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Cardiac myxoma

Mixoma cardiaco

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cardiac myxoma, cardiac magnetic resonance imaging, echocardiogram.

Palabras clave:
mixoma cardiaco, resonancia magnética cardiaca, ecocardiograma.

ABSTRACT

Introduction: the most common primary cardiac tumors are myxomas, of which 75-85% are located in the left atrium and are prevalent in females. They may present with a triad of obstructive, embolism or constitutional symptoms. **Case presentation:** this study presents the case of a patient who, two years before being diagnosed with a cardiac myxoma, presented dyspnea that subsided with the use of a beta-blocker. After a surgical intervention, the symptoms of heart failure were exacerbated, and an echocardiogram showed a mass covering most of the left atrium and protruding through the mitral valve. **Conclusion:** the transthoracic echocardiogram is the first line of imaging for the diagnosis of myxoma. Magnetic resonance imaging (MRI) provides information on the location, size and characterization of the mass, in addition to helping the differential diagnosis with the presence of thrombus, essential data for the diagnosis approach and patient treatment.

RESUMEN

Introducción: *los mixomas son los tumores cardiacos primarios más comunes, en 75-85% se localizan en la aurícula izquierda y suelen tener predominio por el sexo femenino. Se pueden presentar con la triada de obstrucción cardiaca, embolismo o síntomas constitucionales.* ***Presentación del caso:*** *en este trabajo se presenta el caso de una paciente joven, quien dos años antes del diagnóstico comenzó con disnea de medianos esfuerzos, la cual cedió con el uso de un betabloqueador, posterior a una intervención quirúrgica comienza con datos de falla cardiaca, se le realiza un ecocardiograma donde se evidencia una masa que abarca la mayor parte de la aurícula izquierda y protruye a través de la válvula mitral hacia el ventrículo izquierdo.* ***Conclusión:*** *el ecocardiograma transtorácico es la primera línea de imagen para el diagnóstico del mixoma, sin embargo, la resonancia magnética provee información sobre la localización, tamaño y caracterización de la masa, además de ayudar al diagnóstico diferencial con presencia de trombo, datos indispensables para el abordaje y tratamiento del paciente.*

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INTRODUCTION

Primary cardiac tumors in adults have an approximate incidence of 0.001 to 0.3%, with cardiac myxomas being the most common. In comparison, metastatic tumors of the heart are 30 times more frequent.[1,2]

In 75-85% of cases, cardiac myxomas are located in the left atrium. The development begins in the interatrial septum of the fossa ovalis; 22% develops in the right atrium, 2% in the ventricles, and 1% in the valves, specifically attached to the anterior leaflet of the mitral valve.[3-5]

The transthoracic echocardiogram is the first line of imaging for the diagnosis of myxoma. Magnetic resonance provides information on the location, size and characterization of the mass, in addition to helping the differential diagnosis with the presence of thrombus, essential data for the diagnosis approach and patient treatment.[6,7]

CASE PRESENTATION

A 39-year-old Hispanic female with no medical history of chronic diseases was presented with dyspnea grade two and dry cough, per the New

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York Heart Association (NYHA). The patient was treated with a beta blocker, which relieved the symptoms, until two months ago, when she underwent a laparoscopic cholecystectomy. A few days after the procedure, the patient reported shortness of breath, ankle swelling and lipothymy after physical activities. Initial physical exam revealed jugular ingurgitation grade III/IV, lungs with bilateral wheezing, bilateral pleural effusion syndrome with side predominance, the elevation of heart rate, cardiac murmur in mitral foci and lower extremities with symmetric edema +/++++. The electrocardiogram showed the presence of sinus tachycardia with an electrical axis to the right and mitral p wave determining left atrial enlargement *(Figure 1)*.

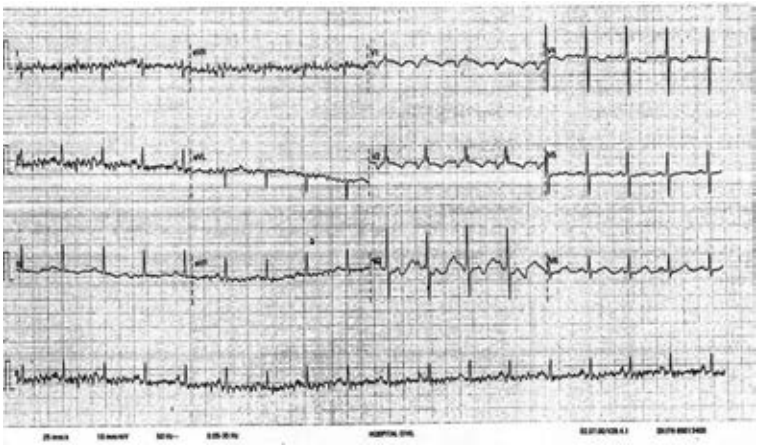

Figure 1: Electrocardiogram. Presence of sinus tachycardia with electrical axis to the right and mitral p wave determining left atrial enlargement.

