Case Reports

Mediastinal Schwannoma Diagnosed Preoperatively as a Cyst

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Mediastinal schwannomas are sometimes diagnosed as pericardial or bronchogenic cysts, if cystic degeneration is extensive. When mediastinal schwannomas are not diagnosed as primary cardiac tumors, the use of cardiopulmonary bypass in their resection appears to be infrequent.

We report the case of a 48-year-old woman who presented with symptoms from a suspected intrapericardial cyst. Multiple diagnostic images revealed a large mass, potentially a proteinaceous or hemorrhagic cyst, in the transverse sinus behind the ascending aorta and against the left main trunk. After complete resection with the use of cardiopulmonary bypass, the mass was identified as a benign extracardiac schwannoma. More than 3 years postoperatively, the patient had no relevant symptoms. We discuss the preoperative diagnosis, the method of resection, and our broad strategy for dealing with such a case. ***(Tex Heart Inst J 2014;41(1):76-9)***

We describe the case of a woman whose symptomatic intrapericardial schwannoma was diagnosed as a large cyst preoperatively. When mediastinal schwannomas are not diagnosed as primary cardiac tumors, their resection seldom seems to require the use of cardiopulmonary bypass (CPB). We discuss the diagnosis and treatment of our patient's condition and our broad strategy, which could apply in similar cases.

Case Report

A 48-year-old woman with occasional chest discomfort was referred to our hospital. Her medical history included pericardial effusion, high blood pressure, and schizophrenia. Chest radiographs showed an enlarged mediastinal shadow with a cardiothoracic ratio of 0.582. An electrocardiogram revealed sinus rhythm. Transthoracic echocardiograms showed a small pericardial effusion, a mass compressing the left atrium, and no specific chamber enlargement. An intrapericardial cyst was suspected, and it was decided to monitor her condition.

Several months later, the patient emergently presented with chest discomfort and palpitations. Computed tomograms (CT) showed a 50 × 51 × 50-mm mass in the transverse sinus (Figs. 1A and 1B). The mass was surrounded by the ascending aorta anteriorly, the aortic arch superiorly, the right pulmonary artery posterosuperiorly, and the left atrium posteroinferiorly. The main pulmonary artery was to its left, and the superior vena cava to its right. The relative density of the mass was approximately 40 Hounsfield units (HU). Although the images of the structure suggested a cyst, the HU value indicated that the mass contained highly proteinaceous fluid or that it was a hemorrhagic cyst. This ruled out the diagnosis of a simple cyst and prompted a more critical evaluation. Magnetic resonance images (MRI) with T2 weighting revealed high signal intensity characteristic of a cyst (Fig. 1C); T1-weighted images showed a lack of internal enhancement, indicating no solid component (Fig. 1D). A coronary angiogram showed an elongated, arch-shaped left main trunk (Fig. 1E). The diagnosis was a pericardial or bronchogenic cyst, potentially proteinaceous or hemorrhagic.

The patient's history of schizophrenia made further evaluation unrealistic. Because the tumor was large and was causing symptoms, the patient was scheduled for surgery. She was placed under CPB and cardiac arrest. The main part of the tumor was behind the ascending aorta; the front part was seen on both sides of that vessel. To resect the tumor safely, we had to transect the ascending aorta. The tumor was encapsulated, with

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slight adhesion to the surrounding tissue. Care was taken not to damage the left main trunk, which was adjacent to the tumor. Two feeding arteries were seen between the aortic arch and the tumor (Fig. 2A). Complete resection was achieved and the ascending aorta was reconnected.

The resected tumor was filled with dark, bloody fluids and had a yellow solid part (Fig. 2B), which was determined histopathologically to be a schwannoma. Spindle-shaped cells proliferated, with interstitial edema, hemorrhage, and hyalinization. Some regions were rich in cells (Antoni A type) with nuclear palisading and Verocay body (Fig. 2C). Other regions were poor in cells (Antoni B type). Immunohistopathologic study with S-100 stain was positive for spindle cells (Fig. 2D). There was no sign of malignancy. The patient's postoperative course was uneventful. More than 3 years postoperatively, she had no recurrence of the preoperative symptoms.

