Case Report

Role of Transesophageal Echocardiography in the Recurrent Biatrial Myxoma of Uncommon Origin

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ABSTRACT

After surgical excision of myxoma recurrence usually happens adjacent to the initial origin site. We report a case of recurrent myxomas in a young male patient that had biatrial recurrence with one tumor originating very unusually from the base of the anterior mitral leaflet. Intraoperative transesophageal echocardiography was instrumental in localizing the site of the origin of left atrial myxoma from the base of the anterior mitral leaflet and in detecting an additional myxoma attached to the wall of the right atrium.

Keywords: Anterior mitral leaflet, recurrent biatrial myxoma, transesophageal echocardiography

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INTRODUCTION

Cardiac myxoma is the most common primary benign cardiac tumor of adults. It usually presents as a solitary tumor attached to the fossa ovalis of the left atrium (LA).[1] The recurrence rate of myxoma after surgical excision varies from 3%–12%; however, it usually occurs adjacent to the initial origin site.[2] We present a case of recurrent multiple biatrial myxomas in a young male patient with one of the two recurrent myxomas originating unusually from anterior mitral leaflet (AML) while the other one from the right atrium. Intraoperative transesophageal echocardiography (TEE) examination was instrumental in localizing the site of origin of one of the recurrent myxomas from the base of AML and in detecting additional myxoma attached to the superior vena cava- right atrial junction.

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CASE REPORT

A 17-year-old male, a known case of post biatrial myxoma excision and left middle cerebral artery infarct with right-side hemiparesis; presented to our institute during the follow-up visit with recurrence of myxoma in the left atrium. He didn't have any constitutional or other symptoms related to myxoma. His right hemiparetic limb power had improved to grade 4/5. There was no associated syndrome, skin lesion, endocrine abnormality, or any family history of myxoma; ruling out the familial forms of myxoma. His follow-up blood investigations were within normal limits. Transthoracic echocardiographic (TTE) examination showed a heterogeneous mass measuring 2 × 2 cm in the left atrium with no obstruction or regurgitation across the mitral valve suggesting the recurrence of myxoma in the LA. He was planned for excision of the left atrial myxoma.

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On the day of surgery, preoperative TTE confirmed the preoperative diagnosis of left atrium myxoma attached to the interatrial septum (IAS) with the normal functioning mitral valve, tricuspid valve, and normal-sized LA.

In the operating room, after instituting routine American Society of Anesthesiologists (ASA) monitoring, peripheral intravenous access, and invasive arterial line monitoring were established under local anesthesia. The induction of general anesthesia was accomplished using fentanyl and titrated doses of propofol. Injection vecuronium was used to facilitate endotracheal intubation. Maintenance of anesthesia was done using continuous infusion of fentanyl, the inhalation of isoflurane-air-oxygen mixture, and intermittent doses of vecuronium. After securing the airway, the transesophageal echocardiographic (TEE) examination performed using the Vivid E9 echocardiography machine (GE, USA), revealed a heterogeneous left atrial mass originating from the junction of IAS and base of AML [Figure 1]. In addition, another similar heterogeneous echogenic mass (1 × 1 cm) was observed attached to the wall of the right atrium [Figure 2]. Both the atrioventricular valves and the ventricular functions appeared normal. There was no thromboembolism in the proximal pulmonary arteries and the pulmonary acceleration time (PAT) was within normal limits suggesting normal pulmonary artery pressure. In order to prevent embolization of the right atrial mass, a central venous catheter was inserted carefully into the right internal jugular vein under ultrasound guidance and TEE monitoring as per our institute protocol.

