Int Cardiovasc Res J.2017;**11**(2):84-86.icrj.11189

International Cardiovascular Research Journal

www.ircrj.com

Mitral Annulus Myxoma Extending into Left Atrium and Left Ventricle with Severe Mitral Regurgitation as a Pre-Operative Finding, a Rarity

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ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 10 Aug 2016
Revised: 09 Nov 2016
Accepted: 04 Jan 2017

Keywords:
Mitral Regurgitation
Myxoma
Emergency

ABSTRACT

Cardiac tumors constitute 0.2% of all tumors. Primary cardiac tumors occur infrequently with an incidence of 0.0017-0.19% as shown by autopsies performed in non-selected populations. Among these tumors, cardiac myxomas are most commonly encountered, with left atrial myxomas being more prevalent than right atrial ones. The classic triad of symptoms, of which at least one is present in a patient with atrial myxoma, are obstructive traits including dyspnea and syncope, constitutional symptoms such as fever and anorexia, and thromboembolic events. Surgical resection confers almost definitive treatment with recurrence rates being as low as 3%. A 50-year-old woman referred to the Emergency Unit with a sudden episode of chest heaviness and shortness of breath. There was no significant physical examination finding and all routine lab investigations were normal. She underwent an angiography that revealed tight left anterior descending artery stenosis. An angioplasty was also performed, but she had an episode of pre-syncope immediately. Then, echocardiogram was performed that showed a large left atrial myxoma causing severe mitral regurgitation. Thus, urgent open heart surgery was planned. The myxoma was identified and excised, the mitral annulus resolved, and normal flow was restored. The patient was then discharged home and followed up for 2 months with no complaints. In the current study, we reported a rare case where mitral regurgitation was caused by a left atrial myxoma. Our report highlighted the diverse clinical spectrum of myxoma and emphasized the need for early echocardiographic diagnosis to aid in identification of myxoma followed by early surgical intervention.

1. Introduction

Cardiac tumors constitute only 0.2% of all tumors found in humans (1). Primary cardiac tumors occur infrequently with an incidence of 0.0017 - 0.19% as shown by autopsies performed in non-selected populations (2). They arise from normal heart tissue, while secondary tumors originate from other parts of the body and are 20 to 40 times more frequent. Cardiac myxomas are the most common benign form of the rare primary tumors, of which 75 - 80% occur in the left atrium and 10 - 18% in the right atrium. Most left atrial myxomas are attached to the interatrial septum (3). However, valvular originations of such tumors are very rare (4). The exact incidence of myxomas originating from the mitral valve is not clear. A previous study reported it to be 1.5% (1 case among 68 myxoma cases) (5).

Left atrial myxomas show excellent prognosis following surgical excision, with a 3% recurrence rate. They can, however, trigger fatal thromboembolic events if left untreated, making timely accurate diagnosis essential (6). Progression of these tumors can simulate symptoms of mitral stenosis, infective endocarditis, and collagen vascular disease (7). Theclassic triad of symptoms, of which at least one is present in a patient with atrial myxoma, are obstructive traits including dyspnea and syncope,

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constitutional symptoms such as fever and anorexia, and thromboembolic events (8).

In this study, we present a rare clinical case of a mitral valve annular myxoma extending into the left atrium and ventricle in a 50-year-old woman with severe mitral regurgitation as a pre-operative finding.

2. Case Presentation

A 50-year-old woman presented with a sudden episode of chest heaviness and shortness of breath to the Emergency Unit in our tertiary care hospital. There were no neurological or any other constitutional symptoms. There was also no history of significant weight loss over the past month. Her medical history was unremarkable, except for well-controlled hypertension and hyperlipidemia. The patient had no history of previous cardiac pathology. Chest auscultation was clear with normal heart sounds. Routine lab investigations were normal, as well. She underwent an angiography, which showed tight left anterior descending artery stenosis for which an angioplasty was performed. Following angioplasty, she had an episode of pre-syncope. Thoracic radiogram was normal. An echocardiogram was also performed, which showed a large left atrial myxoma with half of it protruding through the mitral valve into the left ventricle, causing severe mitral regurgitation. Left ventricle was not dilated with normal ejection fraction. There was insignificant pericardial effusion around the heart. Thus, urgent open heart surgery was planned. Under cardiopulmonary bypass, the left atrium was opened via the superior septal approach. Very friable and soft atrial myxoma was identified arising from the anterolateral potion of the annulus of the mitral valve (Figure 1). The base was identified and the myxoma was excised completely and was then delivered out of the heart (Figure 2). Afterwards, the mitral annulus was repaired with interrupted 4.0 Prolene sutures and mitral valve apparatus was preserved. After all, mitral regurgitation resolved completely with normal flow of blood through the heart.

Histology of the specimen showed abundant hyaline and myxoid stroma containing small groups of tumor cells compatible with the diagnosis of atrial myxoma. There were no postoperative complications and the patient was discharged home after 10 days in perfect health with no breathing abnormalities. She was followed up for the next 2 months as an outpatient and did not suffer from any complications.

