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Stenosing Intracardiac Mass of the Mitral Valve: A Case Report and Review of the Literature

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Benign intracardiac tumours are the most common of the 5% of primary tumours and account for 90% of intracardiac tumours. Myxoma, which is the main benign tumour, rarely localizes to the mitral valve, in the order of 1-5%, associated with severe symptoms and enormous complications.
The objective is to report a rare and severe case of mitral valve myxoma resulting in severe obstruction of the valve orifice.

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Presentation of Case: A 65-year-old Moroccan woman, without profession, the diabetic patient presented with progressive dyspnoea, in whom transthoracic echocardiography (TTE) completed by transesophageal echocardiography (TEE), a cardiac magnetic resonance imaging (MRI), a cerebro-throracoabdominopelvic Positron emission tomography-scan (PET-scan) showed a cauliflower-shaped mass embedded in the small mitral valve, evoking the diagnosis of myxoma, confirmed by the anatomopathological examination. A lumpectomy with mitral valve plasty was performed in association with coronary artery bypass surgery for a tri truncal lesion. The evolution was marked by an improvement in the clinical and echographic state.

Discussion: Myxoma is the first benign tumour encountered in women between the 3rd and 6th decade, whose diagnosis is evoked by a TTE, better by a TEE or more, by a computed tomography (CT scan) or even a cardiac MRI which specifies the visualization of the soft parts with all the internal details of the myxoma, whose confirmation is carried out on the histopathological analysis of the operating room.

Conclusion: The management was based on complete resection of the tumor associated with mitral valve plasty.

Keywords: Stenosing intracardiac mass; mitral valve; intracardiac myxoma.

1. INTRODUCTION

Intracardiac masses are represented by pseudotumours, which are the most common, and cardiac tumours, which are rare. Secondary cardiac tumours are 20 times more common than primary tumours, which account for only 5% [1,2]. Four and twenty percent of primary tumours are benign and 10% are malignant. Of the benign tumours, myxoma is the most common, accounting for 20-40% of intracardiac masses [3]. The cardiac myxoma often has a smooth, irregularly shaped, heterogeneous surface and is embedded in the endocardium by a stalk of less than one centimeter; 85% pedunculated and 15% sessile [1,3]. It occurs twice as frequently in women as in men and is often asymptomatic. The presence of symptoms depends not only on the size of the tumour but also on its location, thus requiring mainly surgical management. The most frequent location is the left atrium in 83% of cases and the least frequent location, associated with serious complications such as obstruction or unexpected sudden death in 67%, is the mitral valve in 1-5% of cases [4]. We report the case of a myxoma of the mitral leaflets causing obstruction of the valve orifice.

The objective of our study is to report a rare and severe case of mitral valve myxoma resulting in severe obstruction of the valve orifice.

2. PRESENTATION OF CASE

A 65-year-old Moroccan woman, without profession, diabetic on metformin 1000mg divided into two doses, was admitted in September 2020 in the cardiology department, for aggravated dyspnea. Clinical examination and electrocardiogram (ECG) were normal.

The Holter ECG showed ventricular and supraventricular hyperexcitability. Cardiac ultrasound showed an echogenic mass embedded with calcareous magma in the small non-vascularized mitral valve on color Doppler measuring 53 x 45 mm, with a gradient on the mitral valve of 12 mmHg and a very large left atrium, in pulmonary hypertension. Transesophageal ultrasound showed a cauliflower mass embedded in Posterior 1, 2 (P1, P2) responsible for the mitral stenosis with attachments to the left atrium and ventricular wall.

