Case Report

Left atrial myxofibrosarcoma: Diagnostic approach through imaging techniques

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A B S T R A C T

Primary cardiac sarcomas are rare. A 63-year-old woman presented with progressive symptoms of fatigue, palpitations, and dyspnea. Transthoracic echocardiography detected a mass in the left atrium, fixed and probably attached to the interatrial septum suggestive of myxoma. Transesophageal echocardiography confirmed the existence of a large lobulated mass in the left atrium measuring 45 mm × 25 mm in diameter. Subsequent cardiac magnetic resonance imaging showed a mass of heterogeneous appearance, with broad base of implantation on the posterior wall of the left atrium. The extension study with multidetector computed tomography showed superficial infiltration of the atrial wall, without involvement of the mitral valve or pulmonary veins and no extension to other extracardiac structures. The patient underwent surgery resecting a bilobed mass, smooth and yellowish, strongly attached to the posterior wall of left atrium. The pathologic study was consistent with the diagnosis of myxofibrosarcoma.

Learning objective: Primary cardiac tumors are among the most challenging disease entities to diagnose because of their rarity and highly variable and usually nonspecific clinical presentation. Although the definitive diagnosis of a cardiac tumor, whether benign or malignant and the tumor type is provided by histological examination, imaging techniques are essential for confirming the existence of a cardiac mass and characterizing the tumor before obtaining a sample for pathologic examination.

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Introduction

Primary tumors of the heart are rare, with a reported prevalence of 0.001–0.03% in autopsy series [1]. About 75% of all primary cardiac tumors are regarded as benign neoplasms, with cardiac myxoma accounting for at least half of them. Of the remaining 25%, that are considered to be malignant neoplasms, the majority are sarcomas, angiosarcoma being the most common variety [2]. Myxofibrosarcoma is a tumor of mesenchymal origin that occurs most commonly in the extremities of the elderly [3]. However, primary myxofibrosarcoma rarely arises in the heart, with only a few cases reported in international literature [4,5]. Myxofibrosarcoma of the heart may involve any of the 4 cardiac chambers, but most frequently affects the left atrium. It may arise from myocardium or valve tissue. Low-grade tumors invade local tissue including valvular structures. High-grade tumors also invade locally, but may also metastasize to contiguous and distant sites. Left atrial myxofibrosarcoma may simulate left atrial myxoma in its clinical presentation.

Case report

A 63-year-old woman with a history of hypertension and asthma presented with progressive symptoms of fatigue, palpitations, and dyspnea. Cardiac auscultation exhibited pure heart tones with no murmurs or added sounds. The rest of the physical examination as well as electrocardiogram, chest radiography, and analytics were normal.

Transthoracic echocardiography detected a mass in the left atrium, fixed and probably attached to the interatrial septum suggestive of myxoma. Transesophageal echocardiography confirmed the existence of a large lobulated mass in left atrium measuring 45 mm × 25 mm in diameter (Fig. 1). The insertion site was large, poorly defined by this technique, up to the roof of the left atrium. The mass had diastolic protrusion through the mitral valve leaflets without being obstructive and showed heterogeneous echo contrast uptake.

A subsequent cardiac magnetic resonance imaging (MRI) showed a mass of heterogeneous appearance, with broad base of implantation on the posterior wall of the left atrium. The mass showed isointense signal relative to the heart muscle on T1-weighted sequences, suggesting the absence of fat or hemorrhage, and markedly hyperintense on STIR sequences, so it was a structure with a high content of water (Fig. 2). In the first-pass perfusion
sequence the mass was highly perfused. Delayed enhancement sequences showed a central hypointense zone, suggesting intratumoral necrotic areas surrounded by foci of increased uptake.

A noninvasive coronary study with multidetector computed tomography (MDCT) was also performed before surgery, the result of which was normal. In addition, the mass was visualized attached to the posterior wall of the left atrium, presenting lower attenuation than the myocardium, with heterogeneous zones inside (Fig. 3). The extension study showed superficial infiltration of the atrial wall, without involvement of the mitral valve or pulmonary veins and no extension to other extracardiac structures.

