Non-Familial Left Atrial Myxoma in an Adolescent: A Rare Mimic of Mitral Stenosis

Navin Agrawal, DM, Soumya Patra, Ashish Agarwal, Kamal Gupta, Sunil Srinivas, Rajni Sharma and Manjunath CN

Department of cardiology Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru 560069, Karnataka, India

ABSTRACT

We are presenting a rare case of large non-familial left atrial myxoma in a fourteen year old girl which presented clinically similar to severe mitral stenosis which was the first clinical differential diagnosis which was considered initially. She had symptoms of progressively worsening orthopnea and exertional dyspnea since the last 2-3 months (NYHA class III symptom status). The final diagnosis which was made after echocardiography was surprising and the girl was diagnosed to have a large atrial myxoma which was causing obstruction of the mitral valve preventing normal inflow across the mitral valve. The patient had no family history of being diagnosed of a cardiac tumor nor there were any cutaneous manifestations in the form of lentigenes or any symptoms suggestive of and systemic or cerebral embolization of tumor fragments. The patient subsequently underwent surgical excision of the tumor which was eventually confirmed to be an atrial myxoma histopathologically. Presence of atrial myxoma in an adolescent patient is an extremely rare occurrence and has seldom been reported.

Keywords: Atrial myxoma, non familial echocardiography

BACKGROUND

Cardiac tumours can present with symptoms related to obstruction, embolism, and constitutional symptoms which might be a manifestation of cytokine production. Left atrial myxoma is a rare mimic of mitral stenosis and can present in a clinically identical way to mitral stenosis. LA myxoma is usually a disease of the elderly and has been reported very rarely in the young but this although being an extremely rare presentation should always be kept in mind as a cause of pulmonary venous hypertension in patients presenting as orthopnea and exertional dyspnea.

Corresponding address:
Dr. Navin Agrawal
Sri Jayadeva Institute of Cardiovascular Sciences & Research,
Jaya Nagar 9th Block, BG Road,
Bangalore 560069, INDIA
Mobile: 9739084640
E-mail: drnavinagrawal@gmail.com

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We are presenting a rare case of large non-familial left atrial myxoma in a fourteen year old girl which presented clinically in a way similar to severe rheumatic mitral valvular heart disease which was also the first clinical differential diagnosis which had been considered in her.

Case Description

A fourteen year old girl presented with orthopnea and exertional dyspnea since the last four months which had been progressively worsening in the last 2–3 months from class I to NYHA class III symptom status. There was no history of any chest pain, palpitations, dizziness or syncope. The patient had a history of on and off low grade fever and malaise since the last few months. There was no family history of any cardiac illness or any similar symptom.

The patient presented in the out-patient department and the initial clinical evaluation was very suggestive of mitral stenosis which is the obvious diagnosis when a patient of the adolescent age group presents with orthopnea and

exertional dyspnea in an underdeveloped or developing country with widespread distribution of rheumatic fever ad rheumatic heart disease. The girl had an early diastolic sound suggestive of an opening snap but was actually a tumor plop and a prominent mid-diastolic murmur.There was ECG evidence of left atrial enlargement but there were no sign of any arrhythmias or conduction blocks. The blood investigations revealed normal hemoglobin levels and normal white blood cell counts.The presence of an LA myxoma was just a theoretical possibility in the minds of the clinician who had initially evaluated the patient as the possibility was extremely unlikely in an adolescent.

The echocardiographic images came as a real surprise for us. The echo revealed the presence of a large atrial myxoma in the atrial cavity causing obstruction of the mitral valve in diastole by prolapsing through the valve apparatus (Figure 1–2, Videos 1–4).

