Images in Rheumatology

Cystic Lung Disease in Sjögren's Syndrome

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We examined a 40-year-old woman with a 4 year history of Sjögren's syndrome (SS) according to the validated European criteria[1], who presented with increasing dyspnea and cough over the last 3 months, worsening of fatigue, and bilateral parotid enlargement. She had lost 8 kg of weight over the last 6 months. Laboratory studies revealed negative antinuclear antibody testing on murine liver and a titer of 1:20 on HEp-2 cells. Anti Ro/SSA precipitins were present, whereas La/SSB and dsDNA (*Crithidia luciliae*) antibodies were absent. Western immunoblotting recognized 52 kDa Ro/SSA. A polyclonal gammopathy was present; the gammaglobulin value was 4.05 g/dl and rheumatoid factor was 34 IU/ml (normal 0–24).

A lip biopsy specimen showed confluent lymphocytic infiltrates of salivary glands. Chest radiographs showed bibasilar reticulonodular opacities and computer tomography (CT) scans showed thin-walled cysts and interstitial abnormalities (Figure 1). A restrictive pattern was seen on lung function testing (low FVC, low FEV1, and normal FEV1:FVC). The total lung capacity was slightly diminished (85% of predictive value) with an increased residual volume (120% of predictive value) suggesting air trapping. The carbon monoxide diffusing capacity was almost unchanged. An open lung biopsy showed interstitial and peribronchiolar lymphoplasmacytic infiltrates, emphysema, and bullae (Figure 2). Polymerase chain reaction analysis of immunoglobulin heavy chain and T cell receptor gene rearrangement did not show predominant clonal salivary gland or lung lymphocyte populations.

Several infectious, neoplastic, and inflammatory diseases can cause pulmonary cysts[2]. A variety of lung lymphoid disorders, including lymphocytic interstitial pneumonia (LIP), often accompany SS[3], and LIP is one of the causes of lung cysts[4]. Rarely, acquired lung cystic disease may be seen in the clinical context of other inflammatory autoimmune disorders.

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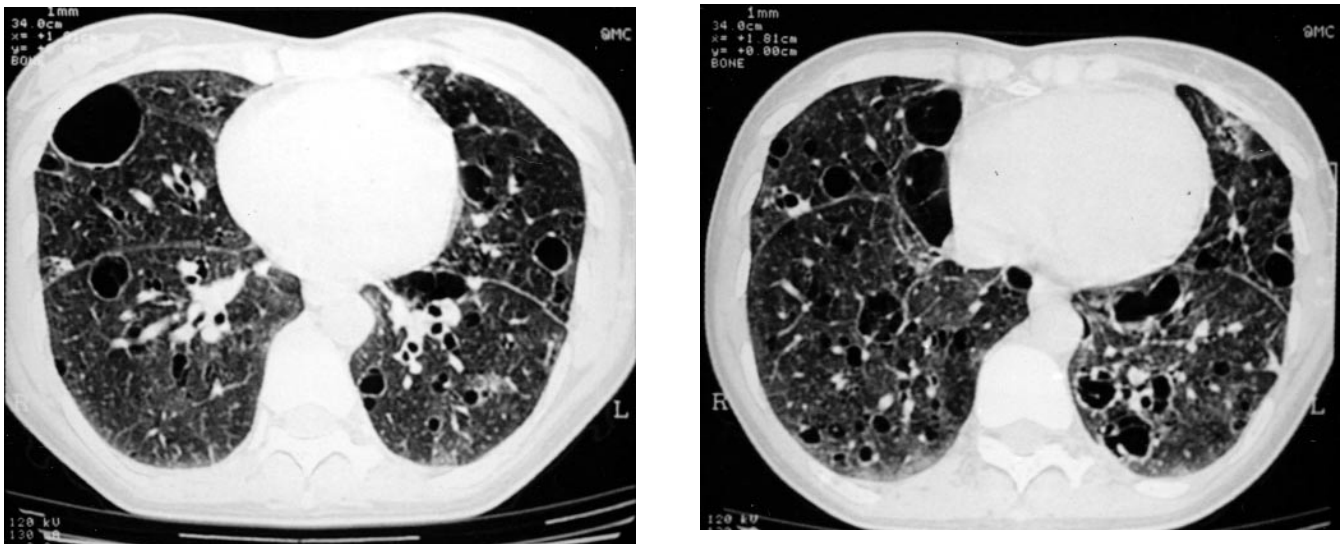

Figure 1. High resolution (1 mm collimation) CT of the thorax obtained without contrast material shows thin walled cysts (0.5–3.2 cm diameter), ground glass opacities, and small nodules in both lower, right middle, and left upper lobes.

