Rheumatology : Current Research
ISSN: 2161-1149

Bogatkevich, Rheumatology 2012, S:1
DOI: 10.4172/2161-1149.S1-e001

Editorial Open Access

Lung Involvement in Scleroderma

Galina S Bogatkevich*

Division of Rheumatology, Medical University of South Carolina, USA

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, itends 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

1. Bolster MB, Silver RM (1999) Assessment and management of scleroderma lung disease. Curr Opin Rheumatol 11: 508-513.
2. Hachulla E, Launay D, Mouthon L, Sitbon O, Berezne A, et al. (2009) Is pulmonary arterial hypertension really a late complication of systemic sclerosis? Chest 136: 1211-1219.
3. Ludwicka A, Ohba T, Trojanowska M, Yamakage A, Strange C, et al. (1995) Elevated levels of platelet derived growth factor and transforming growth factor-beta 1 in bronchoalveolar lavage fluid from patients with scleroderma. J Rheumatol 22: 1876-1883.
4. Ohba T, McDonald JK, Silver RM, Strange C, LeRoy EC, et al. (1994) Scleroderma bronchoalveolar lavage fluid contains thrombin, a mediator of human lung fibroblast proliferation via induction of platelet-derived growth factor alpha-receptor. Am J Respir Cell Mol Biol 10: 405-412.
5. Shi-wen X, Pennington D, Holmes A, Leask A, Bradham D, et al. (2000) Autocrine overexpression of CTGF maintains fibrosis: RDA analysis of fibrosis genes in systemic sclerosis. Exp Cell Res 259: 213-224.
6. Bolster MB, Ludwicka A, Sutherland SE, Strange C, Silver RM (1997) Cytokine concentrations in bronchoalveolar lavage fluid of patients with systemic sclerosis. Arthritis Rheum 40: 743-751.
7. Kinsella MB, Smith EA, Miller KS, LeRoy EC, Silver RM (1989) Spontaneous production of fibronectin by alveolar macrophages in patients with scleroderma. Arthritis Rheum 32: 577-583.
8. Chatterjee S (2011) Pulmonary hypertension in systemic sclerosis. Semin Arthritis Rheum 41: 19-37.
9. Kiatchoosakun S, Ungkasekvinai W, Wonvipaporn C, Tatsanavivat P, Foocharoen C, et al. (2007) D-dimer and pulmonary arterial hypertension in systemic sclerosis. J Med Assoc Thai 90: 2024-2029.
10. Denton CP, Cailes JB, Phillips GD, Wells AU, Black CM, et al. (1997) Comparison of Doppler echocardiography and right heart catheterization to assess pulmonary hypertension in systemic sclerosis. Br J Rheumatol 36: 239-243.
11. Mathai SC, Hummers LK, Champion HC, Wigley FM, Zaiman A, et al. (2009) Survival in pulmonary hypertension associated with the scleroderma spectrum of diseases: impact of interstitial lung disease. Arthritis Rheum 60: 569-577.

***Corresponding author:** Galina S Bogatkevich, Division of Rheumatology & Immunology, Medical University of South Carolina, Charleston, SC 29425, USA, Tel: (843) 792-7631; Fax: (843) 792-7121; E-mail: bogatkev@musc.edu

Received February 17, 2012; **Accepted** February 18, 2012; **Published** February 20, 2012

Citation: Bogatkevich GS (2012) Lung Involvement in Scleroderma. Rheumatology S1:e001. doi:10.4172/2161-1149.S1-e001