SCIENTIFIC LETTER

VENTRICULAR MYXOMA: AN UNUSUAL PERIOPERATIVE FINDING

ABSTRACT

Myxomas are benign cardiac tumors predominantly located in the left atrium and are more common in women aged 30 to 60 years. This article presents the case of a 36-year-old woman with a complex medical history who was diagnosed with an incidental finding of an intraventricular myxoma. Imaging studies, including echocardiogram and MRI, revealed cardiac masses. During surgery, lesions were found in the right atrium and a mass in the left ventricle. Pathology confirmed the diagnosis of cardiac myxoma, suggesting a possible association with Carney syndrome due to the patient's history. This case highlights the rarity and complexity of myxomas, underscoring their significance in clinical practice and the necessity for a multidisciplinary approach to facilitate early diagnosis and treatment.

Keywords: *cardiac masses, myxoma, Carney syndrome.*

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INTRODUCTION

Myxomas are benign intracardiac tumors of primary origin; 75% are located in the left atrium (mitral annulus or edge of the fossa ovalis), followed by the right atrium (20%), and simultaneously atrioventricular (5%). Most are asymptomatic, although patients may have obstructive, embolic, and constitutional symptoms.[1,2] Its presentation is more frequent in women from the fourth to the sixth decades of life.[3] About 10% correspond to Carney complex or "myxoma syndrome", generally in young female patients with unusual skin freckles; they are usually ventricular, multiple, and recurrent, associated with non-cardiac myxomatous tumors and endocrine tumors.[4] However, the exact etiology remains unclear. The following is a clinical case of an intraventricular mass that underwent cardiac surgery.

CLINICAL CASE

The patient is a 36-year-old woman with a medical history of hypothyroidism, bilateral hypoacusis, hypoalbuminemia, bilateral superficial venous insufficiency, and chronic pigmentation of the lower limbs under study; surgical history of drainage of a collection in the left buttock and cesarean section; history in first-degree relative (brother) with elephantiasis and bilateral hypoacusis. The patient was admitted to the health institution for presenting with an abscessed cellulitis in the left buttock. With no other findings on physical examination on admission, she underwent drainage surgery. In the postoperative period, she presented grade I edema in the lower limbs, so studies were requested; among them, a transthoracic echocardiogram showed a mass dependent on the interventricular septum, with preserved right and left ventricular function, type II diastolic dysfunction, left atrium with mild dilatation, mitral sclerosis with traces of insufficiency, and mild tricuspid insufficiency, with systolic pressure of the pulmonary artery calculated at 33 mmHg.

Cardiac magnetic resonance imaging was then requested, which identified a mass attached to the interventricular septum, measuring 14 × 14 mm, with a thickening similar to that detected in the right atrium. The presumptive diagnoses were lymphoma, myxoma, or papillary fibroelastoma.

Elective surgery was indicated for resection of the intracardiac mass and pathological study of the specimen.

The pre-anesthetic assessment showed that the patient was ASA 3, with no predictors of difficult airway, high surgical risk, with EuroSCORE II, and an in-hospital mortality risk of 0.62%. In the echocardiographic evaluation, a round intraventricular left mass was visualized, and another smaller one was in the right ventricle.

Under noninvasive vital signs and invasive blood pressure monitoring, conventional anesthetic induction was performed with lidocaine (1 mg/kg), propofol (2 mg/kg), fentanyl (4 mcg/kg), and rocuronium (1.2 mg/kg). Direct laryngoscopy (Cormack-Lehane I) and orotracheal intubation were performed. Anesthetic maintenance was performed with a balanced technique (sevoflurane and remifentanil). Volume-controlled pulmonary ventilation was indicated, with a tidal volume of 6 mL/kg and an FiO2 of 0.45. The surgical procedure was started with the patient on extracorporeal circulation.

Intraoperative transesophageal echocardiography showed preserved left ventricular function, without valvulopathies, the presence of multiple masses, one of 3 cm^2 pedicled and adhered to the interventricular septum, another of smaller size (0.4 cm) in the right ventricle adhered to the septum, infiltration and a possible mass in both inferior and superior vena cava towards the cresta terminalis posterior to the base of the pedicle in the interventricular septum (*Figure 1*).

Surgery was performed by median sternotomy with findings of thickened cardiac tissue, vessels with thickened adventitia, two lesions with irregular borders of 5 × 6 × 3 cm in the right atrium, at the superior cavo-atrial junction and extending towards the appendage, and another lesion on the lateral aspect of 3 × 3 cm; these lesions were on the epicardium but infiltrated the adjacent tissues. No lesions were observed inside the right atrium; in the

