

A 26-Year-Old Woman With Systemic Lupus Erythematosus Presenting With Orthopnea and Restrictive Lung Impairment



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A 26-year-old white woman diagnosed with systemic lupus erythematosus (SLE) presented with left shoulder pain and a three-pillow orthopnea. Lupus was diagnosed at age 21 years when she developed arthritis, and she has been maintained on prednisone (2.5 mg) and mycophenolate (500 mg bid). In the course of evaluating her new symptoms, imaging revealed a small left pleural effusion with exudative characteristics on a diagnostic thoracentesis, but there was no evidence of infection. Her immunosuppression treatment was increased to 1,000 mg bid of mycophenolate and 20 mg of prednisone. Three months later, she presented to our clinic with worsening six-pillow orthopnea, such that she usually slept with 45° truncal elevation on a recliner at night. She was unable to lie flat. Her dyspnea was worse in the mornings, and she described having to “gasp” for air.

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KEY WORDS: autoimmune lung disease; diaphragmatic dysfunction; pulmonary involvement in SLE; shrinking lung syndrome

Physical Examination Findings

The patient was afebrile, tachypneic (respiratory rate, 26 breaths/min), and anxious, with an oxygen saturation of 94% on room air. Chest auscultation was clear, with slightly diminished breath sounds at the bases. In the supine position, severe abdominal paradox was noted during inspiration.

Diagnostic Studies

Laboratory evaluation showed anemia (hemoglobin, 7.8 g/dL), leukopenia (WBC count, 2,580 k/ μ L), and microscopic hematuria and proteinuria. Inflammatory markers were elevated, with erythrocyte sedimentation rate of 85 mm/h and C-reactive protein level of 6.3 mg/dL. Serology showed a strongly positive antinuclear antibody of 12.7 (normal < 1.5 optical

density ratio) and a markedly elevated anti-dsDNA (365 IU/mL). Antibodies against extractable nuclear antigens and myositis-specific antibodies were negative. Complement levels were normal (C3, 99 mg/dL; C4, 10 mg/dL). The chest radiograph and CT scan of the chest (Figs 1, 2) showed a small left pleural effusion and small lung volumes, with otherwise normal lung parenchyma. Pulmonary function tests (PFTs) (Table 1) revealed a restrictive ventilatory defect, with an FVC of 1.17 L in the sitting position (23% predicted), which decreased by 27% to 0.85 L in the supine position. The maximal inspiratory pressure and maximal expiratory pressure were 57% and 40% predicted, respectively. Evaluation of the diaphragms under fluoroscopy showed reduced downward excursion of both diaphragms, with the right diaphragm being the most impaired.

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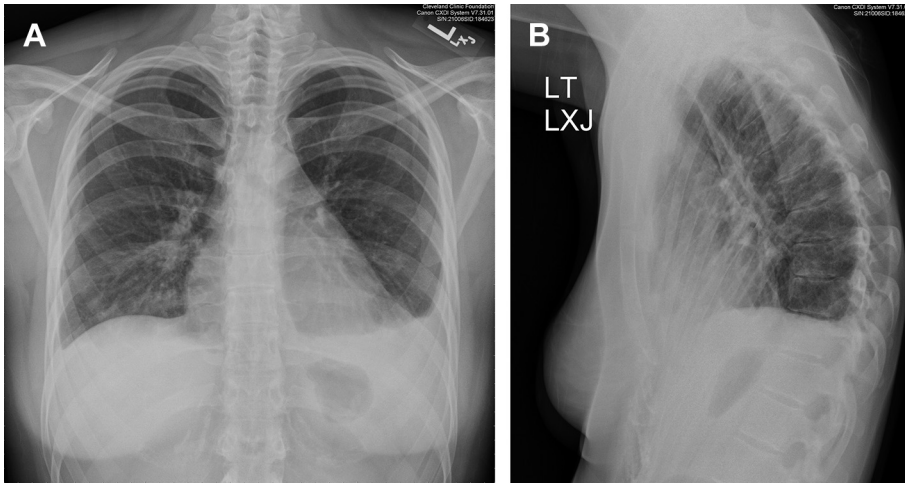


Figure 1 – Chest radiograph, posteroanterior (A) and lateral (B) views, revealing a small left pleural effusion.

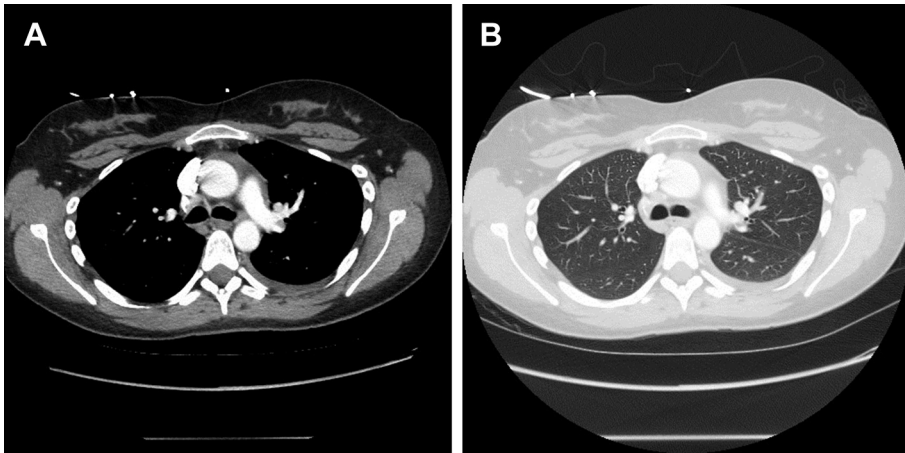


Figure 2 – CT scan of the chest showing a small left pleural effusion (A) with low lung volumes (B).

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