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Obstruction as Unusual Presentation of Cardiac Tumor - Two Cases

Alfredo Mario Naranjo Ugalde[1]*, Gilberto Bermúdez Gutiérrez[1], Elsa Feitas Ruisanchez[2], Caridad Verdecia Cañizares[3], Eutivides Aguilera Sánchez[2], Raquel Maciques Rodríguez[2]

[1]Cardiosurgery Department, William Soler Pediatric Cardiocenter, Havana, Cuba.
[2]Cardiopediatric Department, William Soler Pediatric Cardiocenter, Havana, Cuba.
[3]Oncopediatric Department, William Soler University Pediatric Hospital, Havana, Cuba.

***Corresponding Author:** Alfredo Mario Naranjo Ugalde, Pediatric Cardiocentro William Soler, Ave. 100 y Perla, Altahabana, Boyeros, CP 10800, Havana, Cuba.*

Abstract

Primary tumors of the heart are rare and 80 to 90% of cases are benign. Due to its histological nature, in pediatric age, the most frequent is rhabdomyoma that is associated with tuberous sclerosis, followed by fibroma, teratoma, vascular tumors, myxoma, more frequent in adults and pericardial teratoma. Size, growth rate, and location determine clinical manifestations, time of diagnosis and associated morbidity or mortality.

Echocardiography is essential for diagnosis and decision making.

Symptoms of obstruction to blood flow through the valve orifices, the possibility of causing serious arrhythmias or the possible embolization of or part of the tumor are decisive for the surgical indication.

Two patients are presented, an infant with a diagnosis from the newborn period of tumor in the left outflow tract with clinical expression after the first month of life, which motivates his surgical intervention. The histological study determined that it was a rhabdomyoma.

The second case is a schoolboy whose heart murmur, hepatomegaly was detected with a history of syncope, and when performing an echocardiogram, a large right atrial tumor was detected. After the histological study, a high-grade endocardial myxoid fibrosarcoma was diagnosed, for which chemotherapy treatment was required.

The symptoms prior to the detection of cardiac tumors are not related to their histological nature.

Keywords: *Tumor, heart, surgery.*

Introduction

Development of surgery with extracorporeal circulation after the 1950s, made possible surgical treatment of intracavitary cardiac tumors. The subsequent development of non-invasive echocardiographic diagnostic techniques contributed to early diagnosis and treatment planning. (1-3)

Primary cardiac tumors are infrequent at any age, their incidence varies between 0.0017 and 0.28%. Only 14% of all cardiac tumors appear in children under 16 years old. (4) Those can be primary or secondary (metastatic). (5)

Rhabdomyoma, which is associated in more than 60% of cases with tuberous sclerosis, is the most frequent type in children; followed by fibroma, teratoma, vascular tumors (lymphangiomas), myxoma (more frequent in adults), lipoma, leiomyoma, neurofibroma, ganglioma, and pericardial teratoma. (3, 6, 7)

The clinical presentation does not depend on the type of tumor, a benign tumor can have a malignant behavior due to its growth rate and compromised cardiac function. The clinical debut is determined by size, location of the tumor, its friability, and the possibilities of fragmentation and embolism, such as myxoma, or as the result of the action of substances

released by the same tumor. Heart murmurs, arrhythmias, cyanosis, respiratory distress, and acute heart failure can be identified as the main signs and symptoms of onset. (2, 4, 8)

Between 10 to 25% of primary heart tumors are malignant. (2, 3) Cardiac sarcomas, unlike cardiac myxomas, are similar to those found in other soft tissue locations. Three categories have been described, the most frequent angiosarcomas in the right atrium; tumors based on endomyocardial tissue derived from smooth muscle (leiomyosarcomas) or fibroblastic differentiation, generally sarcomas of the left atrium and those derived from striated muscle or rhabdomyosarcomas, of ventricular location and appearing in pediatric age. They are further classified according to cell type, the presence of necrosis and mitotic activity. (8)

A definitive diagnosis is made by echocardiography. (9) Surgical intervention is indicated in patients who develop symptoms due to hemodynamic compromise. Resection of a benign tumor is usually safe. In the case of malignant tumors, if total resection is not possible, cardiac function should be restored as much as possible and individualized oncological treatment should be incorporated. (9, 10)

The William Soler Pediatric Cardiocenter is the national reference center for the treatment of congenital heart defects. In 34 years, 19 patients with cardiac tumors have undergone surgery. Two patients whose clinical debut were symptoms as a result of obstruction to blood flow are presented.

Clinical Cases

Patient 1

A newborn male was received on suspicion of congenital heart disease due to a heart murmur. Echocardiogram reported a tumor mass with a thick pedicle in the left outflow tract, generating 68 mmHg gradient without clinical signs of hemodynamic repercussion. It was suspected rhabdomyoma. Figure 1.

