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Shrinking lung syndrome – a rare manifestation of systemic lupus erythematosus

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Introduction: Systemic lupus erythematosus (SLE) is a heterogeneous autoimmune disease. Shrinking lung syndrome (SLS) is a rare pulmonary complication of SLE and other connective tissue diseases – e.g. Sjögren's disease, characterised by dyspnea, restrictive ventilatory defect, and diaphragmatic dome elevation, mimicking e.g. infections.

Case description: In 2022, a 32-year-old man had a severe left-sided pneumonia with pleural abscess, treated with ciprofloxacin and therapeutic video-assisted thoracoscopy, with good clinical response. Pleural fluid cultures were negative, including for *Mycobacterium tuberculosis*.

In June 2024, the patient developed arthritis, myalgia, and progressive exertional dyspnea with elevated inflammatory markers (C-reactive protein: 85 mg/l, erythrocyte sedimentation rate: 50 mm/h), high-titer antinuclear antibodies (ANA 1 : 1,280, homogenous pattern), and borderline proteinuria (0.51 g/24 h). Glucocorticosteroid (GC) therapy (prednisone 0.5 mg/kg/day) led to a transient clinical improvement.

The patient was admitted to the Department of Connective Tissue Diseases at the National Institute of Geriatrics, Rheumatology and Rehabilitation in October 2024 for suspected SLE, reporting progressive dyspnea, unintentional weight loss (10 kg in 4 months), and myalgia. Diagnosis of SLE was confirmed based on 2019 American College of Rheumatology/European Alliance of Associations for Rheumatology criteria (23 points): ANA > 1 : 80, arthritis, pleural and pericardial effusion, presence of lupus anticoagulant, proteinuria, and anti-dsDNA antibodies. High disease activity was assessed (SELENA-SLEDAI 12).

Due to progressive dyspnea, severe restrictive ventilatory defect (forced vital capacity [FVC]: 28%, total lung capacity [TLC]: 37%), and right pleural effusion with hemidiaphragm elevation, SLS associated with SLE was suspected. The patient received methylprednisolone pulses (in total 5 g i.v.), followed by oral prednisone (40 mg/day) and hydroxychloroquine (400 mg/day). After infection exclusion, treatment with anti-CD20 monoclonal antibody rituximab (2 × 1,000 mg, 14-day interval, first treatment cycle) and mycophenolate mofetil (p.o. 2 g/day) was initiated in March 2025. This led to a significant clinical improvement (SELENA-SLEDAI 0) and pulmonary recovery (FVC 65%, TLC 58%) by August 2025. In November 2025, GCs were discontinued.

Conclusions: This case shows that SLE can have atypical organ manifestations, complicating diagnosis. A rare autoimmune rheumatic complication – SLS, should be considered in progressive dyspnea with restrictive lung deficit. Prompt immunosuppressive treatment, including B-cell depleting therapy, can improve lung function and induce remission.