Case Report, Takla et al.

Interstitial Lung Disease

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Case Presentation

A 76-year-old woman with a history of heart failure presented with 3 days of non-productive cough and increased dyspnea both at rest and with exertion. The patient had been admitted for community-acquired pneumonia 6 months earlier and has required supplemental home oxygen since that illness. She denied prior history of smoking or significant occupational/environmental pulmonary exposures. On clinical examination, she was afebrile with an oxygen saturation of 93\% on room air. Pulmonary auscultation revealed mild respiratory crackles at the lung bases. A high resolution CT of the chest was performed (Figs. 1-3).

\textbf{Figure.} Axial high-resolution computed tomography (CT) images through the chest (\textbf{A and B}) show septal thickening with a subpleural distribution and traction bronchiectasis. Fibrotic changes with honeycombing are noted at the posterior lung bases (\textbf{B}). Coronal reformatted CT image (\textbf{C}) demonstrates the basilar and subpleural distribution of the findings seen above. Mosaic ground glass attenuation is also noted.
Key Clinical Findings

- Increased dyspnea with non-productive cough
- Bibasilar inspiratory crackles

Key Imaging Findings

- Interstitial lung disease with a peripheral basilar distribution

Differential Diagnoses

- Usual interstitial pneumonia
- Non-specific interstitial pneumonia
- Interstitial lung disease associated with connective tissue disease
- Asbestosis

Discussion

Interstitial lung disease (ILD) encompasses a broad category of pulmonary diseases affecting the interstitium of the lung. Progressive dyspnea and cough are common presenting symptoms. The clinical assessment of patients with suspected ILD includes a thorough history, physical examination, and pulmonary function testing. The radiologic evaluation includes chest radiographs and high-resolution CT. The information obtained from clinical and imaging evaluations may often yield a leading diagnosis without the need for conformational surgical lung biopsy.

Progressive dyspnea, non-productive cough, and restrictive pattern on pulmonary function testing raise clinical suspicion for an interstitial process. The radiologic findings may confirm the presence of an interstitial process, and the specific findings and distribution may further narrow the differential diagnosis. In the context of high-resolution CT showing septal thickening with basilar and subpleural distribution, honeycombing, and traction bronchiectasis, the primary differential diagnosis includes usual interstitial pneumonia, interstitial lung disease associated with connective tissue disease, and asbestosis. A final diagnosis is often not possible by high-resolution CT alone. However, correlation of the imaging appearance with patient’s medical history, occupational/exposure history, and progression over time can often yield a final diagnosis.

Usual Interstitial Pneumonia/Idiopathic Pulmonary Fibrosis.

Idiopathic interstitial pneumonias are primarily classified by histopathologic patterns and criteria. These patterns correlate with fairly characteristic findings on high resolution CT imaging of the chest. Among the idiopathic interstitial pneumonias, usual interstitial pneumonia (UIP) is the most common histologic and imaging pattern. Idiopathic pulmonary fibrosis (IPF) is the prototypical and most common entity that corresponds to the morphologic pattern of UIP.

Patients with UIP/IPF are usually 50 years or older at the time of diagnosis. A history of smoking is associated with increased risk of IPF. Patients often do not respond to treatment with corticosteroids, resulting in a relatively poor prognosis. Patients with UIP/IPF have a median survival ranging from 2 to 4 years after initial diagnosis.\(^1,2\)

On chest radiographs, UIP/IPF may appear normal or demonstrate decreased lung volumes with reticular markings in an apicobasilar distribution with more advanced disease. High-resolution CT imaging shows reticular opacities with a basilar and peripheral (subpleural) predominance, honeycombing, and traction bronchiectasis. Ground glass opacities may also be seen.\(^1,2\)

Non-Specific Interstitial Pneumonia.

