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CASE REPORT

Six-Year Follow-Up of an Octogenarian With a Left Atrial Myxoma: Case Report

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Abstract

Myxoma is the most common primary cardiac tumour, especially in adults. However, reports on this nosological entity in patients over the age of 80 are scarce in the medical literature. Echocardiography is a useful diagnostic tool for detecting cardiac myxomas. These tumours are usually surgically removed soon after diagnosis, limiting knowledge of their natural evolution. In this report, we describe the case of an 86-year-old woman who had been followed irregularly for arterial hypertension since 2016, when an echocardiogram showed a left atrial myxoma adhered to the interatrial septum, initially without impairment of the mitral valve (MV) or obstruction of the pulmonary veins (PV). The patient refused surgical removal at the time. Six years later, she was hospitalized for urinary sepsis. A new echocardiogram revealed an increase in the size of the myxoma, which occupied around 80% of the left atrium (LA), obstructing the PV and MV. New suggestion of surgery was refused by the patient.

Introduction

The first description of a cardiac tumor was made by Columbus in 1559. Among these tumors, metastatic ones are 40 times more frequent. Primary cardiac tumors are rare, accounting for 5% of all cases, with 75% being benign.[1,2] Myxomas comprise approximately 50% of these benign tumors in adults aged 30 to 60 years, with a higher prevalence in women (65%). However, there are few reports in the medical literature involving patients over 80 years of age.[3] Although myxomas can affect any cardiac chamber, approximately 80% occur in the left atrium (LA), 20% in the right atrium, and less than 2% in the ventricles; on rare occasions it is found in more than one cardiac chamber.[4]

From an epidemiological perspective, myxomas can be classified into two categories: sporadic myxomas, which are predominantly solitary and account for approximately 95% of cases, and familial myxomas, which represent about 5% of occurrences and exhibit distinctive characteristics compared to sporadic myxomas.[1,5] These characteristics include onset at younger ages, occurrence in less common sites, multicentric lesions, and associations with rare pathological conditions, along with significantly higher recurrence rates. Therefore, long-term monitoring is crucial to elucidate the recurrence patterns of these tumors.[5]

Patients with myxomas may remain asymptomatic for extended periods, with diagnosis occurring incidentally, or they may present with mild to severe symptoms due to the tumor's slow and variable growth. Therefore, myxomas should be considered in the differential diagnosis of patients with suspected mitral valve (MV) disease. Their location, mobility, architecture, and size (greater than 5 cm or weighing more than 70 grams in the LA) can result in symptoms related to MV obstruction, pulmonary vein (PV) obstruction, compression of the interatrial septum, hematologic alterations, weight loss, and other systemic symptoms.[6-9]

The diagnosis is established through imaging, typically using an echocardiogram (an accessible and cost-effective method) as well as through histological analysis of the tumor following surgical resection. However, the absence of histological confirmation does not invalidate the diagnosis made through imaging.[7] Surgical resection remains the treatment of choice, with a recurrence rate of approximately 3%.[1,6,10,11]

Keywords

Myxoma; Heart Atria; Case Reports

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The natural history of non-operated cardiac myxomas is rarely reported in the medical literature, as they are typically removed shortly after diagnosis, likely due to the associated risks of obstruction and embolism.[10,11]

The authors present the case of an 86-year-old woman diagnosed with a left atrial myxoma, who was followed irregularly over a six-year period and did not undergo surgery due to her refusal.

Clinical case

An 86-year-old woman with a history of hypertension, treated with losartan + hydrochlorothiazide (100/25 mg), irregularly followed as an outpatient since 2016. A routine echocardiogram revealed a left atrial myxoma, but she refused surgical removal. After six years without medical follow-up, she was admitted to the emergency department with sepsis of urinary origin. Physical examination revealed poor general and nutritional condition, a weight of 48 kg, fever (37.9°C), decreased vesicular breath sounds at the right lung base, tachycardia, and normotension. Laboratory blood tests showed hypochromic microcytic anemia and leukocytosis with neutrophilia. The electrocardiogram revealed a heart rate of 118 bpm, a normal axis, atrial tachycardia, and isolated ventricular extrasystoles (Figure 1). A chest radiograph indicated a minimal pleural effusion on the right and an enlarged cardiac silhouette (Figure 2A). Chest computed tomography showed moderate right pleural effusion, small pericardial effusion, and cardiomegaly due to a large expansile mass in the LA (Figure 2B). The patient was admitted for specific treatment (Ceftriaxone 1g and intravenous hydration). On the third day of hospitalization, she developed dyspnea and lower limb edema. A cardiology consultation diagnosed decompensated heart failure (HF). The echocardiogram showed an increase in the size of the myxoma compared to 2016, significant dilatation of the LA at the expense of the heterogeneous and fixed, well-defined, voluminous mass, without vascular flow on color Doppler study, measuring (11.4x8.5x6.8 cm) and occupying more than 80% of the LA, obstructing the PV outflow and compressing the interatrial septum, with MV impairment and left ventricular ejection fraction of 60% (Figure 2C and D).