The patient underwent surgery observing a lobulated mass of 5 cm × 3 cm, smooth and yellowish, strongly attached to the posterior wall of left atrium. The whole mass tumor and its base of implantation on the atrial wall were resected (Fig. 4). The patient did well postoperatively, being discharged 8 days after surgery.

The pathologic study found that the tumor consisted of spindle or stellate morphology cells which adopted a diffuse pattern, sometimes in a swirling shape. There were areas with marked cytologic atypia and a high number of mitosis and areas of focal necrosis (Fig. 5). Immunohistochemistry showed negativity for CD34, CD31, and calretinin. This was consistent with the diagnosis of myxofibrosarcoma. Pathology confirmed that the atrial wall was superficially infiltrated but the borders of the surgical piece were clean.

The entire mass had been completely excised, the borders were not affected, and the extension study was negative, so initially she did not receive any adjuvant treatment. Fourteen months after surgery, an echocardiographic control revealed a recurrence of the tumor in the same location, which was confirmed by cardiac MRI and MDCT. A new surgery was rejected and the patient started on chemotherapy treatment with ifosfamide and Adriamycin, achieving the disappearance of the tumor on imaging tests. Sixteen months later, she experienced a new recurrence of the tumor, newly receiving several chemotherapy cycles with ifosfamide. The tumor decreased in size but did not disappear. Forty-six months after diagnosis, the patient is alive with few symptoms of the disease.

Discussion

Primary cardiac tumors are among the most challenging disease entities to diagnose because of their rarity and highly variable and usually nonspecific clinical presentation. Although the definitive diagnosis of a cardiac tumor, whether benign or malignant and
Fig. 3. Cardiac multidetector computed tomography. (A) Oblique four-chamber view showing the big mass in the left atrium without affecting the pulmonary venous drainage. (B) Oblique two-chamber view shows the mass infiltrating the posterior wall of the atrium, which receives its vascular supply.

Fig. 4. Surgery pictures. (A) View of the mass after performing the left atriotomy. (B) Bilobed mass of 3 cm × 5 cm in diameter once resected.

Fig. 5. Pathological study. (A) Tumor is composed of myxoid cells adopting a diffuse pattern with some larger cells, alternating with zones of necrosis. (B) At higher magnification these cells are large with dense cytoplasm, abundant and irregular nucleus, chromatin rough and frequent mitotic figures (arrows).
the tumor type is provided by histological examination, imaging techniques are essential for confirming the existence of a cardiac mass and characterizing the tumor before obtaining a sample for pathologic examination [6].

Echocardiography is the imaging technique most commonly used as a first approximation for the diagnosis of cardiac tumors, for its speed, high availability, and low cost. It can identify the location of the mass, its insertion site, size, shape, and mobility, in addition to detecting the presence of associated hemodynamic alterations [7]. However, it has several limitations, mainly resulting from problems of acoustic window and a relatively narrow field of view [8].

Cardiac MRI offers several advantages over echocardiography in the study of cardiac tumors, such as the characterization of lesion composition (fat, cyst, fibrosis, calcium, etc.), the definition of myocardial involvement, the study of their vascularization, the assessment of the infiltration grade, and the extension to extracardiac structures by providing a larger field of exploration. Therefore it is considered the preferred technique in the detection and characterization of cardiac tumors [9].

Cardiac MDCT is also an appropriate technique for the characterization of cardiac tumors, but it is particularly useful for assessing the degree of local invasion and spread to adjacent structures and to detect metastatic lesions [10].

In this case, because of the location and type of mass, the most likely diagnosis was atrial myxoma. However, its large size, broad base of implantation in the posterior wall of the atrium, heterogeneous appearance with necrotic areas inside, and great vascularization were more characteristic features of malignant tumors, so it was not possible to rule out a possible sarcomatous etiology, which was subsequently confirmed on histologic examination.

Conflict of interest

All authors have no conflict of interest that should be declared.

References