Resection of an intrapericardial neurofibroma: A presumed anterior mediastinal thymoma

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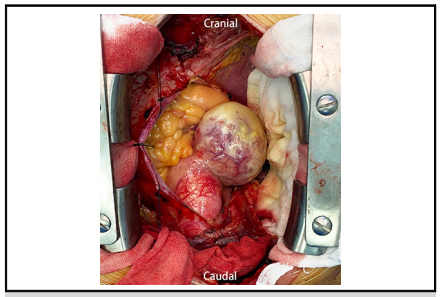

Resection of the pericardial neurofibroma located at the aortic root.

CENTRAL MESSAGE

Correct diagnosis and safety of surgical resection for an intrapericardial neurofibroma located at the aortic root is crucial for a 64-year-old woman without Von Recklinghausen disease.

CLINICAL SUMMARY

A 64-year-old woman sought medical attention at Guangdong Provincial People's Hospital reporting 6 months of intermittent palpitations, chest tightness, and weakness. With no cardiovascular history or familial neurofibromatosis, physical examination revealed fatigue but no cutaneous signs associated with neurofibromatosis. Cardiovascular examination appeared normal, with stable vital signs and normal blood pressure. Laboratory results were unremarkable. A computed tomography scan done in a local medical center gave an ambiguous diagnosis of mediastinal tumor. A contrast magnetic resonance imaging scan was prescribed and it revealed a large, round, anterior mediastinal mass (47 × 49 mm) that showed a high signal on T1-weighted sequences and mixed low signal on T2-weighted sequences (Figure 1, *A* and *B*). The tumor was located within the pericardium and a diagnosis of cystic thymoma was given by the radiologist. Positron emission tomography revealed a large cystic lesion in the anterior mediastinum with a central photopenia (Figure 1, *C*). The pericardium might have been involved and a diagnosis of thymoma was given. There was no evidence of local invasion or distant metastasis. Echocardiography suggested an anterior mediastinal mass adjoining to the aorta but no abnormalities within the pericardium (Figure 1, *D*).

Based on the above information, location variations and possible diagnoses were summarized. Possible location: anterior mediastinal mass with or without pericardial involvement or intrapericardial mass with or without neighboring involvement. Corresponding diagnoses were cystic thymoma/teratoma/lymphoma and pericardial cyst/teratoma/fibrous tumor.

A surgical resection of the tumor was performed via median sternotomy. During the operation, a cystic mass was found to be tightly adhered to the aortic root anteriorly within the pericardium. No pulse was palpable on the surface of the mass and the possibility of thymoma is less for an intrapericardial tumor. Considering the cystic nature and the tight adhesion with the aorta, an en bloc resection was probably unfeasible unless aorta suturing or even

