Lung Involvement in Scleroderma

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Pulmonary complications are the leading cause of mortality in scleroderma (systemic sclerosis, SSc). Two major pulmonary manifestations of SSc are interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH). ILD occurs in SSc patients with either limited or diffuse cutaneous disease, while PAH usually occurs as a late complication in patients with limited cutaneous SSc [1]. However, in some patients, especially aged 60 years or more at SSc diagnosis, PAH can develop early (within 5 years of the first non-Raynaud manifestation) and can be equally prevalent among patients with either limited or diffuse SSc [2].

SSc-ILD is characterized by excessive deposition of extracellular matrix proteins within the tissue and space around the air sacs leading to progressive scarring of the lung. The fibrosis reduces the efficiency of alveolar gas exchange and may lead to hypoxemia and respiratory failure. The mechanisms leading to SSc-ILD remain unknown. A variety of inflammatory and fibrogenic mediators, such as transforming growth factor (TGF)-β [3], thrombin [4], connective tissue growth factor (CTGF) [5], interleukin-8 [6], fibronectin [7], tumor necrosis factor (TNF)-α [6], and many others have been reported to be involved in pathogenesis of SSc-ILD. The diversity of the potential mediators suggests multiple pathogenic mechanisms of SSc-ILD and requires complex analysis of the changes that occur in each patient during development of lung disease.

SSc-PAH, defined as a mean pulmonary arterial pressure greater than 25 mm Hg, is characterized by increased pulmonary vascular resistance due to remodeling and occlusion of the pulmonary arterioles. The pathogenesis of SSc-PAH involves an obliterative vasculopathy characterized by pulmonary vascular smooth muscle hypertrophy, adventitial fibrosis in the small pulmonary arterioles, and in situ thrombosis [8,9]. In patients with SSc-ILD, PAH can be a consequence of extensive pulmonary fibrosis, where the fibrotic process leads to major reduction in the cross-sectional area of pulmonary vascular bed due to obliteration of alveolar capillaries and narrowing of arterioles [10]. Although the severity of ILD is an indicator of survival for SSc patients, the development of fibrotic parenchymal lesions may stabilize over time; PAH, however, tends to progressively worsen [11]. When PAH and ILD are combined, survival of SSc patients is even further reduced [11]. A better understanding of pathogenic mechanisms leading to PAH and ILD in scleroderma patients is needed to develop an effective targeted therapy. This issue of Rheumatology: Current Research is focused on pathogenesis and clinical features of ILD, PAH, and other less frequent pulmonary complications of SSc.

“When you can’t breathe, nothing else matters” – motto of The American Lung Association.

References


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