CASE REPORT

Cardiac tumor of infrequent location. A case report.

Karen Ferreyro-Espinosa, MD, Diego B. Ortega-Zhindón, MD, Ignacio Salazar-Hernández, MD, Karen Aguilar-Alapisco, MD, Serafín Ramírez-Castañeda, MD, and Octavio Flores-Calderón, MD.

Department of Cardiothoracic Surgery, General Hospital of Mexico "Dr. Eduardo Liceaga ". Mexico City, Mexico.

Primary cardiac tumors are uncommon, with a general incidence of 0.0017% to 0.27% of the series. Seventy-five percent are benign and 25% correspond to malignant tumors. Myxoma is the most common primary tumor in adults. The left atrium is the most common location. We present a 17-year-old female with a diagnosis of a tumor on the left ventricle outflow tract, with successful

Key words: **Cardiac tumor; Myxoma; Left ventricle.**

Los tumores cardiacos primarios son poco comunes, la incidencia reportada es de 0.0017% a 0.27%, el 75% son estirpes benignas y el 25% corresponden a tumoraciones malignas. El mixoma es el tumor primario más frecuente en adultos, siendo la localización más habitual la aurícula izquierda. Presentamos femenino de 17 años con diagnóstico de tumor del tracto de salida del ventrículo izquierdo, con resección quirúrgica exitosa. En un principio se pensó que pudiera ser un fibroma, resultando ser un mixoma.

Palabras clave**: Tumor cardiaco; Mixoma; Ventrículo izquierdo.**

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Cardiac tumors are a rare clinical condition with significant variability in pathological structure and clinical presentation [1]. A wide diversity of symptoms can be observed, cardiac and non-cardiac related. Therefore, a high level of clinical suspicion must be maintained in diagnosis [2]. As reported in literature, primary cardiac tumors represent from 0.0017% to 0.27% of the series [3]; out of them 75% are benign while the remaining 25% correspond to malignant tumors [4]. Myxoma is the most common primary tumor in the adult population, and rhabdomyosarcoma heading up the list in pediatric patients, followed by fibroid and teratoma [2].

In 1934, the first cardiac tumor was diagnosed by Barnes, who was able to identify a cardiac sarcoma by using electrocardiography and biopsy of metastatic lymph [5]. Beck in 1936 successfully resected an external teratoma of the right ventricle. More extensive intracardiac resections have been feasible thanks to the extracorporeal circulation pump [6]. The first resected left atrial myxoma was in 1952 by Bahnson using caval occlusion, but with unsuccessful outcome. By using the cardiopulmonary bypass, Crafoord resected the first left atrial myxoma in 1954 [7].

Corresponding author: Dr. Karen Ferreyro Espinosa.
email: karenferreyro_21@live.com.mx

We present here a case of myxoma on an extremely rare site of location, the papillary muscle of the mitral valve.

CLINICAL CASE

A 17-year-old female patient with sudden pain, as well as a diastolic murmur in the aortic focus was admitted and studied at our institution. An echocardiogram was performed. A multilobed tumor was reported protruding through the left ventricular outflow tract, causing dynamic stenosis at the level of the aortic valve, with systole dimensions of 41mm x 17mm and with multiple lobes; in addition, to an aneumoid image of 6.7 mm x 9.8 mm in the ventricular septum, the aortic valve with dynamic insufficiency due to tumor protrusion through leaflets, with a maximum gradient 139 mm Hg, mean gradient 81 mm Hg (**Fig. 1**).

