A Giant Left Atrial Myxoma Causing Acute Heart Failure

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Abstract
Clinical presentation of cardiac myxomas is very heterogeneous and varies from asymptomatic to life-threatening complications. We report an uncommon clinical presentation of a giant left atrial myxoma by acute heart failure in a 52-years old male. Echocardiography showed a significant restriction of flow through the mitral valve with serious impact on the right cardiac cavities. Appropriate medical therapy and early surgical excision of the tumor allowed good clinical evolution and complete regression of congestive heart failure.

Keywords: Giant Left Atrial Myxoma; Mitral Valve; Echocardiography

Introduction
Primary cardiac tumors are rare, most of them are benign, myxoma being the most common one. Clinical presentation is very heterogeneous. We report a case of a giant left atrial myxoma causing acute heart failure.

Case Report
A 52-years old male presented in cardiology department with dyspnea and orthopnea. Physical examination found pulmonary oedema with bilateral massive lower limb oedema and hepatojugular reflux, allowing the diagnosis of acute congestive heart failure. Electrocardiogram found a sinus rhythm with multiple supraventricular extrasystoles. Transthoracic echocardiography was performed and identified a large left atrial mass suggesting a left atrial myxoma occupying almost completely the left atrial cavity, adhering to the interatrial septum and prolapsing into the left ventricle in diastole, causing significant restriction of flow through the mitral valve (Figure 1). There was a dilation and dysfunction of the right ventricle with tricuspid regurgitation and pulmonary hypertension. A surgical excision of the tumor was decided. Preoperative coronaryography showed no significant coronary artery lesion. The patient was treated by diuretics few days before surgery.

Figure 1: Transthoracic echocardiography showing a giant left atrial myxoma prolapsing into the left ventricle in diastole, causing significant restriction of flow through the mitral valve.
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Figure 1: 4 cavity view of transthoracic echocardiography showing a massive left atrial mass during systole (A), that prolapses into the left ventricle during diastole causing significant restriction of flow through the mitral valve (B).

Surgery was done under general anesthesia by median sternotomy. Extracorporeal circulation was installed between an arterial cannulation in the ascending aorta and venous bicaval cannulation. We used a vertical bialtrial transeptal incision which allowed us to approach the tumor that was very large and adherent to the interatrial septum (Figure 2). A complete excision of the tumor with the implantation site in the interatrial septum was performed (Figure 3). Assessment of the mitral valve showed no insufficiency nor lesion, the septal defect was closed by direct suturing. Tricuspid valve was regurgitant with a significant dilation of the tricuspid annulus, therefore we performed a tricuspid annuloplasty using a 32 mm Carpentier-Edwards Classic annulus. The postoperative course was uneventful. Histological examination of the specimen confirmed the diagnosis of myxoma. The patient left the hospital in a good clinical condition.

Figure 2: Operative view after interatrial septal incision showing the left atrial myxoma (arrow) adhering to the interatrial septum and occupying the left atrial cavity.

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Discussion

Primary cardiac tumors are rare, 80% of them are benign, myxoma being the most common one responsible for about 50% of the cases. 75 - 80% of myxomas are located in the left atrium, 15 - 20% in the right atrium and more rarely in the ventricles or multicentric. Myxomas are considered to arise from multipotent mesenchymal cells which have persisted in the heart since the embryonal development period [1]. These cells are very often localised in the fossa ovalis area of the interatrial septum [2].

Myxomas are more common among women and often occur between 30 and 60 years of age. They usually occur sporadically, although family cases of multifocal location associated with Carney’s syndrome are described [3].

Clinical presentation varies from asymptomatic or nonspecific symptoms to life-threatening complications such as stroke, acute heart failure, or even sudden death due to complete atrioventricular valve obstruction or coronary artery embolization [4,5]. Among other things, clinical presentation of patients with left atrial myxoma strongly depends on tumor size, fragility and localization [4].

Transthoracic echocardiography has shown 95% sensitivity in the detection of myxomas [6]. In some cases where diagnosis is difficult, or when a differential diagnosis with a malignant tumor is required, computed tomography or magnetic resonance imaging may be necessary. In addition to confirming the diagnosis, they allow the establishment of the best surgical technique [7]. Transeosophageal echocardiography, particularly intraoperatively, seems to provide more accurate information and helps to better delineate the surgical margins, particularly in small myxomas or when there is a poor transthoracic window [8].

We report here a rare case of acute heart failure secondary to a left atrial myxoma. In the few cases described in the literature, different pathogenic mechanisms have been proposed [9]. Atrial myxoma tissue can release cytokines which have cardiodepressive effect such as interleukin IL-6 and IL-8, which may cause myocardial tissue inflammation and biventricular dysfunction. Overproduction of IL-6 and IL-8 may also induce myxoma cell fragment adhesion to coronary artery endothelium, leading to myocardial ischemia and infarction [10,11]. Left atrial myxoma can also cause mitral insufficiency by atrial and annular dilatation, or mitral stenosis by obstructing left ventricular filling during diastole.

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Surgical resection is the only curative treatment, it should be performed early when complications occur and is coupled with excellent long-term survival. The common surgical approach is a median sternotomy with extracorporeal circulation. Less invasive approach through a right mini-thoracotomy or partial hemi-sternotomy can be used [12]. Complete resection of the myxoma with the implantation site in the interatrial septum or cardiac wall is mandatory to prevent recurrence.

Although tumour recurrence is rare, imaging follow-up is recommended since the recurrence rate is significantly high in the first 10 years after surgery [8].

**Conclusion**

Acute heart failure is an uncommon clinical manifestation of atrial myxoma. Early surgical extirpation is the only available curative option with excellent results. Regular follow-up is recommended regarding the risk of recurrence after surgery.

**Bibliography**