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Myxoma Biatrial: A Case Report And Review of The Literature

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1. Abstract

Cardiac tumors are rare, with myxomas being the most common primary tumor in adults, typically benign and found in the left atrium. Myxomas are usually sporadic but can be familial or syndromic. They are more frequent in women and are often asymptomatic, with diagnosis made incidentally. Imaging techniques such as echocardiography, cardiac tomography, and magnetic resonance imaging aid in diagnosis and surgical planning, which is generally curative. This case highlights a rare biatrial myxoma in an asymptomatic elderly patient.

2. Background

Cardiac tumors are rare, with primary tumors having an estimated incidence of 0.017%-0.19%, of which 75% are benign. Myxomas, representing 50% of adult primary tumors, are typically sporadic (90%) but can be familial or syndromic. They are more frequent in women and commonly found in the left atrium, though rarely in both atria [1] [Table 1].Biatrial myxomas are usually independent but can be interconnected by a pedicle or related to the foramen ovale, with symptoms depending on the tumor's location and size, though many patients remain asymptomatic and are diagnosed incidentally. Imaging methods help define the anatomy and guide surgical treatment, with resection being the curative option, effectively preventing embolic complications and minimizing recurrence.This case illustrates the extremely rare incidental diagnosis of a biatrial myxoma in an asymptomatic elderly patient, and the use of cardiovascular imaging methods for diagnosis and therapeutic approach in this clinical context.

3. Case Presentation

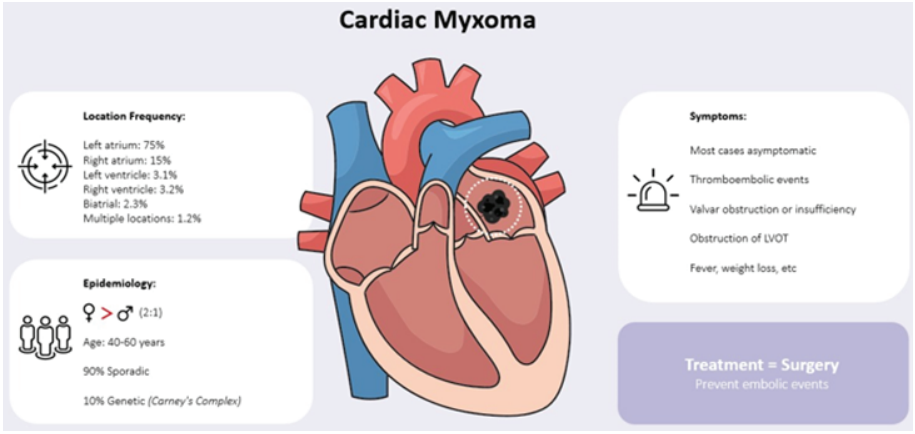
A 74-year-old female, asymptomatic with a history of uncomplicated hypertension treated with losartan, hydrochlorothiazide and amlodipine. The patient underwent routine cardiologic exams for clinical follow-up.The electrocardiogram evidenced a first-degree atrioventricular block with no other alterations.A Transthoracic Echocardiogram (TTE) revealed a mobile echogenic image in the Left Atrium (LA), pedunculated, adhered to the interatrial septum, in the region of the fossa ovalis, measuring approximately 3.1x1.4 cm, suggestive of atrial myxoma, without signs of obstruction to the mitral transvalvular flow. The maximum gradient between LA and Left Ventricle (LV) was estimated at 7.0 mmHg and the mean at 2.0 mmHg. A pediculated echogenic moving image was also observed in the Right Atrium (RA), adhered to the interatrial septum, in the region of the oval fossa, measuring approximately 3.6 x 1.7 cm, suggestive of atrial myxoma, without signs of obstruction to the tricuspid transvalvular flow. The maximum gradient between RA and Right Ventricle (RV) was estimated at 5.0 mmHg and the mean at 2.0 mmHg (Figure 1).A Transesophageal Echocardiogram (TEE), perfomed pre operatively to aid in surgical planning, showed extensive hyperechogenic images in both atria, with ill-defined limits, adjacent to the interatrial septum, suggestive of myxomas. Both presented prolapse movement into the ventricular cavities, however, without signs of obstruction of the atrioventricular valve flows (Figure 2).Cardiac Magnetic Resonance imaging (CMR) (Figure 3) and Coronary Angiotomography (CT) (Figure 4) were performed. The studies confirmed the echocardiographic findings and the CMR allowed myocardial tissue characterization. The CMR images showed a mass of heterogeneous appearance, with mild hypersignal on DOUBLE IR sequence, hypersignal on T2 and mild first-pass perfusion (Figure 3).Surgery was performed to excise the tumor and repair an iatrogenic atrial septal defect via AD (patent foramen ovale wasn't present). Intraoperative findings revealed the tumors with a small and narrow pedicle adhered to the oval fossa (Figure 5). The histopathological examination confirmed the diagnosis of myxoma.The patient presented symptomatic sinus bradycardia in the immediate postoperative period, requiring a temporary pacemaker for 72 hours. She was discharged on the 6th postoperative day without further complications.In the clinical follow-up, to date (three months after surgery), the patient is asymptomatic, with echocardiographic control without alterations.