In the context of the morphologic pattern of UIP, non-specific interstitial pneumonia (NSIP) should be considered. While the characteristic imaging findings of NSIP are somewhat different from UIP, there is a considerable overlap between the two conditions. NSIP shows a similar pattern of subpleural reticular opacities with traction bronchiectasis. However, NSIP tends to lack the apicobasilar gradient and honeycombing. Ground glass opacities are a more prominent feature of NSIP compared to UIP. Clinically, patients with NSIP tend to be slightly younger than patients with UIP (age 40-50).
The most important clinical distinction between UIP and NSIP is prognosis. The histologic pattern of cellular NSIP (predominantly inflammatory without fibrosis) has a survival rate of nearly 100%, and the histologic pattern of fibrotic NSIP is associated with a 5-year survival rate of 45-90%. Compared to UIP, NSIP also shows a favorable response to treatment with corticosteroids and cytotoxic agents.

Connective Tissue Disease Associated Interstitial Lung Disease.

Connective tissue diseases are a group of inflammatory autoimmune-mediated processes that may affect multiple organ systems, including the lung parenchyma and chest cavity. Rheumatoid arthritis and progressive systemic sclerosis are two of the more common connective tissue diseases that may result in interstitial lung disease with reticular opacities in a basilar distribution. The interstitial lung disease that develops with these entities may show a histologic pattern of UIP or NSIP, with UIP being more common in rheumatoid arthritis, and NSIP more common in progressive systemic sclerosis.

The prevalence of interstitial lung disease in rheumatoid arthritis is variable, ranging from 5 to 40%. Interstitial lung disease is usually a late complication of the disease. Other common thoracic manifestations of rheumatoid arthritis include pleural thickening or pleural effusion. Rarely, cavitary necrobiotic rheumatoid nodules may be seen.

Approximately 80% of patients with progressive systemic sclerosis will develop interstitial lung disease, and up to 97% of patients with progressive systemic sclerosis will have esophageal involvement of the disease, leading to esophageal dysmotility. On imaging, the esophageal involvement manifests as a patulous and fluid-filled esophagus. The coexistence of these common manifestations is highly suggestive of interstitial lung disease associated with progressive systemic sclerosis.

Asbestosis.

Asbestos is a non-combustible and durable silicate mineral that has been commercially developed for many purposes. The most common commercial applications include insulation material and brake pads/linings. The two main classifications of asbestos fibers are serpentine and amphiboles. As the name implies, the serpentine fibers are curly and flexible, while the amphiboles are straight, needle-shaped fibers. While both forms may lead to asbestos-related lung disease, the amphiboles are considered more toxic. Exposure to asbestos fibers by inhalation can lead to various manifestations of asbestos-related lung disease, including pleural effusion, pleural plaques (with or without calcification), diffuse pleural thickening, and/or malignant mesothelioma. Patients with asbestos exposure are also at increased risk of primary bronchogenic carcinoma.

Asbestosis is a form of asbestos-related lung disease that leads to interstitial fibrosis. Asbestosis is clinically and histologically similar to idiopathic pulmonary fibrosis. On high-resolution CT imaging, asbestosis will show subpleural septal thickening and traction bronchiectasis in a similar distribution as seen with UIP/IPF; honeycombing is seen in more advanced cases. Concomitant asbestos-related pleural disease may be seen in 75-83% of patients with asbestosis. However, in the absence of pleural abnormalities, asbestosis and idiopathic pulmonary fibrosis cannot be differentiated by imaging alone.
Diagnosis

Usual interstitial pneumonia/idiopathic pulmonary fibrosis

Summary

High-resolution CT is an important component in the evaluation of interstitial lung disease. When used in conjunction with a thorough history, physical examination, and pulmonary function testing, high-resolution CT imaging may eliminate the need for surgical lung biopsy. Knowledge of subtle differences in the imaging of various interstitial lung diseases may help narrow the differential diagnosis. However, there is significant overlap in the imaging appearance of various interstitial lung diseases, and a final diagnosis by imaging alone is not always possible. Rather, the high-resolution CT findings must often be interpreted in the context of patients medical, occupation, and exposure history.

References


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