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Case Report

Surgery Section

Recurrent Left Atrial Myxoma in a Young Patient: A Rare Entity

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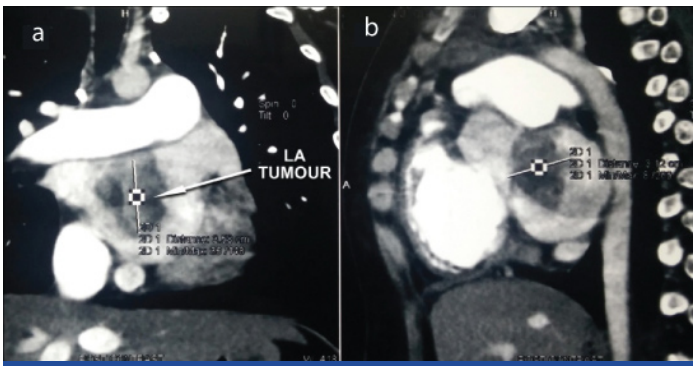
ABSTRACT

Recurrence of atrial myxoma arising from the site other than inter-atrial septum is quite rare, which is more common in familial than sporadic cases. We here in present a case of 15-year-old young female who presented with recurrence of left atrial (LA) myxoma from unusual site – posterior LA wall after 3 years without any constitutional symptoms, which is the hallmark of recurrence. Complete removal of underlying atrial septum with atrial wall for recurrence prevention is the dictum in primary operation for tumour removal.

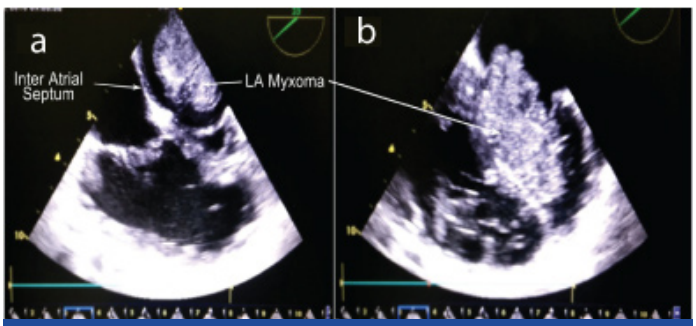
Keywords: Cardiac tumour, Inter-atrial septum, Sporadic

CASE REPORT

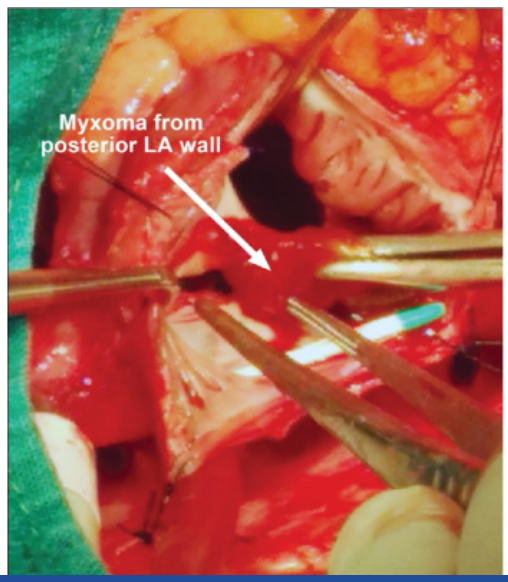
A 15-year-old young female presented with a history of sudden onset of dyspnea on exertion (NYHA class III), easy fatigability and nonproductive cough for 3 months. She had history of Left Atrial (LA) myxoma excision three years back. At the first presentation, three years back patient presented with Dyspnea on exertion. Her 2D echo revealed 38x 33mm large pedenculated freely mobile LA mass arising from inter-atrial septum causing severe mitral inflow obstruction and severe Mitral Regurgitation (MR) with gradient of 34/22 mmHg. So, LA Myxoma removal with excision of Inter atrial septum and closure with untreated autologous pericardial patch was done at her first presentation. There was no positive family history. Physical examination revealed tachycardia with regular heart rate at 113 beats per minute, pedal oedema, and hepatomegaly. Electrocardiogram was inconspicuous. The patient's all haematologic, coagulation and biochemical analyses were normal. Chest radiography revealed moderate cardiomegaly. Two dimensional echocardiogram demonstrated moderate mitral regurgitation, a big LA echogenic mass attached to the inter-atrial septum protruding into the Left Ventricular (LV) cavity causing inflow obstruction with severe TR, severe Pulmonary Arterial Hypertension (PAH) and normal ventricular function. Trans-Esophageal Echocardiography (TEE) pre-operatively confirmed a large pedunculated mass in the LA. The Contrast Enhanced Computed Tomography (CECT) of thorax was performed for confirmation of the diagnosis, which revealed large lobulated mass in the LA attached to lower part of Interatrial Septum (IAS), prolapsing into LV causing severe LV inlet obstruction- suggestive of LA myxoma, with changes of pulmonary hypertension [Table/Fig-1]. There was no history of peripheral or pulmonary embolization. On table TEE after induction showed large LA mass arising from the posterior LA wall, instead of IAS [Table/Fig-2]. After opening, there was a 6x4x3 cm mass; which was pedunculated, glistening, pinkish red and of a gelatinous consistency [Table/Fig-3]. The mass was found to originate from the LA posterior wall between the pulmonary veins protruding into the LV through the mitral valve. The tumour was resected completely with the stump attached to the posterior LA wall, which was shaved off by the trans-septal approach. The IAS was normal and fully endothelized without any tumour extension. The operative and postoperative courses were uneventful. The histology confirmed an identical myxoma as the primary mass. Postoperative echocardiographic examination showed no abnormalities [Table/Fig-4a-c]. She was discharged after 8 postoperative days without any complications. The patient is currently on follow up with medical treatment.