Table 1: Location according to the frequency of cardiac myxomas

Cardiac myxomas location	Frequency
Left atrium	75%
Right atrium	15%
Right ventricle	3,1%
Left ventricle	3,2%
Biatrial	2,5%
Multiple locations	1,2%

4. Discussion

From 1967 to 1998, only 19 cases of biatrial myxomas were reported in the world literature, with a slight increase in its incidence in the last 20 years, possibly due to the evolution of cardiac imaging techniques.Myxomas are more frequent in women and are generally diagnosed around the fifth decade of life. The most frequent location is the LA, at the level of the fossa ovalis [2].Once diagnosed, surgical treatment should be briefly considered due to the known complications described in untreated patients [3].Multiple location myxomas, especially biatrial, are extremely rare (2.5%) and are usually not properly diagnosed, being mostly confused with unilateral myxomas.It is relevant to consider the association of cardiac tumors with Multiple Endocrine Neoplasms (MENs) at the time of diagnosis and treatment. Associations with complex genetic syndromes that include multiple endocrine neoplasms type 1 and 2, von Hippel-Lindau syndrome, neurofibromatosis type 1, and Carney Complex (CC) are described.Familial autosomal dominant syndromes, such as CC, have an atypical anatomic distribution, with onset around



Cardiovascular Images

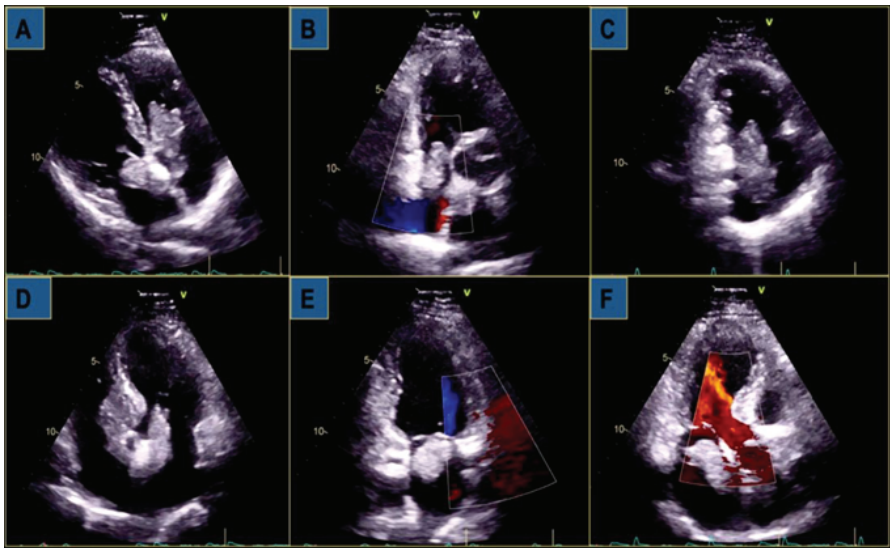


Figure 1. Transthoracic Echocardiographic (TTE) Images depicting the biatrial myxoma in the modified parasternal long axis view (A) and transversal short axis view of the great vessels (B). TTE images of the myxoma in the right atrium (right ventricule inflow view) (C). Biatrial myxoma in the apical 4-chamber view (D) and myxoma in the left atrium in the apical 2-chamber view (E) and longitudinal view (F).