3. Discussion

Cardiac valve tumors are rare, with fibroelastomas followed by myxomas being the most common ones. Other valvular masses include organized thrombi, valvular calcifications, and valvular abscess. These masses are distinguishable on echocardiography. Atrial myxomas are the most commonly encountered curable tumors of the heart. They occur in patients aging 15 to 80 years although the reported average age is 50 years. Indeed, females are three times more likely to develop this tumor, and usually exhibit sporadic forms, while male patients are more likely to present with familial forms of the tumor (9). Incidentally, our patient was female and aged 50 years.

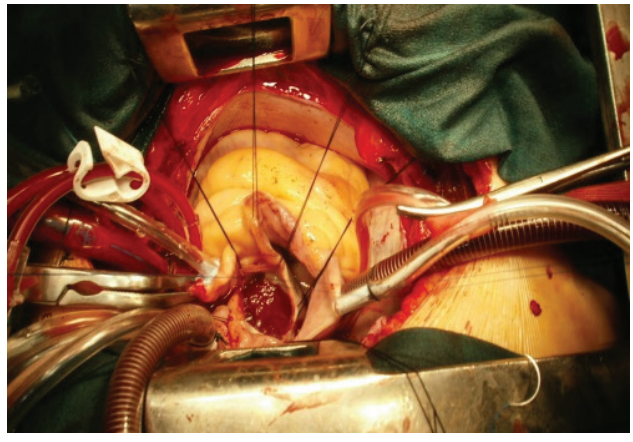

Figure 1. The Myxoma Attached to the Mitral Annulus during Surgery

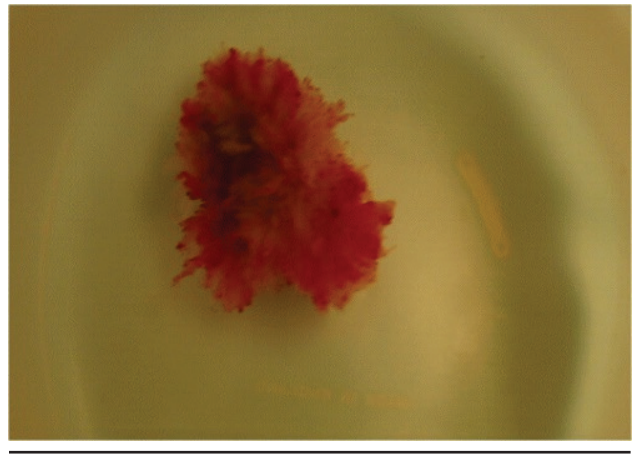

Figure 2. The Large and Friable Myxoma Excised from the Mitral Annulus

Myxomas are usually brown-colored, solitary, pedunculated lesions with a smooth or lobulated surface. They may be oval, rounded, or irregularly shaped and their consistency varies from firm to gelatinous. Additionally, their mobility depends on their degree of attachment to the site of origin and the length of their stalk. Histologically, they resemble multipotent mesenchymal cells of the subendocardium (10).

This case report showed a cardiac myxoma arising from the mitral valve annulus, causing severe mitral valve regurgitation. Mitral valve myxomas are a rare cause of mitral regurgitation. They can originate from anterior or posterior leaflets or the annulus and are usually localized to the atrial side of the mitral valve (11). They produce symptoms upon mechanical interference with cardiac function or embolization. Valvular disease is thus among the top differential diagnoses of mitral valve myxomas.

The anatomical location and size of the mass determines the clinical manifestations, which can be grouped into three main types: embolic, obstructive, and constitutional. Myocardial and visceral infarctions are embolic exhibitions, while obstructive ones can be mistaken for valvular stenosis due to their close mimicry of the pathological presentation. A high index of suspicion aids in diagnosis. In our case, obstruction of the mitral valve orifice by the tumor mass was most likely causing the symptoms of dyspnea and syncope.

Mitral regurgitation was a sequel of annular dilatation.

Transesophageal echocardiography is used for diagnosis because of its greater sensitivity compared to transthoracic echocardiography. Upon successful diagnosis, appropriate surgical treatment must be carried out to prevent fatal systemic embolization. Surgical excision of the tumor appears to be curative and any rare instances of recurrence can be attributed to incomplete excision of the tumor, growth from a second focus, or intracardiac implantation of the tumor during excision.

Our choice of the superior septal approach was based on its good exposure of the left atrium and mitral valve as well as the available expertise. Upon excision of the tumor, the mitral annulus was repaired since there was no significant valvular destruction or tumor adherence to indicate the need for valve replacement.

Echocardiography played a huge role in diagnosis of myxoma in our case since the pre-operative finding of mitral regurgitation caused by a left atrial myxoma was a rare one. The need for early echocardiographic examination is thus emphasized.

3.1. Consent

The patient's identity has been concealed. The authors declare there is no conflict of interests. The authors have no financial interests related to the content of the manuscript.

Acknowledgements

There is no acknowledgement.

Authors' Contribution

Saulat performed the surgery and provided administrative support.

Financial disclosure

The authors have no financial interests related to the material in the manuscript. Tariq came up with the study concept and design and supervised the study. The data were interpreted and the manuscript was drafted by Atiya, Omar, and Hamza.

Funding/Support

Department of Cardiothoracic Surgery, AKU.

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