On the 12th day prior to hospital discharge, the patient and her family were approached regarding the option of myxoma resection surgery, but they refused the procedure again. Consequently, a decision was made to pursue clinical treatment and regular outpatient follow-up. The patient was discharged from the hospital in a stable and asymptomatic condition. At a follow-up consultation 15 days after discharge, she remained asymptomatic.

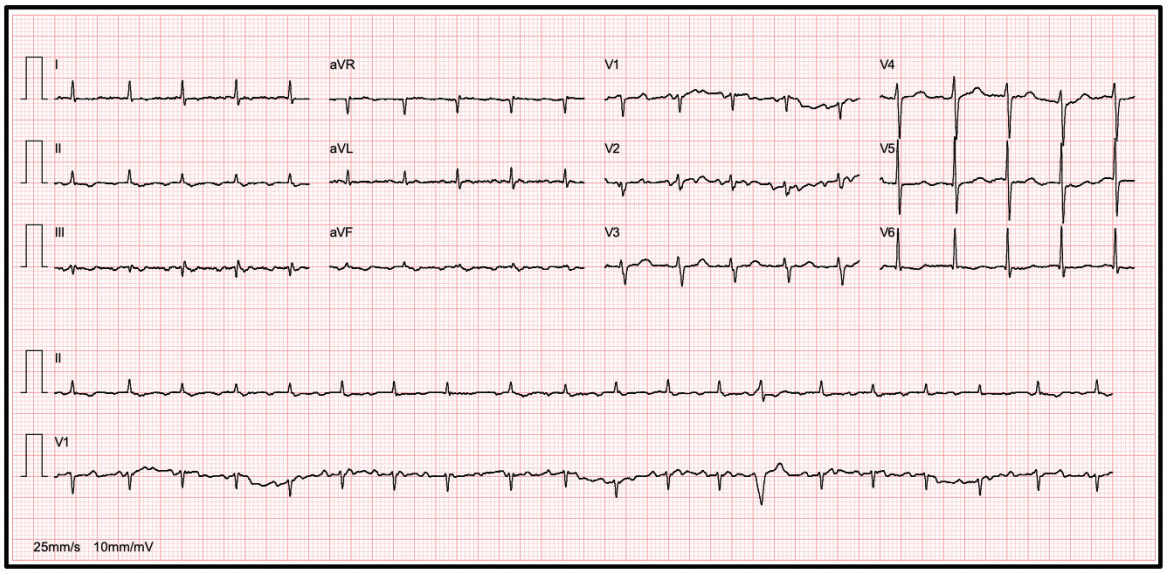

Figure 1 – 12-Lead Electrocardiogram: Heart rate: 118 bpm, atrial tachycardia, normal axis, isolated ventricular extrasystole.

