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Severe Mitral Valve Dysfunction: A Giant Left Atrial Myxoma Was the Guilty

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Abstract

BACKGROUND: Left atrial (LA) myxoma's clinical manifestation depends on the size, location, mobility, and architecture of the tumor. The clinical presentation consists of cardiac (67%), embolic (29%), and systemic (34%) symptoms, even sudden cardiac death.

CASE PRESENTATION: We present an interesting case and images of a patient with severe mitral valve dysfunction and syncope, who was diagnosed with a giant mass in the left atrium and left ventricle as myxomas. Open heart surgery was performed and the mass was detected and successfully removed. The histopathology confirmed the diagnosis of myxoma of the left atrium.

CONCLUSION: Myxoma may be challenging to diagnose, but with imaging techniques, such as transthoracic (TTE), transesophageal (TEE) echocardiograms, when a high index of clinical suspicion exists, early diagnosis and therefore curative intervention in the majority of the cases is more likely.

Introduction

Myxoma is the most common primary cardiac tumor, consisting 50% to 80% of the tumors and is most located in the left atrial (LA) arising from the interatrial septum near the foramen oval [1]. LA myxoma's clinical manifestation depends on the size, location, mobility, and architecture of the tumor [1], [2]. The clinical presentation consists of cardiac (67%), embolic (29%), and systemic (34%) symptoms, even sudden cardiac death [2]. Non-specific symptoms such as fever, weight loss, and fatigue may also occur resulting from the pro-inflammatory cytokines produced by the tumor [2]. Imaging techniques including transthoracic (TTE), transesophageal (TEE) echocardiograms, computed tomography (CT) scans, and magnetic resonance imaging (MRIs) can be helpful [3], although histology is generally required for diagnosis. Echocardiography is the screening and diagnostic method of choice [2]. Surgical removal is the method of choice for the treatment of myxoma. Excision helps to reduce potential complications, such as obstructive and embolic phenomena that are more likely to occur as myxoma increases in size [1], [3].

Case Presentation

In this present report, we would like to present some interest intraoperative images of a patient diagnosed with a huge mass in the left atrium and left ventricle, presumably myxomas being the most heart tumor. The patient underwent median sternotomy, bicaval cannulation, and cardioplegic arrest in an antegrade fashion. After the left atrium opening a huge mass was detected (Figure 1a and b). It was excised from the interatrial septum and taken off without any complications. The shape of the mass detects that its growth was restricted by the mitral valve (Figure, 1c, green arrow). The ventricular part of the myxoma was black and ischemic (Figure 1d, black arrows) also due to restriction caused by the mitral valve; picture similar to the "strangulated hernia." The mitral valve was checked in detail to evaluate any regurgitation caused by this huge mass. Then, at the end of the intervention, an intraoperative transesophageal echocardiography was performed to evaluate the interatrial septum and of course the mitral valve function. The histopathology report revealed a myxoma of the left atrium. The patient fully recovered in follow-up without any symptomatology.

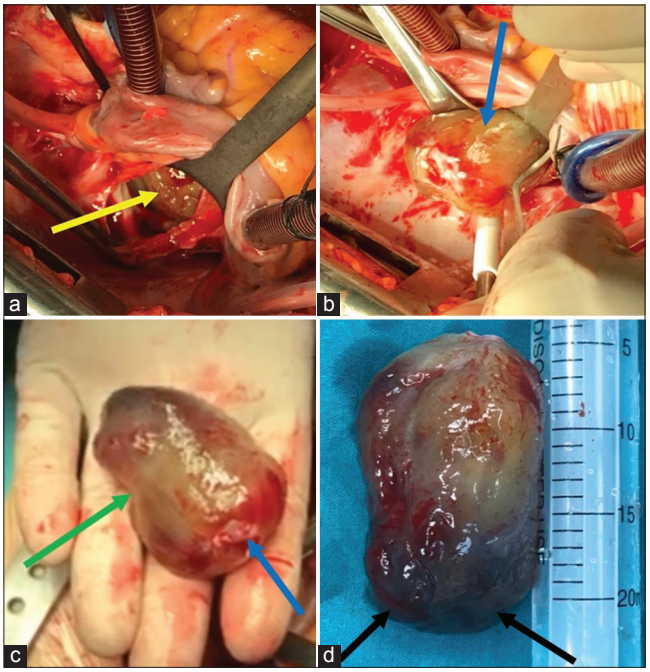


Figure 1: (a) The left atrium is opened revealing the mass (yellow arrow). (b) The mass is excised from the interatrial septum and taken off (blue arrow). (c) The area of the mass previously in touch with the mitral valve is restricted (green arrow), when compared to the rest shape of the mass due to the mitral valve. (d) The ventricular part of the mass is black and ischeamic (black arrows) due to restriction caused by the mitral valve.

Comment

Myxoma is the most frequent intracardiac tumor, but several other pathologies such as intracardiac thrombus, vegetations, and benign or malignant tumors that may develop in the heart chambers must be excluded [1].

Primary cardiac tumors occur infrequently with an incidence of 0.0017–0.19% in autopsy series performed in non-selected populations [4]. Myxoma has an incidence of 0.5/million/year [1] and is more common among women frequently between ages 30–60 years with a female-to-male ratio of 2:1 [1], [4].

The clinical presentation consists of cardiac (67%), embolic (29%), and systemic (34%) symptoms, even sudden cardiac death [2]. In one study, a patient with LA myxoma suffered from multiple cerebral mycotic aneurysms due to the myxoma [5]. Due to the variety of non-indicative findings at first presentation, myxoma is initially suspected in only 5% of patients [2].

The morphological characteristic of the mass, such as the size and the location of myxomas are correlated with their clinical features. According to one study, large LA myxoma size is more related to constitutional symptoms, congestive heart failure, with syncope and auscultatory findings suggestive of mitral valve disease, while smaller myxoma size and irregular surface are associated with embolization [6]. What is more, the tumor surface appearance is also strongly related to its symptoms. The presence of an irregular or villous surface is significantly associated with cerebral and peripheral embolisms, neurologic symptoms, and in general embolic complications, and cardiac auscultation abnormalities, while smooth-surfaced myxomas tend to appear with constitutional symptoms [2].

Myxoma may be challenging to diagnose, but with imaging techniques, such as TTE or TEE, when a high index of clinical suspicion exists, early diagnosis and therefore curative intervention in the majority of the cases is more likely [1], [4]. Once it has been identified, early surgery is required, reducing complications, as myxomas increase rapidly in size causing obstructive and embolic phenomena, even sudden death [1], [3]. Surgical excision of primary benign cardiac tumors, such as myxoma has an excellent prognosis with a long-term recurrence rate of below 5% in sporadic cases, while a careful follow-up is also suggested to monitor for recurrence, particularly in familial cases [1], [4]. Minimally invasive as well as robotic surgical approach has been used, but is technically challenging [7], [8].

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