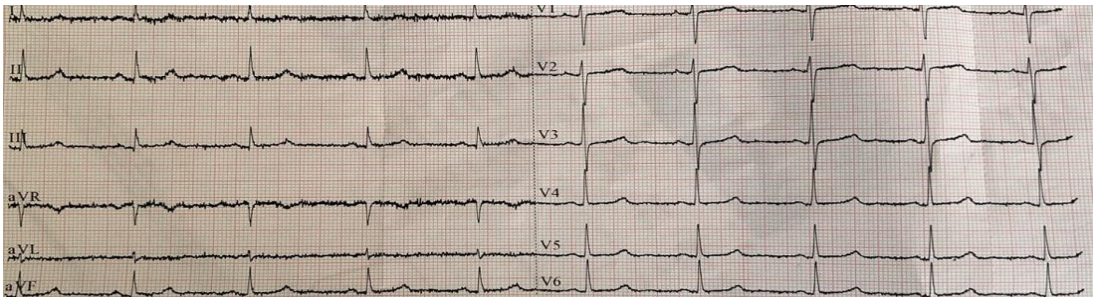

Fig. 1. Normal 12-lead electrocardiogram

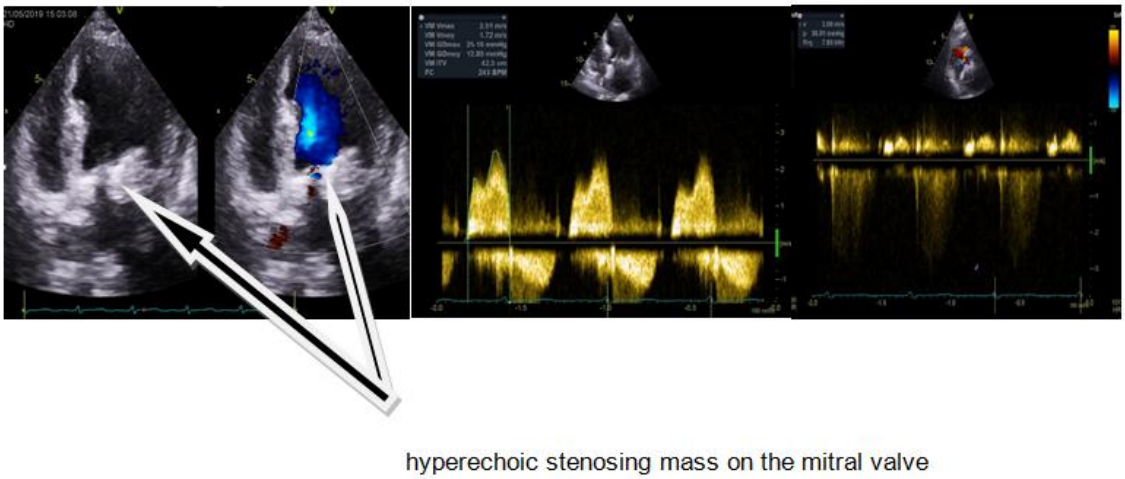

Fig. 2. An echogenic mass in the small mitral valve indicating mitral stenosis

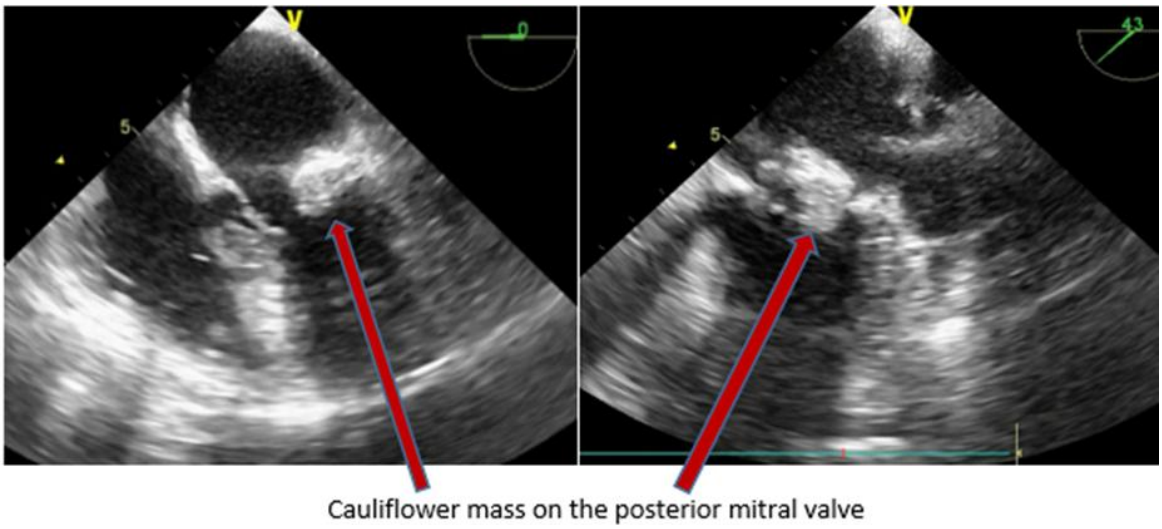

Fig. 3. Cauliflower mass embedded in P1, P2

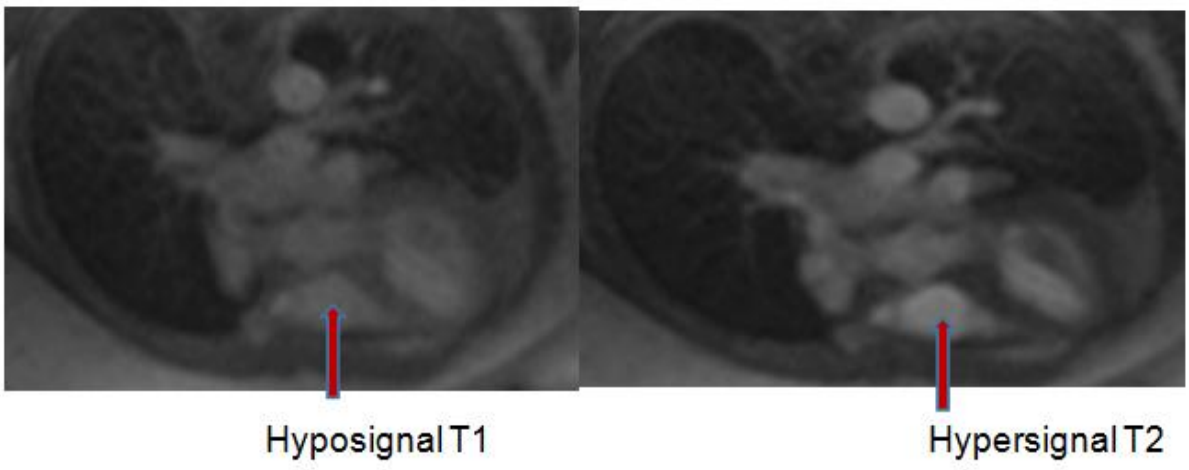

Fig. 4. Cardiac MRI, showing a tumour without infiltrating the left atrial wall

On cardiac MRI, a tumour process, creating a barrier to filling without infiltrating the wall of the left atrium, of the left ventricle with T1 hyposignal, T2 hypersignal and heterogeneous enhancement especially on its peripheral part was objectified.

An extension work-up of the mass was performed: a throracoabdominopelvic computed tomography (CT scan) was performed, showing only calcification of the mitral valve and then completed by a cerebral-throracoabdominopelvic Positron Emission Tomography, showing an intracardiac mass compared to the mitral valve, calcified hyper-metabolic associated with a hyper-metabolic area adjacent to the mitral valve without visualization of the lymph nodes.

