Case report - Pulmonary

Pulmonary Langerhans’ cell histiocytosis presented with recurrent pneumothorax

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Abstract

Pulmonary Langerhans’ cell histiocytosis is a relatively unusual, interstitial lung disease. Several organ systems may be involved, including the lung, bone, liver, lymph nodes and brain. It is known to occur preferentially in heavy smokers, and the cases such as ours presenting pneumothorax as the major clinical manifestation are rare.

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1. Case

A 29-year-old male was admitted to our hospital due to chest pain. Chest X-ray showed a left-sided pneumothorax and high resolution computed tomography (HRCT) showed numerous, variably sized, thin-walled nodules and cysts in both lung fields (Fig. 1). The patient did not have any past history of diabetes, hypertension or tuberculosis. However, he had a 10-year history of smoking approximately 1 pack per day. He had quit smoking two years prior to admission, although he was still exposed to cigarette smoke continuously in the office because of smoking by co-workers. Two years ago, he had been admitted to our hospital for exertional dyspnea. He underwent thoracoscopic lung biopsy and the diagnosis was eosinophilic granuloma. The patient improved without steroids or other special treatment.

During follow-up, right-sided pneumothorax was developed but was improved by tube thoracostomy. However, it recurred 20 days later and thoracotomy was performed. Bullae were not detected in the apex of the right upper lobe. However, in the area approximately 3 cm below the apex of the superior segment of the right lower lobe, one bulla, approximately 0.5 cm in size, was detected and bullectomy was performed.

In the right side, pneumothorax did not recur until now. However, pneumothorax recurred three times in the left side. We recommended an operation but the patient refused. On the fourth recurrence, this time surgery was decided.

Similarly to the right side, thoracotomy was performed and bullae were not detected in the apex of the left upper lobe or in the apical portion of the left lower lobe. Two bullae, approximately 1 cm in size, were detected in the posterior segment of the left upper lobe and in the lower part of the superior segment of the left lower lobe, and bullectomy was performed.

In histological examination, diffuse infiltration of Langerhans’ cells mixed with eosinophils was detected. Immuno-
Their cavitary character may lead to airway wall damage. Airways and ultimately transforming into fibrotic nodules. LCH begins with granulomas growing along the small airways. Langerhans' cells contain variable numbers of lymphocytes, plasma cells, eosinophils, alveolar macrophages and fibroblasts. Pulmonary LCH begins with granulomas growing along the small airways and ultimately transforming into fibrotic nodules. Their cavitary character may lead to airway wall damage. Pneumothorax results from a rupture of subpleural cystic lesions or from pleura destruction by adjacent formed granulomas [5], and has the characteristic of recurrence. In most cases of primary pneumothorax, bullae are formed primarily in the apex of the upper lobes. However, in our case, bullae were not detected neither in the upper lobes nor the superior segment of the lower lobes, but the difference of detection was shown in the slightly lower area, which supported to speculation that the bullae were formed by different mechanisms from the majority of primary pneumothorax.

In the chest X-ray, micronodular or reticulonodular and interstitial infiltration findings are most frequently shown, the infiltration appears as bilateral and symmetric in most cases. Primarily, it involves the upper or middle lobe, and with lesion progression, nodular lesions decrease gradually, cystic changes become prominent, and cysts approximately 2 cm in size may be developed. HRCT is a very useful and sensitive test for the diagnosis of pulmonary LCH. The most common abnormalities on HRCT are cysts and nodules. In the initial period of the lesion, nodular changes are detected primarily, while cystic changes or fibrosis are detected as major lesions in the late period. In most cases, together with small peribronchial nodular opacities, diffuse and irregular shaped cystic spaces are detected in the middle or upper lobes, and the finding of such lesions strongly suggests pulmonary LCH.

The diagnosis of LCH can be made by the detection of Langerhans’ cells under light microscope, positive for S-100 staining, positive for CD1 antigen, or the cases showing intracytoplasmic Birbeck granules under electron microscope [6].

About half the patients have a favorable clinical outcome either spontaneously or with corticosteroid therapy, or partial or complete resolution of radiographic abnormalities. Approximately one third of cases show acute deterioration and progression to respiratory insufficiency [5].

Smoking cessation is essential for the treatment and chemotherapeutic agents can be used in patients with progressive disease that is unresponsive to steroid or with multiorgan involvement [4].

We report our experience of a rare case of pulmonary LCH that has been reported to cause recurrent pneumothorax.

References