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Left Atrial Myxoma: A Case Report and Review of Literature

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Abstract Primary cardiac tumors are an infrequent cause of symptoms mimicking valvular heart disease, congestive heart failure, arrhythmias, conduction defects and pulmonary disease. The rarity of this condition can lead to significant delay and cause patient distress. Here, we present a case of an unusual prolapsing left atrial myxoma in a 61-year-old female who presented with several months of dyspnea. The patient endorsed weight loss and night sweats during this period and treatment with antibiotics and steroids for pulmonary symptoms had not provided relief. Seven months after her initial presentation, an echocardiogram was performed revealing a 2.85 x 5.44 cm left atrial mass on the interatrial septum. The mass was resected by cardiothoracic surgery with complete resolution of her symptoms.

Keywords: *myxoma, benign tumor, cardiac tumors, left atrial tumor*

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1. Introduction

The incidence of primary cardiac tumors varies between 0.0017 to 0.19 % [1]. Majority are benign and nearly half of them represent myxomas [1]. Most cardiac myxomas originate from interatrial septum near fossa ovalis and about 75% are in the left atrium [1]. The clinical features depend on tumor characteristics and vary widely based on obstructed ventricular filling, embolization and tumor cytokine production [1,5]. Diagnosis of this condition can be cumbersome because of its vague presentation. Once diagnosed, surgery provides cure and close follow-up is needed to prevent recurrence [1,5,6].

2. Case Presentation

A 61-year-old Caucasian female with a past medical history significant for hypertension, chronic pulmonary obstructive disease, diabetes mellitus, and major depressive disorder presented to her primary care physician with the chief complaint of cough and shortness of breath. For several months, her shortness of breath persisted, and she was treated on an outpatient basis with steroids and antibiotics without much relief. The patient also endorsed an unquantifiable amount of weight loss and night sweats during this period. Seven months following her initial presentation, an outpatient transthoracic echocardiogram (ECHO) was performed, revealing a 2.85 x 5.44 cm left atrial frond-like mass on the interatrial septum that prolapsed through the mitral valve. Right ventricular systolic pressure was markedly increased and severe pulmonary hypertension was noted. The findings prompted hospital admission and the patient was seen by the cardiothoracic surgery team for further management.

On hospital admission, she was hemodynamically stable. Physical examination including cardiopulmonary exam was unremarkable. Blood counts, electrolytes, liver function tests and coagulation studies were within normal limits. Electrocardiogram (EKG) and chest x-ray were also unremarkable. Coronary angiogram performed on an outpatient basis a year prior due to anginal symptoms and risk factors such as diabetes and hypertension revealed normal coronary arteries. Repeat ECHO confirmed the size of the left atrial mass protruding through the mitral valve (Figure 1, Figure 2, Figure 3). Cardiothoracic surgery recommended surgical resection of the myxoma via a median sternotomy. Patient underwent resection and tissue biopsy confirmed atrial myxoma. Her shortness of breath and cough resolved after surgery and she was discharged in stable condition two days post-op. No symptoms reported on one-month clinic follow up.

3. Discussion

Atrial myxomas commonly present after the third decade of life [1,2,3]. These tumors are endocardial in origin, and the cells giving rise to the tumor are considered to be multipotent mesenchymal cells that persist as embryonic residues during septation of the heart [1]. These tumors generally range from 1 to 15 cm in diameter; Most are 5 to 6 cm. The clinical features depend on their location, size, and mobility. Most patients present

with the classic triad of myxoma symptoms: embolism, intracardiac obstruction, and constitutional symptoms [1,3]. Patients classically present with left sided heart failure (such as dyspnea on exertion, orthopnea or pulmonary edema) secondary to decreased filling of left ventricle [2] and pose a diagnostic challenge for the clinicians due to the variability in their clinical presentations [4]. These tumors often produce interleukin-6 resulting in direct constitutional symptoms [1,5], clinically mimicking mitral valve disease, bacterial endocarditis or connective tissue disorders. For our patient, differentials included left atrial thrombus versus infective endocarditis versus atrial myxoma. Patient did not have fever or chills, elevated white count or risk factors for infective endocarditis. Blood cultures were negative. Transthoracic echocardiogram was performed which commented on the appearance of the mass which was most consistent with a myxoma. Had this been infective endocarditis, management would have been slightly different as we would have started intravenous antibiotics and possibly have consulted surgery for resection given the size of the vegetation. In 30 to 45% of patients, myxomas present with embolization of tumor fragments resulting in devastating cardiovascular consequences [2]. The cerebral arteries can be involved in almost 50% of the cases, and represent 0.5% of ischemic infarcts [2]. Because of significant morbidity associated with these tumors, prompt diagnosis is important because the outcome may be lethal if left untreated.

