Prolonged Air Leakage Caused by Mesenchymal Cystic Hamartoma of the Lung

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A 16-year-old girl was transferred to the department of thoracic and cardiovascular surgery because of a spontaneous pneumothorax with prolonged air leakage. Chest computed tomography demonstrated a cystic lesion measuring 2×3 cm and involving the left upper lobe. Left upper lobectomy was performed via video-assisted thoracoscopic surgery. A pathologic examination of the specimen revealed a mesenchymal cystic hamartoma. Despite the rarity of pulmonary mesenchymal cystic hamartoma, it should be considered a potential cause of pneumothorax for patients with a large pulmonary cyst. Further, surgical resection must be considered because serious complications such as hemothorax, hemoptysis, and malignant transformation have been reported.

Key words: 1. Pneumothorax 2. Hamartoma 3. Lobectomy (lung)

Case Report

A 16-year-old female was transferred to the department of thoracic and cardiovascular surgery due to a spontaneous pneumothorax with prolonged air leakage. The patient had mild dyspnea and dry cough for two weeks. Closed thoracostomy was performed at the other hospital and was treated for 3 days. However, the chest X-ray revealed no improvement in the symptoms and lung expansion. Therefore, she was transferred to our hospital for surgery. She had no specific medical history and was a non-smoker. The initial vital signs were stable. A physical examination revealed that the breathing sounds were decreased in the left lung field. The chest X-ray showed a collapsed left lung with a large cyst (Fig. 1A). Even after closed thoracostomy, a large amount of pneumothorax and collapsed lung were seen. When she was admitted in our hospital, we first applied a low-pressure suction pump and observed a little improvement in the lung expansion (Fig. 1B). Over a period of five days, the chest X-ray showed a little day-by-day improvement, and the patient did not complain about the symptoms any more. However, she still had air leakage through the chest tube. Computed tomography (CT) of the chest demonstrated pulmonary edema caused by re-expansion and a large cystic lesion (measuring 2.0×3.0 cm) in the left upper lobe (Fig. 2). It seemed to be connected between the cyst and the bronchus, but on the bronchoscopy, the cyst was observed to be a solitary lesion. Therefore, we decided to surgically control the air leak and performed video-assisted thoracoscopic surgery. A 4-cm working window and two 10-mm ports were made. We could define the site of the air leakage site on the cyst, as observed
Fig. 1. Simple chest X-rays. (A) Collapsed left lung, right-shifted mediastinum, and large pulmonary cyst (black arrow) were observed on the first chest X-ray. (B) The left lung expanded more after the application of low-pressure suction. However, residual pneumothorax on the left apical area was still observed.

Fig. 2. (A – D) Computed tomography revealed a solitary cystic lesion in the left upper lobe of the lung. It seemed to be connected to the left upper lobe bronchus, but the bronchoscopy revealed no communication between the cyst and the bronchus.
on the chest CT. The cyst was close to the superior pulmonary vein. We attempted to close the cyst by manual sutures after opening the roof of the cyst because of the difficulty of wedge resection. However, several vessels were close to the cyst, there were multiple air leakage sites, and the densely adhered multiple nodules made primary closure impossible. Therefore, the left upper lobectomy of the lung was performed. A pathological examination of the specimen revealed a mesenchymal cystic hamartoma (MCH). On the macroscopic examination, the cut surface showed a cystic lesion (size: 2.0×1.8 cm) and the cystic lesion was 1.0 cm away from the bronchial resection margin (Fig. 3A). The microscopic examination revealed that the cyst was lined by respiratory epithelium and the wall was composed of oval-to-spindle-shaped cells suspected to be mesenchymal cells (Fig. 3B, C). A small intramural nodule was observed focally. The diagnosis was based in features such as multiple cysts, prominent cystic changes in the lung parenchyma, mesenchymal cell wall, and respiratory epithelium. The patient exhibited good progress after surgery. The chest tube was removed on postoperative day 6, and the patient was discharged on postoperative day 7 without any complications.

**Discussion**

The term ‘hamartoma’ means a benign, focal mass that is composed of tissue normally found at that site, but growing in a disorganized manner. Therefore, MCH of the lung implies a benign mass composed of mesenchymal lung tissue and that this hamartoma has cystic components [1]. MCH is a very rare tumor differentiated from primitive mesenchymal cells and is thought to be a low-grade sarcoma of the lung.
Prolonged Air Leakage by Hamartoma

[1-3]. Only 17 cases of MCH had been reported worldwide up to 2011. MCH of the lung was first described in 1986 [2]. MCH can be characterized by the nodules of primitive mesenchymal cells that slowly increase in size and then become cystic lesions when they are larger than 1 cm. By primitive mesenchymal cell proliferation, bronchioles or alveolar ducts lose their own structural stability and this can cause a nodule to transform into a cyst [3]. Cysts consist of two layers; one is the inner layer, which is thin and lined with respiratory epithelium, and the other is the cambium layer composed of primitive mesenchymal cells beneath the epithelium [2,4,5]. After the cysts are developed, new nodules appear continuously [3]. MCH has to be distinguished from other lung diseases, such as pleuropulmonary blastoma, cystic adenomatoid malformation, lymphangioleiomyomatosis, bronchopulmonary sequestration, bronchiectasis, and metastasis of endometrial stromal sarcoma, and from other carcinomas [5]. However, the cyst or solitary mass lesion of the lung on the chest X-rays are difficult to define before surgical excision. Some clinical and radiological features can help to diagnose the MCH of the lung from other conditions: multiple bilateral lesions, a combination of nodules and cysts, absence of fever, absence of weight loss, and absence of diffuse lung disease [5,6]. Usually, MCH has good prognosis, but rarely, it can transform into a malignant tumor [7,8]. Further, MCH can be accompanied with various symptoms, such as hemoptysis, hemothorax, pneumothorax, dyspnea, croup, tachycardia, and thoracic pain [4]. Hemorrhage from the systemic arteries into the cysts causes hemoptysis or hemothorax. A rupture of sub-pleural cysts can cause pneumothorax or hemothorax [7]. This implies that in spite of the rarity and good prognosis of MCH of the lung, it should be considered a cause of pneumothorax for a patient with a large pulmonary cyst. Further, when the patient is thought to have MCH of the lung, surgical resection must be considered because it can be accompanied with fatal complications such as hemoptysis and hemothorax or can transform into a malignant lesion. In this case, the patient showed pneumothorax as the complication of MCH. She had prolonged air leakage but did not show any other complication such as hemothorax or hemoptysis. Further, she had prolonged air leakage, and surgical treatment was the meaningful choice because of the chances of fatal complications and a malignant transformation of MCH.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

References

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