

## Giant Left Atrial Myxoma and Mitral Valve Obstruction: a Case Report

*Mixoma Atrial Esquerdo Gigante e Obstrução da Valva Mitral: Relato de Caso*

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### Abstract

Myxomas are rare entities that represent the majority of benign cardiac tumors in adults. Their clinical presentation may vary by dimension and location, and they can occur in asymptomatic individuals as an incidental finding of cardiac mass from imaging exams. This article reports the case of a female patient with no known comorbidities who was admitted to the emergency department with acute heart failure. Transthoracic echocardiography revealed a mass in left atrium corresponding to a giant myxoma partially occluding the mitral valve, which required urgent surgical intervention.

### Introduction

Primary cardiac tumors are a rare entity. Autopsy series found an incidence of 0.001 to 0.03%, and 75% of these cases are benign tumors. Myxomas are the most frequent tumors in adults, they account for 50–80% in clinical series and 70% in surgical cases.<sup>1,2</sup>

Myxomas mostly comprise single tumors located in the left atrium (LA; 75–80%), followed by the right atrium (RA; 15–20%) and are more rarely related to other structures such as the ventricles and the valves.<sup>3</sup> They commonly present in female adults between 30 and 60 years, with an average age of diagnosis of 50 years. Grossly, myxomas have a globular aspect, with a smooth or slightly lobulated surface and soft gelatinous consistency, with a mean size of 3–4 cm. These tumors are usually attached to the endocardial surface close to the margin of the fossa ovalis by a fibrovascular stalk, with mobility related to the stalk size. Multiple cardiac myxomas are observed in patients with Carney complex.<sup>4,5</sup>

Clinical manifestations may vary and are related to intracardiac obstruction, systemic embolization, and systemic or constitutional symptoms. Dyspnea and orthopnea resulting from pulmonary venous hypertension are common, as are lipothymia and syncope due to transient obstruction of the left ventricular (LV) filling. Embolic events such as stroke, central retinal artery occlusion, and acute ischemia of the limbs can also occur. Systemic manifestations include fever,

arthralgia, weight loss, fatigue, anemia, and increased inflammatory test levels.<sup>2,3</sup>

The following is a report of a young female patient with a large mass inside the LA diagnosed as a giant myxoma causing obstruction of the left ventricular inflow tract, mitral valve regurgitation, pulmonary hypertension, and significant left chamber dilatation.

### Case report

A 48-year-old woman with no underlying diseases was admitted to the emergency department with progressive dyspnea for 30 days associated with orthopnea, paroxysmal nocturnal dyspnea, and lower limb edema. It was also reported episodes of lipothymia and syncope without neurological deficits. On physical examination she had irregular heart rhythm and presented with a systolic murmur in mitral area (3+/6+) radiated to the left axilla, and crepitant rales in both pulmonary bases. Electrocardiogram showed atrial fibrillation with a heart rate of 90 bpm. Chest radiography revealed an increased cardiothoracic ratio, double density sign, third mogul sign, and pulmonary vascular redistribution (“butterfly pattern”).

Transthoracic echocardiography (TTE) showed severe enlargement of the left chambers (LA diameter, 78 mm; LV systolic and diastolic diameter, 67 and 44 mm, respectively), LV ejection fraction of 62%, moderate mitral regurgitation (+3), and a hyperechoic LA mass measuring 97 × 68 mm, attached to the interatrial septum, filling the entire left atrial chamber, with partial occlusion of the mitral annulus, and acceleration of left ventricle flow toward the mitral valve on color Doppler flow examination. Signs of pulmonary hypertension were also seen with a systolic pulmonary artery pressure (sPAP) of 85 mmHg. Cardiac magnetic resonance (CMR) imaging findings confirmed the presence of a large intracavitary mass of 80 × 60 × 90 mm in the LA (Figure 1), isointense in cine–steady-state free precession (SSFP) sequences, discretely hyperintense in double inversion recovery (IR) T1-weighted sequences, hyperintense in triple-IR T2-weighted sequences, with a discrete heterogeneous perfusion during first pass, indicating a solid lesion without hypervascularization, suggestive of LA myxoma (Video 1).