Atrial myxomas should be considered an important differential especially in younger patients who present with congestive heart failure, embolic phenomena or constitutional symptoms even in the absence of EKG or auscultatory findings. The most frequent EKG finding seen is left atrial hypertrophy [6] and auscultatory findings can be absent in up to 36% of these patients [5]. Echocardiography can provide detailed visualization of cardiac chambers and is useful in work up of cardiovascular symptoms. Transthoracic echocardiography is approximately 90% sensitive in detection of left atrial myxoma [2] and the sensitivity of transesophageal echocardiogram approaches almost 100% [5], making it a valuable tool for diagnosis. Computed tomography [1] and cardiac magnetic resonance imaging can also be helpful in delineating tumor characteristics and therapy [5]. A general approach to a mass noticed on echocardiography includes assessing the age of the patient, localizing the mass (atrium, valvular, ventricular), characteristics (appearance and mobility can help decide whether benign vs malignant), and taking into account the clinical context (fever and chills, history of intravenous drug use would lead to higher suspicion of infective endocarditis) [7].

Myxomas represent a curable form of disease if treated surgically [4]. Surgical resection usually results in complete resolution of symptoms and normalization of interleukin-6, erythrocyte sedimentation rate and C-reactive protein [5]. Long term prognosis is excellent [1,6]. Though recurrence of the myxoma did not occur in our case, it has been shown that recurrence is found in 2-5% of cases [6]. Therefore, the patient should be monitored in the future for either the return of previously experienced symptoms or new symptoms that could be manifesting as a result of tumor recurrence with serial annual echocardiograms depending on recurrence risk [5,6].

