




CARDIAC SYNOVIAL SARCOMA

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ABSTRACT

Cardiac tumors are divided into metastatic and primary origin. Primary cardiac tumors have a very low incidence (0.001-0.03%), of which only 25% are malignant and very aggressive. We present a clinical case of a 60-year-old female patient with dyspnea, palpitations and dizziness. Transthoracic echocardiogram showed a heterogeneous mass in the left atrium, so it was decided to resect it. The patient had a torpid postoperative course and dies 48 hours later.

Keywords: *Primary cardiac tumor, Cardiac malignancies, Synovial sarcoma*

INTRODUCTION

Cardiac tumors are divided into metastatic and primary origin. Primary cardiac tumors have a very low incidence (0.001% to 0.03%), of which only 25 % are malignant and very aggressive.⁽¹⁾ Synovial sarcoma is more common in the soft tissue of limbs, but it is extremely rare to find it in the cardiac location.⁽²⁾ Prognosis is poor, and survival sits at around 6 months, even with surgery.⁽³⁾

CASE REPORT

This is the case of a 60-year-old woman admitted to the coronary unit with progressive dyspnea FC III of 3-week evolution. Admission EKG sinus rhythm 85 L/m preserved P and PR. Narrow QRS, normal ST-segment, and isolated ventricular extrasystoles. Ultrasensitive troponin 22.8. Transthoracic echocardiogram: LV normal EF 60%-65%. Left atrium with severe dilatation, heterogeneous mass with areas of hypoechogenicity, mobile, with implantation base in the interatrial septum and occluded mitral valve 4.8 x 3 and 5.6 x 3.8 cm TEVI (**Figure 1**). Moderate mitral stenosis. Normal RA and RV. Coronary angiography: normal coronary arteries. Given the patient's symptoms and the ECG characteristic of the mass added to frequency, diagnosis of suspected left atrium myxoma is achieved. At that moment it was not considered to perform NMR given how certain the diagnosis is, on one hand, and because she is a surgical patient either way according to the heart team.

The indication for surgery was more than clear, because left atrium tumors are considered emergencies, that is, the patient should be operated right after diagnosis is achieved.

Surgery was performed through median sternotomy, cannulation of both cavas with Pacific

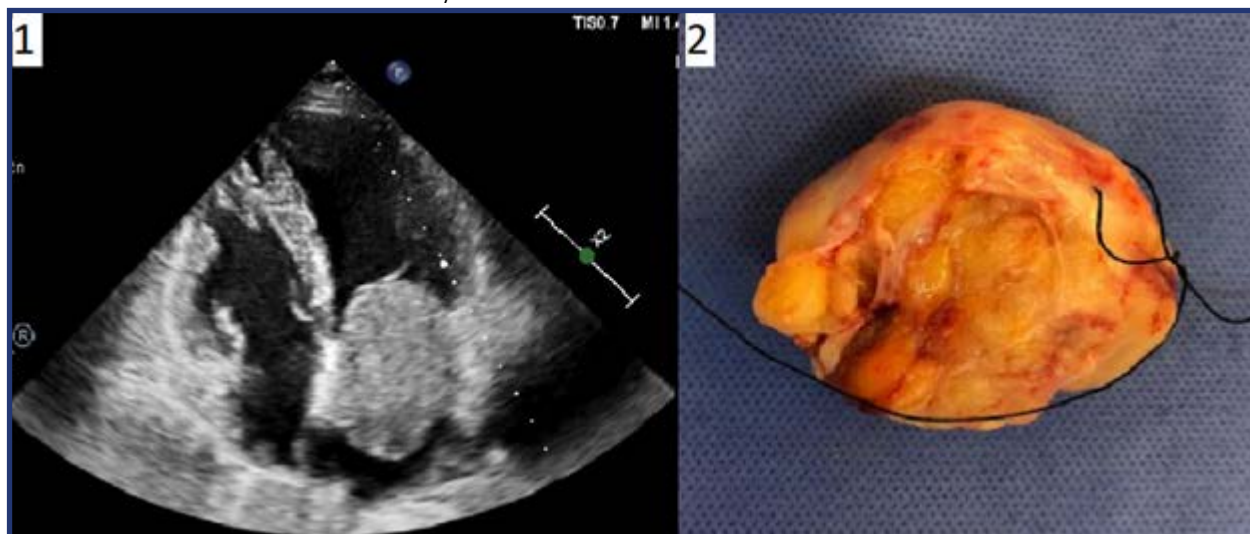
cannulas and ascending aorta with a 22-Fr aortic cannula. After entering ECC, the RA is opened and both the right atrium and ventricle are examined and turn out to be tumor-free. Afterwards, the aorta is clamped and cardioplegia is infused (Bretschneider's solution).

The Guiraudon approach is attempted where it is confirmed, before opening the fossa ovalis, that there is not a less renitent area for the opening given the mass was pressing 100% of the fossa ovalis and the septum. It is decided to make an incision on the left atrial roof, distal to the arteria anastomotica auricularis magna 2 cm away from the left atrial appendage orifice with a 11 blade scalpel, going proximal, that is, towards the interatrial septum.

