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Rare Myxoma Arising from Posterior Wall of Left Atrium

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Cardiac myxoma is an extremely rare condition, with an annual surgical incidence of 0.5 per 1,000,000 and an estimated incidence of 0.0017–0.19% at autopsy [1,2]. Although rare, it is the most common benign primary tumor of the heart [3]. The treatment is urgent surgical removal, and the post-treatment prognosis is excellent with low operative and postoperative mortality [4]. Diagnosis of cardiac myxoma is difficult as it presents with varying non-specific symptoms [4]. Such patients are at risk since untreated cardiac myxoma can lead to sudden cardiac death or embolization.

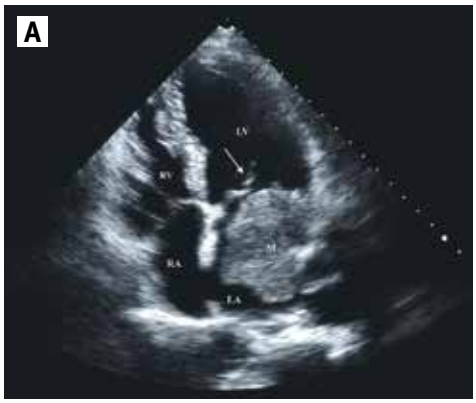
Confirmation of a diagnosis of cardiac tumor is most often by echocardiogram but can also be by magnetic resonance imaging (MRI), computed tomography (CT), or angiography [3,4]. True diagnosis of myxoma can be reached only by histology after excision of the mass.

Seventy-five percent of cardiac myxomas are found in the left atrium, and of the left atrial myxomas 80% arise from the fossa ovalis [4]. Only 5% of left atrial myxomas (or 3.75% of all cardiac myxomas) are found attached to the left posterior wall of the left atrium [4]. Care must be taken at surgical removal to ensure that the entire mass is removed as any fragment presents a risk of embolization [3]. This is especially important if the myxoma ruptures during removal.

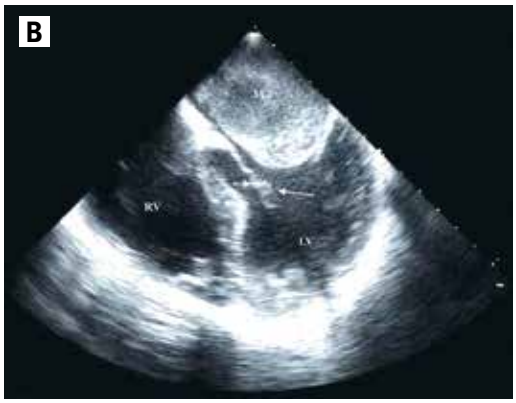
***The first two authors contributed equally to this study**

PATIENT DESCRIPTION

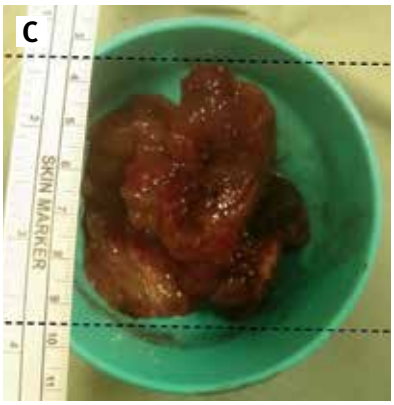
A 51 year old female presenting to the emergency room of Sheba Medical Center, Tel Hashomer, complained of shortness of breath, exertional dyspnea, dizziness and heart palpitations. Physical examination revealed tachycardia and tricuspid regurgitation, without other significant findings. Echocardiogram revealed a mass in her left atrium of approximately 44.5 x 56.5 mm [Figure A and B] and confirmed severe tricuspid regurgitation as well as severe pulmonary hypertension. She was scheduled for emergent surgical removal. During surgery the mass ruptured, requiring extra care to ensure that no fragment of the mass was left in the left atrium. Gross examination of the mass revealed a smooth pedunculated mass of 60 x 50 x 30 mm [Figure C], with a stalk 20 mm in diameter, arising from



[A] Image of left atrial myxoma on trans-thoracic echocardiogram. The white arrow indicates open mitral valve during diastole. Of note is the small gap between the mitral valve and the myxoma allowing blood flow, albeit highly restricted.

