CASE REPORT

Cardiac Rhabdomyosarcoma

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ABSTRACT

Globally, the prevalence of primary cardiac tumors is low, with a predominance of benign tumors. Malignant tumors, even more rare, present high mortality and low survival rates despite surgery or chemotherapy. Their clinical diagnosis is inaccurate, but there are very reliable diagnostic tools. This report describes the case of a woman with no cardiovascular risk factors or significant family history who presented with moderate exertion dyspnea as the only relevant symptom. Imaging studies showed a mass in the left atrium. Following surgical resection of the tumor, histological analysis revealed cardiac rhabdomyosarcoma. Chemotherapy was indicated in order to improve survival. Also, a review of the literature was made on the diagnosis and management of malignant primary cardiac tumors.

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Key words > Rhabdomyosarcoma – Neoplasm - Heart

Abbreviations >

TEE	Transesophageal echocardiography	**TTE**	Transthoracic echocardiogram

INTRODUCTION

Primary cardiac tumors are an important condition whose prevalence is low. They predominantly constitute benign tumors, as atrial myxomas, which are generally easy to manage and present a low mortality rate. (1) Malignant primary tumors have lower prevalence, sarcomas being one of the most common types, and rhabdomyosarcoma one of the most important due to its poor prognosis and survival. Hence, its early diagnosis might contribute to increase survival rates or obtain greater benefits for patients. (2) This report describes the case of a woman who, after a series of studies and surgical resection, was diagnosed with cardiac rhabdomyosarcoma.

CASE REPORT

We report the case of a 67-year-old woman with type 2 diabetes mellitus, referred for evaluation of dyspnea on moderate exertion of one year duration, unchanged over time and with a negative family history.
Clinical examination showed vital signs within normal range, and no clinical evidence suggesting heart disease.

A transthoracic echocardiography (TTE) (Figure 1) to assess cardiac function revealed a heterogeneous image similar to a tumor. A transesophageal echocardiography (TEE) indicated to obtain an improved image and structural involvement, showed clearly a multilobed heterogeneous mass occupying 65% of the total left atrium.

Once the cardiac tumor was diagnosed, the degree of infiltration into adjacent structures was assessed with a CT scan (Figure 2).

A cardiac catheterization to determine mass vascularization revealed circumflex artery-dependent tumor blush.

Cardiac surgery was performed to remove the tumor for analysis; during surgery, an infiltrating tumor of irregular cerebroid aspect was observed in the roof and lateral walls of the left atrium. After tumor resection (most of the tumor to avoid total involvement of cardiac anatomy), the histological study diagnosed a highly malignant rhabdomyosarcoma, confirmed by immunohistochemistry positive for muscle actin, vimentin and desmin, and by the presence of rhabdomyoblasts.

DISCUSSION

Columbus of Padua was the first to describe a cardiac tumor in 1559, and Barnes was the first to indirectly make a premortem diagnosis of sarcoma in 1934. (3)

Over time, further advances in technology with highly precise and sensitive diagnostic tools, such as echocardiography first and computed tomography and magnetic resonance imaging later, improved the diagnostic procedures. (4)

To manage this case it is important to know that primary cardiac tumors have an incidence of 0.0017-0.35%, 70% of which are benign and 30% malignant tumors. Sarcomas present an incidence of 2-5% (5),

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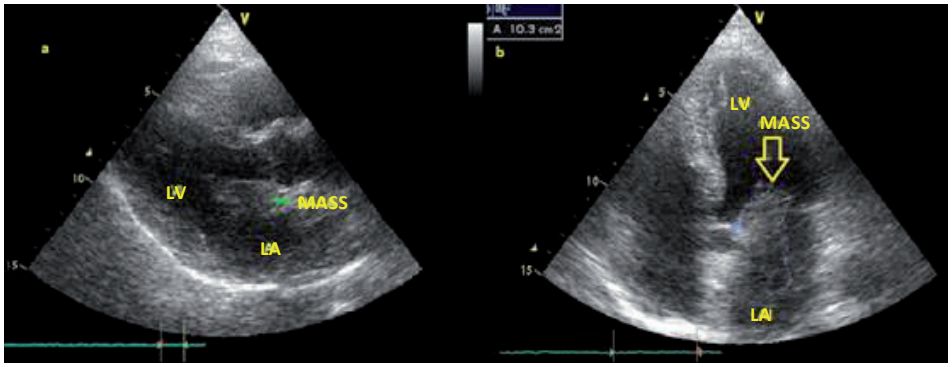

Fig. 1. Transthoracic echocardiography. **a.** Long axis parasternal section. **b.** Long-axis four-chamber view. The image suggests a large mass that involves the atrial septum, anterior mitral valve, and anterior aortic leaflet. **LA:** Left atrium. **LV:** Left ventricle.