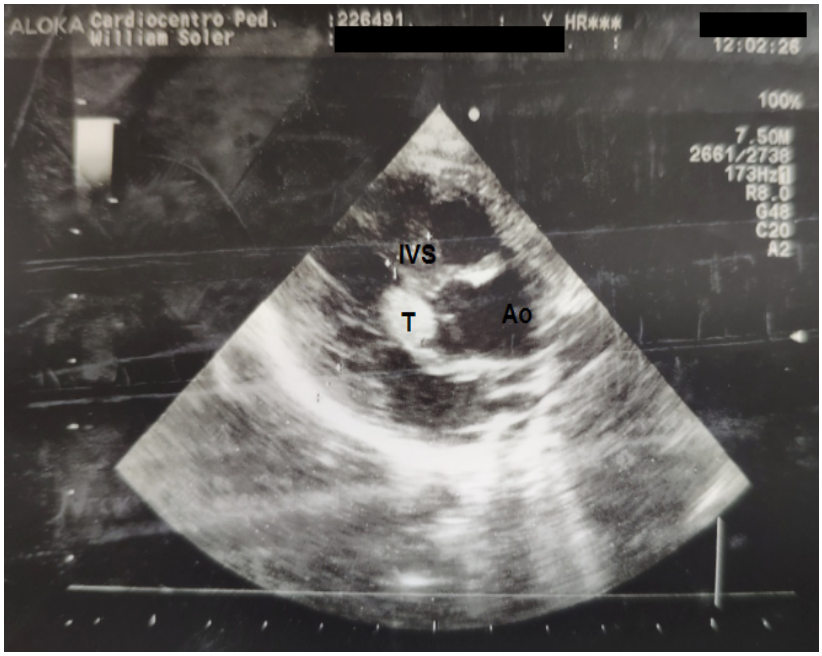

Fig1. Echocardiogram. Patient 1
IVS: Interventricular septum, T: tumor, Ao: aorta

During the observation, he began with profuse sweating and polypnea, with an increase in the intensity of the heart murmur and in an echocardiogram at two months of age, tumor growth is detected that occupies the left ventricular outflow tract and a gradient of 84 mmHg

Surgical treatment and exeresis are then decided. The procedure was performed with the use of extracorporeal circulation for 31 minutes and an anoxic arrest of 14 minutes through the aorta. A round, fibrous, whitish, smooth formation is identified, with a wide pedicle attached to the interventricular septum less than 3 mm from the aortic annulus. Figure.2.

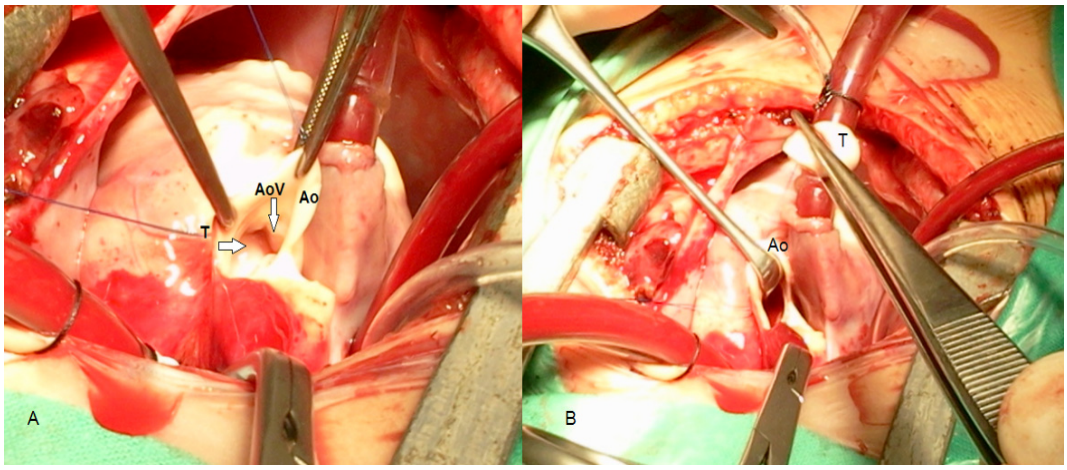

Fig2. Surgical view. Patient 1
T: tumor, AoV: aortic valve, Ao: Aortic wall.

A posterior transesophageal echocardiogram detected the absence of a tumor and slight aortic insufficiency. Six years later, the patient remains without tumor recurrence and without increased valve insufficiency. There is not tuberous sclerosis-associated.