Surgery included complete excision of both masses using cardio pulmonary bypass (CPB). The intraoperative finding confirmed a myxoma attached to the free wall of the right atrium. The incision of the poly tetra fluoro ethylene (PTFE) patch used for atrial septal defect closure in the previous surgery, revealed that the left atrial myxoma was attached to the base of the AML and IAS. After excision of the left atrial myxoma, the defect in the AML was repaired using primary suture. The IAS was closed using a fresh PTFE patch. The patient was weaned off CPB without the use of any inotrope. The post CPB, TEE examination revealed a normally functioning mitral valve [Video 1]. He was extubated after 4 h of elective mechanical ventilation and discharged from the intensive care unit after 2 days of uneventful stay. The histopathological report of the excised mass revealed both masses as myxoma with a component of normal mitral valvular tissue in the left atrial mass.

DISCUSSION

Myxoma, a tumor of mesenchymal origin is the most common primary benign cardiac tumor with the an incidence of 0.0017%–0.19%.[1] They occur most commonly in the left atrium (75%–85%) followed by right atrium (15%–25%) and only rarely in other parts like ventricles, pulmonary veins, and atrioventricular valves.[3,4] When arising from the left atrium, it mostly arises from the fossa ovalis with less than 10% from other parts of the left atrium.[5] The reported incidence of myxoma originating from the atrioventricular valves is about 1.5%, which necessitates valvular repair or replacement.[4,6]

Myxoma arises either as a sporadic lesion or as a part of familial syndromes like Carney's complex or lentigines arial myxomas blue nevi (LAMB) syndrome. Sporadic myxoma is common in females. It has a bimodal distribution pattern during the third and sixth decade, whereas familial myxomas occur more commonly in younger male patients. Myxoma can be either solitary or multiple. When multiple it can be localized in a single chamber with multicentric origin or involve multiple chambers either both atrium or combination of atrium and ventricle. Right atrial

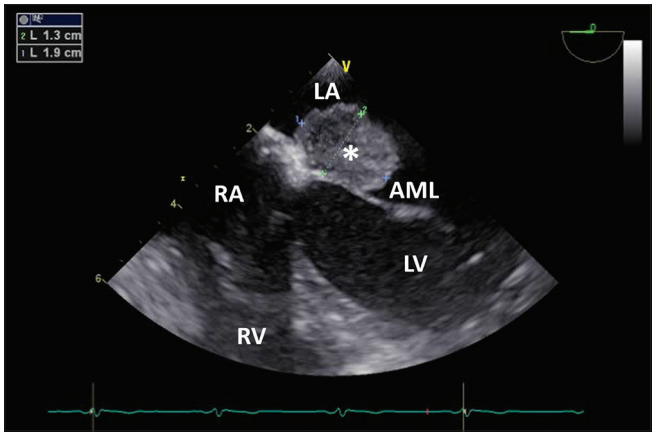

Figure 1: Mid-esophageal 4 chamber view demonstrating left atrial myxoma of size 1.3 × 1.9 cm originating from the junction of Inter-atrial septum and Anterior Mitral leaflet. (LA-Left Atrium, LV-Left Ventricle, RA-Right Atrium, RV-Right Ventricle, * - Myxoma)

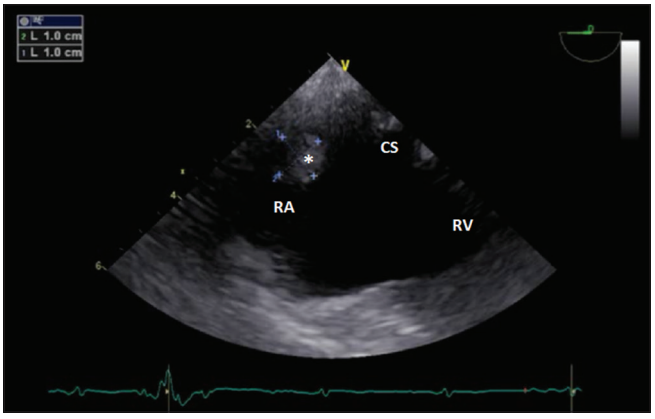

Figure 2: Right atrium focused mid-esophageal 4 chamber view demonstrating myxoma of size 1 × 1 cm attached to the superior wall of right atrium near the superior vena cava opening. (RA-Right Atrium, RV-Right Ventricle, CS-Coronary Sinus ostium, * - Myxoma)

myxomas are more solid and sessile as compared to left atrial myxomas, which has an attachment to the septum.[7,8]