The patient was admitted to the intensive care unit for hemodynamic compensation, and then scheduled for urgent surgery. Surgical procedure was performed through median sternotomy, with cardiopulmonary bypass (CPB) under moderate hypothermia of 28°C. After opening the left atrium, it was visualized a mass filling the left atrial chamber, with an infiltrative aspect over the posterior wall, attached to the interatrial septum. The tumor was resected carefully, followed by a posterior segmental annuloplasty of the mitral valve for

### Keywords

Echocardiography; Magnetic Resonance Imaging; Myxoma.

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annular dilatation repair. Irrigation with saline solution was then performed to remove any debris.

Surgical specimen of 172 g was sent for anatomopathological examination, showing myxoid aspect and gelatinous consistency, without calcification or necrosis. Histological analysis concluded that it was a cardiac myxoma without signs of malignancy. The patient was discharged from hospital uneventfully after 10 days of postoperative and remained asymptomatic at late follow-up, with an 1-year TTE showing signs of reverse cardiac remodeling (LA diameter, 45 mm; LV end-diastolic diameter, 53 mm; and LV end-systolic diameter, 34 mm), trivial mitral regurgitation (+1), sPAP of 30 mmHg, and no evidence of tumor recurrence.

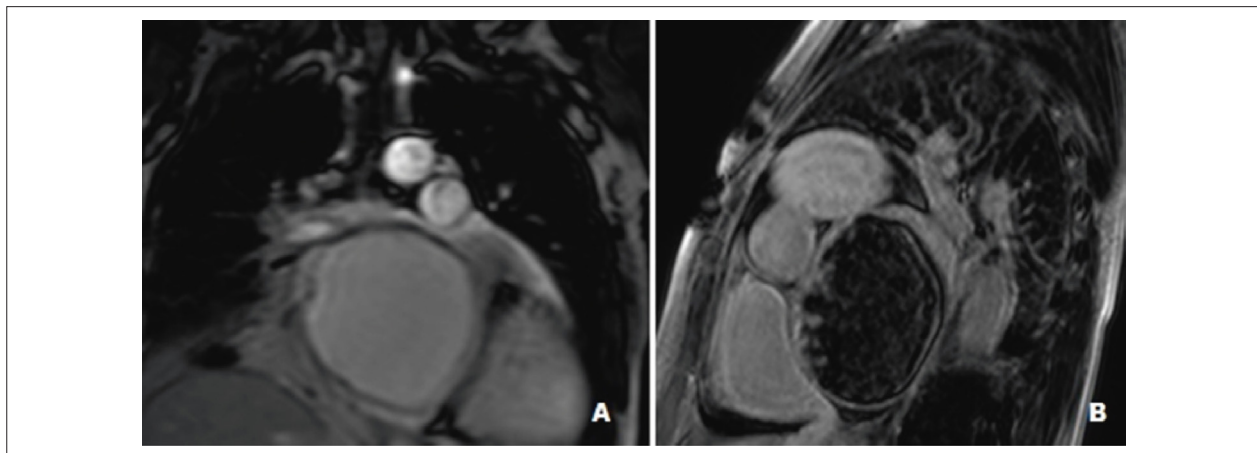
### Discussion

Cardiac myxomas are usually found in young female adults and originate predominantly in the LA. Signs and symptoms of pulmonary venous hypertension must be differentiated from those resulting from heart valve diseases, congestive heart failure, and arrhythmias.<sup>6</sup>

