

Variable Presentations of Left Atrial Myxomas: A Case Series

Muhammad Uwais Ashraf¹, Asif Hasan², Mohd. Aslam³, Juwairia Ashraf⁴

^{1,2,3}Department of Medicine, J N Medical College, AMU, Aligarh, UP, India

⁴AKTC, AMU, Aligarh, UP, India

Abstract: Left atrial myxomas are commonly missed clinically and often lead to grave consequences. A high degree of suspicion is required for diagnosing myxomas clinically. We present here a series of three cases which presented with absolutely different clinical signs and came out to be myxomas on echocardiography. The first case was a female who presented with shortness of breath and anasarca and was being clinically treated as COPD with cor pulmonale, the second was a 70 year old female who had presented with dyspnea on exertion and was being treated as mitral stenosis, and the third case was a research scholar who was being treated as bronchial asthma.

INTRODUCTION

Intracardiac myxoma is the most common benign tumor of the heart. Most of the myxomas (75%) are located in the left atrium, although, they may also arise from right atrium (18%); right ventricle (4%); and left ventricle (3%) [1]. Cardiac myxomas usually originate from the region of the fossa ovalis but may also arise from a variety of locations within the atria [2]. Approximately 5% of myxoma patients show a familial pattern of tumor development. Most true myxomas arise only from the mural endocardium despite isolated reports that they arise from the cardiac valves, pulmonary vessels and vena cava. Although asymptomatic patients with myxomas have been reported, most present with one or more effects of a triad of constitutional, embolic and obstructive manifestations. Cardiac myxomas provoke systemic manifestations in 90% of the patients, characterised by weight loss, fatigue, fever, anemia, raised ESR etc. We present here, four case reports of left atrial myxomas, all having different clinical presentations. It requires a high degree of suspicion to diagnose a cardiac myxoma because of the variable presentations of this disease. Most of our cases had manifestations of other more common cardiac diseases and hence few of them were diagnosed quite late in the course of the illness.

SUMMARY OF THE CASES

Case 1: A 24 year old female, presented to us with complaints of shortness of breath on exertion for six months. It was gradually progressive and was followed by anasarca for three months. On examination, JVP was raised, pedal oedema and ascites were present, S₁ was loud and split, P₂ was also loud. No murmur could be appreciated. Echocardiography revealed a mass arising from left atrium and protruding via mitral valve (Fig. 1).



Fig.1

Case 2: Another patient, a 70 years old female, presented to us with complaints of dyspnea on exertion grade II and palpitations. On general examination, no significant finding was noted. On cardiovascular examination, S₁ was loud, P₂ was also loud, a diastolic murmur was heard, similar to the murmur of mitral stenosis. A provisional diagnosis of mitral stenosis was made. On echocardiography, a left atrial mass, of the nature of a myxoma was detected (Fig. 2)



Fig. 2

Case 3: Another patient, a 26 year old female research scholar, presented to us with complaints of shortness of breath on exertion for two years. Patient was being treated in the pulmonology OPD as a case of bronchial asthma. On increasing symptoms, she presented to the coronary care unit. At presentation, her blood pressure was 100/60 mmhg; pulse rate was 110 per minute; patient had severe orthopnea; respiratory rate was 36 per minute; pedal edema was present; on chest auscultation, bilateral decreased air entry was found with dullness on percussion, suggestive of bilateral pleural effusion; on cardiac auscultation, muffled heart sounds were present. EKG showed low voltage complexes. A provisional diagnosis of pericardial effusion was made and pericardiocentesis was done which revealed an exudative picture. ATT was started on the assumption of tuberculous pericardial effusion. There was no improvement in the patient's condition. When echocardiography was done, it revealed a left atrial tumor (Fig 3). Patient was referred to higher centre, where she was operated, however, she succumbed to her illness after one day of the surgery.