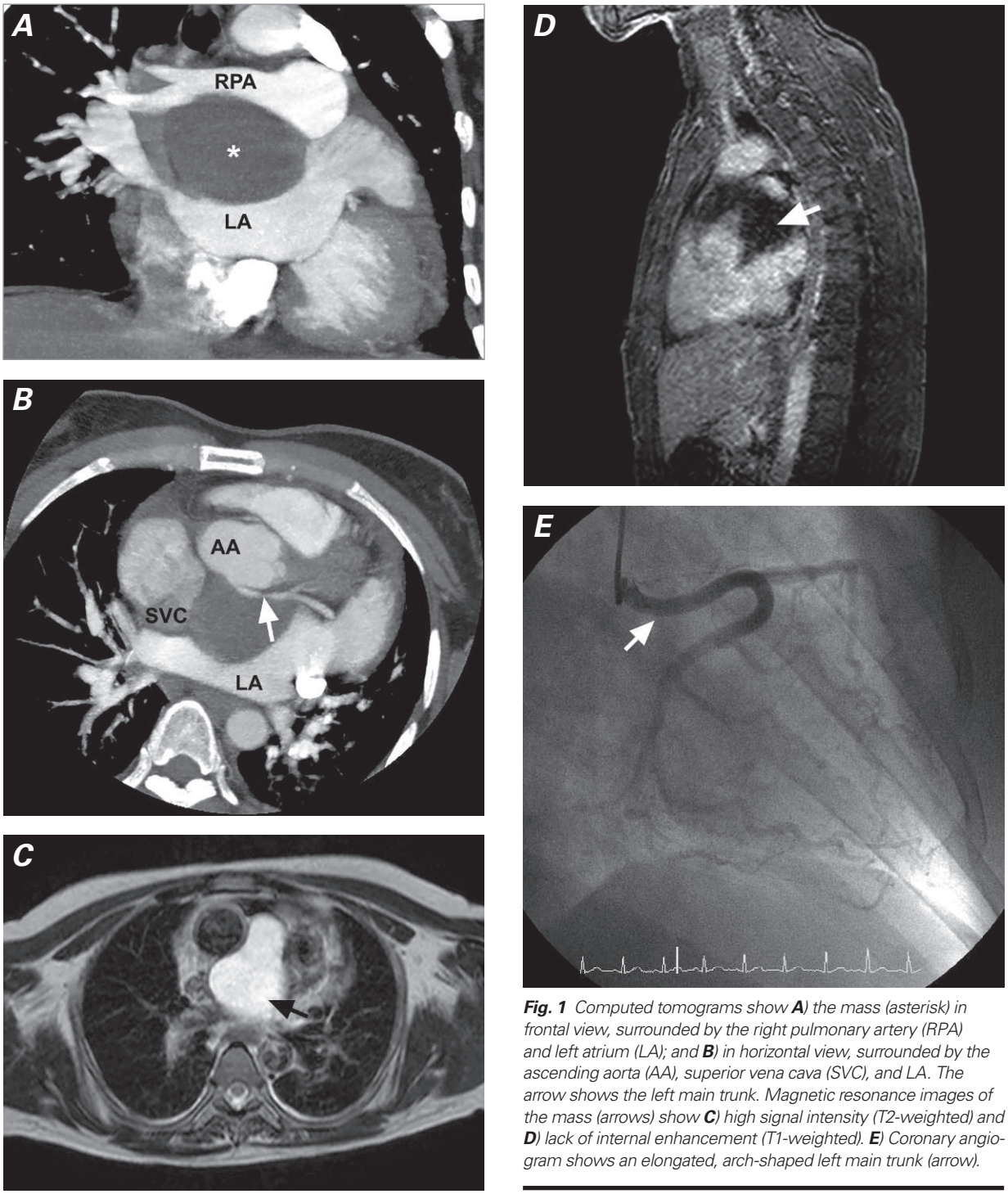

Fig. 1 *Computed tomograms show* ***A****) the mass (asterisk) in frontal view, surrounded by the right pulmonary artery (RPA) and left atrium (LA); and* ***B****) in horizontal view, surrounded by the ascending aorta (AA), superior vena cava (SVC), and LA. The arrow shows the left main trunk. Magnetic resonance images of the mass (arrows) show* ***C****) high signal intensity (T2-weighted) and* ***D****) lack of internal enhancement (T1-weighted).* ***E****) Coronary angiogram shows an elongated, arch-shaped left main trunk (arrow).*