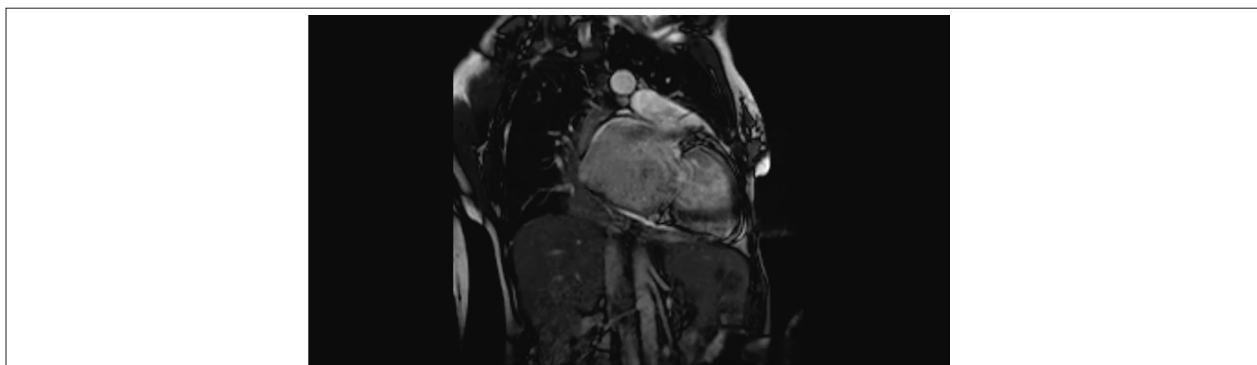
Doppler echocardiography is the most common method used for diagnosis, since it can assess the myxoma size, form, and mobility, as well the location where it is attached.<sup>5,7</sup> CMR can be used additionally to provide valuable information, regarding the differential diagnosis of cardiac tumors. The most common presentation is a mass isointense at T1-weighted and hyperintense at T2-weighted imaging, with foci of hypointensity at one or two of these sequences.<sup>8</sup> Contrast echocardiography can also be used to allow mass differentiation by vascularization analysis.<sup>9,10</sup>

Surgical treatment is normally indicated and should be considered urgent in cases with potential risk of systemic embolization or valve orifice occlusion, a situation seen in atrial myxomas of great dimensions.<sup>2</sup> In present case, the patient underwent surgery before 24 hours since admission, due to clinical deterioration caused by the large myxoma and its hemodynamic consequences. If available, the use of intraoperative transesophageal echocardiography is recommended to optimize the surgical results.

Cajjar et al. suggested some strategies to prevent tumor recurrence, which include minimizing cardiac manipulation to



**Figure 1** – (A) Large intracavitary mass in the left atrium measuring 80 × 60 × 90 mm, isointense in cine–steady-state free precession sequences and discretely hyperintense in T1-weighted double inversion recovery sequences. (B) Hyperintense T2-weighted triple inversion recovery sequences with discrete heterogeneous first-pass perfusion indicative of a solid lesion without hypervascularization and suggestive of a left atrial myxoma.



**Video 1** – Real-time cine cardiac magnetic resonance (CMR). Long-axis two-chamber view showing a giant mass in left atrium partially occluding the mitral valve.

avoid embolization until aortic clamping; resecting completely the myxoma and the adjacent cardiac tissue and the septum, if necessary; removing carefully tumor debris with massive irrigation and suction; and resecting the myxoma intact whenever possible.<sup>3</sup> In our case, it was not feasible to resect the tumor intact due to the extensive infiltration into the atrial wall; however, precautions were taken to remove all debris.

Prognosis after surgery is excellent. Local recurrence is unusual, which is probably related to inadequate tumor resection.<sup>2</sup> The authors believe that the surgical approach should be performed urgently when there are signs of severe hemodynamic impairment.

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## Authors' contributions

Research conception and design: Lucas FC and Veronese ET; data collection: Lucas FC; data analysis and interpretation: Lucas FC, Veronese ET, and Brandão CMA; manuscript writing: Lucas FC, Veronese ET, and Brandão CMA; and critical review of the manuscript for important intellectual content: Brandão CMA, Pomerantzeff PMA, Rochitte CE, and Jatene FB.

## Conflict of interest

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