Right Atrial Myxoma: Unusual Location; Uncommon Association

Abstract

We are presenting a case of the right atrial myxoma found in a case of rheumatic heart disease. During transthoracic echocardiography for the evaluation of a suspected rheumatic valvular heart disease, a diagnosis of severe mitral stenosis with severe mitral regurgitation with the right atrial thrombus was made. On transesophageal echocardiography, a pedunculated mass in the right atrium was confirmed. In the course of surgery, it was found that there is a tumor originating from the right atrial appendage (RAA) which was confused with clot on echocardiography. Rheumatic heart disease and myxomas have different etiopathogenesis, and this coexistence has not been reported. Further, right atrial myxoma arising out of RAA is also being reported for the first time.

Keywords: *Myxoma, rheumatic heart disease, thrombus*

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Introduction

The incidence of primary cardiac neoplasm ranges from 0.17% to 0.19%.[1,2] Three-fourth of these primary cardiac tumors are benign and only one-third malignant. Myxomas make 50% of all benign tumors in adults and stand as the most common primary cardiac tumor of the heart.[3]

Approximately, 75% are located in the left atrial cavity, 23% in the right atrial cavity, and about 2% in a ventricular cavity.[2,4] Majority, almost 94% myxomas are solitary, but biatrial and multicentric myxomas have also been reported. There is female sex dominance, and peak incidence is between third and sixth decade.

Myxomas originate from the interatrial septum close to margin of fossa ovalis. However, they can also originate in descending order of frequency, from the posterior atrial wall, the anterior atrial wall, and the atrial appendage.[2] We report a case of the right atrial myxoma arising from the right atrial appendage (RAA) in a patient of rheumatic heart disease scheduled for mitral valve replacement. Although myxomas of the left atrial appendage have been reported, this is a first reported case of myxoma of RAA.

Case Report

A 32-year-old female patient weighing 44 kg, presented with complaints of loss of appetite, weight loss, fatigue, and recent increase in the severity of dyspnea for 8 months. On examination, there were engorged neck veins, moderate hepatomegaly, and mid diastolic murmur in mitral area. On transthoracic echocardiography (TTE), she was diagnosed to have severe mitral stenosis, severe mitral regurgitation, severe tricuspid regurgitation (TR), and right atrial thrombus. Mitral valve replacement with tricuspid annuloplasty and evacuation of thrombus was planned. All routine preoperative hematological and biochemical investigations were carried out and were within normal limits. Chest X-ray revealed enlarged cardiac silhouette with prominent bronchovascular markings. Electrocardiograph revealed atrial fibrillation with ventricular rate of 108/min. She was taking tablet digoxin 0.25 mg daily 5 days a week and tablet torsemide 10 mg daily for the last 8 months.

In the operation theater, left radial artery was cannulated, and right internal jugular vein was catheterized by triple lumen central venous catheter. Anesthesia was induced with midazolam 4 mg, fentanyl 200 µg, and thiopentone sodium 125 mg. Tracheal intubation was facilitated by vecuronium bromide 6 mg, and positive pressure ventilation was started. Anesthesia was maintained with isoflurane and 50% oxygen in air. Nasopharyngeal temperature and bispectral index

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monitoring were started. Intraoperative transesophageal echocardiography (TEE) confirmed most of the findings of TTE except the findings of the right atrium. It revealed a pedunculated, well-defined mass arising from the right atrium [Figures 1 and 2, Video 1], near inlet of superior vena cava and spontaneous echogenicity of the right atrium. Anestheisa during cardiopulmonary bypass (CPB) was maintained with propofol infusion, intermittent doses of vecuronium and fentanyl.

After heparinization, aortic cannulation, and bicaval cannulation was performed. CPB was started, and aortic cross clamp (ACC) was applied. The right atrium was opened, and the mass was felt as an organized tough structure originating from RAA. The tissue was excised, and RAA amputated. Then, interatrial septum was opened, and mitral valve replacement was done with bileaflet prosthetic mitral valve (St Jude Medical Reagent™ Heart Valve). Interatrial septum was closed after deairing. Weaning from CPB was done. Adrenaline infusion was started at the rate of 0.05 µg/kg/min. CPB time was 78 min and ACC time was 42 min. Heparin neutralization was done with slow infusion of protamine 120 mg over 10 min. After the conclusion of surgery, the patient was transferred to intensive care unit. After regaining consciousness and confirming adequate tidal volume, the patient was weaned from ventilator, and finally, the trachea was extubated after 4 h.

The excised mass was of 4 cm × 5 cm size [Figure 3]. Histopathological examination of the mass confirmed myxoma.

