure with comparison of cyst, cavity, bleb/bulla). A multidisciplinary approach is of utmost importance and helps in avoiding biopsy in certain cases.

Findings and Procedure details:

Potential etiologies for cystic lung disease can be initially divided into those which have cysts as the dominant feature, and those which feature cysts as ancillary findings with more prominent additional parenchymal abnormalities. Each of these categories can be further subdivided based on the distribution of disease. Within these categories, possibly etiologies can be frequently differentiated based on additional radiographic features or clinical history. While chronic cough and dyspnea remain nonspecific features of cystic lung diseases, spontaneous pneumothorax may be the initiating event leading to diagnosis of diffuse cystic lung disease. Chest xray remains a nonspecific modality with either showing normal findings or increased lung volumes and a nonspecific reticular pattern. CT or HRCT is the modality of choice for assessment of lung volume, size, wall thickness, shape and distribution of pulmonary cysts, and associated findings like nodules, septal thickening, pleural effusion, lymphadenopathy and extrathoracic abnormalities.
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**Etiologies with Cysts as Dominant feature:**

**Diffuse or Random Distribution:**

Lymphangioleiomyomatosis

- Lymphangioleiomyomatosis is a multi-system disease that is most often associated with Tuberous Sclerosis, but can occur sporadically, classically in childbearing age females. In most cases this disease manifests with multiple, uniformly distributed round thin-wall (2-5 mm) cysts throughout the lungs. Associated findings include interlobular septal thickening, hyperinflation, patchy ground glass opacities from recurrent hemorrhage and recurrent pneumothoraces. Sporadic-LAM also manifests with lymphatic abnormalities that can lead to chylous pericardial, pleural effusions, thoracic duct enlargement, diffuse thickened mediastinum with decreased density, and lymphangioleiomyoma in the thorax and mediastinum. HRCT appearance is classified as characteristic or compatible with this diagnosis. A characteristic HRCT appearance requires multiple (> 10) thin walled cysts with normal or increased volumes and no significant interstitial abnormality. Compatible HRCT are defined by fewer (>2 but < 10) typical cysts. Additionally, several articles have shown that assessment of serum VEGF-D levels can be useful in differentiating sporadic lymphangioleiomyomatosis from other cystic lung diseases.

Langerhans Cell Histiocytosis: See below
Birt-Hogg-Dube syndrome:

- A multisystem genetic disorder resulting from a mutation in the FLCN gene, classic manifestations include multiple pulmonary cysts, renal tumors and various cutaneous lesions. Although they can be randomly distributed, or appear so when manifesting as only a few lesions, classically the pulmonary cysts in Birt-Hogg-Dube are lower lung predominant. Cysts may be quite large and may be septated, which is more common in larger cysts. Cysts often display variable morphology, often presenting as oval or lentiform in shape and are often perifissural. As with LAM, recurrent pneumothoraces are an unfortunately common occurrence.

Lymphocytic Interstitial pneumonia - See below

Pulmonary amyloidosis.

- Although a rare presentation of amyloidosis, two distinct subtypes of pulmonary amyloidosis have been characterized, the nodular form, and the diffuse form. As the name suggests, nodular pulmonary amyloidosis is most commonly characterized by diffuse randomly distributed nodular densities, with sharp or lobular contours, sometimes displaying internal calcification and usually less than 15mm in diameter. Very rarely pulmonary cysts may also form, however these often form in conjunction with nodular densities elsewhere in the lung. A clue to this diagnosis is often that the patient is asymptomatic, only rarely presenting with associated cough. The more rare subtype, diffuse pulmonary amyloidosis in contrast more commonly manifests with symptoms. Nonspecific interstitial opacities are the hallmark of this subtype with interlobular septal thickening a common associated feature. Calcification is less common in this subtype, though may still occur.