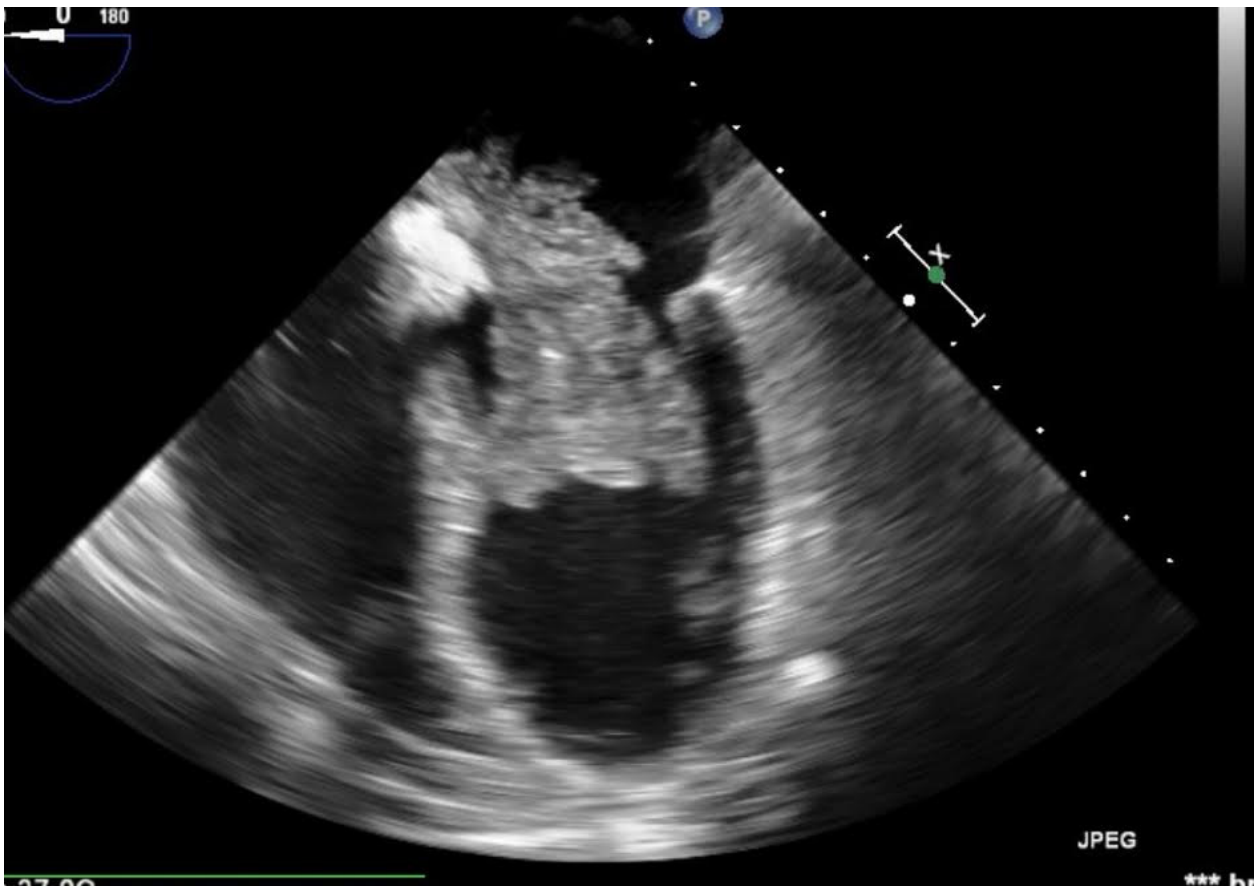

Figure 1. 2D Echocardiogram showing atrial myxoma prolapsing through the mitral valve

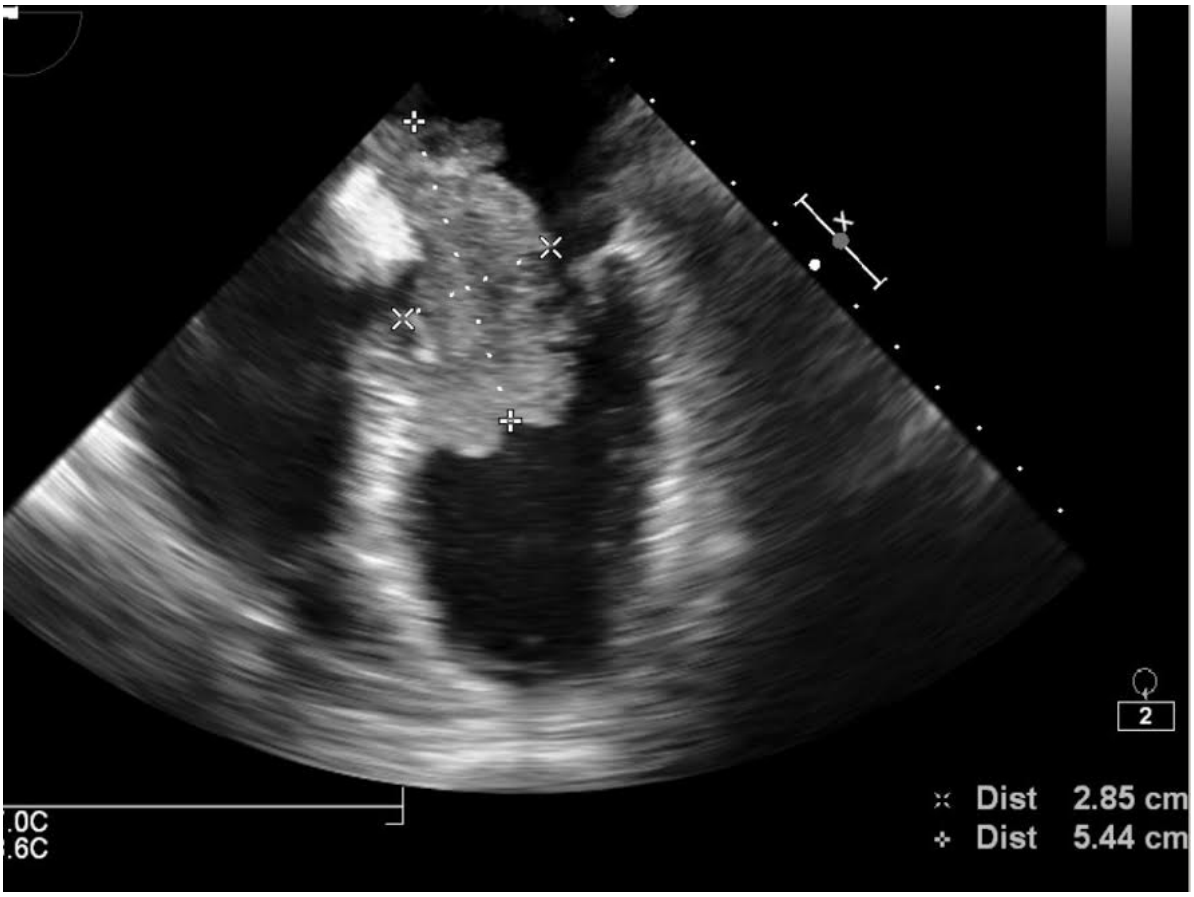

Figure 2. 2D Echocardiogram showing a 2.85 x 5.44 cm atrial myxoma attached to the interatrial septum and protruding through the mitral valve

