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ERASMUS SYNDROME: ASSOCIATION OF SILICOSIS AND SYSTEMIC SCLEROSIS

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BACKGROUND

The pathogenesis of Systemic Sclerosis (ES) involves genetic and environmental factors. Increasing evidence relates occupational exposure to crystalline silica or solvents in the development of ES. Studies indicate a history of occupational exposure in approximately 75% of male patients with ES.

CASE REPORT

A 49-year-old male patient who had a history of occupational exposure to sandblasting for 8 years was diagnosed with Pulmonary Silicosis in 2002, based on occupational history, dyspnea on medium exertion (Modified Medical Research Council Scale: 3), chronic productive cough, tomography image presenting pulmonary fibrosis and biopsy containing silica particles in polarized light. After 15 years he developed inflammatory polyarthralgia in hands, wrists and elbows, Raynaud's phenomenon, skin thickening, swollen hands, dysphagia for solids and weight loss of 15% in 3 years. Laboratory tests: negative serologies for HIV and hepatitis B and C, slightly elevated erythrocyte sedimentation rate (37, normal range <25), antinuclear antibodies 1/640 (coarse speckled nuclear pattern), positive rheumatoid factor (54, normal range <20), weakly positive anti-CCP (34, normal range <20), anti-RNP positive (174, normal range <20), non-reactive anti-Scl70. New thoracic computed tomography showed again fibrosis, this time exhibiting dilation of pulmonary artery trunk (3,5cm). Spirometry evidenced severe mixed respiratory disorder. Upper digestive endoscopy revealed mild non-erosive distal esophagitis. Esophagography presented contrast retention in the valleculae, reduction of segment gauge in the transition at the middle and upper thirds of the esophagus, the other segments slightly dilated. Radiograph and ultrasound scan of hands and wrists without erosions or signs of arthritis. Nail capillaroscopy showed an advanced stage SD pattern, compatible with systemic sclerosis or overlapping syndromes.

CONCLUSION

Erasmus Syndrome was described in 1957 and consists of an association between exposure to silica and ES development. Compared with other connective tissue disorders, ES associated to silica is the least common. It is indistinguishable from idiopathic ES, but has a high predisposition for pulmonary involvement combined to anti-Scl70 antibody positivity. It is more common in short and intense silica exposures (5-10 years) and survival in these patients is lower than in the idiopathic ES group. This case presents an association between Pulmonary Silicosis and subsequent development of autoimmunity with criteria for ES.