

Clinical Image

Shrinking Lung Syndrome in Primary Sjögren Syndrome

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A 48-year-old woman presented with a three months history of dyspnea, fever, and pain and swelling in her hand and wrist joints. Her past medical history was unremarkable. Detailed clinical history revealed she had dryness of the eyes and mouth for three years. On examination, she had a fever of 38 C, tachypnea (18/minute), reduced breath sounds in the left lower lung zone, and arthritis in her wrists, metacarpophalangeal, and proximal interphalangeal joints. On laboratory, erythrocyte sedimentation rate (68; range, 0-20 mm/hr) and C-reactive protein (108; range, 0-5 mg/L) were increased. Antinuclear antibody (ANA) testing was positive in high titers (1/1600, granular staining) along with a positive anti-Ro/SS-A. The rest of the laboratory tests, including urine analysis, anti-ds-DNA, anti-Sm, and complement were normal. On chest X-ray, there was an elevated hemidiaphragm and loss of volume on the left lung with normal parenchyma (Figure 1a). Computed tomography (CT) showed thinning of the diaphragmatic crura (Figure 1b). Fluoroscopy revealed diaphragmatic dysfunction on the left side (Figure 2). Pulmonary function tests were consistent with a restrictive pattern (FVC: 65%) and carbon monoxide diffusion capacity (DLCO: 57%) was reduced. Schirmer's test was positive (<5 mm in 5 minutes for both eyes). Further minor salivary gland biopsy was showing diffuse lymphocytic infiltrations with a focus score of 4. The patient was diagnosed as primary Sjögren's syndrome (pSS) and shrinking lung syndrome (SLS) and started on azathioprine (AZA; 2mg/kg/day) and prednisone (0.5 mg/kg/day). Significant clinical improvement on her dyspnea and arthritis observed within a few days of the treatment and complete radiologic improvement achieved in a month (Figure 1c, d).

SLS is a rare pulmonary manifestation of inflammatory rheumatic diseases, which is mostly reported in systemic lupus erythematosus. Dyspnea, pleuritic chest pain, progressive decrease in lung volumes, the elevation of the diaphragm, and the absence of significant parenchymal and pleural disease on thorax CT are the main findings of SLS. The underlying pathology of SLS is unclear, but researchers suggested the following as possible mechanisms: 1) myopathy or myositis affecting the diaphragm or intercostal muscles, 2) diaphragmatic dysfunction secondary to pleural adhesions, 3) phrenic neuropathy, and 4) pleuritic pain-related diaphragmatic dysfunction through reflex inhibition of diaphragmatic activation (1). Neurological involvement is a well-known complication of pSS, and a wide variety of neurological symptoms could be seen in the course of the disease. In a small study, performed in pSS patients with neuropathy, reported that about 5% of these patients had phrenic nerve palsy (2). Therefore, we may hypothesize that phrenic neuropathy, operating alone or in combination with the ones mentioned above, could explain the SLS in the current case. Currently, there is no established treatment for pSS associated SLS but, similar to our report, corticosteroids in combination with immunosuppressives (AZA, cyclophosphamide or rituximab) have been suggested in the case series. In conclusion, SLS should be considered in the differential diagnosis of pSS with dyspnea, or pleuritic chest pain, and patients should be started immunosuppressants as they provide a rapid and effective response on the condition.

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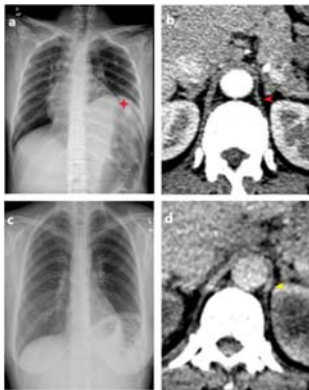


FIG. 1. a) Elevated hemidiaphragm and loss of volume on the left lung with normal bilateral lung parenchyma on chest X-ray. b) Thorax axial CT image is showing the thinning of the diaphragmatic crura (red arrowhead) at the time of presentation. c) The improvement in the lung volume and diaphragm and d) diaphragmatic crura (yellow arrowhead) after immunosuppressive treatment.

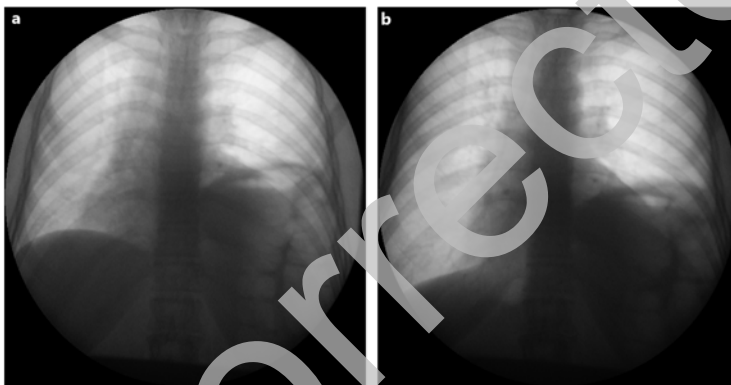


FIG. 2. Fluoroscopy imaging of the lung is showing reduced left diaphragmatic excursion. Images are obtained in a) Expiration and b) Inspiration.