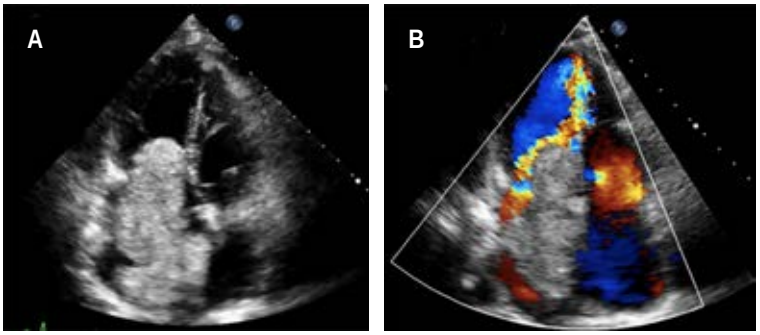

Figure 2: Transthoracic echocardiogram. **A)** Four-chamber diastole showing a mass in the left atrium protruding through the mitral valve into the ventricle. **B)** Color Doppler in four chambers.

A transthoracic echocardiogram showed a preserved ejection fraction. A mass was identified in the left atrium of approximately 52 × 29 mm in systole with slight protrusion into the anterior mitral valve leaflet and left ventricle during diastole, producing severe mitral stenosis. A concentric remodeling of the left ventricle without mobility alterations and both atriums with normal size was observed *(Figure 2)*.

DISCUSSION

Approximately 10% of primary cardiac tumors are malignant, and 90% are benign.[2] Myxomas are the most common benign cardiac tumors, accounting for about 50% of the primary cardiac neoplasms.[2]

Cardiac myxomas can develop at any age, but individuals older than 40 are at an increased risk. Furthermore, females are at a higher risk for cardiac myxomas, with a female-to-male ratio 3:1.[3,7,8]

The World Health Organization defines a cardiac myxoma as a neoplasm composed of stellate to plump, cytologically bland, mesenchymal cells set in a myxoid stroma.[9]

The classic triad of cardiac myxoma are: 1) symptoms due to cardiac obstruction, 2) symptoms due to cerebral or peripheral embolism, and 3) constitutional symptoms. The clinical spectrum depends on the location, size, and mobility of cardiac myxoma.[2,4] 10-33% of the patients are asymptomatic.[7]

The embolic events are present in 30-40% of cases. Intracardiac flow obstruction occurs in approximately 50% of cases, with dyspnea, palpitations and syncope being the most common symptoms. Constitutional symptoms are present in 20-60% of cases.[2,7]

Circulatory vascular collapse and signs of heart failure (dyspnea, paroxysmal nocturnal dyspnea, orthopnea, acute pulmonary edema and pulmonary hypertension) may occur. In those with ventricular location, outflow tract obstruction with syncope and sudden death may occur.[3,7,8] It can also cause damage to the subvalvular and valvular apparatus structures.[9]

The constitutional symptoms, characterized by fatigue, fever, myalgia, arthralgia, lethargy, anorexia, and weight loss, could correlate to the