Tracheobronchial Papillomatosis:

- The most common benign tumor of the tracheolaryngeal airway, with a bimodal age distribution, frequently presents with multiple cystic lung lesions. The lesions themselves are squamous papillomas most widely thought to occur in association with HPV infection, particularly with types 6 and 11. Often these occur in association with adjacent nodules. Most frequently the distribution is random, however, with more extensive disease, the cysts can exhibit a predilection for the dependant portions of the lung. Additional associated findings include bronchiectasis, mucus plugging and distal atelectasis.

Cystic Primary and Metastatic disease

- An atypical presentation for pulmonary malignancy, cystic pulmonary primary malignancy and metastases have been described in many cancer types. Unlike in
malignant lesions elsewhere in the body, these can appear develop internal air, related to expulsion of necrotic content. Additional contributing factors to cyst formation include alveolar wall disruption by invading tumor cells, and development of neoplasm in pre-existing cystic lesions. Typically patients will present with a history of cancer or have additional findings to suggest malignancy as a possibility.

Pulmonary interstitial emphysema

- Most commonly associated with neonates, and highly associated with mechanical, positive pressure ventilation, this condition is thought to arise from rupture of alveoli resulting in infiltration of air into the lymphatics and interstitium. Often diagnosis can be made based solely on chest radiographs. Cystic spaces in this etiology can commonly be serpiginous in shape and may cluster around pulmonary artery branches.

Cystic Fibrohistiocytic tumor of the lung

- An extremely rare tumor, cystic fibrohistiocytic tumor of the lung has been described as both a primary benign lung neoplasm or as a metastatic lesion in patients with similar appearing benign fibrohistiocytic tumor of the skin. Radiographically these appear as either solitary or multiple cystic lesions, sometimes in association with scattered or adjacent nodules, which makes it difficult to distinguish from the more common Langerhans cell histiocytosis, which also presents this way.

Cystic Mesenchymal Hamartoma

- Another rare benign neoplasm, cystic pulmonary mesenchymal hamartomas typically present as multiple bilateral cysts and nodules which are randomly distributed. As with cystic fibrohistiocytic tumor of the lung, this can be difficult to distinguish from Langerhans cell histiocytosis. Although these lesions, like all hamartomas are classically benign, there have been reported cases of sarcomatous degeneration arising from these lesions.

Chronic Coccidioidomycosis and Paracoccidioidomycosis

- Although Coccidioidomycosis (endemic to the San Joaquin Valley) and Paracoccidioidomycosis (endemic to South America) are distinct entities caused by different endemic, thermally dimorphic fungi, their pulmonary manifestations are similar. Pulmonary findings are a common manifestation of these infections, and in the acute setting are characterized by Reticulonodular densities, ground glass attenuation, consolidation or cavity formation. Lymph node enlargement is also a common finding. Chronic infection may lead to fibrosis, predominantly in a perihilar distribution or rarely (reported in approximately 10% of cases) cyst formation where the cysts are typically scattered. Treatment for both of these mycoses involved a 3-6-month course of anti-
fungals which the pulmonary parenchymal findings usually demonstrating improvement by 3 months.

**Hereditary syndromes: Limited cysts**

Marfan syndrome:

- Marfan syndrome is a type of inherited connective tissue disorder the result of defect in the fibrillin 1 gene with a reported prevalence of approximately 6/100,000. Patients are typically of tall stature with disproportionate extremity length and are highly predisposed to developing aortic root dilation and resultant dissection which is the most common cause of death. Multiple organ systems can be affected which, in addition to the skeletal and cardiovascular manifestations include ocular symptoms, including lens dislocation, and pulmonary findings, including lung cysts and bullae, as well as spontaneous pneumothoraces. Typically patients will have known or suspected diagnosis at the time of imaging, however it is important to realize that lung cysts in these patient are a common finding related to their underlying genetic disorder and not necessarily the result of a second superimposed disease process.

NF-1

- Neurofibromatosis type 1 is the most common phakomatosis, affecting multiple organ systems, a complete discussion of which is beyond the scope of this poster. Most commonly pulmonary manifestations are related to neurofibroma formation and may involve the chest wall or mediastinum. Less commonly cysts or bullae may form which demonstrate an upper lung predominance. Unlike emphysema, these cysts have a well-defined wall. Fibrosis, and pulmonary arterial hypertension may also occur.

