Lung cavities in Sjögren’s syndrome

Clinical images

A 57-year-old woman presented with history of progressive dyspnea, fatigue and arthralgia for 5 years. A computed tomography of chest demonstrated numerous thin-walled pulmonary cysts and patchy bilateral ground-glass opacities (Figure 1A and B). Work-up with infectious laboratory include HIV, serologic testing for autoimmune antibodies and vasculitides, yielded negative results except anti-nuclear antibody, anti-Ro/SSA and anti-La/SSB antigen-antibody. Lymphoid interstitial pneumonia (LIP) secondary to Sjögren’s syndrome was diagnosed by open lung biopsy. Patient was subsequently started on prednisone with clinical and radiologic improvement.

LIP is a rare form of interstitial lung disease which is characterized histopathologically by infiltration of the interstitium and alveolar spaces of the lung by lymphocytes, plasma cells and other lymphoreticular elements. Etiology of LIP is unknown, likely manifestations of an underlying infection or involves derangements in the immune system, Sjögren’s syndrome is associated with one-fourth of reported cases of LIP. Radiographic finding of LIP has a varied appearance. It can present as reticular opacities or nodular process. As the infiltrative process continues, alveolar spaces become involved, and a mixed pattern of ground glass and consolidative opacities appears. Cystic changes are seen in endstage disease as in this case.

Differential diagnosis of diffuse lung cystic lesions including pulmonary lymphangioleiomyomatosis, pulmonary Langerhans’ cell histiocytosis, lymphoid interstitial pneumonia, Birt–Hogg–Dubé syndrome, amyloidosis, light chain deposition disease, honeycomb lung associated with advanced fibrosis. This clinical

Figure 1. (A, B) CT images of the lung demonstrating groundglass opacities and scattered pulmonary cysts.
image will raise awareness of LIP and guide us in differential diagnosis of cystic lung diseases.

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References