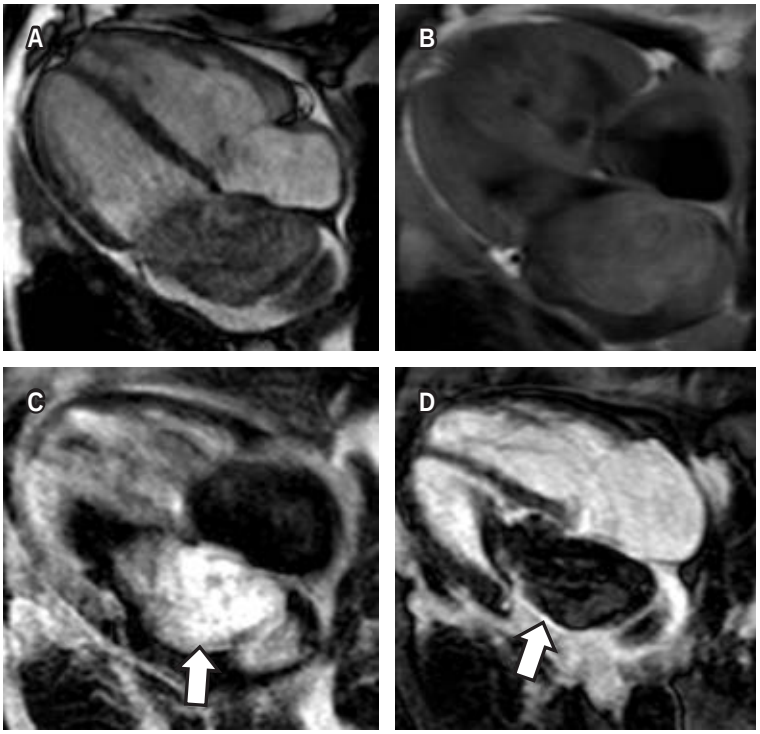

Figure 3: Cardiac magnetic resonance imaging showing left atrial mass. **A)** Cine (SSFP) four chambers. **B)** T1 FSE I-R four chambers. **C)** STIR T2W FSE I-R four chambers showing hyperintense mass suggesting high fluid content. **D)** T1 GRE I-R (late enhancement). White arrows point to hyperintense mass and areas of late enhancement in C and D, respectively.

interleukin-6 cytokine and vascular endothelial growth factor secreted by the myxoma.[2,3]

The thromboembolic event results from tissue fragmentation, detachment of the tumor as a whole and dissemination of overlaying thrombi from the tumor surface, which can result in stroke or ischemic complications.[3,9]

Upon physical examination, a proto-diastolic murmur -also called a tumor plop- may be present and simulates a mitral opening click. In addition, the penetration of the tumor into the ventricle through the valve generates a rumble due to the obstruction of blood flow.[10]

Electrocardiography abnormalities are common but also non-specific, present in 20-65% of the patients. It most commonly shows left or right atrial enlargement or non-specific ST segment abnormalities.[7] In some cases, conduction abnormalities can be presented, including a complete block of the left or right branch, grade one atrioventricular block and atrial fibrillation.[8]

Echocardiography is the first-line imaging modality for myxomas, with up to 95% sensitivity in diagnosing atrial myxoma.[6-8] According to echocardiography findings, there are two different anatomic appearances of atrial myxomas. The first appearance is solid and round with a non-mobile and smooth surface. The second appearance is polypoid, soft, an irregular shape and friable surface, and this one is associated with a higher incidence of embolization.[11] The most common differential diagnosis of atrial myxoma on echocardiography is intracardiac thrombus, which has a homogenous appearance instead of the myxomas' heterogeneous appearance.[6,11]

An MRI provides information with respect to localization, insertion site, and size of the mass. Myxomas typically show a heterogeneous appearance in MRI due to areas of necrosis, hemorrhage or calcification.[11]

On T1-weighted cine images, cardiac myxoma shows as a hypointensity mass relative to the myocardium; meanwhile, on T2-weighted images, it shows as a hyperintensity mass. Gadolinium-enhanced MRI demonstrates contrast enhancement due to high neovascularization, which is an important discriminator from a thrombus *(Figure 3)*.[11]

On cardiac tomography, cardiac myxoma usually appears as a hypodense filling defect in the cardiac cavities. Due to repeated episodes of hemorrhage, dystrophic calcification is common, which tends to be more common in right atrial myxoma.[3]

Tumor biopsy, with histological assessment, remains the gold standard for confirmation of the diagnosis.[6] Cardiac myxoma can be sessile or pedunculated, gelatinous in consistency, and the surface may be smooth, villous, or friable, with a pale gray, white, yellow or brown appearance.[1,5]

The treatment of choice is the prompt, complete surgical excision of the myxoma with its surroundings to avoid tumor recurrence, which is generally curative.[6,7] As rare complications of the surgical excision due to atriotomy, scar can appear atrial fibrillation, atrial flutter or supraventricular arrhythmias.[7]

CONCLUSIONS

Myxoma is the most common primary cardiac tumor, with 75-85% located in the left atrium and predominantly affecting females. They may present with the triad of cardiac obstruction, peripheral embolism or constitutional symptoms.

Depending on the degree of cardiac obstruction, it can cause sudden death. In this case study, the patient had a mass that covered almost the entire left atrium with protrusion into the left ventricle through the mitral valve. However, medication with a beta-blocker decreased the transmitral reflux, attenuating the symptoms of heart failure until its exacerbation after the surgical procedure.

Transthoracic echocardiography is the first line of imaging for the diagnosis of myxoma. However, magnetic resonance imaging provides information on the location, size and characterization of the myxoma. In addition, MRI helps with the differential diagnosis with the presence of a thrombus, indispensable data for the diagnosis approach and treatment of the patient.

The gold standard for diagnostic confirmation is biopsy. The treatment of choice is surgical resection of the tumor, which can be curative with a 10-year survival in 90% of patients. Although cardiac myxomas are rare, the early diagnosis can lead to a better prognosis of the patients.

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