CASE REPORTS

A rare association of left atrial myxoma with rheumatic mitral stenosis

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ABSTRACT

Primary cardiac tumors are very rare. Atrial myxomas are the most common primary tumors and are commonly located in the left atrium. Myxomas can cause mitral valve inflow obstruction and can present with clinical signs and symptoms of mitral stenosis and the possibility of myxoma should be considered in differential diagnosis of mitral stenosis. Left atrial myxoma is rarely associated with rheumatic mitral stenosis. Transesophageal echocardiography plays a major role in the diagnosis of this tumor. We report a case of an extremely rare association between left atrial myxoma and rheumatic mitral stenosis. Because of its location and mobility, a thrombus in the left atrium may have a similar appearance to left atrial myxoma. However, a careful inspection of features of the left atrial mass may allow a diagnosis of left atrial myxoma. In our case, the site near the atrial septum, heterogeneous echogenicity, and detection of a stalk connecting the myxoma to the atrial septum were inspected. Such findings essentially exclude thrombus.

Key Words: Rheumatic mitral stenosis, Atrial myxoma, Transthoracic echocardiography, Transesophageal echocardiography

1. INTRODUCTION

Atrial myxomas are the most common primary cardiac tumors and are located in the left atrium in 75%-80% of cases. They originate from multipotent mesenchymal cells of the subendocardial region. No etiological association appears to exist between Atrial myxoma and coexisting rheumatic mitral stenosis. Myxoma can manifest with symptoms due to obstruction of blood flow, constitutional symptoms or thromboembolic events. Due to these clinical symptoms, it can mimic endocarditis and occasionally mitral stenosis. Transthoracic echocardiography (TTE) has an excellent sensitivity of 95% in detecting myxomas, and the sensitivity increases to 100% when TTE is followed by transesophageal echocardiography (TEE). Diastolic prolapse of a left atrial mass between the mitral valve leaflets can be observed using TTE, which is a specific feature of left atrial myxoma (LAM). The attachment of the free mass to the interatrial septum with a stalk allows its prolapse between mitral valve leaflets and is a characteristic feature of LAM. TTE is inconclusive if prolapse of the mass cannot be demonstrated, the site of attachment cannot be seen, or there is high likelihood of a thrombus because of a coexisting mitral valve disease. TEE is superior to TTE in such situations. We report an extremely rare case of rheumatic mitral stenosis presenting with symptoms of shortness of breath and with a left atrial mass diagnosed as LAM through TTE and subsequently confirmed

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through pathological examination.

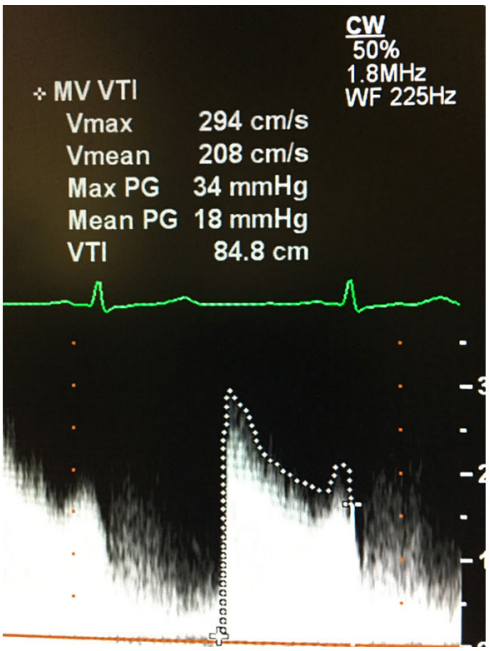

Figure 1. Continous wave doppler: showing diastolic mean transmitral pressure gradient suggestive of severe mitral stenosis