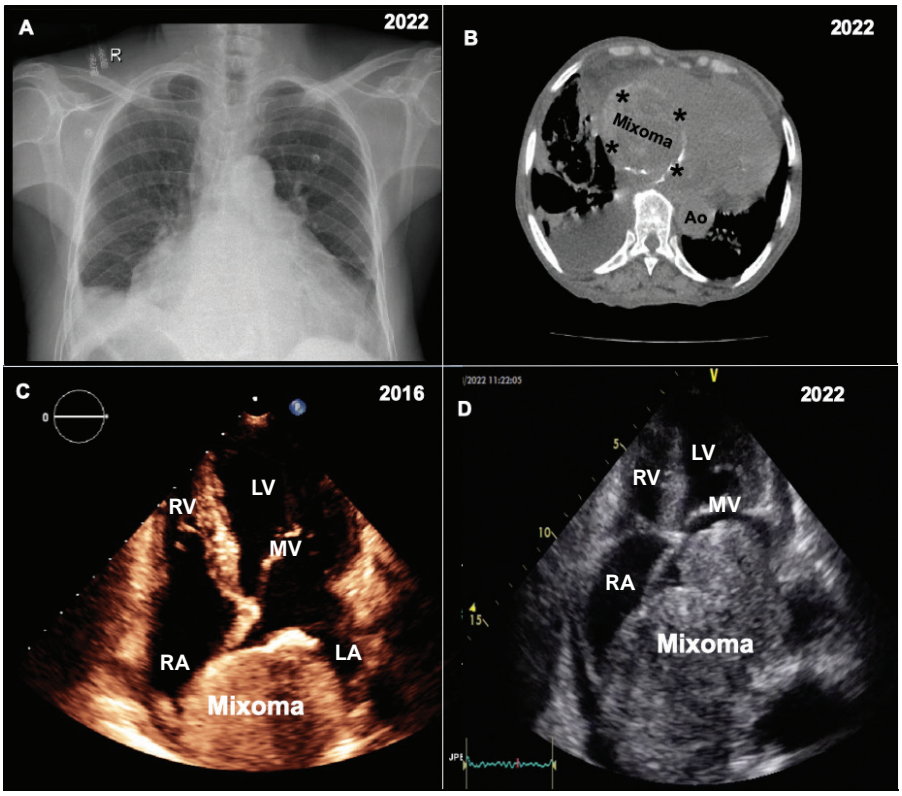

Figure 2 – A) Chest X-ray: An increase in the cardiac silhouette and aortic knob is observed, with blurring of the right costophrenic recess and a double contour sign of the LA. B) Computed Tomography of the Chest: Axial non-contrast slice showing cardiomegaly due to a large myxoma. C and D) Bidirectional Transthoracic Echocardiogram: Apical four-chamber view showing a giant myxoma in the LA, adhered to the interatrial septum without significant obstruction of the MV; the myxoma in 2022 is larger than in 2016, with MV involvement.
RV: right ventricle; LV: left ventricle; MV: mitral valve; LA: left atrium; RA: right atrium.

Discussion

The growth rate of cardiac myxomas before diagnosis is not well understood,[3,9] as most are surgically removed primarily due to the risks of embolism (particularly when appendiculated) and obstruction (due to tumor growth).[1,5,10] This creates a significant gap in understanding the true evolutionary growth process of myxomas over time.

In this case, the growth of the myxoma was monitored through comparative echocardiographic imaging over a six-year period, and it was considered to exhibit slow growth with a non-catastrophic clinical progression, consistent with previous reports on giant myxomas.[3,4,7]

In general, the nonspecific symptoms associated with myxoma contribute to the difficulty of early diagnosis, with most cases being discovered incidentally and at advanced ages.[4,8,10]

The clinical presentation of myxomas is variable; however, the classic symptoms are associated with obstructive, embolic, and constitutional cardiac issues. These may lead to MV obstruction and obstruction of the drainage ostia of PV, resulting in weight loss, pulmonary edema, orthopnea, malaise, syncope, and palpitations. Additionally, systemic embolism can occur, potentially leading to neurological and pulmonary complications as well as sudden death.[7,11]

The definitive diagnosis of a myxoma is established through histological examination following surgical resection, which allows for the identification of myxoid stroma. This stroma is characterized by an oval nucleus, a slightly prominent central nucleolus, and abundant eosinophilic cytoplasm with poorly defined edges.[2,6,7]

Other imaging modalities, such as cardiac computed tomography and magnetic resonance imaging, provide

superior resolution for detecting and differentiating myxomas from other cardiac tumors, including rhabdomyomas, fibromas, lipomas, and papillary fibroelastomas.[10] Despite this, the echocardiogram remains an essential tool due to its accessibility and low cost, exhibiting high specificity and 100% sensitivity for myxoma diagnosis. It allows for the evaluation of various characteristics such as: location, size, mobility, fixation, relationships with adjacent structures, and the presence of potential tumors in other cardiac cavities.[8,11]

The present case illustrated the natural evolution of a left atrial myxoma over a six-year period. The myxoma, which was fixed and adhered to the interatrial septum, remained untreated due to the patient's refusal and initially did not involve the MV or PV (2016). However, by 2022, a follow-up transthoracic echocardiogram revealed an increase in the size of the myxoma and partial obstruction of the drainage ostia of the PV and MV, leading to ineffective left atrial filling and subsequent hemodynamic repercussions.

The slow evolution of myxomas may explain the adaptive/compensatory mechanisms that develop, which are influenced by the absence of specific symptoms over an extended period.[3,9]

This scenario raises several important questions: When is the appropriate time to indicate the resection of myxomas? Should resection be recommended for all myxomas, even when they are small and asymptomatic? The reality is that the risks associated with embolization and obstruction of the LA outflow tract are significant, and surgical intervention should be considered. In the present case, the patient and her family were informed about the importance of the surgery and the inherent risks of not proceeding with the proposed resection; however, they chose to maintain their decision to refuse the operation.

Other cases of myxomas have been diagnosed and successfully treated surgically worldwide.[1,3,6,7,9,11] While diagnosis during the eighth decade of life is infrequent, the prognosis is excellent in most instances.[3,9,11]

The use of intravenous hydration and antibiotic therapy as the foundation for treating the patient hospitalized for sepsis with a urinary focus increased circulating volume. This, combined with the obstructive nature of the myxoma, precipitated pulmonary edema and decompensation of HF, which was successfully managed in a timely manner.

Conclusion

This case is one of the few that provides imaging documentation of the clinical follow-up of an atrial myxoma, which was possible due to the patient's refusal of surgery. Resection of the myxoma following diagnosis typically means that the natural growth evolution over time remains unknown.

Author Contributions

Conception and design of the research, writing of the manuscript: and critical revision of the manuscript for intellectual content Vicente MBA, Amaro T, Fernandes E

Acquisition of data and analysis and interpretation of the data: Vicente MBA.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This study was approved by the Ethics Committee of the Gabinete de Ensino Pós-Graduação e Pesquisa - Clínica Girassol under the protocol number GEPPCG-02/23LDA. All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.

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