Case Report

Atrial Myxoma: A Rare But Treatable Cause of Breathlessness

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The primary cardiac tumors are extremely rare but among them the benign myxoma is the commonest and that usually arise in left atrium. The classic triad of clinical manifestations are due to embolism, intracardiac obstruction and mainly constitutional symptoms which are non-specific i.e. malaise, fever, anorexia, arthralgia and weight loss and thus may create a challenging diagnostic dilemma, however the size, site and mobility of tumor plays a crucial role in symptomatology. The patient had shortness of breath that closely mimicked the pre-existed COPD and systemic hypertension, however this discomfort developed within a very short span of time with the patient adopting a supine posture necessitating a2D ECHO. The ECG/ D-dimer etc. were essentially normal. These types of masses also need to be differentiated from an intramural thrombus. The cardio thoracic vascular surgeon (CTVS) after an initial work up, successfully resected the benign myxoma by median sternotomy and the patient got marked relief of her symptoms.

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Introduction

Cardiac tumors are extremely rare, with an incidence of about 0.02% in autopsy series.[1] It was first diagnosed in 1952 by Goldberg.[2] These tumors are broadly classified into primary when they originate from the heart, and are called secondary when there is direct extension from the neighboring structures or metastasis from the distant primary source.[3] Primary cardiac tumors can further be classified into benign and malignant on the basis of histopathology. Primary cardiac tumors are less often found, whereas secondary involvement of the heart and pericardium occurs in about 20% of patients with end-stage metastatic cancer.[3] Among the primary cardiac tumors, the benign myxoma is most common and usually found intra-cavitary and mainly affects the atrial chamber, that too from the inter-atrial septum near the fossa ovalis as a commonest site.[2] Despite non-specific symptoms, its clinical presentation mostly depends upon tumor size, anatomical location, mobility and relationship with the surrounding structures.[1,4] The prognosis is excellent with the surgical resection of myxoma; therefore, a good clinical suspicion with accurate workup is critical in timely management to prevent the long-term morbidity and permanent damage to the cardiac valves and their associated apparatus.[4] The present case became interesting because of a typical clinical presentation of breathlessness in an aged patient having dual co-morbidities, the accidental detection of myxoma with timely surgical management due to an efficient team effort working at a tertiary rural Medical College.

Case Report

A 72-year-old female was admitted to the respiratory medicine ward with the chief complaints of breathlessness (mMRC grade-III) with positional variation while lying down for 1 month. She also had generalized weakness for 15 days, followed by fever with cough and expectoration for the last 8 days. Her general condition was poor with BP-130/80 mm Hg, Pulse 108/min, RR 24/min, SpO2 95% and BMI of 21 kg/M^2. The pallor, Icterus, cyanosis, clubbing, lymphadenopathy and pedal edema were absent and JVP was not raised. The respiratory system on auscultation revealed bilateral rhonchi throughout the chest without other adventitious sounds. The cardiovascular, gastrointestinal and CNS were normal;

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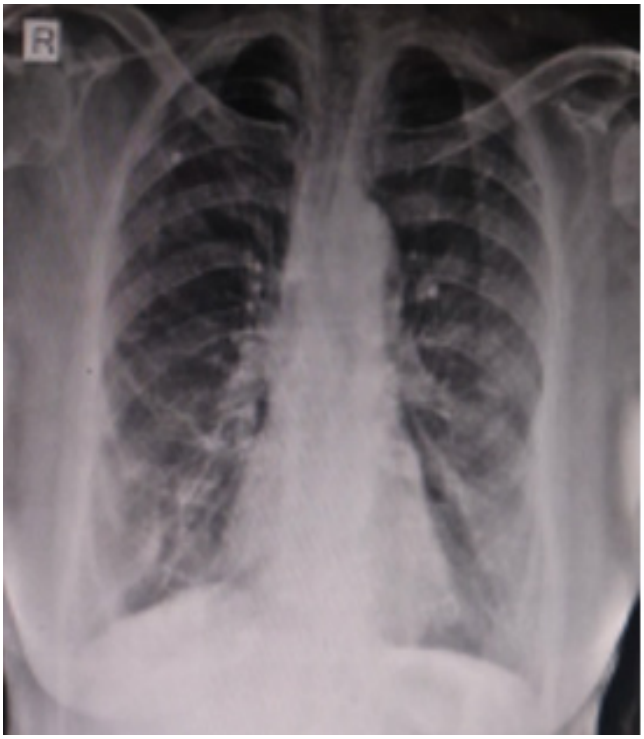