Ehlers-Danlos Syndrome

- Multiple subtypes of Ehlers-Danlos syndrome have been described, many of which are so rare that the clinical implications have not been fully characterized. Type Iv, also called vascular Ehlers-Danlos syndrome, though more typically associated with aneurysm formation and dissections, has been reported to have pulmonary complications including hemoptysis, multiple cavitary lesions, and pulmonary parenchymal cysts which may spontaneously occur and regress.

Proteus Syndrome

- Proteus syndrome is a congenital syndrome characterized by multiple hamartomatous lesions, hemimegalencephaly, hemihypertrophy and lipodystrophy. As with other multisystemic syndromes, the diagnosis is usually known or suspected based on clinical
findings at the time of imaging. Pulmonary manifestations may occur at an early age with cases of rapidly progressive cystic lung disease having been reported in the literature in the pediatric population, which may lead to respiratory compromise. The underlying mechanism for this development is not known, although it has been hypothesized that it shares a similar mechanism to that of LAM.

**Common PITFALLS:**

Centrilobular emphysema:

The most prevalent subtype of pulmonary emphysema, centrilobular emphysema may appear similarly to cystic lung disease, particularly in the early stage when the process is confined to the secondary pulmonary lobule. The underlying pathology involves alveolar damage, sparing the centrilobular vascular structure with accounts for the "central vessel sign" characterized as lucency surrounding an intact pulmonary vessel. The pulmonary vessels in the emphysematous regions may be small then in spared areas due to hypoventilation and subsequent ventilation-perfusion matching. Unlike cystic lung disease, the areas of emphysematous transformation display hyperexpansion, an imperceptible (as opposed to thin) wall, and will commonly coalesce to form larger hypodense regions.

Cystic bronchiectasis

- Cystic bronchiectasis is a subtype of bronchiectasis and can occur as the result of numerous etiologies. As the name implies, this entity is characterized as cystic bronchial dilation extending to the pleural surface. Differentiation from true cystic lung disease is made by identifying connections between the cystic appearing airspaces or more characteristic appearing bronchials, often greatly assisted through the use of multiple imaging planes.

Pneumatoceles

- Pneumatoceles are most commonly associated with ventilator induced lung injury or as a post-infectious finding, particularly with Staphylococcus aureus. These can be difficult to distinguish from true cystic lung lesions once fully formed, however clinical context provides the largest clue to diagnosis. Early on in the course of development these lesions are typically surrounded by consolidation. Pneumatoceles may persists beyond the resolution of symptoms and may become secondarily superinfected or may rupture leading to pneumothorax.

**Upper Lobe Predominant:**
Langerhans Cell Histiocytosis:

- Classically occurring in younger adults who either have a history or smoking or are current smokers, Langerhans cell histiocytosis is a rare disorder that can manifest with multiple cystic lung lesions as a primary lung process or as part of a multi-system disorder. Although this is often included in the differential diagnosis for diffuse cystic lung lesions, it is more commonly seen as an upper or mid lung predominant disease, with sparing of the costophrenic angles. There is a spectrum of findings from early to late disease with the early phase characterized by a more nodular appearance, and a more cystic appearance in the late stage. The cysts can be bizarre in shape. Typically these findings coexist, which is frequently a clue to the correct diagnosis. Irregular margins of the cysts are present with adjacent fibrotic changes.

EXTRAPULMONARY FINDINGS: Bone lesions(< 20% patients), diabetes insipidus and skin lesions are the most common extrapulmonary manifestations. Definitive diagnosis of PLCH requires surgical lung biopsy with identification of Langerhans cell granulomas.

