

**MYXOMA: OTHER DIAGNOSIS OF HIGH MITRAL GRADIENT THAN MITRAL STENOSIS**

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**ABSTRACT**

Myxoma is the most common primary tumor of the heart and can arise in any of the cardiac chambers. This paper reports a 65 -year-old hypertensive and menopausal woman, hospitalized for exertional dyspnea since ten months with significant weight loss. Transthoracic echocardiography showed a left atrial myxoma, prolapsing across the mitral valve into the left ventricle causing a mean gradient of 24 mmHg with severe pulmonary hypertension (97 mmHg). The patient underwent successful resection of the myxoma with good outcome.

**KEYWORDS:** Myxoma, Cardiac tumors, mitral valve gradient, systemic embolization,**INTRODUCTION**

Myxomas are benign cardiac tumors that are mostly (75%) located in the left atrium. Most myxomas present with one or more of the constituents, embolic or obstructive manifestations,<sup>[1]</sup> and can mimic severe mitral valve stenosis and cause severe pulmonary hypertension.<sup>[27]</sup>

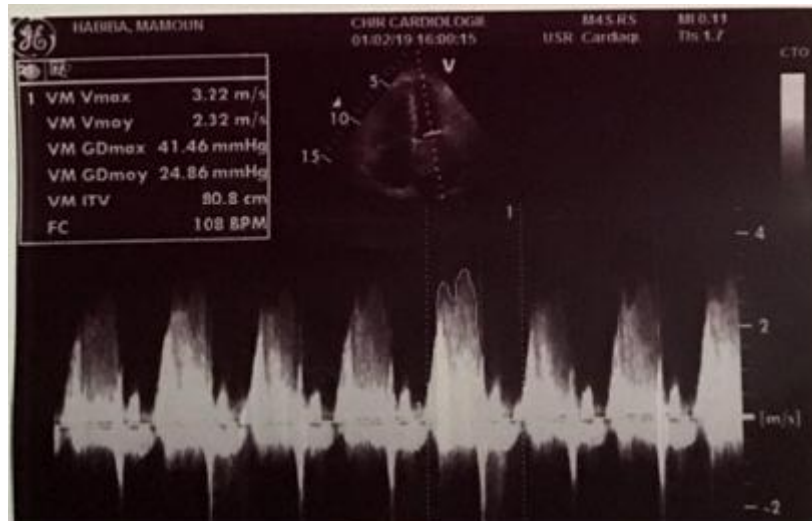
**CASE REPORT**

A 65-year-old, hypertensive and menopausal woman was hospitalized for exertional dyspnea and palpitations since ten months with significant weight loss (about 9 kilograms). The history reveals a notion of a transient ischemic attack that occurred 3 years ago, neglected by the patient.

Clinical examination found a stable hemodynamically patient with a dyspnea stage III (NYHA). Burst of B2 at the pulmonary focus with a systolic tricuspid murmur and a diastolic mitral murmur. No signs of heart failure were revealed. Electrocardiogram (EKG) showed normal sinus rhythm.

Transthoracic echocardiography showed a left atrial polypoid mass (27×25mm) with irregular contours, adherent to the upper third of the interatrial septum (**Figure 1**). The tumor was seen prolapsing across the mitral valve into the left ventricle, causing a gradient on the mitral valve of 24mmhg (**Figure 2**), with a severe pulmonary hypertension (97mmhg).

**Figure 1: Transthoracic echocardiography 4 cavity view shows a left atrial myxoma measuring 27\*25mm.**



**Figure 2: A mean gradient of 24mmhg on the mitral valve due to the prolapsing myxoma across it.**

Chest CT confirms the tissular nature of the mass with no signs of pulmonary embolism.

Due to the risk of embolism and obstruction of the mitral valve by the mass, the patient underwent successful resection of left atrial myxoma, under cardiopulmonary bypass, with simple consequences. Pathology report subsequently confirmed the diagnosis of myxoma.

## DISCUSSION

Myxomas are the most common primary cardiac tumor (more than 50%).<sup>[1]</sup> Myxomas can sit in the atria (either at the mitral annulus or the fossa ovalis border of the interatrial septum), ventricles or on the mitral valve, with a clear predilection for the left atrium (more than 75%).<sup>[2,3,4]</sup>

Histologically, myxoma is a benign tumor with a friable gelatinous component resulting from the embryonic residue of mesenchymal cells sequestered in the fossa ovalis of the interatrial septum.

