Posterior mediastinal neurilemmoma accompanied by intrapulmonary sequestration in the left lower lobe

A case report

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Abstract

Rationale: The presence of a mediastinal neurilemmoma accompanied by intrapulmonary sequestration is a rare occurrence. The clinical symptoms of a neurilemmoma depend on the site of the tumor. Diagnosis of pulmonary sequestration mainly depends on the presence of aberrant feeding arteries.

Patient concerns: A 78-year-old woman was admitted to our hospital with a mediastinal space-occupying lesion of 50 years. Computed tomography and magnetic resonance imaging showed 2 roundish low-density shadows in the left posterior mediastinum.

Diagnosis: The pathological findings of the upper cystic mass support the diagnosis of neurilemmoma. A branch of aorta was found supplying blood to the lower mass; it was considered a pulmonary sequestration.

Interventions: Left-sided thoracotomy was planned to remove the chest space-occupying lesions. Two masses were completely removed. Severe adhesion between the left lower lobe and the diaphragm was successfully separated, the aberrant feeding vessel was properly managed, and the lower lobe was resected completely.

Outcomes: The patient experienced remission of symptoms, had no significant postoperative complications, and was discharged from the hospital.

Lessons: Special attention should be paid to neurological involvement of the neurilemmoma and the fragile feeding arteries of the intrapulmonary sequestration. Early diagnosis and treatment are important in such cases.

Abbreviation: CT = computed tomography.

Keywords: neurilemmoma, pulmonary sequestration, thoracotomy

1. Introduction

Neurilemmoma, also known as neurilemmoma or schwannoma, is a common benign neurogenic tumor originating from Schwann cells in the peripheral epithelium. Neurilemmoma accounts for approximately 29% of intraspinal tumors, often presents as a single tumor, and is most commonly found in patients aged 20 to 50 years. The clinical symptoms of a neurilemmoma depend on the site of the tumor. Fewer than 9% of neurilemmomas occur in the mediastinum.[1]

Pulmonary sequestration is a rare congenital bronchopulmonary malformation of the foregut characterized by cystic, nonfunctioning lung tissue without normal communication with the bronchial tree.[2,3] Pulmonary sequestration has 2 types: intrapulmonary and extrapulmonary sequestration. Pulmonary sequestration accounts for approximately 0.15% to 6.40% of all congenital dysplasias of the lungs.[4] The pathological basis of this malformation is that a portion of lung tissue developed into a separate lesion during the embryonic period. The diseased lung tissue has an aberrant arterial supply but no respiratory function.[5] Pulmonary sequestrations have been located in the left lower lobe (71.53%) and right lower lobe (25.97%).[5] Diagnosis of pulmonary sequestration mainly depends on the presence of aberrant feeding arteries.
Herein, we report a case with a mediastinal neurilemmoma accompanied by pulmonary sequestration in the left lower lobe. Two large cystic masses were found occupying most of the left thoracic cavity. The clinical features of the patient were recorded and the management and outcome are discussed below.

2. Case presentation

This study was approved by the Ethics Committee of Chinese-Japan Union Hospital of Jilin University. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from the individual participant included in the study.

A 78-year-old woman presented with a history of a mediastinal space-occupying lesion of 50 years. Chest tightness and shortness of breath had increased for 2 weeks. The preoperative plain computed tomography (CT) and contrast-enhanced magnetic resonance imaging scan of the chest showed 2 round-like low-density shadows next to the left posterior mediastinal spine (Fig. 1). The smaller lesion (∼95.6 × 87.8 × 65.7 mm) was located next to the left side of the spine between the lower edge of the T8 and T11 vertebral body. The CT value of the small mass was about 16.0 HU, the density almost uniform, and strip calcification was observed. The larger shadow (∼116.1 × 104.0 × 123.5 mm) was located at the T1 to T8 vertebral levels.

Figure 1. Contrast-enhanced MRI (A) and CT scan (B) revealed 2 round-like low-density shadows next to the left posterior mediastinal spine. Arrows I and II showing the upper and lower mass, respectively. CT=computed tomography, MRI=magnetic resonance imaging.
with an uneven density inside and a cystic necrosis area (∼10.0–22.0 HU of CT value) in the center. The adjacent T4 and T5 vertebral bodies, their appendixes, and the left third, fourth, and fifth ribs were abnormal in shape, with osteosclerotic margins, and varying degrees of bone resorption.