A large, whitish mass attached to the septum is revealed, which clearly did not have the appearance of a myxoma, but of a tumor that was infiltrating the fossa ovalis, the interatrial septum, the posterior mitral valve in P3, the medial commissure, 3 cm of the left atrial posterior wall and part of the left ventricular posterior wall. The tumor is resected, part of the left atrial posterior wall, the entire interatrial septum, and the mitral valve. Part of the LV wall and the mitral annulus were left with remains of the tumor since the myocardium was clearly infiltrated by the mass. This was followed by complete resection of the mass that obviously lacked oncologic validity, but did multiply OR mortality. Repairs of the resected area are made with a pericardial patch and mitral replacement using mechanical prosthesis No. 25 (**Figure 2**). With clamp times of 61 minutes and 85 min of extracorporeal ECC is exited.

The patient evolves with severe SIRS characterized by massive clinical bleeding, and high requirement of vasopressors. Patient is re-examined due

FIGURE 1. 1. Echocardiographic image on TTE. Mass observed in the left atrium with invasion of mitral apparatus (Image from the Hospital Fernández Echocardiography Department, courtesy of Dr. Silvia Makhoul). **2.** a 5.3 x 4.3 x 2.5 cm nodular, solid mass, of lobulated external surface, yellowish brown in color can be seen here.



to clinical bleeding without any findings of surgical bleeding; she dies 48 hours after surgery. Anatomical pathology: malignant mesenchymal tumor that due to histopathological signs plus the immunohistochemical pattern reveal, in the first, place, synovial sarcoma. (Vimentin positive, BCL-2 positive, TLE1 positive, K167 > 30%).

CONCLUSION

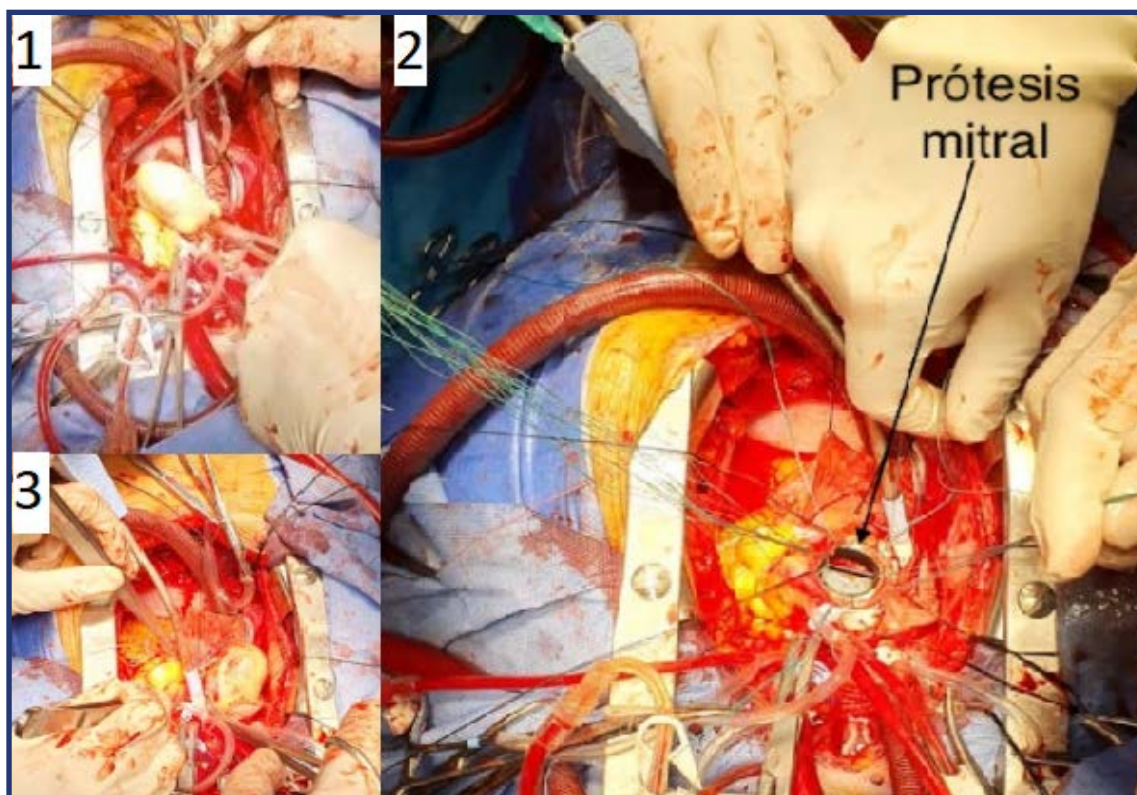
Although when a malignant cardiac tumor is suspected an MRI is suggested, in this case it was not performed, which was not an error per se given that the surgical indication would not have changed. Still, it would have helped estimate the postoperative outcome with greater probability.

What's more, cardiac sarcomas can be treated with preoperative neoadjuvant chemotherapy, although prognosis is still poor. They are highly aggressive and survival of < 1 year even with surgical treatment.⁽⁴⁾

In this case, in a patient with dyspnea at rest, indication of neoadjuvant chemotherapy would have been an illusion based on the clinical signs and not on reality.

Although the incidence rate is low, malignant cardiac tumors should be taken into account when an intracardiac mass is seen in the images, especially if no pedicle can be seen or if the tumor appears to be attached to the mitral valve. Its proper study improves the decision-making process.

FIGURE 2. Intraoperative images. 1 and 2. Mass resection. 3. Replacement of mechanical mitral valve.



Conflicts of interest

The authors have no disclosures to declare.

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