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Occupational silica exposure as a potential trigger of systemic sclerosis: a case report and diagnostic dilemma

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Introduction: Systemic sclerosis (SSc) is an autoimmune connective tissue disease that may progress with multi-organ involvement. Both genetic predisposition and environmental factors are implicated in its pathogenesis. However, their exact roles remain incompletely understood.

Case description: A 59-year-old non-smoking man with a non-contributory family history who has worked as a foundry worker for 35 years, with exposure to silica-containing dust, was admitted to the clinic due to pain and swelling of the fingers, progressive skin tightness, Raynaud's phenomenon, pitted scars on the fingertips, exertional dyspnea and dysphagia. The patient reported symptom onset approximately 16 years earlier, with arrhythmia and conduction disorder requiring pacemaker implantation. One year earlier, the patient was diagnosed with dyspnea at the pulmonological department. Chest computed tomography (CT) revealed numerous enlarged, calcified mediastinal lymph nodes, pleural thickening at the lung apices and multiple, scattered, well-calcified perilymphatic nodules, predominantly in the upper lobes and subpleural regions, along with features of emphysema. Findings were consistent with silicosis given silica exposure. Bronchoscopy and lymph node cytology (BAC) were unremarkable; bronchial secretions and acid-fast bacilli microscopy were negative.

Rheumatological evaluation revealed markedly elevated inflammatory markers, normocytic anaemia, hypoalbuminemia with hypergammaglobulinemia, and an IgG κ protein with an acute-phase oligoclonal pattern. The levels of C3 and C4, as well as tumour, cardiac, and muscle injury markers, were normal. Antinuclear antibody titers were strongly positive (1 : 5,120) with the presence of anti-Scl-70 and anti-Pm-Scl-100 Abs. Urinalysis showed no proteinuria. Nailfold capillaroscopy indicated SSc, corresponding to the late phase according to Cutolo (Figs. 1,2). In addition to silicosis-related findings, CT demonstrated basal ground-glass opacities consistent with SSc-related non-specific interstitial pneumonia (Fig. 3). The patient met the 2013 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for the diagnosis of SSc.

Conclusions: Long-term occupational exposure, including high silicon content, may contribute to the development of SSc. Abnormalities in lung imaging may be both a consequence of SSc and a result of environmental factors. It remains unclear whether the clinical presentation is attributable to chemical exposure (Erasmus' syndrome) or represents pulmonary involvement in the course of overlap syndrome. Treatment decisions, including antifibrotic therapy, require prior evaluation of pulmonary changes.

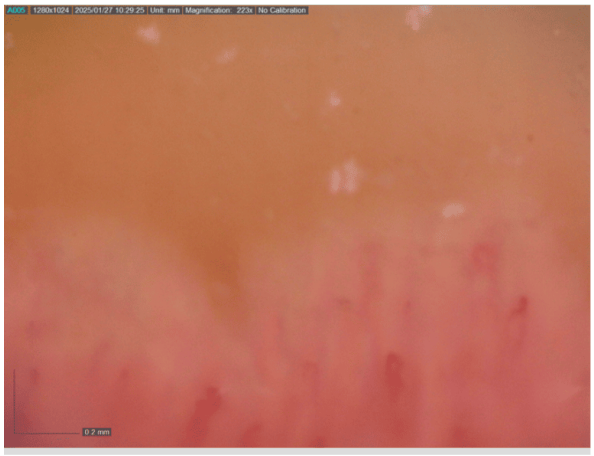

Fig. 1. Nailfold capillaroscopy demonstrating reduced capillary density and disorganised capillary architecture consistent with the late systemic sclerosis pattern.

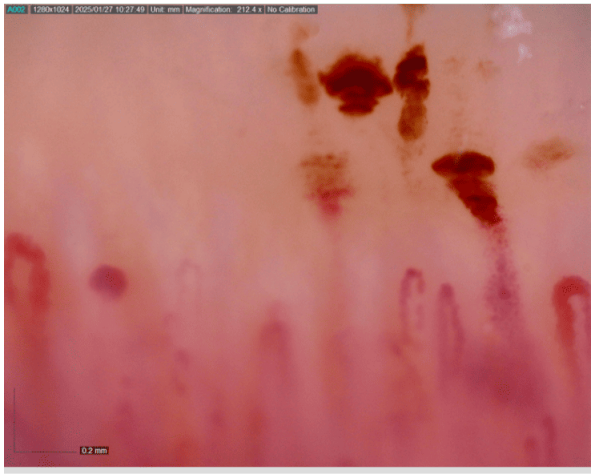

Fig. 2. Nailfold capillaroscopy showing giant capillaries and capillary haemorrhages.

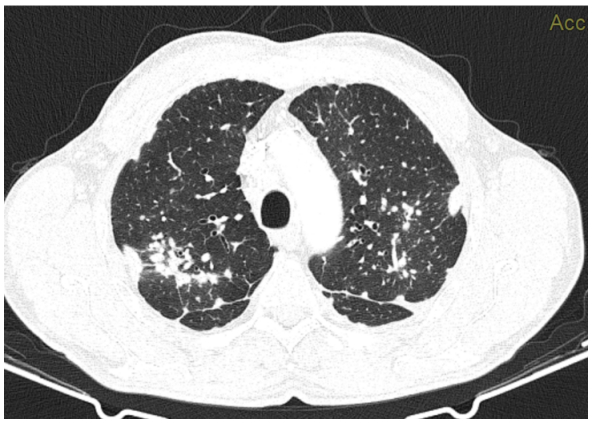

Fig. 3. Axial chest CT showing calcified perilymphatic nodules consistent with silicosis and basal ground-glass opacities suggestive of non-specific interstitial pneumonia.