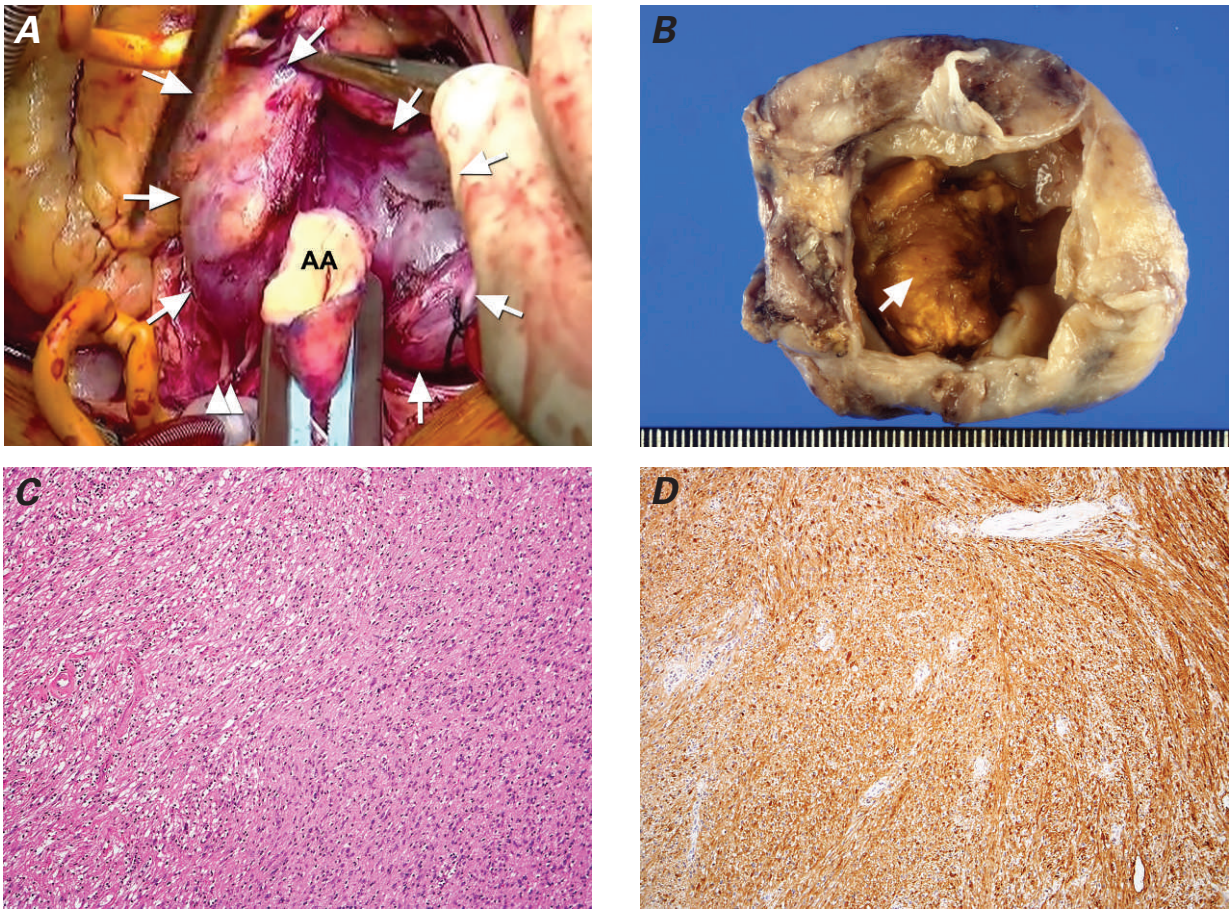

Fig. 2 ***A****) Operative photograph of the tumor (delineated by arrows). The arrowheads indicate the 2 feeding arteries between the aortic arch and the tumor.* ***B****) Macroscopic view of the resected mass shows the yellow solid part of the interior (arrow), determined to be a schwannoma on histopathologic examination.* ***C****) Right half of photomicrograph shows cell-rich Antoni A type tissue; left half shows cell-poor Antoni B type (H & E, orig. ×15).* ***D****) Photomicrograph shows positive staining for spindle cells (S-100, orig. ×15).*

AA = ascending aorta

Discussion

We discuss 3 points regarding this case: the preoperative diagnosis of a cyst, the method of resection, and our broad strategy for dealing with the circumstances.