Patient 2

Nine-year-old male patient with a history of malnutrition, the family doctor detected a murmur and sent him to the cardiologist in his health area, a week after, the patient presented a syncope, and echocardiographic study detects a large cardiac tumor and he is referred to our institution.

The symptom referred to the admission was precordial pain to the effort that yields with the rest and physical examination detected a sistodiastolic murmur in tricuspid focus, no hepatomegaly or edemas. Normal blood complements were performed, electrocardiogram with sinus tachycardia without other alteration, chest X-ray light cardiomegaly.

Echocardiogram: tumor mass of large dimensions located at the right ventricular entrance with an obstructive character, impresses with a myxomatous nature, cystic images are visualized inside the tumor, the mass is related to the septal and anterior tricuspid valves to which it adheres dynamic and sessile. Global cardiac contractility conserved usual coronary pattern, normal aortic arch. Figure 3.

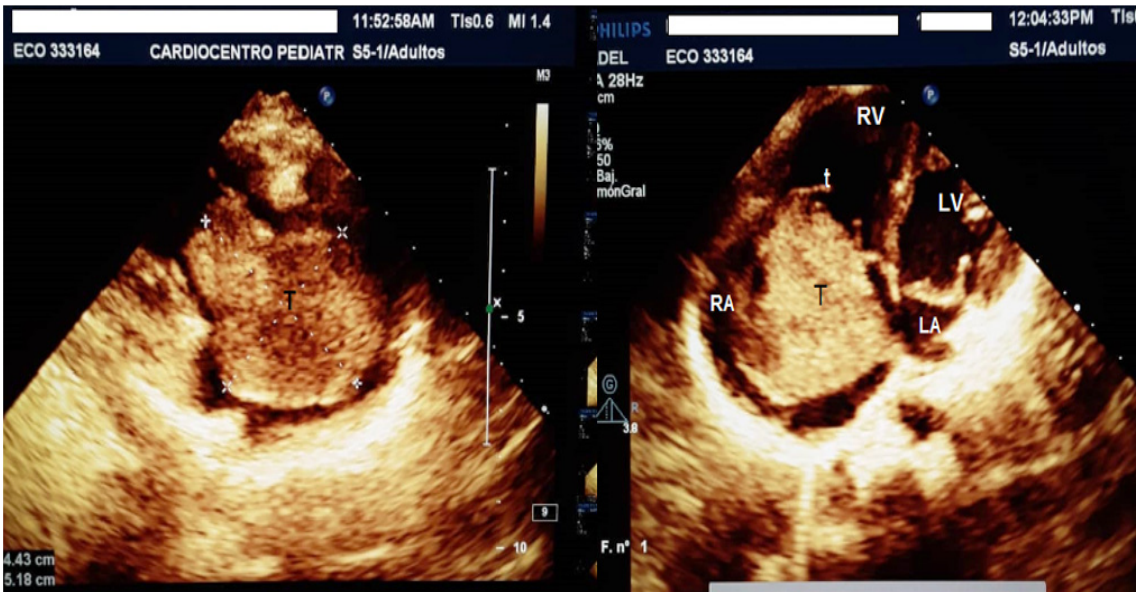

Fig 3. Echocardiogram. Patient 2
T: tumor, t: tricuspid valve, RA: right atrium, RV right ventricle, LA: left atrium, LV left ventricle.

Surgery was performed through the right atrium, with resection of a large, smooth, encapsulated, polylobed tumor with an extracapsular gelatinous lower portion in relation to the tricuspid valve, firmly adhering to the interatrial septum where it was inserted. The endocardial component of the interatrial septum was resected too. Figure 4. There were no trans or postoperative complications. Tricuspid residual insufficiency by echocardiogram was slight.