Complications: Pneumothorax, pulmonary hypertension

_Lower Lobe Predominent:_

Lymphocytic Interstitial pneumonitis

- The result of diffuse lymphocytic infiltration of the lungs, LIP can occur at any age, but is most commonly seen in the 6th decade of life. LIP is associated with a variety of autoimmune or immunocompromising disorders, most commonly Sjogren syndrome, and is frequently found in association with polyclonal or monoclonal IgM gammopathy. The most classical presenting radiographic feature is multiple bilateral thin-walled cysts, which may be diffuse, but are most commonly described as appearing in a mid to lower lung predisposition. Typically these cysts are less than 30mm in diameter. Associated features include thickening of the interstitium and bronchovascular bundles, multiple small nodules, and ground glass changes. Late-stage manifestations include fibrosis and honeycombing, with predisposition for lymphoproliferative disorders and pulmonary amyloidosis

Birt-Hogg-Dube - See Above

_Etiologies with Cysts as Ancillary Feature_

_Diffuse or Random distribution:_
Desquamative Interstitial Pneumonia

- One of the rarer interstitial pneumonias, Desquamative Interstitial Pneumonia (DIP) is typically associated with extensive history of smoking although uncommonly, autoimmune, infectious or drug toxicity may also be associated with this entity. The predominant finding is diffuse ground glass attenuation, with reticular opacities and cyst formation being associated findings. Unlike other cystic lung diseases, cysts are typically surrounded by areas of ground glass density. Honeycombing is an uncommon findings. Due to overlapping imaging findings, lung biopsy may be helpful in differentiating respiratory bronchiolitis interstitial lung disease (RB-ILD) from DIP.

Follicular Bronchiolitis

- Follicular bronchiolitis is defined as hyperplasia of bronchus-associated lymphoid tissue, most commonly occurring in the setting of underlying immunodeficiency or autoimmune disease and is different from lip with peribronchial deposition of the lymphocytes with germinal centres unlike lip where there is a diffuse deposition. The predominant finding is multiple small (3 mm) diameter centrilobular ground glass nodules, sometimes associated with peribronchial nodules. Additionally pulmonary cysts or tree-in-bud opacities may also be seen. Treatment of the underlying immunodeficiency or autoimmune disorder typically leads to resolution of the parenchymal findings.

Light-chain deposition disease:

Light chain deposition disease is a rare entity, which rarely affects the lungs, and primarily, almost universally involves the kidneys, leading to proteinuria and renal failure. This entity commonly associated with multiple myeloma or other lymphoproliferative disorders. Pulmonary manifestations results from light chains depositing in the alveolar walls and small airways. Radiographic manifestations include multiple nodules, lymphadenopathy and cyst formation, without a predominant distribution. Severe pulmonary involvement necessitating lung transplantation has been reported.

Amyloid - See above

Upper Lobe Predominant:

Subacute or Chronic Hypersensitivity pneumonitis

- Hypersensitivity pneumonitis (HP) presents in three distinct phases. The acute form often manifests with nonspecific radiologic findings including homogeneous bilateral ground glass densities, sometimes patchy in distribution with a mid to lower lung
predominance. Common differential considerations for the acute form include infection or edema. Subacute HP results from ongoing pneumonitis beyond the acute phase on the order of weeks to months. Ground glass densities remain a common feature with gradual replacement with more well defined small centrilobular nodular opacities. Mosaic attenuation resulting from air-trapping may begin to develop and a small percentage of patients will begin to develop pulmonary cysts, typically randomly distributed. Once radiographic evidence of fibrosis develops the term chronic HP is used. By this stage nodular densities predominate, though scattered areas of ground glass may persist. There is typically persistent air trapping, as well as evidence of fibrosis which may progress to honeycombing. CT findings at this stage are typically upper or mid lung predominance, which allows differentiation from Usual interstitial pneumonia or nonspecific interstitial pneumonia which are lower lung predominant. Pulmonary cysts may also be present at this stage though they are not a dominant feature, and are located remote to areas of honeycombing, and randomly distributed.

Neurofibromatosis Type 1 - See above.