2. Case report

A 52-year-old Caucasian woman presented with worsening shortness of breath for 1-month in addition to intermittent low-grade fever. She had a history of paroxysmal nocturnal dyspnea, but she denied any syncopal episodes, exertional or rest angina, rash, arthralgia and palpitations. Her medical history was remarkable for mitral valve prolapse and rheumatic fever despite no major travel history outside of the United States. She denied any history of previous infective endocarditis. Family history was noncontributory. Physical examination was remarkable for a heart rate of 135/min, blood pressure of 100/70 mmHg, irregular rhythm, and a grade 1/6 holo systolic murmur and mid-diastolic murmur audible at the left sternal border. Electrocardiogram revealed atrial fibrillation with rapid ventricular response. Chest X-ray revealed airspace opacities with cephalization suggestive of pulmonary edema and congestive heart failure. Laboratory data revealed normal electrolytes, renal function, and thyroid hormone levels. TTE revealed normal left ventricular internal cavity size and an ejection fraction of 55%. Mitral valve assessment revealed thickening of the anterior mitral leaflet with mild calcification, and impaired mobility with hockey stick appearance suggestive of rheumatic mitral stenosis. Mean gradient across the mitral valve was 18.2 mmHg suggestive of severe mitral stenosis with a Wilkin score < 8 (see Figures 1, 2). Color Doppler imaging revealed moderate mitral valve regurgitation with central regurgitant jet (see Figure 3). The left atrium was severely enlarged, and the apical four-chamber view revealed a 3.36 mm × 2.14 mm irregular, hyperechoic, fixed mass, with smooth borders adherent to the atrial septum (see Figure 4). An echo also revealed a hockey stick appearance suggestive of mitral stenosis (see Figure 5). No obvious rheumatic involvement of other valves was noted.

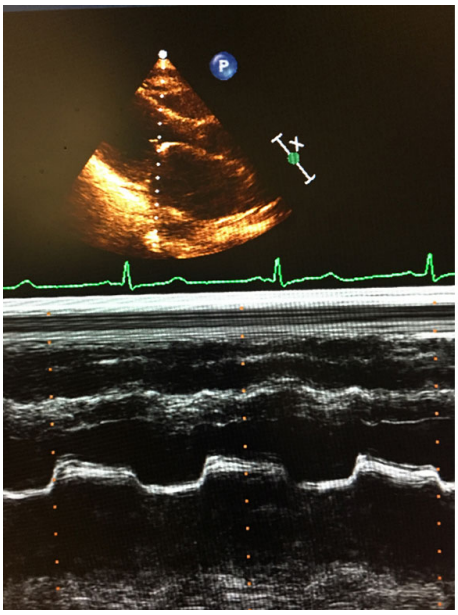

Figure 2. M-mode Echocardiography: Across the mitral valve showing thickening and severe mitral stenosis

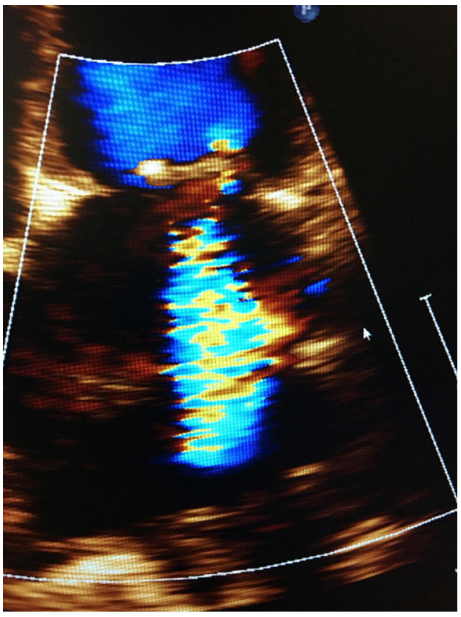

Figure 3. Color Doppler: Across mitral valve showing moderate mitral regurgitation

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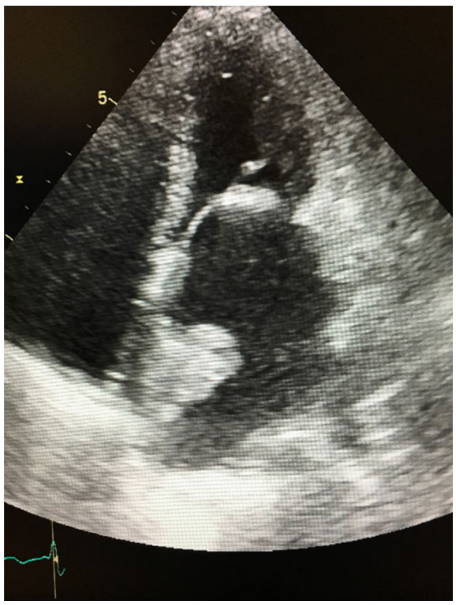