Operation was carried on using a standard approach by open chest surgery. Median sternotomy and CPB were used. Ascending aorta bicaval cannulation, and aortic cross-clamping. Custodiol © was administered as cardioplegia. A left atriotomy was performed during cardiac arrest and a single 15mm x 15mm tumor was found near the left atrial appendage with a myxomatous appearance. Next, an aortotomy was performed, with no evidence of direct involvement of the aortic leaflets by tumor. We continued with a left ventriculotomy finding a tumor at the level of the left ventricle associated with the interventricular septum of 6 cm x 4 cm x 2 cm, multilobed with a free aortic valve (**Fig. 2**). The cardiopulmonary bypass

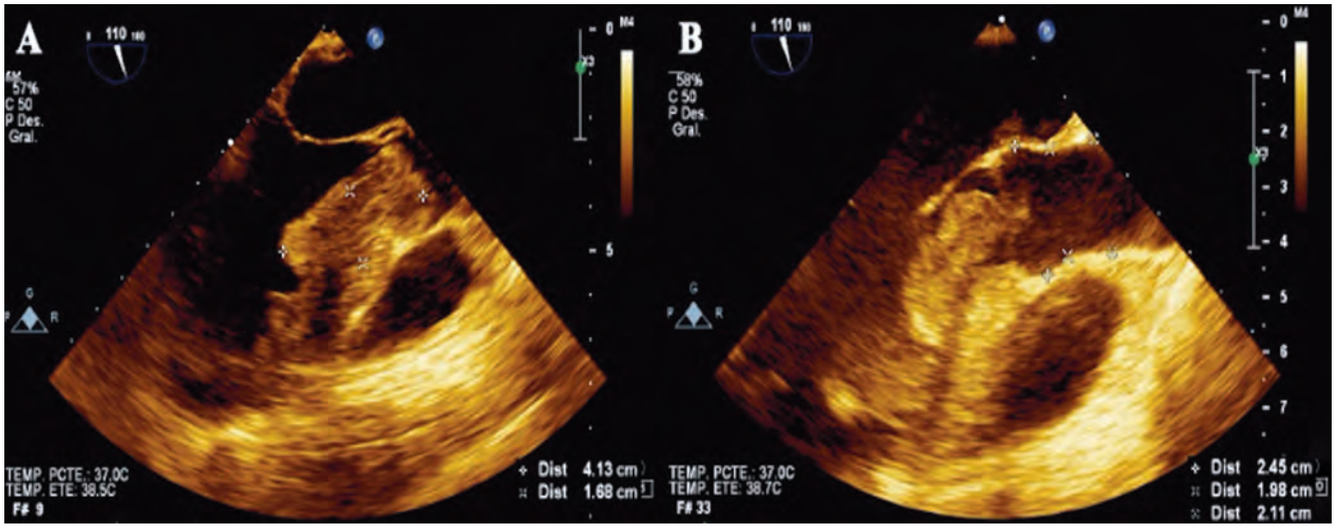

Figure 1. A) Tumor at level of the left ventricular outflow tract. B) Free aortic valve, without tumor involvement.

time and aortic cross-clamping times were 101 minutes and of 86 minutes, respectively. Finally, we proceeded as usual for the rest of operation. The length of stay at ICU was for three days. uneventful, and the in-hospital discharge at 3rd day after operation.

Currently the patient comes to her controls in the outpatient clinic, where histological diagnosis of cardiac myxoma of both tumors is established.

COMMENT

The most frequent location of cardiac tumors is the atrium and right ventricle, followed by the atrium and left ventricle. According to Tzani et al., tumors of the left ventricular outflow tract occupy 4.2% in pediatric patients [2]. In addition, 75% of myxomas develop from the fossa ovalis in the left atrium and only 15 to 20% occur in the right atrium [8].

The average age of onset of myxomas is 50 years. Ninety percent of patients are between 30 and 60 years at the time of diagnosis, being more frequent in women, in a 2.5: 1 ratio [8]. Up to 10% is associated with autosomal dominant genetic mutation with a higher risk of recurrence and the appearance of multiple lesions.