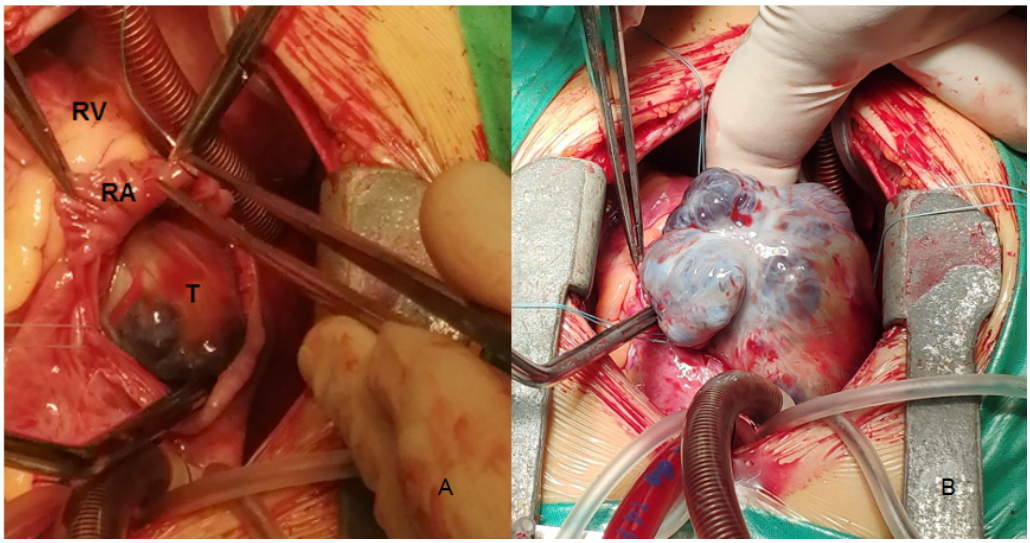

Fig 4. Surgical view. Patient 2.
RV: right ventricle, RA: right ventricle, T: tumor

The patient was transferred to the oncology room on the tenth day postoperatively with favorable evolution. Biopsy reported myxoid fibrosarcoma with moderate cell atypia, presence of 15 to 20 mitoses. There was no invasion of vascular structures. The presence of tumor in the surgical margin was informed. Conclusion: High-grade myxoid fibrosarcoma.

Currently undergoing chemotherapy treatment.

Discussion

Neonatal tumors represent only 2% of pediatric tumors and appear in 1-4 out of 10,000 live births. Evolution differs from those that occur at other ages due to immaturity and rapid cell growth, which could limit therapeutic options. (11)

Patient one had no prenatal diagnosis, the diagnosis was made at birth by murmur and echocardiography. In 15 years, Padalino reports an incidence of surgical indication for cardiac tumors in 8 patients, 3 of them with prenatal diagnosis. (4) In 34 years of work in the CPWS, 19 patients with cardiac tumors have undergone surgery, none with diagnosis prenatal. Patients with a prenatal diagnosis of a cardiac tumor in the CPWS have not required subsequent surgery, probably because they were rhabdomyoma that involves.

In a reported series of cardiac tumors of all ages, the most frequent tumor in the left chambers was mesenchymal and on the right, angiosarcoma. It highlights that the most frequent tumor in pediatric age is rhabdomyoma and also in the left cavities, although unlike the reported case, related to the mitral subvalvular apparatus (7, 12)

The described characteristics of the tumor of patient one coincide with those referred in terms of fibrous texture, smooth surface, white color, related to the ventricular myocardium and the reference to its asymptomatic character except when it obstructs, produces hemodynamic disorder or arrhythmias. (6, 12) At first, the behavior was expectant due to the age of the child and the absence of symptoms, behavior that should be followed until a hemodynamic commitment is demonstrated by the tumor lesion, in addition to ruling out tuberous sclerosis in the evolution; absent in the patient one. (6, 13, 14)

Patients operated on for myxomas show a long-term survival comparable to that of the general population. Total resection for the obstructive rhabdomyoma in LVOT can guarantee a survival that is also comparable to that of the general population. Mortality is mainly related to the histological nature of the tumor, clinic debut, and the duration and general repercussion of these manifestations. (15)

Primary cardiac sarcomas rapidly evolve into myocardial infiltration, obstruction, or distal metastases in the lymph, liver, or lung. Only 1% of the patients with malignant lesions debut with symptoms early. (2) Patient two, with a reported history of poor nutrition, is treated for fatigue and a murmur was detected, then presented syncope. In the latter study, the right atrium was found to be practically fully occupied by the tumor lesion.

As referred in the literature, right lesions manifest with asthenia, edema, peripheral, ascites, hepatosplenomegaly and elevation of jugular venous pressure with prominent a wave. Diagnosis is often late, with an average interval from presentation to correct diagnosis of 2.5 to 3 years, depending on the degree of medical suspicion. (16)

Although incomplete resection of the tumor was not planned by the CPWS team, the biopsy detected tumor at the resection edges. Restoration of normal flow transit is an indication in cases where only partial resection can be performed and completed chemotherapy treatment; heart transplantation has been suggested when it is not yet a metastatic disease. (3, 17-19)

There are many observational studies of cardiac tumors, however, there are no consensus or treatment guidelines for primary malignant lesions of the heart, due to its exceptional nature. According to autopsy studies, metastatic sarcomas in the heart are more common than primary sarcomas. Survival is said to be longer in patients with left lesions and in those receiving postoperative chemotherapy. (2, 9)

The presentation as an obstructive lesion in cardiac tumors is rare or late. Clinical manifestations are not related to the histological nature of the lesion.

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