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A diagnostic and therapeutic challenge of rapidly progressive fibrosing interstitial lung disease: a case report

Marta Tokaj, Karolina Skalska-Ziółkowska

Department of Internal Medicine, Pulmonary Diseases and Allergy, Medical University of Warsaw, Poland

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Introduction: The differentiation between smoking-related interstitial lung diseases (SR-ILD) and interstitial pneumonia with autoimmune features (IPAF) represents a major diagnostic and therapeutic challenge. We present a complex case of a 46-year-old male with a dual phenotype of desquamative interstitial pneumonia (DIP) and smoking-related interstitial fibrosis (SRIF), complicated by high-titer autoantibodies and recurrent pulmonary embolism.

Case description: A 46-year-old male former smoker presented with progressive exertional dyspnea, dry cough, and recurrent fevers. Physical examination revealed digital clubbing and bilateral Velcro-like crackles. Initial high-resolution computed tomography showed diffuse ground-glass opacities (GGO) and lymphadenopathy. Over a three-year observation period, the patient experienced significant functional decline, reaching a restrictive pattern (total lung capacity: 55% predicted) and critical diffusion impairment (diffusion capacity of the lungs for carbon monoxide: 19% predicted). Echocardiography consistently showed no signs of pulmonary hypertension. Surgical lung biopsy confirmed a dual histological pattern of DIP and SRIF. Extensive immunological screening revealed high-titer anti-nuclear antibodies (1 : 2,560) and scleroderma-associated antibodies (anti-Th/To, anti-RP155), yet the patient did not meet the full American College of Rheumatology/European Alliance of Associations for Rheumatology criteria for systemic sclerosis. The clinical course was further exacerbated by recurrent pulmonary embolism and an admission to the Intensive Care Unit (ICU) and mechanical ventilation. Therapeutic strategies evolved from initial glucocorticosteroids to a combined regimen of mycophenolate mofetil, methylprednisolone pulses, and the addition of Nintedanib due to a progressive fibrotic phenotype. This multi-targeted approach was maintained throughout the patient's critical stabilisation following the ICU stay.

Conclusions: This case demonstrates that "overlap" ILD phenotypes can lead to life-threatening respiratory failure requiring intensive care intervention. However, the subsequent clinical improvement and partial radiological regression of GGO highlight the effectiveness of combining potent immunosuppression with antifibrotics. The recovery of a patient requiring intubation underscores the need for aggressive, multi-targeted treatment even in high-complexity, refractory cases.