Children and adolescents with cardiac tumors may be as-

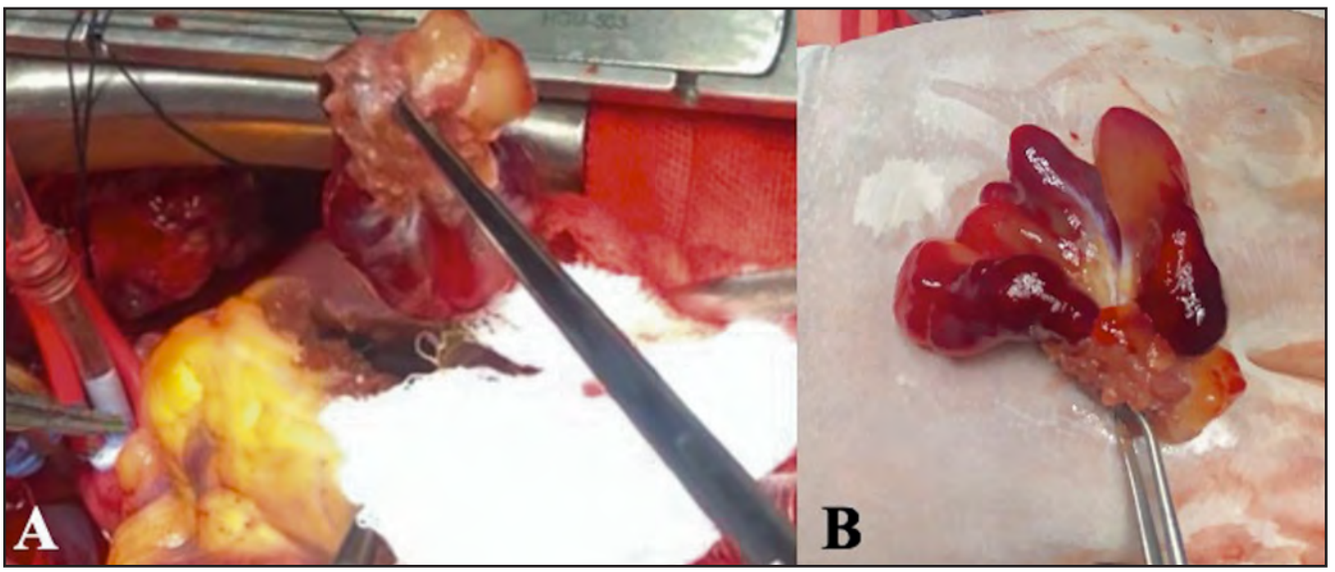

Figure 2. A) Extraction of the tumor through the left ventricle. B) Macroscopic characteristics of the tumor.

ymptomatic or have a range of manifestations depending on the size, location and histological subtype of the tumor. Dyspnea and chest pain are the most frequently referred symptoms. However, they may refer to palpitations that would correspond to ventricular tachycardia in 14.9% of cases. Up to one third of patients can identify a regurgitant murmur, in any case it would indicate the presence of paroxysmal valvular insufficiency caused by the prolapse of the pedicle tumor through the leaflets [2]. They are often diagnosed for the first time after stroke, a peripheral vasculature embolism or a pulmonary artery embolism, caused by detached tumor tissue or by mobilization of thrombotic deposits [3].

Diagnosis and treatment can be challenging. The lack of high specificity in imaging as well as difficulty for biopsy make the diagnosis a challenge [1]. Prior to establishing therapeutic behavior, it is advisable to identify the histological spectrum of the tumor to be treated, since spontaneous regression of rhabdomyomas has been documented in pediatric population, with conservative management as the most appropriate [2,3].

On the other hand, surgical resection should be the first-line in therapeutic options when a pedicle and mobile tumor is evident, since it represents a high risk of embolism and associated complications. The surgical intervention should aim at the total resection of the tumor with disease-free borders, preserving the anatomy and normal function of the heart [2].

Finally, attention is drawn to the location, age and absence of symptoms in the presentation of myxoma in this case herein, in frank contrast with what has been documented in the literature. Since cardiac tumors are so infrequent in the pediatric population, we must always suspect the presence of a previously unidentified cardiac tumor when there is evidence of peripheral embolism. The diagnosis and timely treatment of this pathology is of fundamental importance for the prognosis of these patients.

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