Figure 1: Chest radiograph PA view showing hyperinflated lungs

except that she was a known case of systemic hypertension and COPD for which she was on regular medication for the last 4 years. She has a history of exposure to biofuel during cooking with Chulha. No history of smoking, CAD, diabetes, ATT, surgery and blood transfusion. The routine investigations were essentially normal. The Chest radiograph shows hyperinflated lung fields with increased broncho-vascular markings (Fig. 1). An initial symptomatic and supportive treatment with an antibiotic and bronchodilator with nebulization was started for the acute exacerbation of COPD and there was improvement of oxygenation without BiPAP support. The D-dimer, ECG, ABG and cardiac markers were within normal limits. On the 3rd day of admission, the bronchospasm was relieved, but the patient continued to experience persistent dyspnea in adopting supine position. Hence, a transthoracic 2D ECHO was done that accidentally revealed a large left atrial mobile mass of the size approximately 2.7 x 2.4 cm, likely a myxoma, that was attached to the inferolateral septum and obstructing the mitral valve (Fig 2) with an ejection fraction of 60%. A Cardiothoracic and vascular surgeon's opinion was sought, and surgical resection was performed (Fig. 3) via median sternotomy. The thymus was dissected, a pericardial patch was prepared and partial bypasses were initiated. Root cardioplegia was given to achieve diastolic arrest of the heart, the left atrial myxoma was excised, and the atrium was closed in two layers. Histopathological examination of the excised tissue/mass confirmed features consistent with a final diagnosis of benign atrial myxoma (Fig. 4). The postoperative period remained event-free with an excellent recovery on scheduled follow-up.

Discussion

Cardiac myxoma is the most common benign tumor of the heart, accounting for 50% of all benign cardiac tumors, yet it remains rare with an annual incidence of 0.5 per million.[4,5] Myxoma mainly affects atrial chambers and

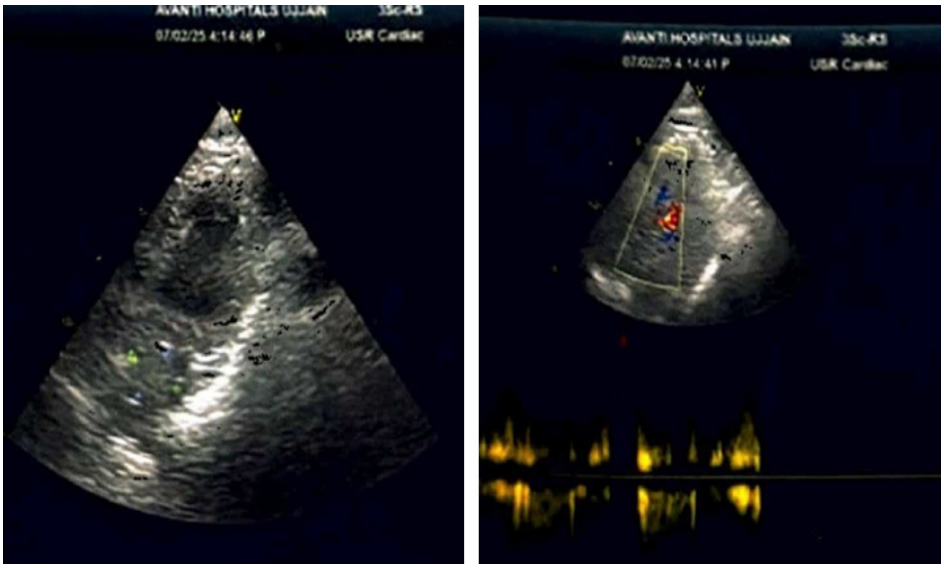

Figure 2: 2D Echo findings showing left atrial mobile mass obstructing mitral valve