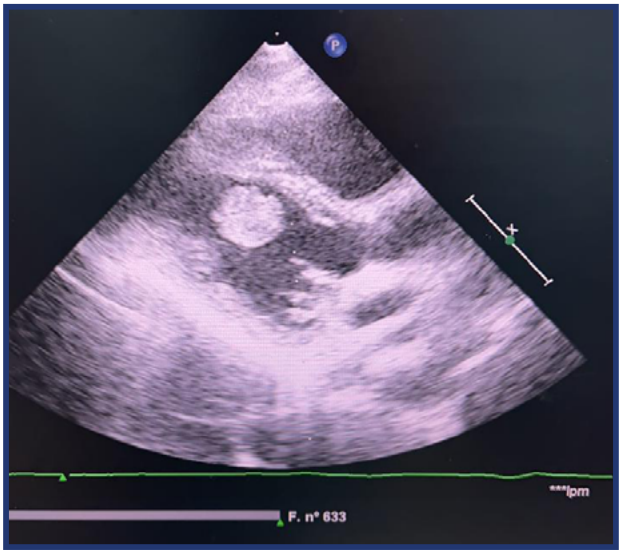
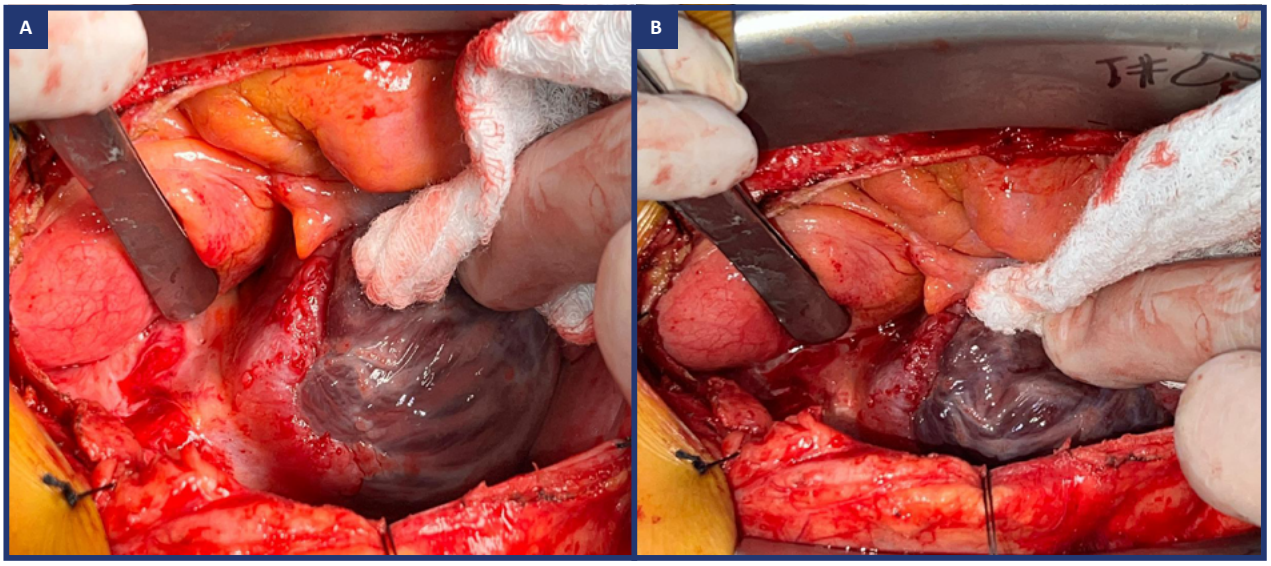


FIGURE 1. Preoperative transthoracic echocardiogram: 3 cm^2 pedunculated mass attached to the interventricular septum.

left ventricle, a mass of 3 × 2 cm was observed adhered to the interventricular septum under the mitral valve, of yellow color and gelatinous consistency. Samples were sent for pathological study (*Figures 2A and 2B*). The lesions in the right atrium were not completely resectable because they infiltrated the entire wall. At the end of the surgical procedure, a bilateral spinal erector block was performed at the T5 level guided by ultrasound with simple bupivacaine 0.5%; the patient was extubated 10 minutes after anesthetic closure, under deep inspiration. The patient was transferred to the intensive care unit with an adequate ventilatory pattern, no pain, and stable vital signs. No intraoperative or postoperative complications were observed, and the patient was discharged from the hospital on the fifth postoperative day. Microscopic analysis showed lesions compatible with cardiac myxoma. Further immunohistochemical evaluation showed a B-cell phenotype (positive for CD20, PAX5, CD79A) of atypical blast lymphoid cells, along with partial positivity for BCL6, IRF4, BCL2, CD30, and C-MYC (<40% of cells). It was negative for CD10, CD5, CD15, ALKc and HHV8, and a very high Ki67 proliferation index (>80%). In situ hybridization for Epstein-Barr virus was positive in a diffuse pattern.



FIGURES 2A AND 2B. Irregular border lesions measuring 5 × 6 × 3 cm at the superior cavo-atrial junction.

DISCUSSION

Intracardiac tumor lesions are infrequent in healthy adults. They can be of primary or secondary origin. The incidence of primary intracardiac tumors is low (between 1.38 and 30 per 100,000 persons per year); 80% are benign, and 20% are malignant. Myxomas account for 50% of benign tumors, followed by lipomatous tumors (21%) and papillary fibroelastomas (16%).

The clinical presentation is variable, ranging from asymptomatic, incidental findings, signs of heart failure and shock, to sudden cardiac death. Influencing factors include the size, mobility, and location of the lesion within the cardiac cavity.[5]

Morphologically, myxomas are usually round, polypoid in appearance, lobulated, and have a smooth surface. Histologically, they contain primitive proliferating cells. On immunohistochemistry, they are positive for vimentin, calretinin, S100, nonspecific enolase, factor VIII, CD31, and CD34.[6]

Approximately 10% of myxomas are associated with Carney syndrome, an autosomal dominant genetic disorder characterized by mutations in the PRKAR1A gene. These tumors tend to present at a younger age, with the involvement of multiple cardiac chambers and have a higher recurrence rate of up to 20%. In addition, thyroid involvement, benign or malignant, has been reported to occur in 25.7% of patients with Carney syndrome and cardiac myxoma.[7-9]

Based on the findings in the literature and the characteristics of the patient presented here (a young woman with thyroid and cutaneous involvement, who was found incidentally to have a ventricular myxoma and participation of the right ventricle, right atrium, and cavo-atrial junction), it can be

concluded that this is an atrial myxoma associated with Carney's complex.

The review and description of this clinical case enable us to update the clinical and paraclinical presentation of this disease, which is a rare and low-incidence situation of interest to the scientific community.

Declarations

The authors declare no conflict of interest.

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