In view of the risk factors for atherosclerosis, a coronary angiography was performed as a preoperative check-up, showing a tritruncal lesion.

A complete lumpectomy under extracorporeal circulation with mitral valve plasty was performed, associated at the same time with a triple aortocoronary bypass.

The surgical specimen was analyzed by histopathology, morphological examination showed a myxoid tumour proliferation compatible with cardiac myxoma with the expression of calretinin by these tumour cells on the immunohistochemical study.

The evolution was marked by an improvement of the clinical and echographic symptomatology with very close echographic controls.

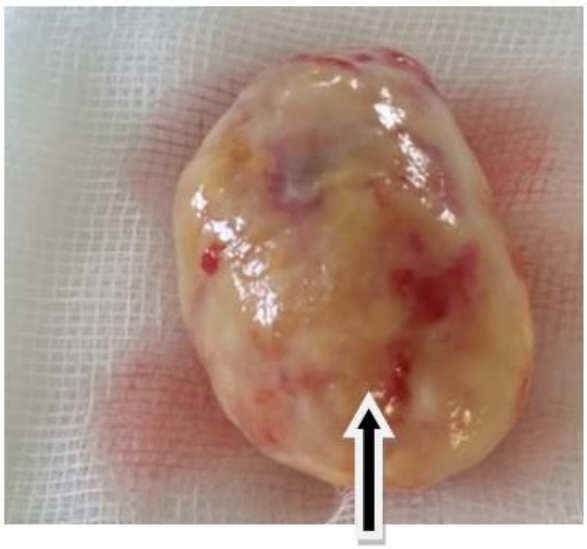

Fig. 5. Round mass resected from intracardiac origin

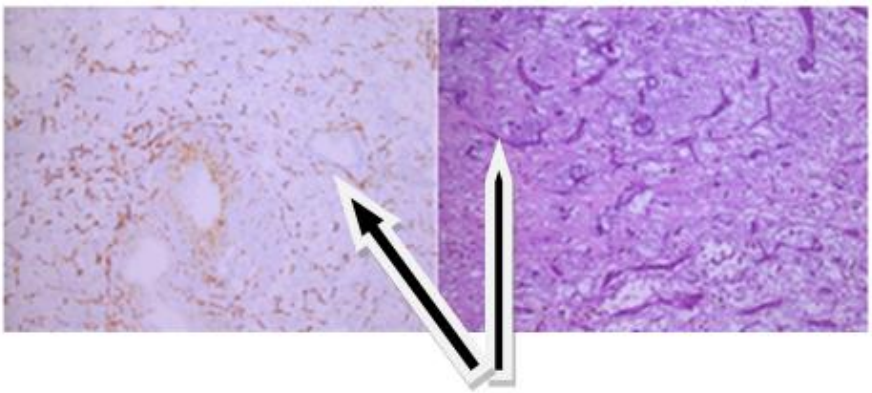

Fig. 6. Immunohistochemical Image On The Left And Morphology On The Right In Favour Of A Myxoma