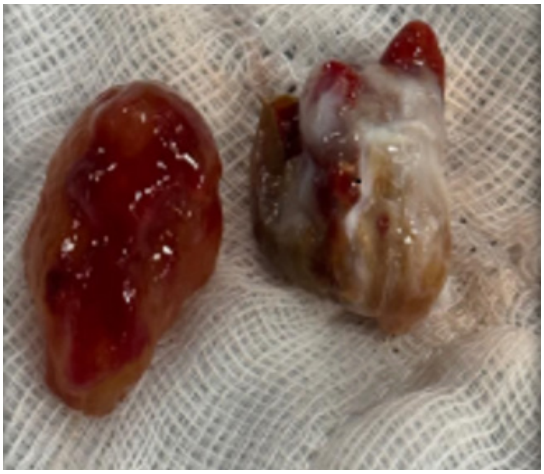

Figure 3: Resected gelatinous atrial myxoma

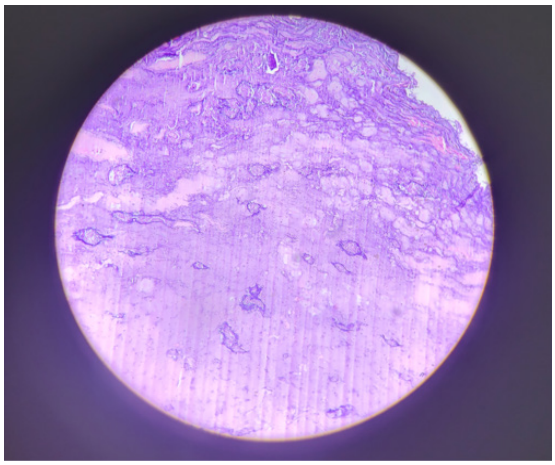

Figure 4: 10x microscopic picture showing inflammatory cell infiltrates within myxoid stroma

75% occur in the left atrium and arise from the interatrial septum near the fossa ovalis, as also happened in our case, which too originated from the inferolateral part of the interatrial septum.[2] The myxoma cells are believed to originate from local pluripotent or multipotent mesenchymal stem cells and during the development of heartsome of these stem cells may remain in the septum that later differentiate into endothelial cells, smooth muscle cells, and other mesenchymal tissues. Somehow, this process explains why myxomas commonly occur in the atrial septum.[6] However, our case was a 72-year-old female who might have remained asymptomatic due to slow growth, size and location of the myxoma.

The myxoma can be familial (rare) or present as sporadic cases that constitute about 95% and are solitary and commonly arise in the left atrium from the fossa ovalis.[2,6] The familial type is characterized by an autosomal dominant mutation of the PRKAR1A gene located on chromosome 17q2. In addition, the familial myxomas present with a part of the syndrome complex (Carney complex) that includes myxomas (cardiac, skin, and/or breast), lentigines or pigmented nevi, and endocrinopathies.[2,3,5,8] In our case, the myxoma was solitary, located in the left atrium, and did not exhibit familial predisposition; thus, it was likely a sporadic origin.

Clinical presentation of myxoma is non-specific and mainly depends upon the size, mobility and location, thus has wide variability in presentation from being asymptomatic to life-threatening.[6,8] The classic Goodwin's triad of clinical manifestations is due to embolism, intracardiac obstruction, and constitutional symptoms.[4,6] The interleukin-6 released by the myxoma could be the reason for the non-specific constitutional symptoms like malaise, fever, anorexia, arthralgia and weight loss[4,7] and may create a challenging diagnostic dilemma; however, except for fever, these were not dominant in our case. In our case, obstructive symptoms were more pronounced with the dyspnea getting aggravated while lying supine. In an elderly COPD patient, these non-specific symptoms initially mimicked an acute exacerbation of COPD,[4,8] but after giving all symptomatic and supportive treatment to our patient, there was only a mild relief in symptoms, so ECHO was done. The ECHO was planned to rule out any cardiac pathology, PAH, etc., though there was no clinical evidence of the same.[4-6,9] Casavecchia G et al have concluded in their review article that cardiac tumors are likely to be misdiagnosed with other cardiac conditions (including rheumatic valvular disease, cardiomyopathies, and congenital heart disease, etc.) and pulmonary conditions (including pulmonary emboli and pulmonary hypertension, etc). He further mentioned that ECHO remained the first choice, yet CT, MRI and FDG-PET/CT may have a contributory role in precisely differentiating benign from malignant tumors.[1] On an occasion intramural thrombus needs to be differentiated from an intracardiac mass lesion by the fact that they do not have resting perfusion on a Doppler ECHO study.[9] The myxoma is further classified on the basis of morphology into solid and papillary types. The obstructive symptoms are more common in the solid type (as in our case), whereas the embolic symptoms are more common in the papillary type.[10]

The treatment of choice is surgical excision of the tumor, and the resected tumor must be sent for histopathological examination to rule out malignant or metastasis nature.[7] As is evident in our case, the histopathological examination of the resected tumor confirmed its benign

nature. The overall surgical result remained excellent with survival and relapse; however, an annual follow-up is recommended, especially in familial type myxoma.[2]

Conclusion

In simple words, cardiac myxoma can potentially be an emergency, as there are many serious complications that can lead to morbidity and mortality. So, early diagnosis and resection are key to survival in these patients. The echocardiography plays a major role in incidental or intentional detection. Clearly, echocardiography can not be routinely performed for all patients presenting with dyspnea and bronchospasm, but cardiac causes and differentials must be kept in mind if the patient is not adequately responding to initial symptomatic therapy. The correct diagnosis and surgical management of this rare condition greatly help in improving outcomes.

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