This explains the strong predominance of its implantation at the septum.<sup>[5, 6]</sup> Atrial myxomas occur predominantly in females with the peak between the fourth and sixth decade of life.<sup>[9]</sup> They are associated with a triad of complications, including obstruction, emboli, and constitutional symptoms (such as fever, weight loss).<sup>[2,8]</sup> The symptomatology remains variable and depends on the location, shape, size and activity of the patient.<sup>[7]</sup>

Patients with left-sided myxomas usually develop signs and symptoms associated with mitral valve obstruction or regurgitation, left-sided heart failure, and secondary pulmonary hypertension. On physical exam, "tumor plop" may be characteristically heard early in diastole.<sup>[10]</sup> This sign depends on the position in which the examination is performed. In fact, cardiac auscultation should be performed in a sitting or standing position allowing the prolapse of the mass across the mitral valve causing the obstruction. The most common symptoms

are dyspnea with exertion followed by orthopnea, paroxysmal nocturnal dyspnea, and pulmonary edema.<sup>[11]</sup>

Systemic embolization is the most common risk increased by left atrial myxomas, particularly in the central nervous system, such as transient ischemic accident (as our patient), hemiplegia, or in retinal arteries as well as viscera...<sup>[12]</sup> Neurologic defects remain the most serious complications.<sup>[13]</sup> Constitutional symptoms include fever, anorexia, arthralgia, weight loss, and can be explained by the release of cytokine IL-6, which plays a major role in the proliferation of myxoma cells and the release of acute-phase reactants.<sup>[14]</sup>

Echocardiography is usually the diagnostic modality of choice. It can characterize the size, number, location, attachment, and mobility of the atrial mass, as well as the extent. The tumor can obstruct the circulation and act as a source of emboli. Transesophageal echocardiography is superior to transthoracic echocardiography in profiling atrial mass (sensitivity of 97%) even though transthoracic echocardiography is simpler and usually first obtained if there is clinical suspicion.<sup>[15]</sup>

The classic ultrasound image is that of a mobile mass attached to the wall by a pedicle. This mass, most often echogenic, has hyper and hypoechoic areas. In fact, the morphology of the myxoma is correlated with the embolic risk. Thus, villous and polypoid tumors are more fragile and embolish more often than those with a smooth and regular surface. The size of the myxoma which can range from a few millimeters to more than 15cm is not correlated with the embolic risk.<sup>[16]</sup> It also studies their hemodynamic impact. More than half of the patients present a pulmonary arterial hypertension (as our patient). This is due to the obstruction of the left atrioventricular blood flow by the myxoma.<sup>[16]</sup>

Up to more than a half of left atrial myxomas show obstructive symptoms,<sup>[3]</sup> but only in 10% of patients will

it cause severe mitral stenosis and severe pulmonary hypertension (as our patient).<sup>[25,26]</sup>

Doppler echocardiography can demonstrate valvular dysfunction and associated outflow obstruction by calculating the mean pressure gradient across the valve in apical views.<sup>[27]</sup> Our patient's mitral gradient measured by pressure half-time method was 24 mm Hg which was consistent with severe functional mitral stenosis. She also had pulmonary hypertension.

The hyper mobile aspect of the mass with a hemodynamic impact is a very urgent indication for surgery in order to avoid embolization.<sup>[17]</sup>

Cardiac magnetic resonance imaging (cardiac MRI), and cardiac computed tomography (cardiac CT), are also valuable evaluation tools, especially in difficult cases with intracardiac thrombus which constitutes the main differential diagnosis. They also help to differentiate from other intracardiac tumors (such as primary sarcoma, primary cardiac lymphoma, large B-cell lymphoma can also mimic atrial myxomas).<sup>[4,18,19]</sup>

Considering the high risk of embolization, prompt surgical excision is the treatment of choice for atrial myxomas. Pericardial patch placement is highly recommended to help constructs defects not fixed by primary closure especially for myxomas implanted on the fossa ovalis.<sup>[20]</sup> The resected tumor is sent to pathology for confirmation, and also to rule out other tumors.<sup>[22]</sup>

The prognosis remains excellent. Mortality rate does not exceed 5% with rapid postoperative recovery.<sup>[2,8]</sup> Late recurrence risk is estimated at 2%.<sup>[21]</sup>

Medical treatment (diuretics, anticoagulation and antiplatelet therapies, antibiotics,...) can be required in complicated cases ( Congested heart failure, arrhythmia and thromboembolic events, infection, ... ).<sup>[23,24]</sup>

## CONCLUSION

Myxoma, although a benign mass, can induce dramatic symptoms and be life-threatening by mimicking severe mitral valve stenosis and severe pulmonary hypertension. Surgical management is urgent, particularly in the presence of morphological characteristics predictive of embolism on echocardiography.

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