Resection of an intrapericardial neurofibroma: A presumed anterior mediastinal thymoma

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Disclosures: The authors reported no conflicts of interest.
Funding Statement: This work supported by the International Science and Technology Cooperation Program of Guangdong (2022A0505050048)

IRB/ERB statement: 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.

Central figure. 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 aortic root is crucial for a 64-year-old woman without Von Recklinghausen disease.

Clinical Summary

A 64-year-old female sought medical attention at Guangdong Provincial People’s Hospital, reporting six 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 are unremarkable. A computed tomography (CT) done in local medical center gave an ambiguous diagnosis as mediastinal tumor. A Contrast magnetic resonance imaging (MRI) was prescribed and it revealed a large round anterior mediastinal mass (47×49mm), which showed a high signal on T1-weighted sequence and mixed low signal on T2-weighted sequences (Figure 1A, 1B). The tumor was located within the pericardium and the diagnosis of cystic thymoma was given by the radiologist. Positron emission tomography (PET) revealed a large cystic lesion in
the anterior mediastinum with a central photopenia (Figure 1C). The pericardium might be involved and a diagnosis of thymoma is 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 1D).

Based on the above information, location variations and its possible diagnoses were summarized. Possible location: 1) anterior mediastinal mass with or without pericardial involvement 2) intrapericardial mass with or without neighboring involvement. Corresponding diagnoses were: 1) cystic thymoma/teratoma/lymphoma; 2) pericardial cyst/teratoma/ fibrous tumor.

A surgical resection of the tumor was performed via median sternotomy. During the operation, the cystic mass was found to be tightly adherent 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 tight adhesion with aorta, an en-bloc resection is probably unfeasible unless an aorta suturing or even replacement was prepared. A 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 aortic wall, the remaining mass was then separated into several parts and removed successively except for the bottom tissue rooted on aorta.

Histopathological analysis identified the tumor as a cystic mass comprising plexiform or sarcomorph spindle cells with elongated wavy nuclei (Figure 2A, 2B).
Immunohistochemistry studies showed that the spindle cells were strongly positive for S-100 (Figure 2B), SOX10 protein (Figure 2C) and negative for NF, SMA, STAT6, CD34, CD31, Desmin, EMA, and Ki67 (Figure 2D). The diagnosis of a neurofibroma was made according to the final pathological result.

The patient was discharged 5 days after operation with an uneventful postoperative recovery. A three-year follow-up shows 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 pericardium, cases of cardiac neurofibroma have been reported by Debonnaire[1] and these patients carry a diagnosis of Neurofibromatosis or Von Recklinghausen disease. However, a sporadic neurofibroma localized at the aortic root within 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 including teratoma or pericardial cyst if within the pericardium. Similar to the case reported by Hedieh Alimi[2], the patient did not show any clinical signs of neurofibromatosis, thus we neglected the diagnosis of neurofibroma at the beginning.

A differential diagnosis of a functional intrapericardial paraganglioma is necessary for
this case of intrapericardial tumor with palpitation. Due an extremely rare prevalence for elderly people and no hypertension history of this patient, we considered a mass effect causing palpitation preoperatively.

The tumor's strong positive stains for S-100[3] and SOX10[4] protein indicated its neurogenic nature, while low proliferative activity, evidenced by scattered Ki-67[5] positivity(Figure 2D), 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 MRI often offering superior local characterization for soft tissue compared to PET or echocardiography. However, imaging might be still insufficient for this critical location concerning potential variability in location, invasiveness and histology. This dilemma requires surgeons more comprehensive and detailed preoperative planning.

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**Figure legend**

**Figure 1.** Thoracic MRI 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 showed in echocardiology study (arrow), AO: aorta, LA: left atrium (D).

**Figure 2.** Hematoxylin-eosin staining of 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.