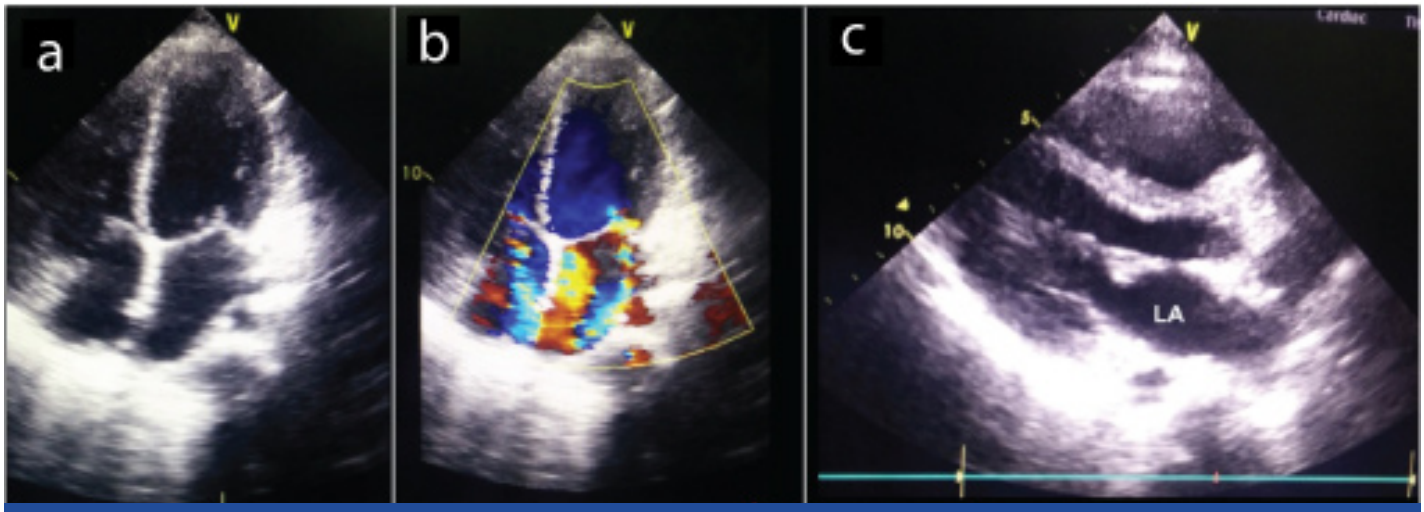
[Table/Fig-1]: CECT Thorax showing LA tumour: a) Coronal plane b) Sagittal plane.



[Table/Fig-2]: On table TEE showing: a) Large LA tumour arising from posterior LA wall with free Inter-atrial septum, b) Tumour going through mitral valve causing inflow obstruction.



[Table/Fig-3]: Intra-op image after opening LA trans-septally showing gelatinous LA mass arising from posterior LA wall.



[Table/Fig-4a-c]: Postoperative Echo showing complete removal of the LA mass with intact IAS: b) Apical 4-chamber view showing excise LA tumour with no residual mass, no flow across IAS, no significant MR; c) parasternal long axis view.