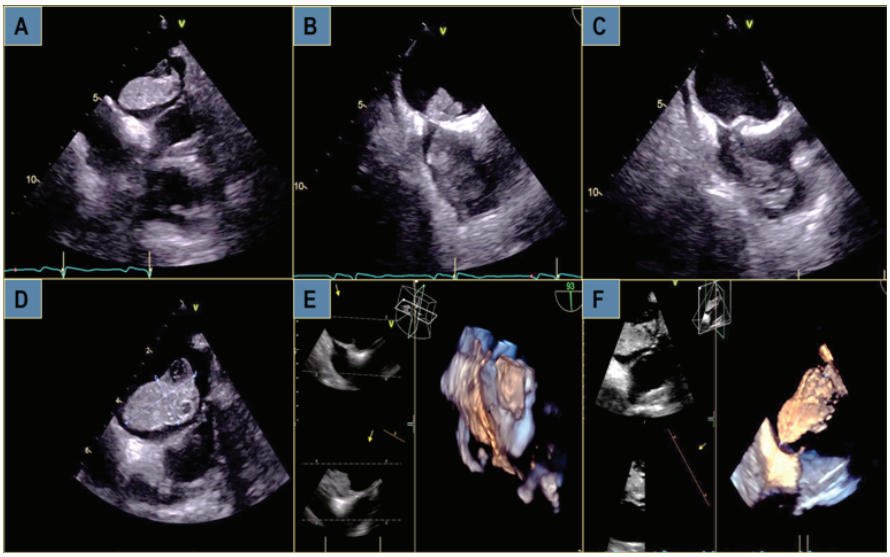


Figure 2. 2D Transesophageal Echocardiographic (TEE) Images depicting the biatrial myxoma (A) and the myxoma in the right atrium (B) and left atrium (C) in the bicaval view. 2D TEE image of the myxoma in the left atrium in the apical 4-chamber view. 3D TEE image of the myxoma in the left atrium in the transgastric 4-chamber view (E). 3D TEE images of the myxoma in the left atrium en face (E) and in bicaval view.

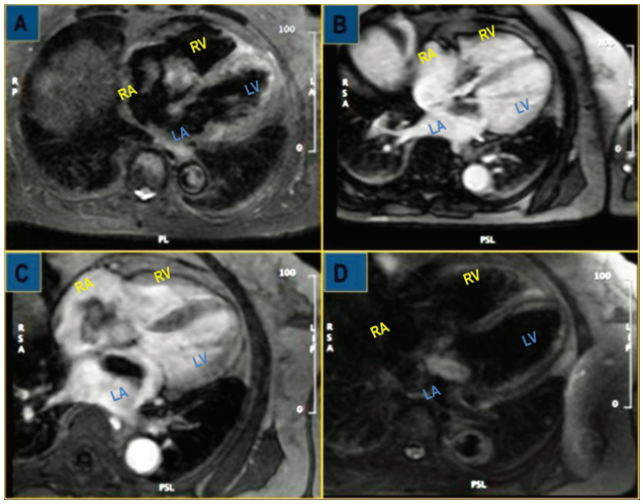


Figure 3. (A) T2 Weighted (Triple IR) image shows a mass in the right atrium with diffuse heterogeneous hypersignal, typical of myxoma. (B) Rest perfusion reveals a mass in the right atrium with minimal and diffuse perfusion, and a mass in the left atrium with no perfusion. (C) Late gadolinium enhancement (LGE) with TI 600ms shows a mass in the left atrium with heterogeneous and patchy enhancement, and a mass in the right atrium with no late enhancement. (D) T2 Weighted (Double IR Fat-Sat) demonstrates a mass in the right atrium with hypersignal and no fat component.

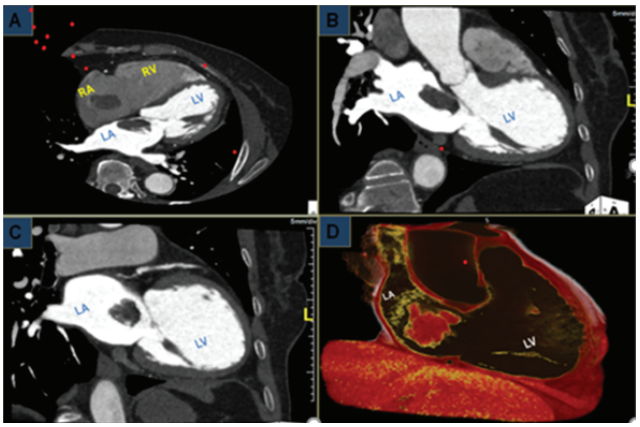


Figure 4. Contrast-enhanced coronary computed tomography images showing mass in LA and RA in the 4-chamber longitudinal view (A) and in LA in longitudinal view of LV outflow tract (B) and 2-chamber longitudinal view (C). Three-dimensional reconstruction in longitudinal view showing myxoma in LA (D).