*Mid Lung predominant:*

Pneumocystis jiroveci pneumonia:

- Pneumocystis jiroveci pneumonia (often referred to as PJP or PCP) is the most common opportunistic in patients with AIDS. The principle finding in this infection is perihilar or mid lung zone predominant ground glass opacity, which when combined with septal thickening may also manifest as crazy paving. The distribution of disease may also be upper zone predominant if the patient is receiving aerosolized pentamidine for prophylaxis. Pneumatoceles are are relatively less common manifestation and may occur in up to 30% of cases. The cystic form of the disease is more common in people receiving aerosolized prophylaxis with upper-lobe predominant thin walled cysts (often pneumatoceles) which predispose the patient to the development of pneumothorax. Gallium-67 scan may be helpful in the evaluation of PJP as it is highly sensitive for the disease, though not specific.
Fig. 1: Lymphangioleiomyomatosis in a 31 year-old female. Axial and coronal images demonstrate uniform, diffusely distributed cysts throughout the bilateral lungs with associated interlobular septal thickening. A chest tube can also be seen along the anterior of the right hemithorax which was placed for treatment of a spontaneous pneumothorax. The pleural effusion, also in the right hemithorax was shown to be chylous.

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**Fig. 2:** Lymphangioleiomyomatosis in a 31 year-old female. Axial and coronal images demonstrate uniform, diffusely distributed cysts throughout the bilateral lungs with associated interlobular septal thickening. A chest tube can also be seen along the anterior of the right hemithorax which was placed for treatment of a spontaneous pneumothorax. The pleural effusion, also in the right hemithorax was shown to be chylous.

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Fig. 3: Birt-Hogg-Dube syndrome in a 59 year-old female. Axial and coronal images show few bilateral randomly distributed pulmonary cysts, some of which are larger than is typical in other cystic lung disease. The lentiform and oval shape is characteristic of Birt-Hogg-Dube.

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**Fig. 4:** Birt-Hogg-Dube syndrome in a 59 year-old female. Axial and coronal images show few bilateral randomly distributed pulmonary cysts, some of which are larger than is typical in other cystic lung disease. The lentiform and oval shape is characteristic of Birt-Hogg-Dube.

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Fig. 5: Birt-Hogg-Dube syndrome in a 59 year-old female. Axial CT image of the abdomen showing a heterogeneously enhancing left renal mass which was later characterized as a renal cell carcinoma, chromophobe-type, which patients with Birt-Hogg-Dube are predisposed to developing.

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Fig. 6: Pneumocystis jirovecii pneumonia in a 43 year-old male. Axial and coronal images display diffuse ground glass density throughout the lungs in this patient with known HIV. There are associated apical cysts which can be seen in up to 30 percent of cases.

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Fig. 7: Pneumocystis jirovecii pneumonia in a 43 year-old male. Axial and coronal images display diffuse ground glass density throughout the lungs in this patient with known HIV. There are associated apical cysts which can be seen in up to 30 percent of cases.

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**Fig. 9:** 24 year old woman with recurrent laryngeal and tracheal squamous cell papillomas. CT scan demonstrates multiple cavitating nodules with basal and posterior predilection. This is compatible with parenchymal extension of tracheobronchial papillomatosis.

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**Fig. 10:** 24 year old woman with recurrent laryngeal and tracheal squamous cell papillomas - CT scan demonstrates multiple cavitating nodules with basal and posterior predilection. This is compatible with parenchymal extension of tracheobronchial papillomatosis

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**Fig. 11:** Mucinous Lepidic Variant Adenocarcinoma of the lung in an 80 year-old female. Axial CT images displaying bilateral, left greater than right ground glass opacity with scattered small pulmonary cysts. Additionally there is interlobular septal thickening primarily on the right corresponding to lymphatic spread.

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Fig. 12: Pulmonary interstitial emphysema in a 16 year-old male admitted following stabbing. Patient initially presented with liver and gastric injury repaired following exploratory laparotomy. Axial and coronal CT images show prominent pneumomediastinum with scattered serpiginous lucencies consistent with pulmonary interstitial emphysema.

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Fig. 13: Pulmonary interstitial emphysema in a 16 year-old male admitted following stabbing. Patient initially presented with liver and gastric injury repaired following exploratory laparotomy. Axial and coronal CT images show prominent pneumomediastinum with scattered serpiginous lucencies consistent with pulmonary interstitial emphysema.

