

## Silicosis and Progressive Massive Fibrosis associated with Systemic Sclerosis related ILD

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Silicosis refers to pulmonary diseases caused by inhalation of free crystalline silica. Silicotic nodules coalesce resulting in progressive massive fibrosis (PMF) in a minority of patients with chronic silicosis. Silicosis is associated with increased mycobacterial/fungal infection, lung cancer, and connective tissue diseases (CTD). Systemic sclerosis (SSc) is found in less than 0.5% of people with silicosis. We present a case of biopsy-proven silicosis and PMF associated with autoantibodies, SSc-related interstitial lung disease (ILD) and small vessel vasculopathy. A 65-year-old man visited the outpatient department with numb fingers, cough with minimal sputum, mild dyspnea on exertion, and febrile sense for a few months. He denied smoking and had worked as a stone cutter for the past 30 years. Serum C-reactive protein and erythrocyte sedimentation rate were mildly increased (1.25 mg/dL, 51 mm/hr). Antinuclear antibody was positive. Scl-70 antibody was over 241 U/mL. Nailfold Capillary Microscopy revealed microangiopathy with scleroderma pattern. Raynaud's scan revealed decreased perfusion in right hand. On the contrast enhanced chest CT showed multiple various-sized nodules and fibrotic consolidations and ground glass opacities in both lungs. Transbronchial lung biopsy was done to confirm silicosis and to exclude infection or malignancy. Histologically, the specimen showed birefringence with polarized crystal deposition in interstitium, fibrotic nodules with collagen deposition and interstitial fibrosis with fibroblastic foci formation. This case shows chronic silicosis with SSc involving lungs and blood vessels. The Erasmus syndrome describes association of SSc following silica exposure. There were a few case reports that had documented the coexistence of silicosis and progressive SSc, and the biopsy-proven cases of silicosis combined with SSc-ILD were not easily found. Hence, We present a case of biopsy-proven silicosis and PMF associated with positive autoantibodies, SSc-related ILD, and small vessel vasculopathy. In case of occupational exposure to silica, we should thoroughly check and follow up not only chest images but also the development of CTD.