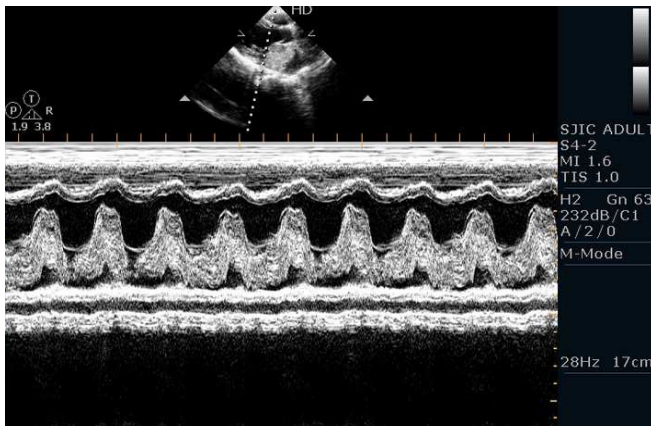

Figure 1: M mode across the mitral valve showing valve obstruction by the left atrial myxoma

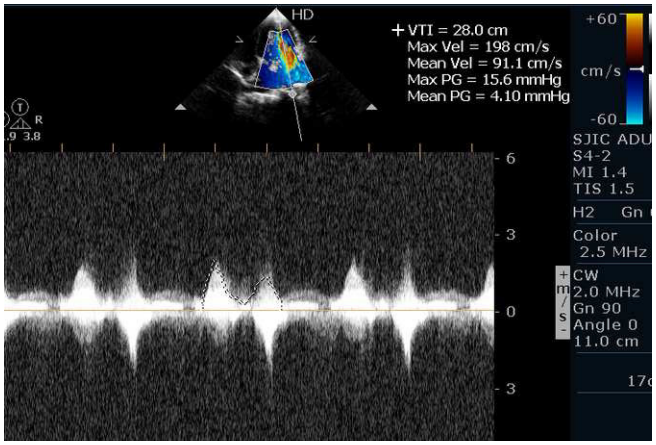

Figure 2: Continuous wave Doppler across the mitral valve showing diastolic gradient across the valve

The patient had no family history of being diagnosed of a cardiac tumor nor there were any cutaneous manifestations in the form of lentigenes which form the most common cutaneous manifestation of a familial myxoma. The patient`s close relatives were also screened for the presence of myxomas.There was no history of any symptoms suggestive of systemic or cerebral embolization of tumor fragments. The patient was subsequently planned for a surgical excision of the large tumor which was subsequently performed successfully and this lead to dramatic reduction of her symptoms and the patient was completely asymptomatic at the time of her discharge and has maintained her asymptomatic status during follow up.

The excised mass was examined histopathologically and was confirmed to be an atrial myxoma. The patient has been stable at 10 months of follow up now and so far there is no evidence of any recurrence of the tumour. The patient is planned for follow up echocardiograms every six months or every year to assess for any recurrence of the tumour in the future.

The patient was not evaluated for the genetic mutation for Carney`s complex predominantly because of financial constraints and unwillingness on the part of the relatives and also with the clinical suspicion being relatively low on account of the absence of the other characteristic features of the syndrome.

DISCUSSION

Cardiac tumor scan present with various symptoms which could include symptoms related to obstruction, embolism, and constitutional symptoms which might be a manifestation of cytokine production. The presenting symptoms are generally related to the site and friability of the tumor than to the tumour type. The rate of embolization is higher with myxomas, especially those with an irregular surface, and papillary fibroelastomas. Embolic phenomena occur in 30–40% of patients most frequently to the central nervous system, kidney, spleen and extremities. Smooth surfaced tumors are more likely to produce valvular obstruction, while polypoid and myxoid ones are more likely to embolise. The treatment of most of the cardiac tumors is primarily surgical except for the lymphomas.

In over 50% of patients left atrial myxomas cause symptoms of mitral valve stenosis or obstruction (dyspnea and orthopnea from pulmonary oedema or heart failure). Right atrial myxomas may obstruct the tricuspid valve and cause symptoms of right sided heart failure.Anemia,

leukocytosis and raised erythrocyte sedimentation rate (ESR) are the most common laboratory findings. Constitutional symptoms(possibly related to interleukin IL-6 production by tumour cells) seen in approximately 20% of patients include myalgia, muscle weakness, arthralgia, fever, fatigue and weight loss. About 20% of cardiac myxomas are asymptomatic; incidental myxomas are usually smaller than 40mm. Abnormal, but non-specific, electrocardiographic changes may be identified in 20–40% of patients which include atrial fibrillation.