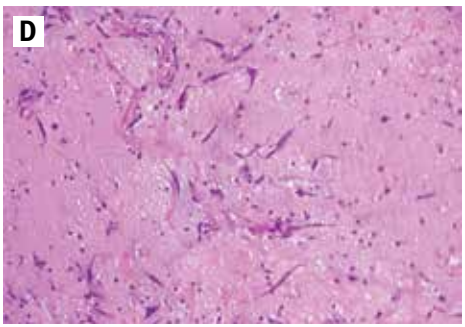


[B] Image of left atrial myxoma on trans-esophageal echocardiogram. White arrows indicate mitral valve. Of note is the small gap between the mitral valve and the myxoma allowing blood flow, albeit highly restricted.



[C] Gross appearance of left atrial myxoma after removal. Note the myxoma had ruptured during the process of removal. Approximate measurement is 60 mm in length. When sent to histology, the tumor was measured and confirmed to be 60 x 50 x 30 mm.

M = myxoma, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle



[D] Histological appearance of left atrial myxoma (hematoxylin & eosin, x200), 75 g of greyish-pink gelatinous tissue fragments. Microscopically the tumor was composed of cords and rings of bland-looking myxoma cells, displaying spindled, stellate and slender nuclei and an eosinophilic cytoplasm. The cells were embedded in a myxoid edematous matrix, which contained scattered vascular channels.

the left posterior wall of the left atrium. The diagnosis was confirmed histologically [Figure D]. Post-surgical examination of the patient revealed normal heart sounds without murmurs, clear lungs, and no signs of deep vein thrombosis.

COMMENT

Cardiac myxoma is a rare and insidious disease. It often presents with non-specific symptoms only, such as fatigue, fever, myalgia, erythematous rash, arthralgia, weight loss, as well as laboratory abnormalities [3]. This makes myxoma difficult to diagnose, and it is often fatal if untreated. It is interesting to note that unlike with most other tumors, death by mechanical obstruction is much more dangerous than metastasis or end-organ failure. Treatment of cardiac myxoma also differs significantly from treatment of other tumors. Whereas most tumors carry the danger of metastasis if there is incomplete removal, incomplete cardiac myxoma removal can result in embolization and subsequent ischemic attack. However, there are still similarities between myxoma and other tumors: incomplete removal carries the danger of cardiac myxoma recurrence [3].

Cardiac myxomas generally arise from the fossa ovalis of the inter-atrial septum [4]. Our patient's myxoma was rarer, arising from the posterior aspect of the left atrial wall. As a result the removal was more difficult, as the stalk of the tumor was more difficult to reach during surgery. Care must be taken to remove the myxoma in its entirety in order to reduce the risk of embolism and recurrence, as mentioned earlier.

Our case illustrates one of the greatest dangers of myxoma – the danger of misdiagnosis. The patient's presenting symptoms of shortness of breath, exertional dyspnea, dizziness, and heart palpitations cover a vast differential of diagnoses. Her X-ray showed no findings indicative of myxoma. Without the echocardiogram, it is most likely that a diagnosis of myxoma would have been missed and the patient would not have received appropriate care. Our case further highlights the necessity for echocardiogram (or other imaging such as CT or MRI) whenever cardiac myxoma is suspected.

In conclusion, cardiac myxoma is a rare type of tumorous growth found in a very small subset of the population. It presents with non-specific symptoms and the treatment is emergency surgery. Echocardiogram is the single most important method to diagnose a cardiac myxoma, as it is non-invasive, cost-effective and highly sensitive. We have presented a case of a rare cardiac myxoma arising from the posterior aspect of the left atrial wall, diagnosed by echocardiogram and confirmed by histological analysis.

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