

# Mitral Mega Mix – a Case Report

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## Abstract

Background and aim of the study: Atrial myxomas are the most common primary cardiac tumors and are generally discovered as incidental findings. When obstructive symptoms become manifest, acute hemodynamic impairment may occur with the necessity of surgical correction. Results: We describe a case of a young woman with progressive dyspnea and syncope, and presence of a huge intracardiac mass. The mass completely obliterated the mitral valve orifice requiring emergent surgery. Immediately following excision and at follow-up normal hemodynamics were restored. Conclusions: Whenever huge intracardiac masses cause hemodynamic compromise, emergent surgery is necessary. Immediate relief is seen following excision.

## Introduction

Atrial myxomas are the most common primary cardiac tumors<sup>1</sup>. Clinical features are dictated by their size, location and mobility. They arise in the left atrium in 75-80% of cases. In the great majority of cases they are discovered as incidental findings, being asymptomatic. However, whenever clinical symptoms become manifest, the most common presentations are embolic, obstructive and constitutional<sup>2</sup>. Obstructive symptoms resemble valve stenosis. Furthermore, impingement on the valve may impair valve function, requiring associated valve repair/replacement. Primary treatment is surgical removal of the myxoma; whenever hemodynamic compromise is present, emergent intervention is necessary<sup>3</sup>.

We describe a case of a huge left atrial myxoma that caused nearly complete obliteration of mitral valve orifice, mimicking mitral stenosis, and requiring emergent surgical excision.

## Case Presentation

A 32-year-old woman presented to the emergency room with dyspnea, palpitations and syncope that had arisen approximately in the previous 24 hours. The patient was hypotensive and with progressive worsening orthopnoea. She underwent a transthoracic echocardiogram that showed a massive mobile intracardiac mass of 6 x 6 cm, fluctuating within the left atrium and determining nearly complete mitral valve occlusion, with severe mitral stenosis (Gmed 11mmHg). **Fig. 1a and 1b**. Associated findings were moderate-to-severe mitral insufficiency (MR), **Fig. 1c**, with moderate left-ventricular dysfunction, ejection fraction (EF) 40%. Furthermore, right ventricular impairment was present, with severe tricuspid regurgitation (TR) and severe pulmonary hypertension (systolic pulmonary artery pressure (sPAP) 60mmHg).

Due to initial hemodynamic impairment requiring inotropic support and worsening respiratory conditions with the need of high-flow oxygen, the patient underwent emergent surgery. Upon chest opening there was evidence of severe biventricular dilation and dysfunction, prevalently involving the right chambers, with severe right atrial distension (central venous pressure (CVP) 25mmHg). A bi-atrial approach was used, in order to better identify the implant of the pedicle. The root of the pedicle, together with the full thickness of approximately 1cm<sup>2</sup> of adjacent interatrial septum were resected. The mass was then completely removed; dimensions were 17 x 7 cm, a solid component together with thrombotic and gelatinous aspects, **Fig. 2**. The resulting atrial septal defect was closed with a direct suture. Tricuspid valve repair with Kay technique was performed. Evidence at transesophageal echocardiogram (TEE) of residual severe MR due to multiple jets was seen (**Fig. 3a**), requiring a second cardiopulmonary bypass run to perform mitral annuloplasty with a complete semirigid ring (St Jude Medical Saddle n30), **Fig. 3b**. Inotropic and mechanical circulatory support were necessary for severe biventricular dysfunction. Immediately following intervention, CVP decreased to 5 mmHg and pulmonary hypertension disappeared. At histologic examination, diagnosis of myxoma was confirmed.

The patient had a regular postoperative course, with recovery of good biventricular function and no pulmonary hypertension. She was discharged home on post-operative day 8.

At follow-up after 3 months the patient was asymptomatic, with good functional class (NYHA I), and could undertake all normal daily activities. A transthoracic echocardiogram showed complete excision of the atrial myxoma, with a good result of both mitral and tricuspid valve repair.

## Discussion

Cardiac myxomas are most frequently detected as incidental findings being prevalently asymptomatic. Clinical manifestations greatly depend on the anatomic location and the size of mass. Whenever myxomas reach significant dimensions they may lead to obstructive manifestation, which are commonly mistaken for valvular stenosis. In fact, in our patient, complete obliteration of the mitral valve orifice lead to dyspnea, orthopnoea and syncope, mimicking mitral valve stenosis. Intracardiac masses responsible for hemodynamic compromise are not a frequent finding; emergent surgery is the gold-standard treatment and, upon mass excision, immediate hemodynamic improvement is observed. Furthermore, both clinical improvement and symptomatology relief are immediately detected.

## Author Contributions

Alessandra Sala: data collection, concept/design, drafting article;

Paolo Denti: concept/design, data analysis/interpretation, critical revision of the article;

Ottavio Alfieri: critical revision of the article;

Alessandro Castiglioni: critical revision of the article;

Michele De Bonis: critical revision of the article.

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## Figure Legends

**Figure 1a :** Transesophageal echocardiography shows a huge left atrial mass that obliterates the mitral valve orifice.

**Figure 1b :** Transesophageal echocardiography shows the left atrial mass that determines nearly complete mitral valve occlusion.

**Figure 1c :** Transesophageal echocardiography showing the left atrial mass which determines associated severe mitral regurgitation.

**Figure 2 :** Macroscopic appearance of the mass following complete excision.

**Figure 3a:** Intraoperative transesophageal echocardiography showing residual moderate-to-severe mitral regurgitation following complete mass excision.

**Figure 3b :** Intraoperative transesophageal echocardiography showing good result of mitral valve repair, with no residual mitral regurgitation following annuloplasty.

### **Video Legends**

**Video 1. Huge left atrial mass** - Transesophageal echocardiography showing a huge left atrial mass that protrudes through the mitral valve, causing mitral valve stenosis and regurgitation.

**Video 2 . Huge left atrial mass determining mitral steno-insufficiency** - Transesophageal echocardiography showing a huge mass that fluctuates within the left atrium, determining nearly complete mitral valve occlusion, with severe mitral valve stenosis.

**Video 3. Mitral annuloplasty result** - Intra-operative transesophageal echocardiography following second cardiopulmonary bypass run showing good mitral annuloplasty result with a complete semi-rigid ring.