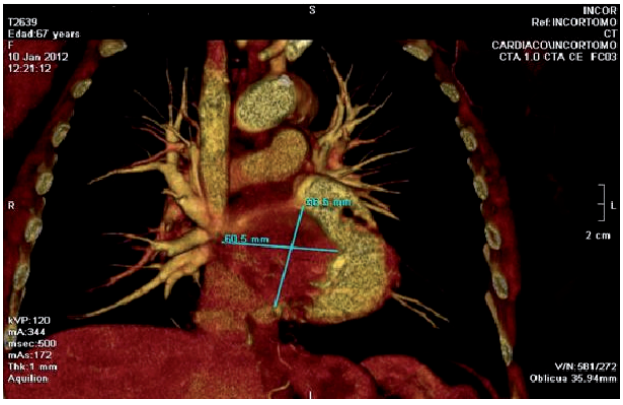

Fig. 2. Multislice helical computed tomography reconstruction showing solid tumor of 60 × 58 × 40 mm in the inferior aspect of the anterior wall of the left atrium.

angiosarcomas and rhabdomyosarcomas being their most common variables, in that order. (2)

Sarcomas differ from benign tumors in the following aspects: (6)

1. They originate mainly in the right chamber, usually in the right atrium, from where they invade other chambers.
2. Their configuration is variable, mostly polypoid.
3. They usually occur in the third, fourth and fifth decades of life.
4. Mostly men are affected.
5. They are characterized by their rapid and infiltrative growth.
6. Sarcomas have a strong tendency for metastasis.
7. Their clinical features vary widely.

Rhabdomyosarcoma is one of the major types of sarcoma in which the left chamber is also involved and it is more common in children and adolescents (75% of cardiac sarcomas in patients under 1 year of age). (7) It may involve several structures (atrial-septal-valvular). (8)

Clinically, sarcomas are asymptomatic until some degree of structural involvement occurs, such as pericardial infiltration causing mostly symptoms of effusion and cardiac tamponade. They may also infiltrate cardiac structures producing arrhythmias or valve lesions leading to obstruction and increased pressures that sometimes involve cardiac function causing symptoms of heart failure. (9)

Symptoms occur in the following order of frequency: dyspnea (61%), chest pain (28%), heart failure symptoms (28%), trepidation (24%), and others (4%). (10)

Initially, diagnosis is made with echocardiography, which attains a high sensitivity of approximately 93% according to some studies. Then TEE is used to determine the degree of structural involvement, with a sensitivity of 96%. It should be mentioned that in these studies, lack of 100% accuracy was due to tumors located in the posterior region of the heart or infiltrating the pericardium, or owing to very small tumors of about 4 × 8 mm. (4, 8)

Angiography is also a diagnostic option when malignancy is suspected due to vascularization or tumor blush, (11) which, by definition, is the angiographic evidence of tumor revascularization during administration of the contrast agent in the catheterization laboratory.

Resection surgery is the course of treatment; (4, 9, 12) and preoperative prognosis depends on variables such as age, level of dyspnea (NYHA) and degree of malignancy. (13) As there is no standard procedure for the surgical technique, most reviews recommend as much surgical resection as possible avoiding loss of cardiac anatomy, while others suggest different alternatives such as autologous transplantation, where the heart is removed to facilitate tumor resection, anatomical repair and reimplantation. (4) There are no comparative studies to demonstrate which is more beneficial.

After resection, adjuvant chemotherapy is recommended because it is believed that it slightly improves survival (9, 12) as the rest of the reviews report a survival rate not higher than 15 months with surgery as the only treatment of malignant tumors (including rhabdomyosarcomas); (13) Bibliographic analysis provides unclear data regarding chemotherapy or radiotherapy prior to surgery to reduce tumor size.

The case of this woman is important considering that these tumors are much more frequent in men of 40-50 years of age and more in children, who are usually clinically deteriorated by metastatic processes

that involve both the left and the right sides of the heart.

CONCLUSIONS

Malignant primary cardiac tumors are a rare but very important condition. Their knowledge would allow early detection, and an adequate understanding of the available diagnostic procedures, such as TTE and TEE, and CT scan and NMR to assess the degree of infiltration, would improve the diagnosis.

Cardiac surgery is the only effective treatment. The surgical technique largely depends on the surgeon's expertise, but it should be borne in mind that the more tumor is removed the better is the patient's prognosis. It is not clearly defined that chemotherapy after resection improves survival, though some reviews indicate that it increases slightly.

RESUMEN

Rhabdomyosarcoma cardíaco

Los tumores cardíacos primarios son una patología de baja prevalencia a nivel mundial, con predominio de los tumores benignos. Los malignos, mucho más infrecuentes, son de alta mortalidad y poca supervivencia a pesar de tratamiento quirúrgico o quimioterápico. Son de diagnóstico clínico impreciso, pero se cuenta con herramientas diagnósticas muy confiables. En esta presentación se describe el caso de una paciente sin factores de riesgo cardiovascular y sin antecedentes familiares de importancia, que solicitó atención médica por disnea a esfuerzos moderados como único síntoma, por lo que se le realizaron estudios por imágenes que revelaron una masa en la aurícula izquierda. Se efectuaron la resección quirúrgica de la masa tumoral y su posterior estudio, en el que se arribó al diagnóstico de rabdomiosarcoma cardíaco. Posteriormente se indicó quimioterapia, con la intención de mejora en la supervivencia. Asimismo, se realiza una revisión de la bibliografía en el diagnóstico y el manejo de los tumores cardíacos primarios malignos.

Palabras clave > Rabdomiosarcoma - Neoplasia - Corazón

Conflicts of interest:
None declared.

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