Less than 5% of myxomas form a component of the myxoma complex[1], which includes abnormal skin pigmentation (lentigenes and blue nevi), calcifying Sertoli-Leydig testicular tumours, cutaneous myxomas, myxoid breast fibroadenomas, pigmented adrenal cortical hyperplasia, pituitary hyperactivity, psammomatous melanotic schwannoma and thyroid tumours. In contrast to patients with sporadic tumours, who have a 1% recurrence rate, about 10% of patients with familial myxomas either have recurrent tumours or develop another tumour in a different location[2]. Patients with myxoma complex have cardiac myxomas at a young age, and are more likely to have multiple tumours or tumours in locations other than the atria. Our patient was unique in having a non-familial cardiac myxoma at a very young age which is rarely seen in young and in more common in males in the younger age group as compared to females in which myxoma is more common in the elderly age group.

Because embolisation is the major complication of myxoma, especially of myxoid, friable, familial ones, identification of first degree relative's of patients with documented myxoma syndrome is important. Intracranial aneurysm caused by embolisation is also a rare, but potentially morbid, complication. The etiology of these aneurysms is unclear but histologic verification of myxoma cellsin arterial walls has been reported.

The symptoms of myxoma as predominantly dependent on the size of the tumour and the extent of disruption of the valve function which is caused. The larger tumours are potentially more symptomatic and mimic the symptoms of mitral stenosis which is very common in the developing countries. The only common difference which is seen in the symptoms caused by myxoma is the presence of postural variations which is caused by the difference in the extent to which the myxoma impinges and obstructs the mitral valve. The site of the myxoma also affects the symptoms and the closer the myxoma is present to the valve the more pronounced are the symptoms. Left atrial myxomas are usually more symptomatic as the compared to right atrial myxomas predominantly because of the larger size of the tricuspid valve and the potential of tricuspid regurgitation to be asymptomatic for longer periods as compared to mitral regurgitation.

As the majority of left atrial myxomas arise from the inter-atrial septum,[3] The tumours can be removed en bloc with a 5 mm margin of normal tissue. The fossa ovalis, where the pre-tumour cells of myxomas are thought likely to exist, should also be excised if possible. Resection of the attachment, and 5 mm of normal tissue including endocardium and underlying myocardium, is generally recommended. Despite anecdotal reports of recurrence after incomplete excision,[4] There are no data that clearly link negative margins at the time of surgery with an increase in the recurrence rate.

CONCLUSION

Atrial myxoma is an extremely rare occurrence in adolescent age group and this case forms a learning opportunity to consider the presence of an atrial myxoma in an adolescent when a patient presents with symptoms of exertional dyspnoea and orthopnea which are classically described in cases with rheumatic heart diseases which are a common in adolescent population in under developed countries where the prevalence of rheumatic heart disease is still rampant. Non familial myxoma is unlikely to is an uncommon occurrence is adolescent and familial myxoma is common in males, hence this case in an adolescent female is an extremely rare and seldom reported occurrence.

VIDEO FILE LEGENDS

Video 1:-Parasternal long axis view showing a large atrial myxoma causing obstruction of the mitral valve in diastole by prolapsing through the valve apparatus

Video 2:-Color Doppler in parasternal short axis view across the mitral valve showing the comparison with the same view without color doppler

Video 3:-Parasternal short axis view at the mitral level showing a large left atrial myxoma bobbing through the valve

Video 4:-Apical four chambered view showing the large left atrial myxoma

Video's are available in online only

REFERENECES

1. Burke AP, Tazelaar H, Gomez-Roman JJ, et al. Benign tumors of pluripotent mesenchyme. In: Travis W, ed. Tumours of the lung, thymus, pleura and heart. Lyon: Springer Verlag. 2004: 260–5.
2. Carney JA. Differences between non familial and familial cardiac myxoma. Am J Surg Pathol . 1985; 9: 53–5.
3. Araoz PA, Eklund HE, Welch TJ, et al. CT and MR imaging of primary cardiac malignancies. Radiographics. 1999; 19: 1421–34.
4. Kosuga T, Fukunaga S, Kawara T, et al. Surgery for primary cardiac tumors. Clinical experience and surgical results in 60patients. J Cardiovasc Surg (Torino). 2002; 43: 581–7.