Figure 4. Apical four chamber view: Showing a solid, heterogeneous hyperechoic mass attached to atrial septum

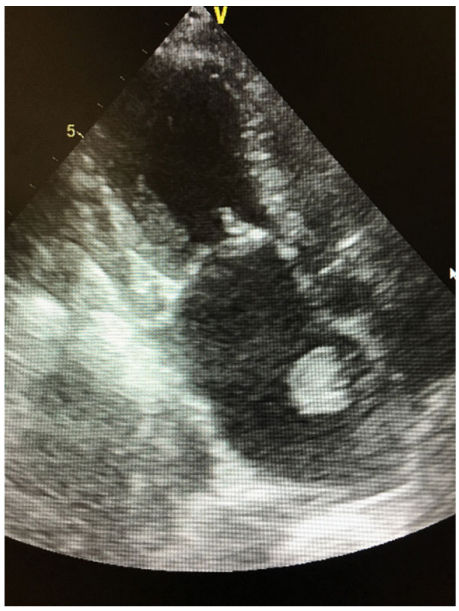

Figure 5. Apical two-chamber view: Showing a hockey stick appearance characteristic of rheumatic mitral stenosis

The initial management of atrial fibrillation included the initiation of therapeutic anticoagulation along with beta blocker and digoxin for rate control because of baseline low blood pressure. Subsequently, after a diagnosis of LAM with rheumatic mitral stenosis which is a very rare association, the patient was transferred to a tertiary care facility. Although the patient had a low Wilkin score, she was not a candidate for mitral annuloplasty because of the associated myxomatous tumor and moderate mitral regurgitation. She underwent mechanical mitral valve replacement, a MAZE procedure for atrial fibrillation and resection of the myxomatous tumor. A biopsy of the tumor later confirmed the myxoma (see Figure 6). The patient was discharged on Coumadin, beta blocker and she remained asymptomatic with an increase in functional capacity at the six month follow-up.

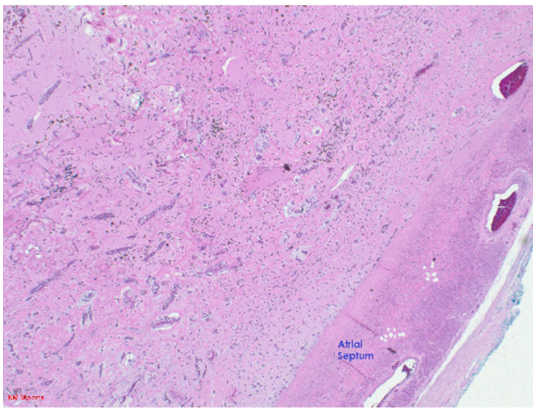

Figure 6. Light microscopy of tumor specimen showing Myxoma attached to Atrial Septum. (H&E, 10×)

3. Discussion and conclusions

Primary cardiac tumors are rare with an incidence ranging between 0.0017%-0.03% in the general population according to case series.[1] Metastatic tumors are 20-30 times more common than primary lesions. In all, 75% of the primary cardiac tumors are benign, and myxomas constitute 50% of these tumors. Sporadic cases of myxomas are more common in women than in men. Approximately 7% of cardiac myxomas are associated with Carney's complex, which is characterized by the association between cutaneous pigmentation, fibromyxoid tumors of the skin, myxomas of the heart, endocrine overactivity and autosomal dominant inheritance.[2] The association between myxoma and rheumatic mitral stenosis is exceedingly rare with very few cases reported in the literature when rheumatic fever was prevalent.[3–6] According to our review of relevant literature, no such cases were reported in in the last 10 years possibly because of a substantial decline in the incidence of rheumatic heart disease. Myxomas are most commonly located in the left atrium, originate from the fossa ovalis, and present with symptoms of thromboembolism[7] or mitral valve outflow obstruction.[8] Constitutional symptoms include fever, weight loss, arthralgias, anemia, elevated globulins and sedimentation rate which could mimic infective endocarditis, particularly with mitral stenosis. Our patient presented with symptoms of dyspnea, fever, and sweating but no weight loss or arthralgias. These symptoms are believed to manifest because of the tumor producing interleukin-6 and to improve after the tumor is removed.[9]