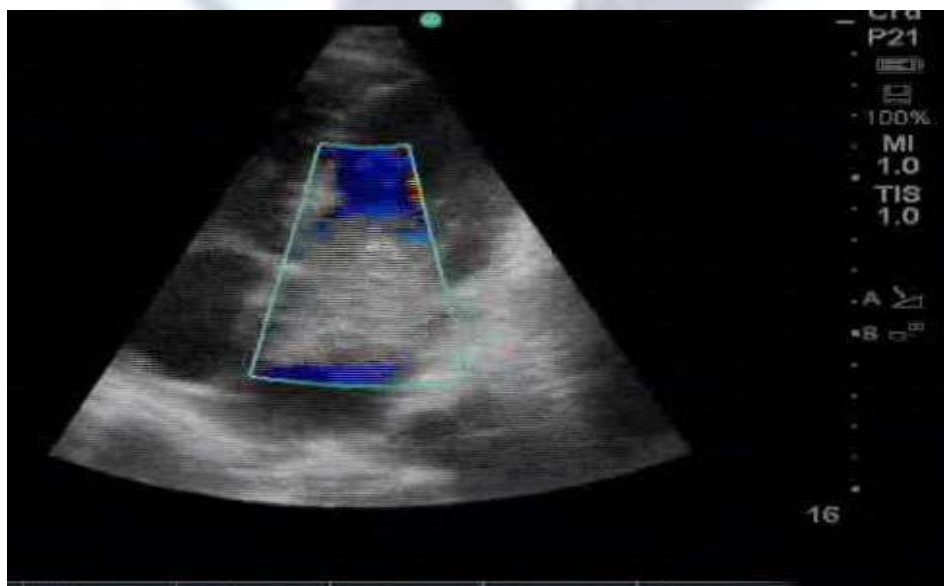


Fig. 3

Case 4: A 50 years old female patient presented to us with complaints of recurrent syncopal attacks. On examination, pulse rate was 66 per minute, blood pressure was 120/86 mmhg, cardiovascular examination was normal. EKG showed right bundle branch block. Echocardiography revealed a dense array of wavy tumor echoes behind the anterior mitral valve leaflet, commensurate with the description of a myxoma (Fig. 4).



Fig. 4

DISCUSSION

Earlier, there was a debate whether cardiac myxoma was a neoplastic entity or an organised thrombus, recent gene expression and immuno histochemical studies have shown that cardiac myxoma is a neoplasm with tumor cells arising most likely from multipotent mesenchymal cells [3]. Despite several documented reports of metastasis to various anatomic sites [4], the typical cardiac myxoma is regarded as a benign neoplasm. Although histopathologically benign, cardiac myxomas can cause chronic systemic inflammation, embolism, or intracardiac obstructions.

The pathogenesis of cardiac myxoma is poorly understood, especially for those that are sporadic. Studies have, however, shed more light on the pathogenesis of familial cases of cardiac myxomas. Carney syndrome accounts for the majority of familial cases of cardiac myxomas and 7% of all cardiac myxomas [5]. It is an autosomal dominant syndrome characterised by myxoma formation in cardiac and several extra-cardiac locations, spotty skin pigmentation, endocrine hyperactivity, and other tumors (such as testicular sertoli cell tumors, psammomatous melanotic schwannoma, pituitary adenoma and thyroid tumors). Clinically, patients with symptomatic cardiac myxomas can have a myriad of clinical manifestations, depending upon the size, location and mobility of the tumor. Our case series has shown, how diverse and confusing can be the clinical presentations of cardiac myxomas.

Around one-third of the patients of cardiac myxomas have abnormalities in the EKG, in the form of evidence of left atrial enlargement, atrial arrhythmias or rarely, conduction disturbances [6,7]. The typical imaging modalities used for diagnostic and pre-operative assessment purposes include echocardiography, CT scan and cardiac MRI. The treatment of symptomatic cardiac myxoma is prompt surgical resection of the tumor [8]. Complete excision is the goal, although it may not be possible in all the cases. Recurrence occurs in around 3% of the tumors, and the risk of recurrence is higher among familial cases [9].

REFERENCES

- [1]. Yeh ET, Courtney L et al. Hurst's; The Heart. Mc Graw Hills publications. 13th edition, volume 2. Page 2025.
- [2]. Cardiac myxoma: its origin and tumor characteristics. Amano J, Kono T et al. Ann Thorac Cardiovasc Surg. 2003 Aug;9(4):215-21.
- [3]. Terracciano LM, Mhawech P et al. Calretinin as a marker for cardiac myxoma. Diagnostic and histogenetic considerations. Am J Clin Pathol 114:754, 2000.
- [4]. Rodrigues D, Matthews N. Recurrent cerebral metastasis from a cardiac myxoma: case report and review of literature. Br J Neurosurg. 2006 Oct;20(5):318-20.
- [5]. Wilkes D et al. Inherited disposition to cardiac myxoma development. Nat Rev Cancer 6:157, 2006.

- [6]. Pinede L, Duhaut P et al. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine (Baltimore)* 80:169, 2001.
- [7]. Acebo E et al. Clinicopathologic study and DNA analysis of 37 cardiac myxomas: A 28 year experience. *Chest* 123:1379, 2003.
- [8]. Rosenberg FM, Chan A et al. Cardiac neoplasms. *Curr Treat Options Cardiovasc Med.* 1:243, 1999.
- [9]. Multiple recurrences of nonfamilial cardiac myxomas: a report of two cases. Mendoza C, Bernstein E, Ferreira A. *Tex Heart Inst J.* 2007;34(2):236-9.