Discussion

Right atrial myxomas are sporadic in 95% of cases, and in 5% cases, it has got an autosomal dominant inheritance. Clinical presentation of a myxoma is intracardiac obstruction to flow from one chamber to other resulting in congestive cardiac failure, embolization, and constitutional symptoms. Right atrial myxomas can present with features of right-sided heart failure. On examination, there may be hepatomegaly, ascites and dependent edema. In our case, the size of myxoma was 4 cm × 5 cm, and probably was sufficient to cause obstructive symptoms in normal sized right atrium. However, overwhelming dilatation of the right atrium due to severe TR could have prevented overt obstructive features. Right-sided myxomatous emboli may cause pulmonary hypertension due to the obstruction of pulmonary arteries. Nonspecific constitutional symptoms such as fatigue, fever, and arthralgia occur due to the production of interleukin-6 by tumor cells.[5] The patient described here shared most of these symptoms although they can be explained by the mitral valve disease too.

The chest radiograph may reveal cardiomegaly or individual chamber enlargement depending on the site of myxoma. Although in our case, there was no such

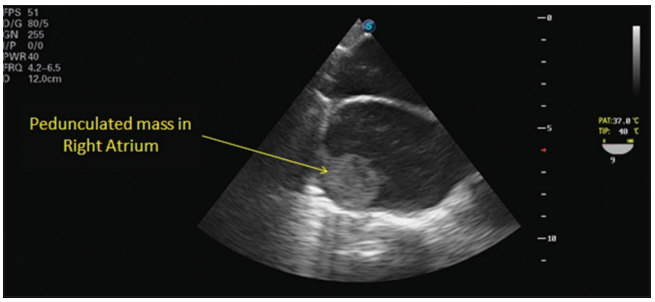

Figure 1: Transesophageal echocardiography view: Mid-esophageal 4 chamber view showing a pedunculated mass in right atrium

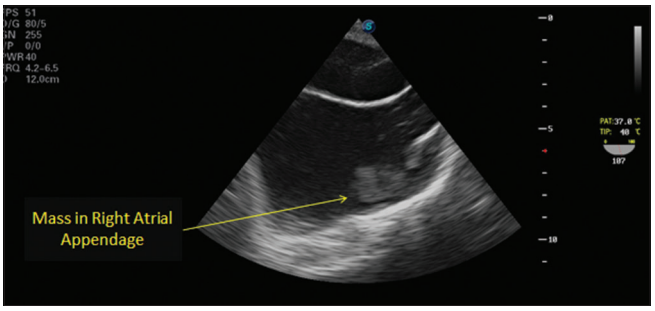

Figure 2: Transesophageal echocardiography view: Mid-esophageal bicaval view showing mass near the inlet of superior vena cava

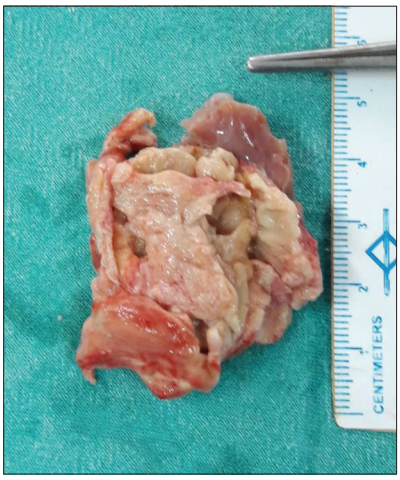

Figure 3: Picture of the excised mass measuring 4 cm × 5 cm in size

feature. Electrocardiographic findings may include nonspecific abnormalities such as cardiac enlargement and bundle branch block, etc., Transthoracic supplemented by TEE has become the most appropriate screening and diagnostic modalities for myxoma.[6,7] TEE provides the best information concerning size, attachment, mobility, and location.[8] In this case also, TTE performed by cardiologist preoperatively diagnosed the mass to be a thrombus. However, intraoperative TEE clearly showed a pedunculated, organized mass which ruled out the presence of thrombus. This finding made the surgeon more comfortable to handle right atrium during bicaval cannulation. Surgical approach to myxoma is different from that of thrombus removal, as myxoma has a tendency to

recur, so complete removal along with appendectomy was necessary. This, in turn, entailed prolongation of ACC and CPB time.

Only 15%–20% myxomatous tumors of the heart arise from the right atrium. Right atrial myxoma usually arises from the interatrial septum close to the border of fossa ovalis. However, unusual locations are reported most frequently in cases of familial myxoma.[9] In our case, there was no such family history. Review of available literature does not reveal any reported case of atrial myxoma arising from RAA.

Patients with myxoma had been reported to have atrial arrhythmias or atrioventricular conduction abnormalities postoperatively in 26% of patients. Moreover, these patients are at risk for recurrence of the myxoma or the development of additional lesions with an incidence of up to 5%, suggesting the need for careful follow-up. Development of a second primary myxoma may be more common in patients with a family history of myxoma.[10]

This case is also unique because of coexistent rheumatic heart disease and right atrial myxoma, as there is no relation between the two as far as etiopathogenesis is concerned. such coexistence is unreported and is of interest to cardiac surgeons and anesthesiologists.

Conclusion

We report for the first time, a right atrial myxoma arising from a very unusual position, i.e., RAA and any myxoma coexisting with advanced rheumatic heart disease. Intraoperative preincision TEE raised the suspicion of the right atrial myxoma by its pedunculated attachment to atrial appendage. We suggest that any intracardiac mass should be evaluated by TEE to rule out the possibility of myxoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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