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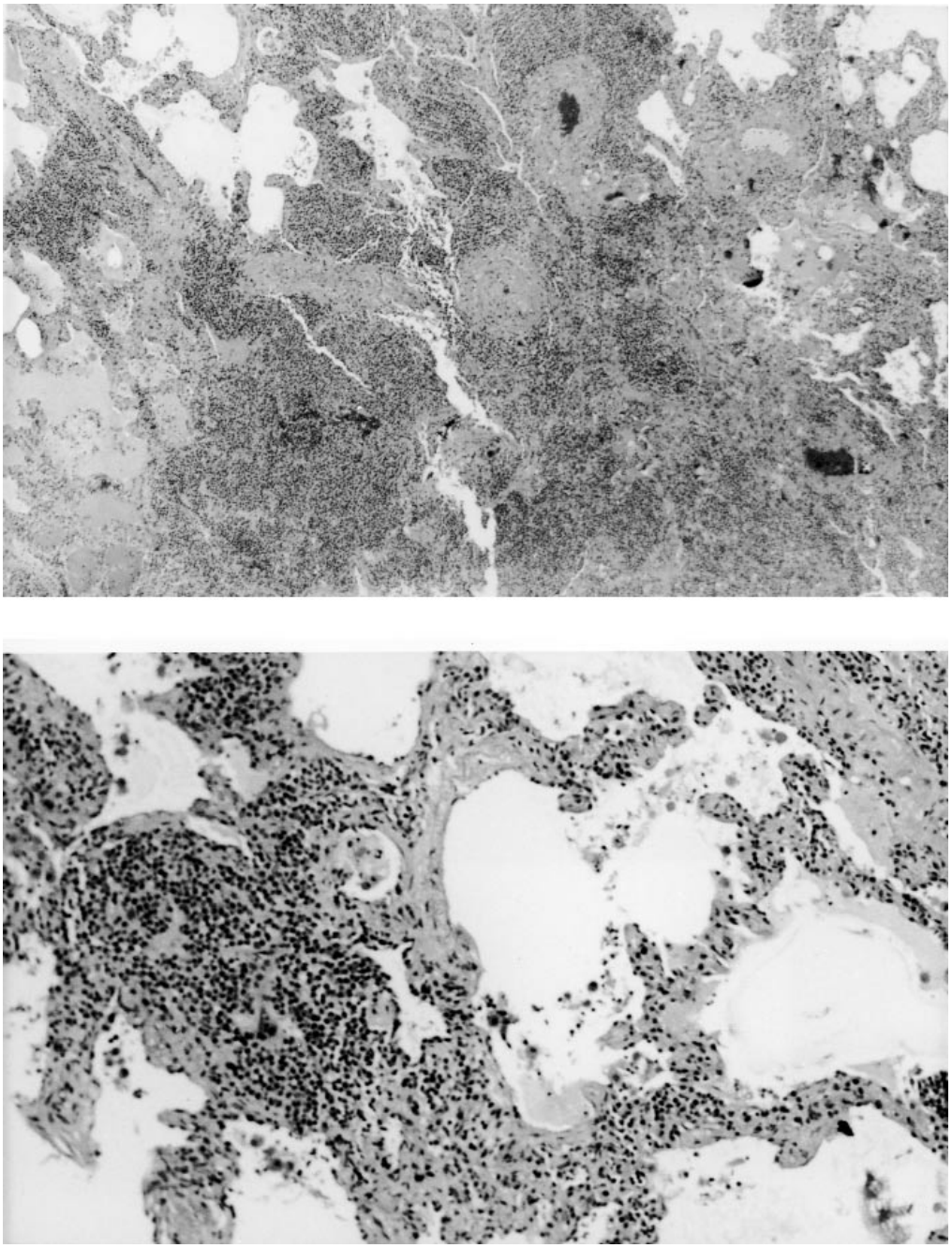

Figure 2. A. Histopathology of lymphocytic interstitial pneumonia. Dense interstitial infiltrate of predominantly mature lymphocytes, visible as well in the alveolar areas adjacent to cysts. B. Higher power shows the interstitial lymphocytic infiltrates (H&E, ×40 in A, ×100 in B).

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