Case report - Pulmonary

Systemic lupus erythematosus with multiple lung cysts

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Received 2 December 2008; received in revised form 23 January 2009; accepted 23 February 2009

Abstract

We report a rare case of recurrent pneumothorax complicated by systemic lupus erythematosus (SLE) with multiple lung cysts. The patient was a 53-year-old woman without a history of smoking. At the age of 29, she was diagnosed with SLE. A computed tomography (CT) scan of the chest showed multiple, bilateral lung cysts. She had recurrent episodes of spontaneous pneumothorax on the same side within a period of two months. Although pulmonary involvement is common in SLE patients, cystic lung disease associated with SLE is extremely rare. We report a case of recurrent, spontaneous pneumothorax complicated by SLE with multiple lung cysts in a 53-year-old woman.

Keywords: Cystic lung disease; Systemic lupus erythematosus; Pneumothorax

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic systemic disease that can involve multiple organs [1], and a number of pulmonary complications have been also reported in patients with SLE [2]. Although pulmonary involvement is not uncommon in SLE patients, cystic lung disease associated with SLE is extremely rare. We report a case of recurrent, spontaneous pneumothorax complicated by SLE with multiple lung cysts in a 53-year-old woman.

2. Case report

The patient was a 53-year-old woman without a history of smoking. At the age of 29, she was diagnosed with SLE at another hospital according to the 1982 American Rheumatism Association diagnostic criteria for SLE [1]. She had been treated with low-dose hydrocortisone and cyclophosphamide, after which she had complete relief from symptoms. At the age of 42, a chest roentgenogram showed an abnormal shadow, and she was referred to our hospital. Because a computed tomography (CT) scan of the chest showed multiple, bilateral lung cysts (Fig. 1), video-assisted thoracic surgery (VATS) biopsy was performed to obtain a definitive diagnosis. During the operation, pleural effusion and pleural thickening were not observed in the right thorax, although multiple, small bullae were found on the right visceral pleura. Three biopsy specimens were obtained.

Bacteriological tests for the resected specimens were negative. Histological examination showed that cystic spaces were empty, without cellular or fluid contents (Fig. 2a). A slight proliferation of smooth muscle cells was found at the edges of the cysts (Fig. 2b, c), but the cells were negative for HMB-45 by immunohistochemistry. Partial recanalized thrombosis was also seen (Fig. 2d), but there was no evidence for granuloma formation, inflammatory cell infiltration of the interstitium, fibrotic lesions, Langerhans cells, eosinophils or reactive mesothelial cells. From the biopsy specimens, cystic abnormalities of the lung, including Langerhans cell histiocytosis, interstitial pneumonia, infection and lymphangioleiomyomatosis, and different types of airway disease, such as bronchiectasis and bronchiolitis obliterans, were all excluded. A definitive pathogenesis for the formation of multiple lung cysts could not be determined from the histology of the biopsy specimen.

We carefully followed the patient after the VATS biopsy. She was maintained with low-dose hydrocortisone and cyclophosphamide and the symptoms were suppressed. The number and sizes of the lung cysts also did not change for 11 years. However, at the age of 53, she presented to the emergency department with a chief complaint of shortness of breath. A chest roentgenogram showed a collapsed lung on the right side. After a chest tube was inserted, the lung promptly expanded. Subsequently, the patient had recurrent episodes of spontaneous pneumothorax on four occasions, on the same side, within a period of two months. Each time, closed thoracotomy was performed because she refused to undergo a curative operation with repeated lung biopsy. On the last occasion, a chest drainage tube was inserted, which showed the co-existence of an empyema. The patient was treated with antibiotics and we successfully performed pleurodesis using tetracycline. She has had no recurrence of pneumothorax since that time.

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Cystic abnormalities of the lung include Langerhans cell histiocytosis, lymphangioleiomyomatosis, emphysema or different types of airway disease, such as bronchiectasis and bronchiolectasis. In addition, honeycomb cysts in pneumoconiosis and carcinoid pneumonia are also included in cystic lung disease. Cystic lung disease associated with SLE is extremely rare and, to date, very few cases have been reported [5, 6].

The pathogenesis of cystic lung disease is unclear. Diffuse interstitial fibrosis, an uncommon finding in SLE, might predispose to pneumothorax due to rupture of cysts or blebs, as suggested by Richards et al. [5]. However, in our case, interstitial fibrosis was not found in the VATS biopsy specimen. Others have suggested that spontaneous pneumothorax may be associated with inflammatory reactions in the lung, and that bullae formation may be a check-valve mechanism caused by inflammatory reactions in the bronchiolar wall [3]. Although the actual pathological changes leading to multiple lung cysts are not entirely clear, it is generally considered that any severe inflammatory process of the lungs can present a potential cystic change. In our case, no inflammatory change or fibrosis was found and we were unable to give any clear explanation for the cystic lung change from the VATS biopsy specimen. However, it possibly arose because the VATS biopsy was performed after embarking on immunosuppressive therapy. Although there was not a clear relationship between SLE and lung cysts in our case, SLE can accompany multiple lung cysts. While not the most common etiologic entity, SLE should be a diagnostic consideration in those patients with pulmonary lung cysts, and one should distinguish the cystic lesions associated with SLE from other cystic lung diseases.

The prognosis for SLE with multiple lung cysts is unknown. In other reports, many patients had synchronous pneumothorax with active SLE [5, 6], but in our case, the cystic lesion had caused recurrent pneumothorax a long time after SLE was diagnosed. Therefore, we should carefully follow the clinical course over the entire life of the patient and initiate appropriate therapeutic intervention as required.

References