Left-sided thoracotomy was planned to remove the chest space-occupying lesions. Surgical exploration showed that 2 large cystic masses occupied most of the left thoracic cavity, compressed the lung tissue, and caused atelectasis. The upper and lower poles of the upper mass were located at the cervical pleura and T8 vertebral body, respectively. Its basal portion was relatively wide, located in the posterior mediastinum at the level of T4 to T5, and compressed the local vertebral bodies and pedicles of the vertebral arches. The lower mass was located within the left lower lobe.

A severe adhesion between the left lower lobe and the diaphragm was carefully separated. The inferior pulmonary ligament was then dissociated to the level of the inferior pulmonary vein. For more intrathoracic space, the cystic fluid of the lower mass was drained with syringe (Fig. 2A). A branch of aorta was found supplying blood to the lower mass (Fig. 2B). Therefore, it was considered a pulmonary sequestration. The lower lobe was then resected completely (Fig. 2C). Deep yellow liquid was drawn out from the upper mass by a syringe. Close adhesions among the upper mass, the cervical pleura, and the posterior chest wall were bluntly separated to expose fully the base of the adhesion. A neurosurgery consultation was invited to participate in the operation, and confirmed that the mass did not infiltrate the spinal canal. The basal portion was then severed and the mass was completely removed (Fig. 2). Since the mass closely adhered to the upper lobe of the left lung, the lung tissue was damaged during blunt separation. After removal of the mass, the damaged lung tissue was sutured.

The pathological findings of the upper cystic mass, including abundant cells with active growth, bleeding, and necrosis, support the diagnosis of neurilemmoma (Fig. 3A). The pathology of pulmonary sequestration in the left lower lobe manifested as interstitial fibrosis in most tissues, glandular cystic dilatation, mild atypical hyperplasia of the epithelium, lymphocytic infiltrations, and thick vascular wall with tortuous dilatation (Fig. 3B).

The patient experienced remission of symptoms, had no significant postoperative complications, and was discharged from the hospital.

3. Discussion

This report describes an elderly patient with a mediastinal neurilemmoma accompanied by pulmonary sequestration. Two large cystic masses were found occupying most of the left thoracic cavity. The surgery was difficult and the risk of complications was high. Severe intrathoracic adhesions were successfully separated.
Two masses were completely removed. The patient experienced remission of symptoms, and had no significant postoperative complications.

Neurilemmomas are benign and sporadic in the vast majority of cases. Most neurilemmomas arise in a paravertebral location from intercostal nerves or sympathetic trunks, grow slowly, and are generally solitary. Neurilemmomas are more common in women. They occur mostly in the posterior mediastinal paraspinal region, and rarely in the anterior mediastinum and middle mediastinum. After a complete resection of a neurilemmoma, the prognosis is good, and the recurrence rate is relatively low.

In the present case, the woman patient had a 50-year history of neurilemmoma. The neurilemmoma was located in the posterior mediastinum at the level of the T4-to-T5 vertebral body, and compressed the local vertebral bodies and pedicles of the vertebral arches. Deep yellow liquid was drawn from the mass by syringe. The tumor was separated carefully to prevent injury to the nerves.

Pulmonary sequestration mostly manifests as an abnormal entry of blood-supplying arteries into the diseased lung tissue via the inferior pulmonary ligament. The aberrant nutrient arteries mainly arise from the thoracic aorta, and less often by the abdominal, intercostal, or subclavian arteries. Pulmonary sequestration is mainly treated by surgical resection of the diseased lung tissues via conventional thoracotomy. The key to successful treatment in surgery is the aberrant feeding vessels. The abnormal feeding arteries are fragile due to lack of a muscular layer, and are generally located in the inferior pulmonary ligament. In addition, multiple aberrant blood vessels may be present. Thus, extreme care is needed when separating the inferior pulmonary ligament and adherions near the diaphragm, and fatal bleeding caused by vascular rupture must be avoided. In the case reported herein, the pulmonary sequestration lesion was located within the left lower lobe. Severe adhesion between the left lower lobe and the diaphragm was successfully separated, the aberrant feeding vessel was properly managed, and the lower lobe was resected completely.

In summary, the presence of a mediastinal neurilemmoma accompanied by pulmonary sequestration is a rare occurrence. Special attention should be paid to neurological involvement of the neurilemmoma and the fragile feeding arteries of the pulmonary sequestration. Early diagnosis and treatment are important in such cases.

Author contributions

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