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Rapidly progressive lymphocytic interstitial pneumonia with plasma cell predominance in Sjögren's disease

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Introduction: Pulmonary involvement is a major systemic manifestation of Sjögren's disease (SjD) and may present as interstitial lung disease (ILD) with various patterns. Lymphocytic interstitial pneumonia (LIP) is an uncommon but characteristic SjD-associated ILD, usually considered slowly progressive. However, LIP may occasionally lead to extensive cystic lung remodeling, severe impairment of gas exchange, and respiratory failure. Data on rapidly progressive, plasma cell-predominant and treatment-refractory LIP in SjD remain limited, and such cases may ultimately require lung transplantation.

Case description: A 47-year-old man with LIP and previously unexplained autoimmune features was subsequently found to have underlying SjD, followed by severe, rapidly progressive ILD. Within two years, serial high-resolution computed tomography (HRCT) scans demonstrated extensive progression to diffuse cystic remodeling with severe parenchymal destruction, affecting approximately 80% of both lungs (Fig. 1). Pulmonary function tests showed moderate restriction (forced vital capacity [FVC]: 54–60% predicted) and severely impaired diffusing capacity (diffusing lung capacity for carbon monoxide [DLCO]: 25–35% predicted, HGB = 14.7 g/dl), consistent with progressive respiratory failure. Clinically, the patient required long-term oxygen therapy and experienced marked exertional desaturation. The disease progressed despite treatment with hydroxychloroquine, cyclophosphamide, dose glucocorticosteroids, azathioprine and rituximab (anti-CD20 monoclonal antibodies). Bronchoscopy was unremarkable, and bronchoalveolar lavage revealed a low lymphocyte fraction. Histopathologic evaluation from CT – guided lung biopsy and mediastinal lymph node sampling excluded malignancy and immunoglobulin G4-related disease and supported the diagnosis of plasma cell-predominant LIP related to SjD. Due to progression and advanced hypoxemic respiratory failure, the patient was referred for lung transplantation evaluation.

Conclusions: This case demonstrates that SjD-associated LIP, although usually slowly progressive, may rarely progress into an aggressive cystic phenotype with profound diffusion defect despite immunosuppressive therapy. It also draws attention to the late diagnosis of SjD in men and confirms that men, although they are less likely to suffer from SjD, may present with a more severe course. In such patients, early recognition of treatment failure and timely referral for lung transplantation are essential to improve prognosis.

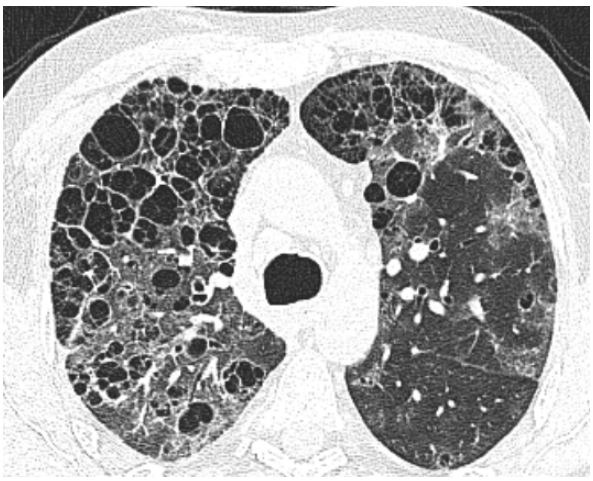

Fig. 1. High-resolution computed tomography showing diffuse cystic lung disease in Sjögren's disease-associated lymphocytic interstitial pneumonia.

Source: Department of Radiology, National Institute of Geriatrics, Rheumatology and Rehabilitation in Warsaw.

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