DISCUSSION

Primary tumours of the heart are rare entity with a prevalence rate of around 0.0017% to 0.003% [1]. Atrial myxoma is most common primary benign tumour in adults, which are most commonly found in the LA (75%). They are found more commonly in females (75%) in sporadic cases, while males are affected more in familial cases [2]. They are also found in right atrium (20%) and rarely in ventricular chambers or valves, found usually between the third & sixth decade [3]. Usually it is attached to inter-atrial septum by a stalk in LA in the region of limbus fossa ovalis, but around 10% of LA myxoma arises from other regions including the posterior LA wall, pulmonary veins or mitral annulus [4]. Although, it is a histologically benign disease, recurrence can occur most likely in about 3% of patients, which is higher in familial cases as compared to sporadic cases. Multiple tumours are found in less than 2.5 % of all myxoma cases [5]. Right atrial myxomas tend to be more solid and sessile than left atrial myxomas, with a wider attachment to the atrial wall or septum. It can arise within a single chamber (multicentric) or involve both atria. They are usually polypoid, oval or round gelatinous mass and can be lobulated or smooth. Length of the stalk and septal attachment decides the mobility of tumour [5,6].

The recurrence is around 3% in sporadic cases; while it is higher in familial and complex types of disorder, with recurrence rate of 12% and 22% respectively [3]. The interval between the formation of the new tumour is usually more than 4 years. Unlike primary myxoma, which is more frequent in women, tumour recurrence appears to be more frequent in men [7]. Familial forms, which are more frequently diagnosed in younger individuals, constitute 10% of all myxomas and have autosomal dominant transmission. Gerbode et al., encountered the first recurrence of a myxoma after removal [8]. Since then, there have been reports of recurrences of myxoma both at the site of the primary tumour and at site distant from the primary site. Exact cause for these recurrences has not been known, but several mechanisms can be suggested for recurrences [9].

1) The original tumour may regrow due to incomplete resection.

2) There may be a familial predisposition for recurrence.

3) Embolic fragments of the original tumour may be implanted in the myocardium spontaneously or due to a previous surgery.

4) A pretumourous focus may be present in another part of the myocardium, leading to recurrence.

Mainly, recurrences are due to multicentric disease rather than inadequate resection or seeding of tumour cells during the primary resection. Familial myxomas may present in form of various syndromic associations like Carney's syndrome or NAME syndrome [10]. Symptoms are produced by either mechanical interference with cardiac function or embolization. Embolism occurs in 30-40% of patients and has been reported in every organ system. Constitutional manifestations occur in approximately 30% of cases of cardiac myxoma, and commonly appear as fatigue, fever, rash, arthralgia, myalgia and weight loss. Haemolytic anaemia occurs in approximately 33% of cases, which is due to mechanical destruction by abnormal flow across the tumour [5].

Recurrence of the myxoma in this case could be due to embolization of previous tumour fragments into other part of myocardium or multicentric disease. Usually constitutional symptoms guide towards the recurrence; however, there were no constitutional symptoms present in our case. Although recurrence of myxoma is rare, in our case it points to an invasive nature of the tumour. We feel, therefore, that excision of the underlying atrial septum with shaving of the part of myocardium underlying stalk is necessary during primary myxoma excision for prevention of recurrence; so excision of the atrial septum & closure by untreated autologous pericardium is the dictum in all atrial myxoma excision cases.

There are difference of opinion regarding the ideal extent of surgical excision, however, it is preferable to excise the tumour with wide cuff of the inter-atrial septal cuff (at least 5mm margin all around) with excision of the pedicle, with or without cauterization of the base to which tumour was attached [5,11]. However, according to some data in literature, excision of the underlying atrial septum or wall is justified in every case, as such a procedure will be more accurate in keeping with the surgical principles of tumour surgery without adding greatly to the technical difficulties of the operation. The defect created in the septum or the atrial wall can be easily closed primarily or with a pericardial or Dacron patch to restore normal anatomical and physiological function [12].

CONCLUSION

Recurrence rate of the most common cardiac, primary benign tumour–atrial myxoma are rare, especially from uncommon sites like; posterior LA wall, mitral annulus & pulmonary veins. Multicentricity & local spread is most common cause of recurrence. Famlial syndrome is a strong possibility in case of recurrence, which is usually suspected in case of clinical features & cardiac lesions suggesting syndromic association. Trans-septal excision with removal of part of myocardial wall to which stalk is attached is mandatory for recurrence prevention.

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