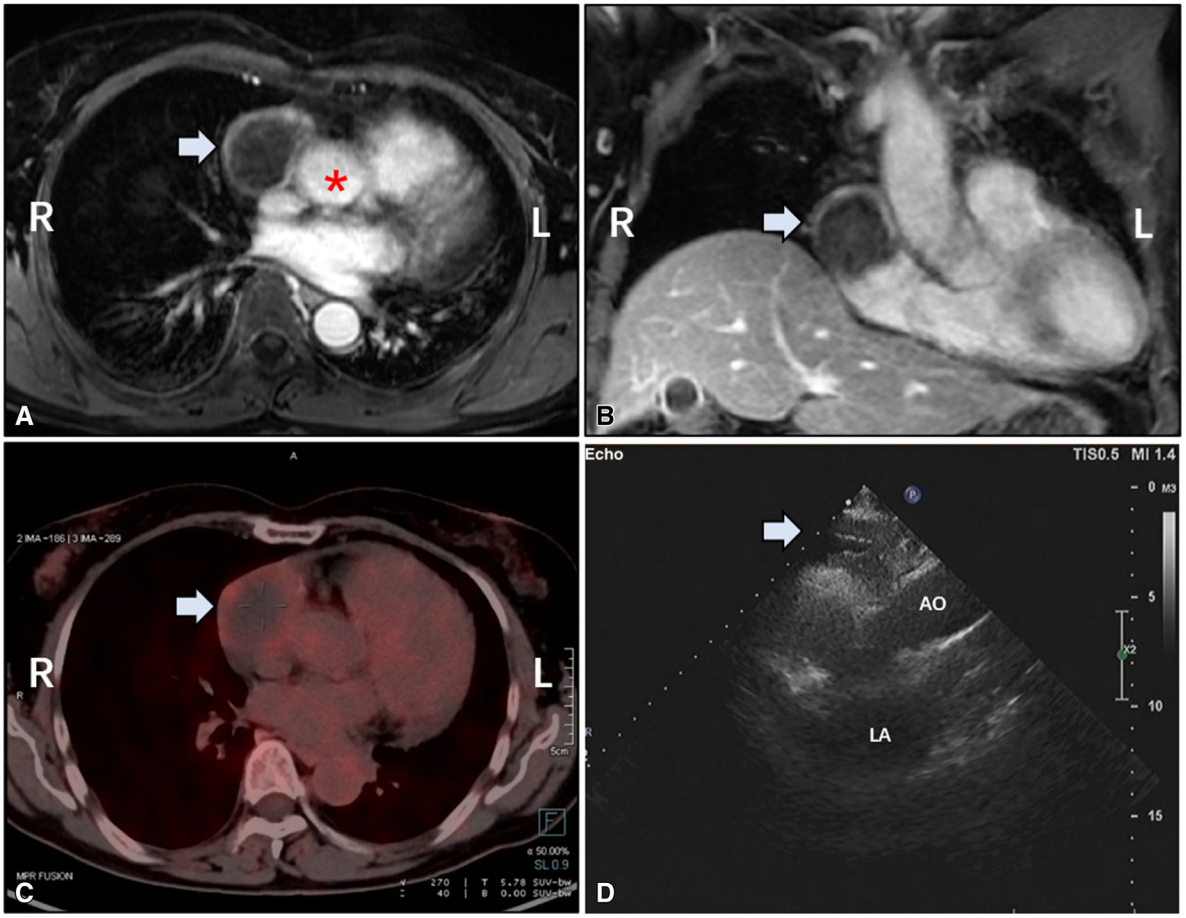

FIGURE 1. Thoracic magnetic resonance imaging scans showed an anterior mediastinal mass (*arrow*) of cystic density from transverse view (A) and coronal view (B). The aortic root (*asterisk*) and the arrow indicates the mass. Positron emission tomography (PET) depicting a solitary well-defined mass with homogeneous enhancement adjacent to the aorta (C). The mass was visible in the echocardiography study (*arrow*) (D). *AO*, Aorta; *LA*, left atrium.

replacement was prepared. Fine needle aspiration produced dark red fluid, alleviating concerns about direct vascular connection to the aorta. The roof of the mass was resected and an intraoperative pathological examination revealed a benign mesenchymal tumor. To avoid potential rupture of the aortic wall, the remaining mass was then separated into several parts and removed, except for the bottom tissue rooted on the aorta.

Histopathological analysis identified the tumor as a cystic mass comprising plexiform or sarciniform spindle cells with elongated wavy nuclei (Figure 2, *A* and *B*). Immunohistochemistry studies showed that the spindle cells were strongly positive for S-100 (Figure 2, *B*) and SOX10 proteins (Figure 2, *C*) and negative for neurofilament, smooth muscle actin, signal transducer and activator of transcription 6, CD34, CD31, desmin, epithelial membrane antigen, and Ki67 protein (Figure 2, *D*). The diagnosis of a neurofibroma was made according to the final pathological result.

The patient was discharged 5 days after the operation with an uneventful postoperative recovery. At 3-year follow-up, the patient remains symptom-free and enjoys a satisfactory quality of life.

DISCUSSION

Neurofibromas are benign peripheral nerve sheath tumors. They can occur sporadically or as a part of syndromic neurofibromatosis. Mostly they are present as lesions around the trunk or within the cranium, although they may also be present within the thorax, mostly in the posterior mediastinum. As for neurofibroma within the pericardium, cases of cardiac neurofibroma have been reported by Debonnaire and colleagues[1] and those patients carry a diagnosis of neurofibromatosis or Von Recklinghausen disease. However, a sporadic neurofibroma localized at the aortic root within the pericardium is extremely rare. To the best of our knowledge, this patient is the first case ever reported.