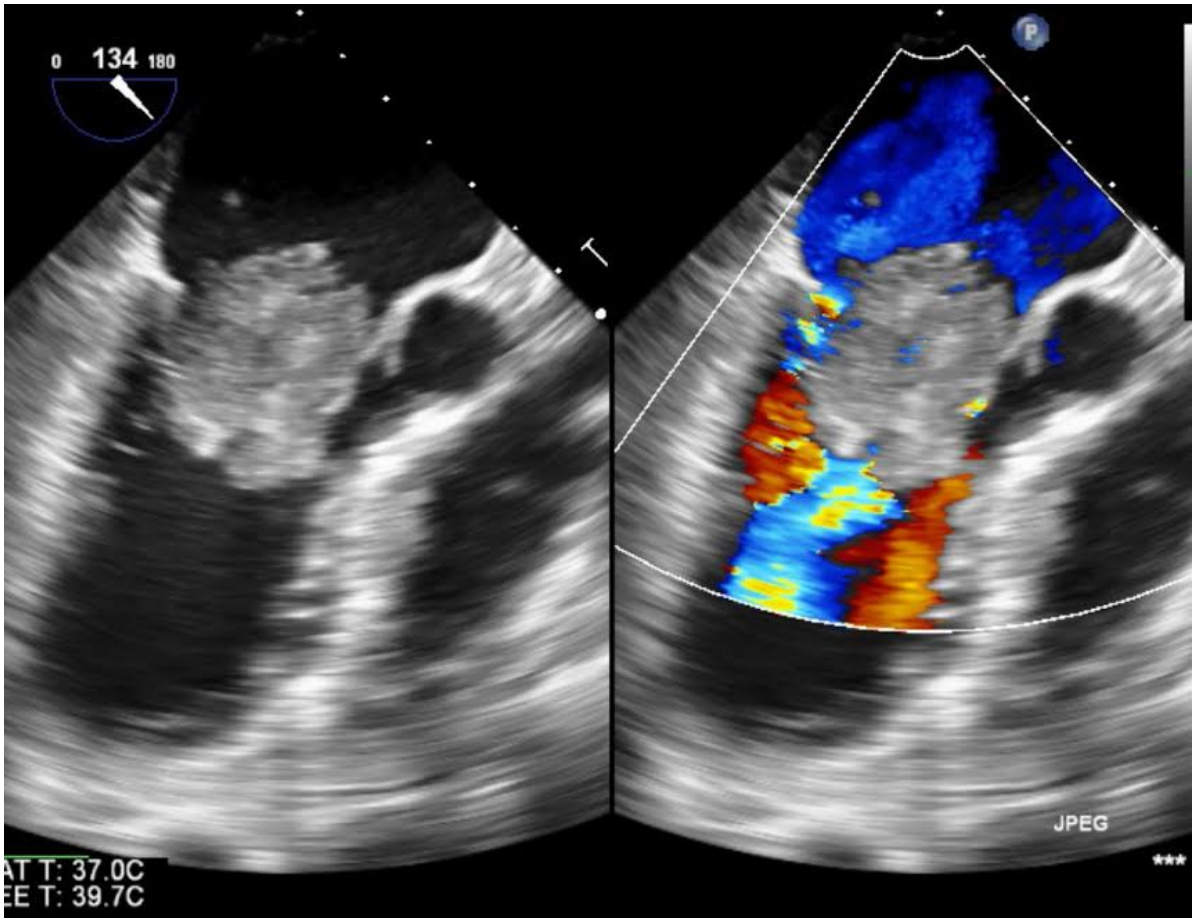

Figure 3. Color doppler image of a 2D Echocardiogram showing left atrial myxoma

4. Conclusion

This case illustrates the importance of including atrial myxomas on the differential for treatment resistant symptoms that are being attributed to cardiopulmonary disease, as atrial myxomas represent a completely curable cause of the associated symptoms.

Rarity of condition can lead to delay in diagnosis and distress for many patients. Physicians should have a high index of suspicion to diagnose this uncommon tumor earlier to avoid unnecessary testing and interventions.

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Declaration of Conflict of Interest

None.

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