Case Report

Cardiac Intimal Sarcoma: A Rare Cardiac Tumor Entity

Sarcoma Cardíaco Intimal: uma Entidade Rara Dentre os Tumores Cardíacos

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Introduction

Malignant cardiac neoplasia is a rare entity with an extremely low incidence that has a nonspecific clinical presentation that may mimic other conditions. It can present arrhythmias and complications such as ischemia due to coronary flow obstruction.[1]

More than 75% of primary cardiac tumors are benign. Of the 25% of malignant tumors, 75% are sarcomas.[1-3] Primary cardiac tumors can have a primary or metastatic origin and are diagnosed by imaging methods. Primary tumors occur more frequently in the left chamber, especially in the atrial myxoma, while metastatic tumors originate from malignant melanomas, which occur more frequently in the right chambers.[1]

Cardiac muscle tumors are quite rare entities. Of malignant tumors, cardiac muscle sarcoma is the most frequent histological type and presents aggressively, with dissemination and local invasion potential as well as a high recurrence rate.[1,2] It used to be diagnosed by macroscopic findings at necropsy; however, with advanced imaging methods, today it can be diagnosed and treated earlier to optimize patient outcomes. Treatment consists of lesion excision associated with radiotherapy and chemotherapy.[1,3]

This report describes the case of a patient with cardiac sarcoma to illustrate its echocardiographic presentation pattern to determine whether the association of images and symptoms can lead to an earlier diagnosis, positively impacting the clinical outcome.

Case report

A 44-year-old man suddenly developed a nocturnal cough and palpitations that progressed over 3 days with orthopnea and dyspnea at rest, which made him seek medical assistance in the emergency department of a referral hospital. Upon admission, he was tachycardic and dyspneic and presented clinical signs of hypervolemia and venous capillary congestion on a chest X-ray. An electrocardiogram showed 2:1 atrial flutter (Figure 1). Drug treatment was started with antiarrhythmics and beta-blockers due to suspected tachycardiomyopathy, but the pace was not successfully controlled. Transesophageal echocardiography was performed to evaluate the possibility of electrical cardioversion, which showed an atrial mass of heterogeneous echotexture and lobulated contour, a pedicle located in the interatrial septum measuring 7.7 × 1.5 cm in its largest diameter. The mass had broad movement and entered the left ventricular cavity and the left atrial appendage, obstructing the mitral valve flow, suggesting an atrial myxoma (Figures 2 and 3). The patient underwent urgent excision of the cardiac tumor (Figure 4). The surgical procedure was technically difficult, with possible tumor residue *in loco*. Postoperatively, the patient presented a bradycardic junctional rhythm and episodes of atrial fibrillation with low ventricular response requiring a permanent pacemaker. The histopathological findings of the surgical specimen was compatible with an intimal sarcoma (Figure 5). Cranial and chest tomography were performed for staging; both showed secondary tumor implants. The patient is being followed up at an oncology center with proposed palliative chemotherapy.

Discussion

Primary cardiac muscle neoplasia is a rare pathology with an incidence in the literature of 0.0017–0.28% that has increased in recent decades.[1,2]

Cardiac sarcomas affect more men than women at a 2.5:1 ratio, usually presenting in the third and fourth decades of life.[3]

Intimal sarcoma tend to develop in the right atrium but is sometimes seen in the left atrium. In most cases, it can be confused with benign conditions such as myxoma or thrombus.[3]

Most sarcomas spread very quickly and cause death of myocardium invasion, blood flow obstruction, and/or distant metastases. Therefore, an early diagnosis is essential to successful treatment.[2] Metastatic disease can be present in up to 50% of patients at diagnosis.[4]

This type of tumor presents arrhythmias and conduction system changes more often than myxoma and secondary tumors due to the edema that can occur around the tumor, which is more common in pedicled than sessile tumors.[1]

Its clinical presentation can start with the onset of heart failure and nonspecific symptoms such as chest pain, syncope, and seizure.[1]

The Doppler echocardiogram has relevant sensitivity and specificity to diagnose intracardiac masses and is more often used due to its greater availability and noninvasive nature. This test assesses the size, location, valve leaflet involvement, presence of intracavitary blood flow obstruction, and signs of cardiac muscle invasion. The presence of pedicled or sessile vegetative lesion in one of the valve leaflets or in the valve apparatus can contribute to the diagnosis of cardiac tumor.[1]

Keywords

Sarcoma; Heart; Heart Diseases; Atrial flutter.