TTE has a high sensitivity in detecting the myxomas but may not identify the tumors smaller than 5 mm in diameter. Therefore, TEE is required to diagnose the smaller tumors.[10] Cardiac magnetic resonance imaging (MRI) can provide more information about tissue characteristics; cardiac and paracardiac morphologies which would aid in surgical treatment. Alternate imaging modality includes real time three-dimensional echocardiography (RT3DE) which allows accurate measurements of the entire volume of the mass in multiple planes and also can evaluate the real time effects of the mass on left ventricular inflow.[11] The treatment is surgical removal. Surgery involves removing the tumor and a wide resection of the pedicle to prevent recurrence. Resection of the tumor with its implantation base and adequate safety margins is necessary. Our patient required surgical removal of the tumor and mitral valve replacement. The tumor pathology showed myxoma cells which are stellate with eosinophilic cytoplasm and indistinct borders (see Figure 7). These cells had pale nuclei with open chromatin. Many areas of extravasated erythrocytes and thrombi were observed (see Figure 8). The biopsied leaflet revealed remarkable fibrosis with areas of nodular calcification. The recurrence rate and operative mortality rates are 1%-3% and 0%-3% respectively.[12] Tumor recurrence is more likely to occur in the first 10 postoperative years, especially in younger patients, patients with smaller tumor size, and those located in ventricle.[13] An important aspect of the surgical technique is to clamp the aorta before resection to avoid embolization of fragments because they are gelatinous and friable masses.

The uncommon association between LAM and mitral stenosis may increase the possibility of a myxoma being incorrectly interpreted as left atrial thrombus.[14] Because of its location and mobility, a left atrial thrombus may have a similar appearance to LAM. However, a careful inspection of the features of the left atrial mass may allow a diagnosis of LAM to be made. In our case, the site near the atrial septum, heterogeneous echogenicity, and detection of a stalk connecting it to atrial septum were inspected which allowed a diagnosis of LAM. Such findings essentially exclude thrombus. Although LAM is uncommon, the serious morbidity and mortality rates associated with LAM and the availability of effective surgical treatment make the need for correct diagnosis critical.

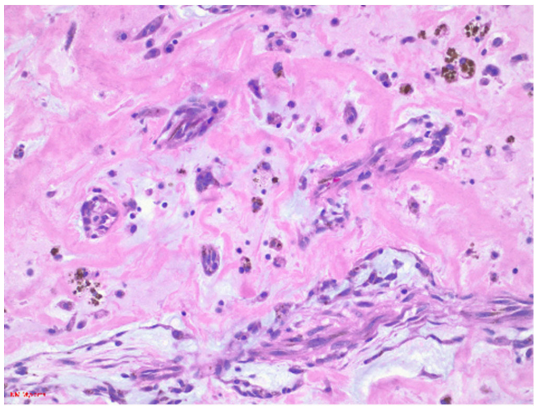

Figure 7. Light microscopy showing Myxoma cells with eosinophilic cytoplasm and indistinct borders. (H&E, 40×)

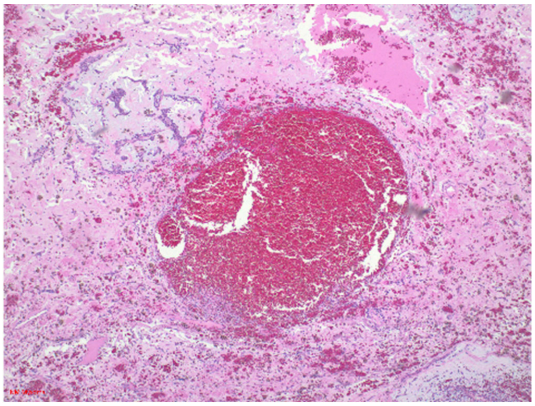

Figure 8. Light microscopy showing areas of scattered erythrocytes and thrombus. (H&E, 20×)

Informed Consent

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

Conflicts of Interest Disclosure

The authors have no conflicts of interest to declare.

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