CASE REPORT

Epicardial cavernous hemangioma: The diagnostic challenge of a middle mediastinal mass

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Abstract
Primary tumors in the middle mediastinum are rare and pose diagnostic challenges. Lymphomas, mediastinal cysts and thymomas most frequently affect this anatomic area. Primary cardiac tumors are rare and constitute a differential diagnosis for the inferior middle mediastinum. Surgical exploration and resection is often mandatory in order to make a definitive diagnosis. Here, we report the case of a 69 year-old women who presented with persistent dyspnea. A complete preoperative workup revealed a large tissular mass adjacent to the right atrium. A diagnosis of a typical epicardial cavernous hemangioma was made following surgical resection.

KEYWORDS
cavernous hemangioma, mediastinal tumor, middle mediastinum

INTRODUCTION

The classification of Fraser et al.1 divides the mediastinum into three compartments: the anterior, middle and posterior mediastinum based on a lateral chest radiograph. The middle mediastinum comprises the heart, pericardium, ascending and transverse aorta, brachiocephalic vessels, vena cava, main pulmonary arteries and veins, trachea and main stem bronchi. Primary tumors in this anatomical region are extremely rare and pose diagnostic challenges. Lesions invading the middle mediastinum such as lymphoma, mediastinal metastatic lymph node, lymph node enlargement due to tuberculosis, cystic lesion, neurogenic neoplasm, ectopic thymoma or primary cardiac tumors may be suspected on chest CT scan.2 Other modalities of imaging can assist in the assessment of mediastinal tumors such as magnetic resonance imaging (MRI), 18 fluorodeoxyglucose (18-FDG) positron emission tomography (PET) and transthoracic echocardiography (TTE).3,4 Albeit being useful tools, imaging and conventional diagnostic techniques are often unable to ensure the precise nature of a mediastinal lesion giving surgical exploration and resection a place of choice in their management.

Here, we present a case of a middle mediastinal mass that posed diagnostic challenges, despite a complete imaging workup leading to surgical exploration and resection.

CASE REPORT

A 69 year-old woman with a medical history of hypertension and endometrial cancer presented at our institution with persistent dyspnea. The patient denied symptoms of chest pain, orthopnea, lower extremity oedema and arrhythmia. Chest computed tomography (CT) scan revealed a 80 x 70 x 40 mm homogeneous tissular mass, adjacent to the right atrium (Figure 1(d)). Transthoracic echocardiography disclosed a tissular lesion around the roof of the right atrium that was not causing extrinsic compression. Left ventricle ejection fraction was 70% without focal contractile abnormality. Pulmonary function tests were also performed and facilitated a concomitant diagnosis of asthma, and explained the patient’s persistent dyspnea. Contrast-enhanced MRI in morphokinetic sequences in the right ventricular outflow tract (RVOT) confirmed a homogeneous, hypervascular and encapsulated mass in the roof of the right atrium, in the pericardium but extra-cardiac, associated with a laminated superior vena cava (Figure 1(a)). 18-FDG PET scan showed...
an isolated low and heterogeneous hypermetabolism of the lesion with a standardized uptake value of 1.8 (Figure 1(c)). At this time, the main diagnostic hypothesis was an intrapericardial thymoma.

As the lesion was deemed resectable, the patient underwent surgical exploration by median sternotomy. After pericardiotomy, a lesion was found in the roof of the right atrium between the laminated superior vena cava and the ascending aorta (Figure 1(b)). The mass emerged from the epicardium without invading the right coronary artery as suggested by preoperative imaging. A cardiopulmonary bypass without cardiac arrest was deemed to be necessary to ensure safe and complete removal of the tumor.

Pathological examination showed a lesion with large vascular structures with regular endothelium enhanced with muscular fibers, making the diagnosis of typical epicardial cavernous hemangioma. Resection margins were negative (Figure 2). The postoperative course was uneventful, and the patient was discharged eight days after surgery. After five years of follow-up, the patient is alive without evidence of recurrence.

**DISCUSSION**

Primary mediastinal masses encompass a wide spectrum of neoplastic, congenital and inflammatory diseases. Despite a complete imaging workup, defining the precise nature of these lesions is difficult and histological confirmation is essential. Therefore, patients with mediastinal masses or cysts usually undergo surgical exploration with eventual resection as it often allows definitive treatment at the same time. Here, we present the case of a patient with a middle mediastinal mass with a challenging preoperative diagnosis. Although uncommon in the middle mediastinum, the mass in our patient was considered a thymoma by its imaging characteristics. More frequently localized in the anterior mediastinum, 4% of thymomas can arise from other locations including the middle mediastinum. Our preoperative diagnosis was plausible. But, as previously reported, a mediastinal mass is not always what it seems and can create some surprises during surgical resection. This was the case in our patient when we discovered a cardiac tumor arising from the epicardium.

Primary cardiac tumors are rare with an incidence estimated at 0.001 to 0.3 per 100,000 patient-years according to large autopsy studies. Approximately 75% of primary tumors of the heart are benign and mostly correspond to atrial myxoma. Hemangiomas comprise 2%–5% of benign tumors and can arise from all layers of the heart including endocardium, myocardium, epicardium and pericardium. Anatomically, they can be located anywhere but are predominantly found in the right ventricle. Less than 100 cases of cardiac hemangiomas and less than 10 cases of epicardial cavernous hemangiomas have been reported in the literature. Symptoms actually depend on the anatomic location and the size of the tumor without pathognomonic signs. Dyspnea, as well as arrhythmias, congestive heart failure or pericardial effusion have been previously reported. However, most are asymptomatic and discovered incidentally by echocardiography, CT scan, cardiac MRI or at autopsy. MRI remains the most specific imaging modality as it is able to detect a heterogenous isointense or hypointense lesion on T1-weighted sequences, and as a hyperintense mass on T2-weighted images. The natural history of cardiac cavernous hemangioma is actually uncertain, regarding both growth rate and the risk of sarcomatous transformation. One case of angiosarcoma arising in a patient with a history of cavernous hemangioma transformation has been previously reported.

In conclusion, to define the precise nature of a middle mediastinal lesion is challenging. Surgical exploration and resection are often mandatory in order to make a diagnosis.
and ensure definitive treatment. Lymphoma, mediastinal cyst and thymoma may affect the middle mediastinum but the clinician should also consider primary cardiac tumors when it comes to the inferior middle mediastinum.

CONFLICT OF INTEREST
The authors declare no conflict of interest related to this work.

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