Resection of Intrapericardial Schwannoma Co-Existing with Thymic Follicular Hyperplasia through Sternotomy without Cardiopulmonary Bypass

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A 35-year-old man was admitted to Korea University Anam Hospital for evaluation of intermittent chest pain. Computed tomography of the chest showed enlargement of a previously identified anterior mediastinal mass and also a well-defined, circumscribed mass in the subcarinal area, surrounded by the roof of the left atrium, right pulmonary artery, and the carina. Complete resection of the intrapericardial tumor was performed through median sternotomy without cardiopulmonary bypass. Pathologic examination identified the tumor as schwannoma, of an ancient type, diffusely positive for the S-100 antigen. Unlike other reported cases, grossly, the tumor did not seem to be involved with any nerve.

Key words: 1. Schwannoma  
               2. Mediastinal neoplasms  
               3. Pericardium  
               4. Sternotomy  
               5. Benign neoplasm

CASE REPORT

A 35-year-old male patient with intermittent chest pain was admitted to Korea University Anam Hospital for further evaluation of a mediastinal mass that was found on computed tomography (CT) at another hospital. He had a history of Graves disease treated with radioactive iodine and methimazole and endoscopic sinus surgery for chronic sinusitis with a nasal polyp. The initial physical examination and chest X-ray were non-specific and electrocardiography showed ST segment elevation in leads V1–3, but no irregular rhythms. CT of the chest showed diffuse enlargement of the anterior mediastinal mass and a well-defined, circumscribed, heterogeneous soft-tissue attenuation mass in the subcarinal area (Fig. 1). It was 2.5×2.5×2 cm in size and was surrounded by the roof of the left atrium, right pulmonary artery, and carina. Echocardiography revealed a hyperechogenic mass in the posterior left atrium, but its relationship with the left atrium was not clear. Considering the possibility of neural or cardiac involvement, cardiac magnetic resonance imaging (MRI) was performed. A well-defined, intermediate-signal-intensity, 2.4-cm mass was noted in the middle mediastinum, at the roof of the left atrium. A relatively preserved regional fat plane was seen.
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Fig. 1. (A) Contrast enhanced chest computed tomography (CT) and (B) three dimensional CT reconstruction showing the well-demarcated margin of the mass and its location between the posterior wall of the left atrium, right pulmonary artery, and the carina. (C) A diffusely enlarged anterior mediastinal mass was also seen in chest CT.

Fig. 2. Intraoperative findings. (A) About 2.5×2.5×2 cm in size round tumor was located just posterior to the roof of the left atrium, and between the aorta and the main pulmonary artery. (B) Macroscopic view of the tumor.

Under general anesthesia, the patient underwent median sternotomy and total thymectomy. Then, the pericardium was opened and the major vessels exposed; cardiopulmonary bypass (CPB) was readied in case it was required. The mass and its surrounding structures were explored carefully. A 2.5-cm round tumor was located just posterior to the roof of the left atrium, between the aorta and the main pulmonary artery. No significant surrounding tissue invasion was observed; therefore, we decided to continue the operation without CPB. The plane between the ascending aorta and the main pulmonary artery was dissected carefully, and a soft, smooth, grey, ovoid tumor with scant vascularity was seen surrounded by the roof of the left atrium, right pulmonary artery, and carina. Unlike the reported cases, the tumor did not seem to involve any nerve grossly and the fat plane kept the tumor in place (Fig. 2). The tumor was removed completely, without injuring the surrounding tissues. After removing the intrapericardial tumor, the thymic mass and its surrounding mediastinal fat tissues were removed as an extended thymectomy.

Pathologically, the intrapericardial tumor was identified as an ancient-type schwannoma, diffusely positive for the S-100 antigen (Fig. 3). The cut surface revealed a well-demarcated white-and-gray fibrotic mass with a multifocal hemorrhagic appearance. The anterior mediastinal mass was reported to be thymic follicular hyperplasia. The postoperative course was uneventful, and the patient continues to do well 2 months postoperatively without any complications.
DISCUSSION

Schwannomas, also called neurilemmomas, are benign encapsulated tumors derived from Schwann cells [1,2]. They are the most common tumor of peripheral nerves and grow from peripheral nerves or nerve roots in an eccentric fashion, with the nerve itself usually incorporated within the capsule [1]. Schwannomas are the most common benign posterior mediastinal neurogenic tumors. These are typically asymptomatic but can produce signs of nerve compression, paralysis, Pancoast’s syndrome, and Horner’s syndrome. Intrapericardial neurogenic tumors are extremely rare, and most arise from the cardiac branches of the vagus nerve or the cardiac plexus. Mediastinal neurogenic tumors, which comprise 19% to 39% of all mediastinal tumors, develop mostly from mediastinal peripheral nerves, sympathetic and parasympathetic ganglia, and embryonic remnants of the neural tube. They are most frequently located in the posterior mediastinum. Of the posterior mediastinal neurogenic tumors, schwannomas are the most common benign tumors. They originate from Schwann cells and affect patients of both sexes, predominantly in the third and fourth decades of life [3]. Mediastinal benign schwannomas are most often asymptomatic, although signs of nerve compression, paralysis, Pancoast’s syndrome, and Horner’s syndrome can occur.

Among mediastinal neurogenic tumors, intrapericardial neurogenic tumors are extremely rare and arise mostly from the cardiac branches of the vagus nerve and the cardiac plexus [4]. Unlike other cases, despite our pre- and intraoperative studies, the tumor in our patient seemed to lack neural involvement, either grossly or microscopically. Neither preoperative cardiac MRI nor echocardiography showed involvement of the tumor with any nerve. Intraoperatively, the tumor was surrounded only by fat tissues and encased between the left atrium, right pulmonary artery, and carina.

Among the many possible surgical approaches, including thoracotomy and video-assisted thoracic surgery, we chose median sternotomy to approach the tumor, considering the tumor’s intrapericardial location and because there was also an anterior mediastinal tumor. The thymic tissue and pericardial fat were removed easily by extended thymectomy, in combination with the removal of the intrapericardial mass. Although CPB has been used in some reported cases for better hemodynamic stability and visualization of the tumor [4,5], it was avoided since the tumor was small and did not show any direct involvement of the surrounding cardiac wall, vessels, or carina. Consequently, the tumor was removed easily, but carefully, with minimal handling of the heart and without injuring the surrounding tissues.

Unlike malignant neurogenic tumors, which usually manifest symptoms of cough, dyspnea, chest wall pain, and nerve compression, most benign neurogenic tumors are asympto-
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matic and are usually discovered incidentally. Our patient had atypical chest wall pain in the left anterior chest, which was evaluated to rule out a cardiac origin. As there was no specific evidence of cardiac pain, chest CT was done. Compared with the reported mediastinal or intrapericardial tumors, the tumor was small, round, and well-demarcated. Despite their small size and absence of signs of cardiac compression, surgical resection is the standard of care for neurogenic tumors, since they can continue to grow and subsequently compress the cardiac chambers, resulting in cardiac dysfunction, arrhythmias, or even thromboembolism [6,7]. Therefore, the tumor in our patient was resected completely, and we expect a good long-term prognosis in keeping with the literature.

We present an intrapericardial schwannoma that was not directly associated with the cardiac wall or the surrounding tissues, combined with a thymoma. Both were removed successfully via sternotomy without CPB.

CONFLICT OF INTEREST

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

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