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**Fig. 14:** Chronic Pulmonary Coccidioidomycosis in a 49 year-old female. Axial and coronal images show a fluid and air containing cyst in the posterior right lung which had been present for many years. The fluid was a new finding and was concerning for superinfection, a known complication.

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Fig. 15: Chronic Pulmonary Coccidioidomycosis in a 49 year-old female. Axial and coronal images show a fluid and air containing cyst in the posterior right lung which had been present for many years. The fluid was a new finding and was concerning for superinfection, a known complication.

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Fig. 16: Neurofibromatosis in a 54 year-old female. Axial and coronal CT images show a large left sided neurofibroma expanding the neural foramen with bilateral small pulmonary cysts which demonstrate an upper lobe predominance.

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Fig. 17: Neurofibromatosis in a 54 year-old female. Axial and coronal CT images show a large left sided neurofibroma expanding the neural foramen with bilateral small pulmonary cysts which demonstrate an upper lobe predominance.

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**Fig. 18:** Centrilobular Emphysema in a 78 year-old female. Axial and coronal CT images demonstrating apical predominate low density regions with no perceptible wall, distinguishing them from pulmonary cysts. The centrilobular vasculature is preserved (arrow) displaying the "central dot sign".

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**Fig. 19:** Centrilobular Emphysema in a 78 year-old female. Axial and coronal CT images demonstrating apical predominate low density regions with no perceptible wall, distinguishing them from pulmonary cysts. The centrilobular vasculature is preserved (arrow) displaying the "central dot sign".

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Fig. 20: Langerhans Cell Histiocytosis in a 62 year-old male smoker. Axial and coronal images display multiple bilateral pulmonary nodules with scattered parenchymal cysts, in an upper-lung predominant distribution, with the typical sparing of the costophrenic angles.

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Fig. 21: Langerhans Cell Histiocytosis in a 62 year-old male smoker. Axial and coronal images display multiple bilateral pulmonary nodules with scattered parenchymal cysts, in an upper-lung predominant distribution, with the typical sparing of the costophrenic angles.

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Fig. 22: Lymphocytic interstitial pneumonia in a 42 year-old female. Axial and coronal images demonstrate mid and lower lung predominant cysts with associated interstitial thickening and ground glass density.

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Fig. 23: Lymphocytic interstitial pneumonia in a 42 year-old female. Axial and coronal images demonstrate mid and lower lung predominant cysts with associated interstitial thickening and ground glass density.

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Fig. 24: Desquamative interstitial Pneumonia in a 56 year-old male. Axial CT image displaying diffuse ground glass opacities as the predominant with multiple bilateral small pulmonary cysts.

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Fig. 25: Follicular Bronchiolitis in a 56 year-old female. Axial and coronal CT images demonstrate multiple small centrilobular ground glass nodules as the predominant feature with rare, scattered cysts.

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Fig. 26: Follicular Bronchiolitis in a 56 year-old female. Axial and coronal CT images demonstrate multiple small centrilobular ground glass nodules as the predominant feature with rare, scattered cysts.

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**Fig. 27:** Follicular Bronchiolitis in a 56 year-old female. Axial and coronal CT images demonstrate multiple small centrilobular ground glass nodules as the predominant feature with rare, scattered cysts.

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**Fig. 28:** Hypersensitivity pneumonitis in a 44 year-old male. Axial and coronal Ct images show development of fibrotic changes primarily in the upper lungs with multiple small cysts. Fibrosis is consistent with the Chronic phase of the disease.

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Fig. 29: Hypersensitivity pneumonitis in a 44 year-old male. Axial and coronal Ct images show development of fibrotic changes primarily in the upper lungs with multiple small cysts. Fibrosis is consistent with the Chronic phase of the disease.

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Conclusion

Pulmonary cysts may present in a wide variety of conditions, which radiology can play a valuable role in differentiating. Evaluation of the distribution of the cysts, and the presence or absence of associated findings in concert with clinical findings can significantly narrow the differential diagnosis and result in better and more timely patient care.
References