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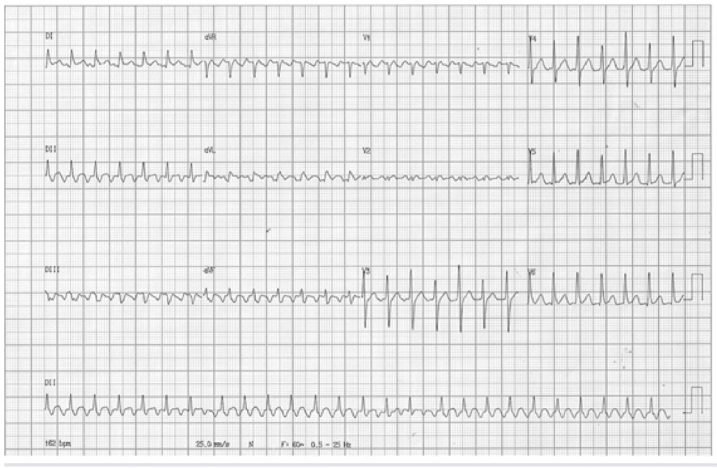

Figure 1 – *Electrocardiogram showing 2:1 atrial flutter.*

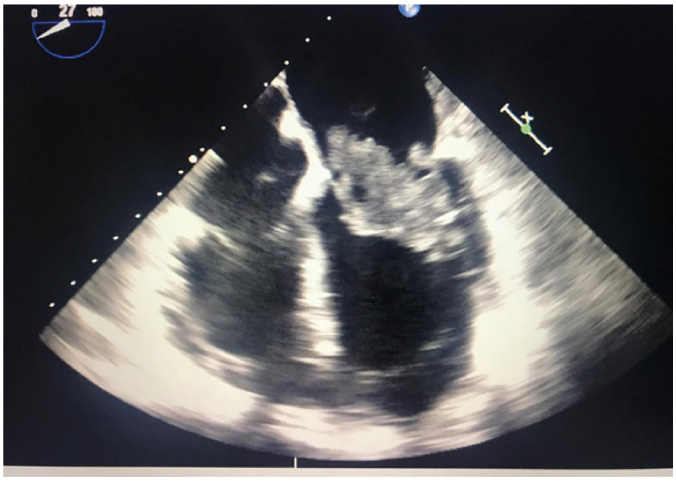

Figure 2 – *Transesophageal echocardiogram in apical four-chamber window view showing a left atrial mass entering the left ventricular cavity.*

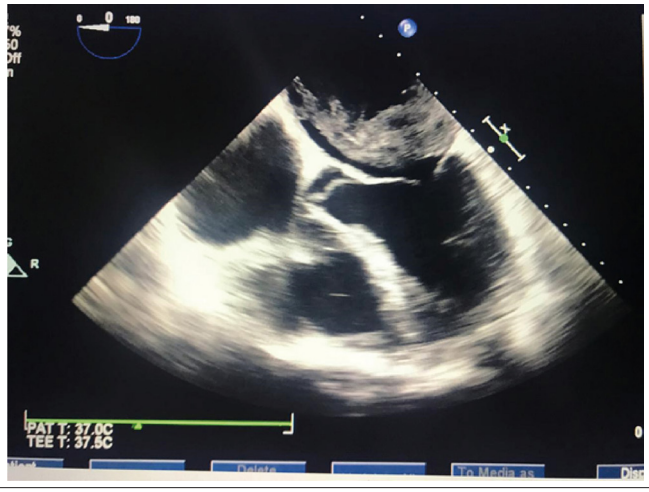

Figure 3 – *Transesophageal echocardiogram in apical four-chamber window view showing a left atrial mass.*