Preoperative Diagnosis. Because of this tumor's location in the transverse sinus and its imaging characteristics, it was diagnosed as a pericardial cyst or bronchogenic cyst. Computed tomograms of a cyst usually show uniform water attenuation and an imperceptible wall.[1] Because the HU values of this mass were beyond the range of simple fluid, the primary diagnosis from the CT results was that this was a proteinaceous or hemorrhagic cyst. This was compatible with the fact that the tumor was filled with dark, bloody fluids. The T2-weighted MRI of a cyst usually shows homogeneous high signal intensity without septation or a mural nodule, and T1-weighted MRI shows low signal intensity with a regular thin wall.[1] Because these standards fit the characteristics of this mass, it was diagnosed as a cyst. Conversely, inhomogeneity in CT appearance can be characteristic of schwannomas, whether benign or malignant,[2] as are inhomogeneous high intensity on T2-weighted MRI and low-to-intermediate signal intensity on T1-weighted MRI.[3] If the cystic degeneration of tumors is extensive, they can be indistinguishable from congenital cysts.[1] Figure 2B shows the yellow solid part, which was the schwannoma itself. Although the size ratio of the inner mass to the outer cystic capsule was not unduly small, we were unable to detect the solid part in any preoperative MRI or CT image.

Method of Resection. Mediastinal neurogenic tumors, including schwannomas, constitute 10% to 25% of all mediastinal masses.[4] In contrast, primary cardiac schwannomas are rarely reported, and some necessitate CPB for resection.[5] Although our patient's neoplasm was in the transverse sinus and adjacent to the left main trunk, it had feeding arteries from the aortic arch and was not of certain cardiac origin. Because most of the tumor was behind the ascending aorta, it was necessary

to transect that vessel. In addition, placing the patient under cardiac arrest was necessary to avoid damaging the left main trunk. This reasoning prompted the use of CPB, which is apparently seldom instituted in the resection of extracardiac mediastinal schwannomas.

Broad Strategy. Although the tumor was large, our patient had no major hemodynamic instability and no venous congestion. Because she had symptoms of chest discomfort and palpitations, she was scheduled for surgery. Even though the tumor looked like a cyst preoperatively, resection was indicated because the mass was large and was causing symptoms. Part of the intention of surgery was to avert future hemodynamic instability and worsening symptoms if the mass were to enlarge. Accordingly, we think that the operation was justified: the mass proved to be a schwannoma, with the potential of malignancy or cardiac tamponade. Performing a preoperative thoracoscopic biopsy was of questionable value, because the solid portion of the mass would have been punctured only by chance.

Malignant pericardial schwannomas have been reported, albeit rarely. Because this tumor proved to be benign, our patient probably has a good long-term prognosis. Incomplete resection would have left the potential for future malignant transformation.

In our symptomatic patient with an extracardiac mediastinal schwannoma initially diagnosed as a cyst, surgery with the use of CPB was appropriate. Because confirmatory preoperative diagnosis can be difficult in such a case, aggressive surgery can be justified even upon the initial diagnosis of a cyst.

References

1. Jeung MY, Gasser B, Gangi A, Bogorin A, Charneau D, Wihlm JM, et al. Imaging of cystic masses of the mediastinum. Radiographics 2002;22 Spec No:S79-93.
2. Cohen LM, Schwartz AM, Rockoff SD. Benign schwannomas: pathologic basis for CT inhomogeneities. AJR Am J Roentgenol 1986;147(1):141-3.
3. Sakai F, Sone S, Kiyono K, Maruyama A, Ueda H, Aoki J, et al. Intrathoracic neurogenic tumors: MR-pathologic correlation. AJR Am J Roentgenol 1992;159(2):279-83.
4. Oldham HN Jr. Mediastinal tumors and cysts. Ann Thorac Surg 1971;11(3):246-75.
5. Hashimoto T, Eguchi S, Nakayama T, Ohzeki H, Hayashi J. Successful removal of massive cardiac neurilemoma with cardiopulmonary bypass. Ann Thorac Surg 1998;66(2):553-5.