3. DISCUSSION

Intracardiac masses attached to the mitral valve are mainly represented by pseudotumours made of vegetation and then thrombus and tumours. Primary cardiac tumours are the least numerous compared to secondary cardiac tumours, representing 5% of cardiac tumours [2]. Their incidence is in the order of 0.0017-0.03% [5] of which the most frequent are benign tumours in 90% of cases with their leader, the intracardiac myxoma in 20-40% of cases [2,3]. Other benign tumours are: fibroelastoma, lipoma, haemangioma, and rhabdomyoma [6]. Malignant tumours are less frequent, in the order of 10%, dominated by sarcoma and primary cardiac lymphoma.

Women are twice as likely to develop myxoma as men, and it occurs from the third to the sixth decade of life [7,8], which explains why in our study it was a myxoma occurring in a woman in her sixth decade. It can have a sporadic origin, which is the most frequent, but it can also be autosomal dominant in 7% of cases, thus giving Carney syndrome, linked to anomalies of the second chromosome. Other manifestations of Carney syndrome include cutaneous myxomas, skin hyperpigmentation, and endocrinopathies [4,9]. In our case, the patient had no pathological history of myxoma in the family and no other myxoma location; The sporadic origin seems to be the most obvious. The area of predilection for cardiac myxoma remains in the left atrium at 75-85% versus 18% in the right atrium. It may sit on the oval borders of the interatrial septum in 64.7%; 11.9% on the mitral annulus and only 1-5% on the mitral valve as in the case presented in our study with blood flow obstruction on the mitral tract [3,10]. The cardiac myxoma has a smooth, irregularly shaped, and usually heterogeneous surface, sessile in 15% of cases and in 85% pedunculated, embedded in the endocardial wall by a stalk of less than 1 cm [1].

The clinical manifestations of myxoma depend on the size, architecture, and location of the tumour [7]. In general, the smaller the size, the more asymptomatic the myxoma, as described by Pinede et al. in 112 cases of left atrial myxoma studied, examined macroscopically, and then correlated by size and symptoms. A correlation between myxoma size greater than 5 cm and the occurrence of symptoms was found to be statistically significant (P = 0.009) [1]. Symptoms include signs related to atrioventricular valve obstruction such as dyspnoea, syncope, angina pectoris, and vertigo suggestive of heart failure, arrhythmia, or pericardial effusion as well as systemic and pulmonary embolization [11,12]. Systemic embolization occurs in 30-40% of cases of stroke-like atrial myxoma [13]. In our case, the obstruction of the mitral valve by the myxoma and the accompanying symptoms were the focus of our clinical case in relation to its size which was greater than 5 cm (5.3cm x 4.5cm) but also its location, present on the small mitral valve.

The diagnosis of atrial myxoma is based on several diagnostic modalities of choice [14]. Transthoracic echocardiography (TTE) is the examination of choice to differentiate between an intracardiac pseudotumour and a pericardial or intracardiac tumour, while specifying its characteristics. However, transesophageal echocardiography (TEE) outperforms TEE in the description and detection of intracardiac tumours or pseudotumours despite their size, as reported by Shyu et al. In which 17 patients were examined for intracardiac tumours using both TTE and TEE. In 14 patients, who had undergone cardiac surgery, TTE had 4 false positives and 2 false negatives in contrast to TEE which had only one false positive and no false negatives. Thus, TEE showed superiority over TTE not only in detecting small masses but also in specifying their locations, fixations, and dimensions.

Other modalities such as CT and cardiac MRI are better than ultrasound due to their main advantage in visualizing with sharp accuracy the soft tissue with all the internal details of the myxoma, as well as identifying its attachment. Confirmation of the diagnosis is based on histopathological analysis of the surgical specimen. In our case, as reported in the literature, the diagnosis was evoked by TTE, TEE, cardiac MRI and even positron emission tomography. Only histopathological examination has accurately confirmed the diagnosis.

The management as reported in the literature [15] is based on complete resection of the tumour under extracorporeal circulation after right atriotomy and septotomy completed by mitral valve plasty. Early mortality is less than 5%, in-hospital mortality is zero. The risk of recurrence is 1 to 4% in sporadic myxoma. It is higher in complex Carney forms. All this is consistent with our clinical case, in which the management was done as in the literature and again in association with aortocoronary bypass

surgery associated with a favorable short- and medium-term evolution.

4. CONCLUSION

Intracardiac myxoma is the most frequent benign tumour whose diagnosis requires the use of several diagnostic tools and surgical management must be early, even more so when the location is valvular because the complications are serious.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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