The recurrence of myxoma after excision occurs in about 3% of sporadic cases and more so in the familial forms (12%) or with complex syndromes (22%).[2] Following the first report of recurrence by Gerbode *et al.* multiple such reports have been published, however, the exact mechanism for its recurrence is not clear.[9] Various postulated hypotheses include incomplete resection of the primary tumor, implantation of the tumor cells into the adjacent myocardium during resection, familial predisposition for multiple tumor occurrence, pretumor foci in another part of myocardium, which grows after the excision of primary myxoma, etc.[10] The usual time gap between the excision of the tumor and recurrence is about 4 years.[11] Some authors recommend the excision of the underlying atrial septum (at least 5 mm margin all around) with the shaving of the part of myocardium underlying the stalk to prevent its recurrence after excision.[12]

In the index case, the recurrence occurred within 2 years of excision. Though the exact site of origin of the myxomas resected at the time of first surgery is not known, the presence of PTFE patch in the IAS suggested that the myxomas originated from the fossa ovalis and adequate excision of the IAS was done. The familial and syndromic forms of myxoma were ruled out by the absence of first-degree relative involvements, skin lesions, endocrine organ involvement, and neurofibromas. The occurrence of multiple myxomas in a young male favored a multicentric disease that started growing after the first surgery. Also, the origin of myxoma from the AML favors either familial form or preoccurrence of tumor foci in the AML rather than the implantation of the cells or inadequate excision during the first surgery.

Myxoma usually presents with symptoms of embolization or mechanical obstruction to the flow in the cardiac chambers. Embolization occurs in about 40% of patients particularly to the lungs and brain but may involve almost every organ. Constitutional symptoms like fever, myalgia, arthralgia, and weight loss may be present in about 30% of cases. Nearly 33% of patients may have features of hemolytic anemia due to the obstruction to the blood flow produced by the tumor.[7] Usually, the appearance of constitutional symptoms helps in diagnosing the recurrence. In the index case, there was no symptom suggestive for the recurrence; however, routine follow-up TTE detected the recurrence signifying its importance during the follow-up period.

Even though TTE plays a vital role in diagnosing myxomas, it may fail to detect the lesions presenting in the far-field like right atrium and the posterior wall of LA (Sensitivity TTE-95% Vs TEE-100%).[13] In the index case insertion of TEE probe prior to the central venous catheterization helped in detecting additional myxoma in the superior vena cava-right atrial junction and allowed the insertion of central venous line under continuous monitoring. The guidewire and central venous catheter were inserted under vision to prevent embolization and hemodynamic instability.

Myxoma arising from the mitral valve is rare. Selkane *et al.* found 3 out of 40 myxomas originating from the MV that required mitral valve replacement in all 3 cases.[14] In the index case, TTE examination didn't reveal the exact origin of the left atrial myxoma and further absence of mitral valve regurgitation or stenosis also suggested the absence of myxoma involving the mitral valve, whereas TEE examination revealed the mass originating from the base of AML and the IAS free from the mass. This helped the surgeon in finding the attachment of myxoma, and its excision followed by repair of the AML. Earlier Turhan *et al.* had reported a case of recurrence of myxoma originating from the AML after successful resection of primary myxoma from the left atrium and recommended serial follow-up of these patients after primary surgery.[15] In the index case, routine follow-up after primary surgery using TTE, aided in the early detection of the recurrence and intraoperative TEE helped in the appropriate management of the case.

CONCLUSION

Cardiac myxomas in young males who had previous multiple myxomas may recur after primary excision. Frequent follow-up of such patients using echocardiography may help in the early detection of recurrence. Intraoperative TEE examination may detect additional myxoma and should preferably be done before central venous catheters insertion to prevent embolization.

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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