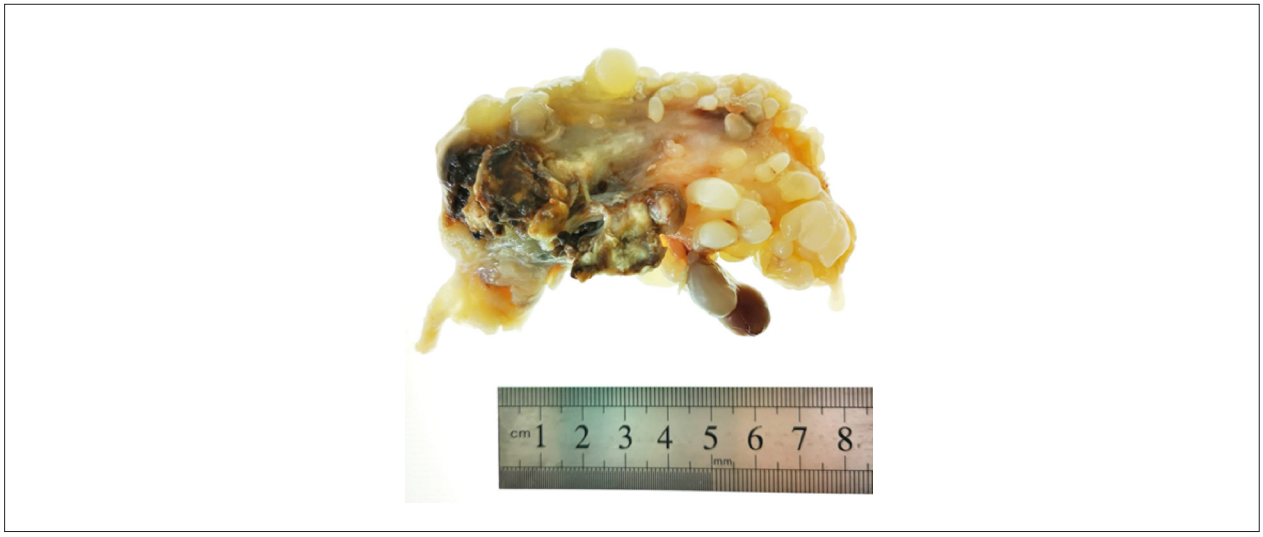

Figure 4 – *Surgical specimen.*

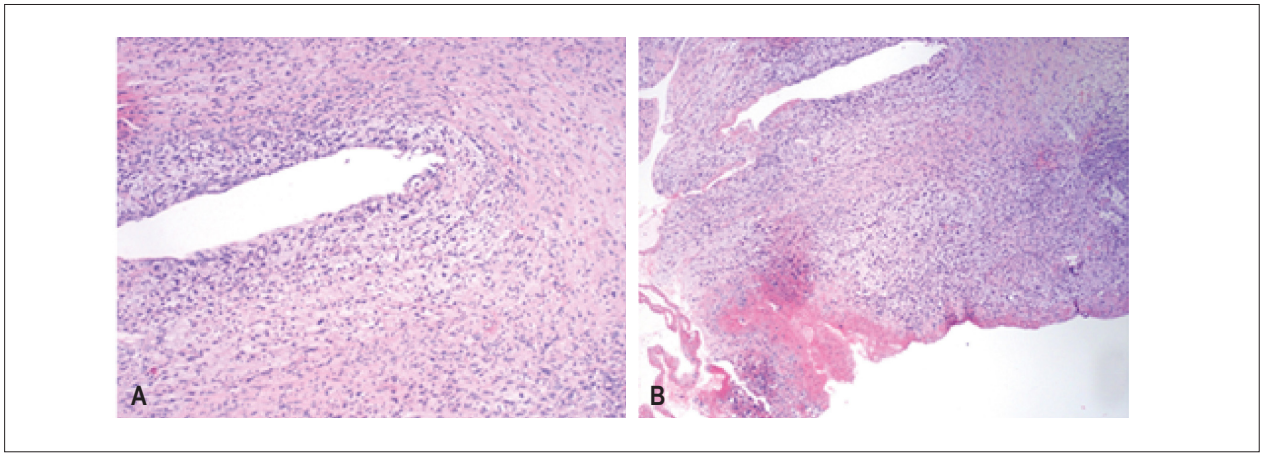

Figure 5 – *A) Low-magnification photomicrograph showing malignant neoplasm with an overlying fibrin layer (below), hypercellular zones, and focal tumor necrosis. Presence of cell densification around the vessel (above), hematoxylin and eosin, 40×. (B) Intermediate-magnification photomicrograph showing round and spindle cell neoplasia with atypia, hyperchromasia, and high mitotic activity in hyaline and myxoid backgrounds, hematoxylin and eosin, 200×.*

Computed tomography of the chest and magnetic resonance angiography provide more accurate information about tumor location, degree of local invasion, and distance and degree of resectability.[1]

Determination of the histological tumor type is important to guiding treatment. In this context, a biopsy is extremely important, which can be incisional, guided by computed tomography; or excisional, by surgical specimen analysis.[1]

Treatment consists of tumor resection, chemotherapy, and radiotherapy, with surgical resection possible in about 30% of cases.[3,5] Even despite complete resection, the disease has a high recurrence rate with a mean survival time of 6–12 months.[2,3,5]

Conclusion

Malignant cardiac tumors are rare entities, but they must be diagnosed early due to their high potential for dissemination and associated complications. In these cases, early clinical suspicion and the use of cardiologic imaging methods is important for timely diagnosis and adequate therapeutic planning.

Authors' contributions

Research concept and design: Andrade RB; data collection: Corrêa LC; data analysis and interpretation: Freitas IA; manuscript writing: Alcantâra ACB; critical review of the manuscript for important intellectual content: Lino DOC, Lima CJM.

Conflict of interest

The authors have declared that they have no conflict of interest.

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