Connective Tissue Disease-Associated Interstitial Lung Disease

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Interstitial Lung Diseases (ILDs) also known as Diffuse Parenchymal Lung Disease (DPLD), include a large group of lung diseases characterized by various patterns of inflammation and fibrosis on high-resolution CT and in lung biopsy.

ILD is one of the most serious pulmonary complications in Connective Tissue Diseases (CTDs), resulting in significant morbidity and mortality with an overall incidence estimated at 15%.

The spectrum of ILD in CTD (ILD-CTD) includes all of the disease classified in the idiopathic interstitial pneumonias [1], but the prevalence differs between the CTD.

ILD can be secondary to CTD, but can also be caused by the medications used to treat them. Several anti-inflammatory and biologic drugs have been associated with the development of ILD [1].

It is now recommended that all patients with CTD are checked for ILD. Early detection is very important for the initiation of therapy. Progressive exertional dyspnea is the most frequently symptom. However, its presence is not reliable in identifying lung involvement because it may also reflect the increased work of locomotion in individuals with arthritis or myositis. In addition, patients with severe systemic disease may be unable to exercise sufficiently to experience dyspnea, even when significant pulmonary abnormalities are present [2].

There are some risk factors for the development of CTD-ILD. Some of these risk factors may be disease-specific and peculiar to a particular CTD, whereas others are host-specific and attributable to individual patient factors [3].

Some patients may not require therapy, and we do not initiate it with asymptomatic or mild ILD and suggest serial monitoring of symptoms and pulmonary function [4,5]. The management of ILD in patients with subclinical disease remains unclear. It is not known whether treatment will alter the course of the disease in these patients.

When deciding whether a patient with CTD-ILD may benefit from immunosuppressive therapy, we evaluate the following factors: rate of disease progression, severity of disease, underlying CTD, likelihood of response based on radiographic and histopathology patterns, patient age, and ability to comply with therapy and monitoring [5].

We believe that multi-disciplinary assessment of patients with CTD-ILD between rheumatologists and pulmonologists improves its diagnosis and management of this important complication of CTD.

References