In this case, thymoma was the primary diagnosis because we assumed the mass was more likely located outside the pericardium. Certainly, other possibilities were not ignored,

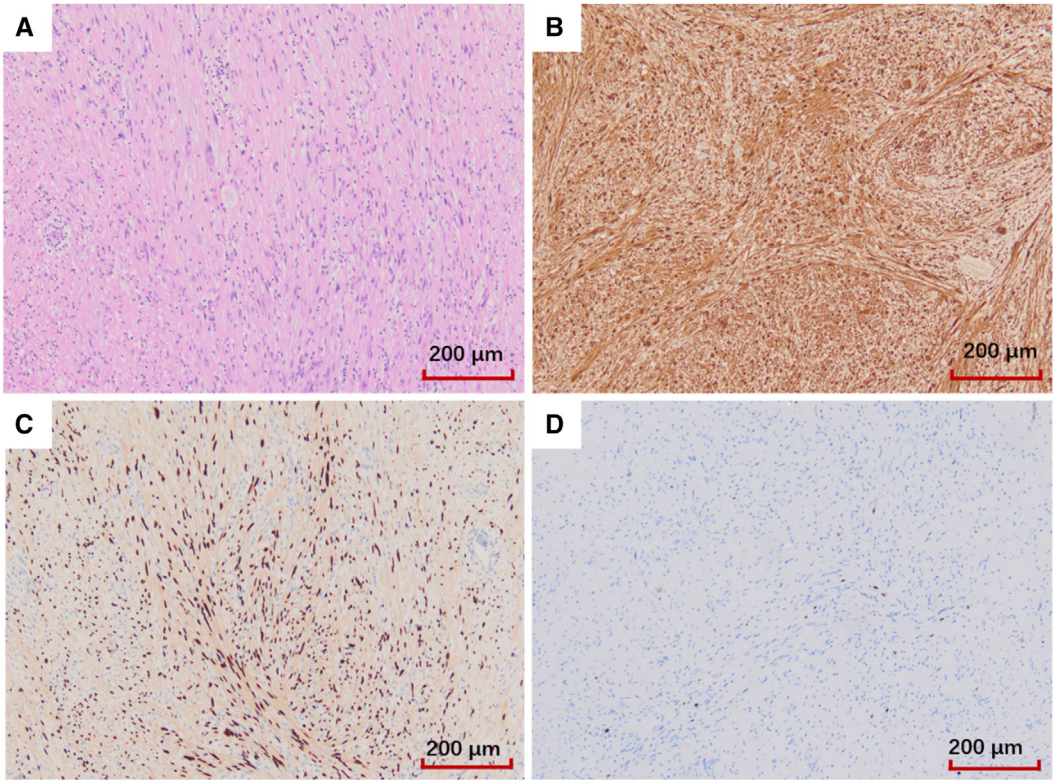

FIGURE 2. Hematoxylin-eosin staining of the pathologic specimen: spindle cells with elongated nuclei in extensive fibrous tissue (A). Immunohistochemical stain of abundant S-100-positive (B), SOX10-positive (C), and Ki-67-negative (D) spindle cells.

including teratoma or pericardial cyst if within the pericardium. Similar to the case reported by Alimi and colleagues,[2] the patient did not show any clinical signs of neurofibromatosis; thus, we rejected the diagnosis of neurofibroma at the beginning.

A differential diagnosis of a functional intrapericardial paraganglioma was necessary for this case of intrapericardial tumor with palpitation. Due to an extremely rare prevalence in elderly people and this patient's lack of hypertension history, we considered a mass causing palpitation preoperatively.

The tumor's strong positive stains for S-100[3] and SOX10[4] proteins indicated its neurogenic nature, whereas low proliferative activity, evidenced by scattered Ki-67[5] positivity (Figure 2, *D*), supported its benign character. Regarding its anatomic location, the tumor likely originated from Schwann cells and fibroblasts within the aortic adventitia rather than the vagus nerve, given its intact pericardial covering.

This case underscores the significance of precise image-based differential diagnoses for mediastinal and pericardial tumors, where mediastinal magnetic resonance imaging often offers superior local characterization for soft tissue compared with positron emission tomograpyhy or echocardiography. However, imaging might be still insufficient for this critical location when considering potential variability in location, invasiveness, and histology. This dilemma requires comprehensive and detailed preoperative planning.

The study was approved by the ethics committee of Guangdong Provincial People's Hospital (No. GDREC2020232H) in January 2020. The written informed consent of the patient for publication of data was provided.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers for this article have no conflicts of interest.

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