25 years of age, tending toward multiple location in 45% of cases, associated with mutation in the type I protein kinase A regulatory subunit gene - PRKAR1A gene [4].To date, it is controversial whether there is an increase in complications and mortality rates in multiple location myxomas.Myxomas are derived from pluripotent mesenchymal cells of the subendocardium4 and often present as polypoid or pedunculated lesions from the endocardium, in contrast to other cardiac tumors, such as lipomas or rhabdomyomas, which are not pedunculated. In addition, most myxomas present as gelatinous, smooth, rounded masses with a shiny surface.In most patients, myxomas are asymptomatic like the patient described in this case report. Accordingly, the patient presented an incidental diagnosis after the age of 70, making the case even more peculiar. The clinical presentation is related to the size and location of the tumor.Myxomas located in the LA, can present similar symptoms to those found in patients with mitral stenosis or tricuspid stenosis and in tumors located in AD.Syncope and sudden death, arrhythmias and

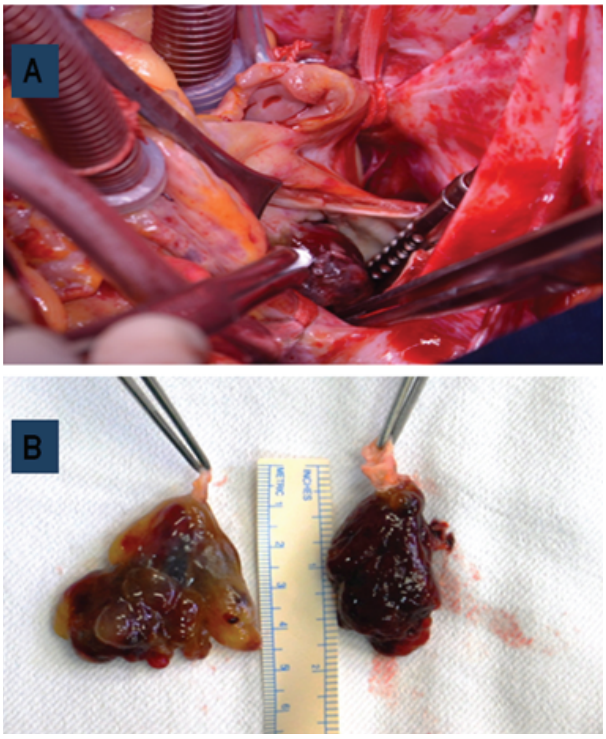


Figure 5. Exposure of myxoma in left atrium during surgical treatment (A). Macroscopy of resected myxomas during surgery (B).

embolization may occur due to detachment of tumor tissue or thrombi associated with its surface [3]. The most frequent symptomatic triad includes heart failure, embolization and nonspecific symptoms. Indirect or nonspecific symptoms are arthralgias, myalgias, asthenia, muscle fatigue, fever, and Raynaud's phenomenon (associated with autoimmune mechanisms due to the increase in serum interleukins from the tumor tissue).Transthoracic echocardiography is currently the best method of cardiovascular imaging because it consists of a noninvasive technique, widely available and relatively low cost [6,2]. TTE allows anatomical, morphological, and functional cardiac evaluation, besides direct visualization of the presence of cardiac masses, their location and possible hemodynamic changes related to the tumor.In the case of this patient, TTE was requested for a better anatomical definition of the biatrial myxoma and its relationship with the atrial septum for surgical planning purposes.CMR, when available, allows tissue characterization and description of the perfusion profile of the tumor mass, providing additional information compared to echocardiography in the sense of guiding the histological origin of the tumor[2].The treatment of choice is surgery, which has become curative in most cases. The main technique is complete resection of the tumor and surrounding tissue in order to avoid recurrence. Although infrequent, recurrence can occur from a few months after surgery to decades after, and rates range from 1% to 5%, and are more frequent when associated with familial myxoma. The familial form should be suspected in any patient with a history of recurrent cardiac myxoma. In these cases, multidisciplinary clinical follow-up of the patient is also indicated, as well as first-degree relatives presenting with a clinical presentation suggestive of mutations in the PRKAR1A gene, due to the high recurrence rate (15-22%) reported in CC cases [4].During the postoperative period, most of the complications reported are associated with supraventricular arrhythmias due to atrial manipulation [3], besides the risk of recurrent interatrial communication in cases of biatrial myxomas.

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