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Case report

Cardiac mass causing pulmonary hypertension: Dilemma resolved with multimodality imaging

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ABSTRACT

Myxoma, the most common cardiac tumor is known to cause mitral valve obstruction and right heart failure with pulmonary hypertension. Patients with this condition often present with dyspnea, peripheral edema, and other signs of right heart failure. Pulmonary hypertension secondary to mitral valve obstruction presents unique difficulties in diagnosis as opposed other secondary causes such as thromboembolic pulmonary hypertension. Multimodality imaging with MRI, CT, and echocardiography can help resolve this dilemma. This case report serves to elucidate the important role of multimodality imaging in the management of patients with such a presentation. We also demonstrate in our case report how surgical removal can be curative by relieving mitral valve obstruction and thereby reversing pulmonary hypertension.

1. Case report

Patient is a 44 year old female with no significant past medical history who presented with dyspnea on exertion for the past 10 days along with bilateral pedal edema. Physical exam on presentation revealed normal vital signs and bilateral pedal edema; cardiac exam revealed an early diastolic murmur at the apex and a systolic murmur at the left sternal border. Lung exam revealed bibasilar rales.

An echocardiogram revealed the following: a cardiac mass with an echolucent center in the left atrium attached to the interatrial septum prolapsing through the mitral valve in diastole causing severe mitral stenosis; a dilated right ventricle with severely reduced systolic function; septal flattening with a D shaped septum suggestive of elevated right ventricular pressure; severe pulmonary hypertension (PA pressure 70–75 mmHg); and a small pericardial effusion (Figs. 1–4). Chest X-ray showed an enlarged cardiac silhouette and pleural effusion.

Based on this clinical presentation and echocardiographic findings, the possibility of pulmonary embolism could not be entirely excluded. To resolve this diagnostic dilemma, cardiac MRI was performed which showed a large irregularly contoured left atrial mass containing multiple ovoid components (largest measuring 4.4 cm × 3.2 cm). The mass was

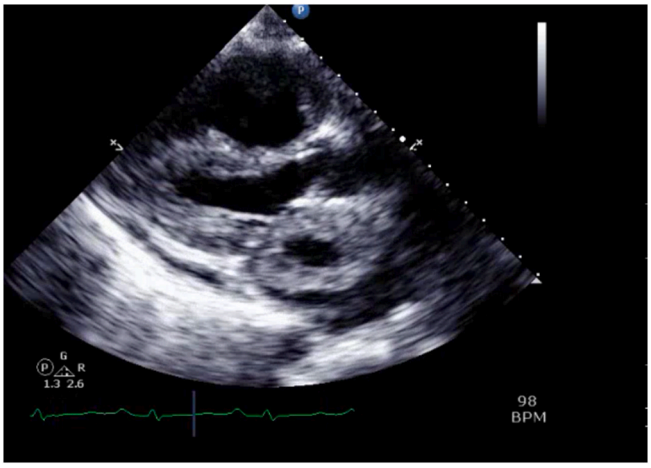

Fig. 1. Parasternal Long axis View showing myxoma, dilated RV, septal flattening.

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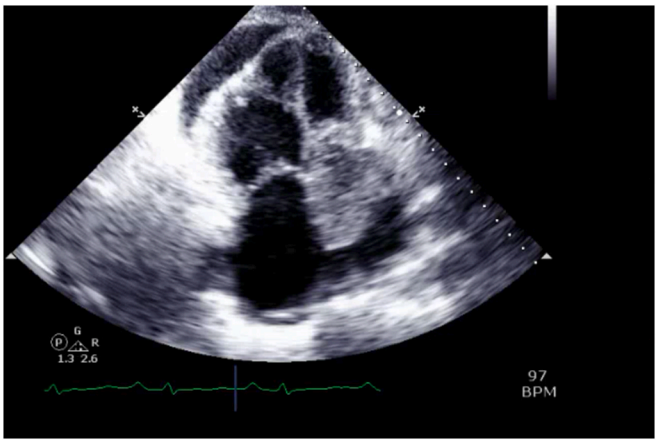

Fig. 2. Apical 4 chamber view showing myxoma causing mitral obstruction, dilated RV with severe dysfunction.

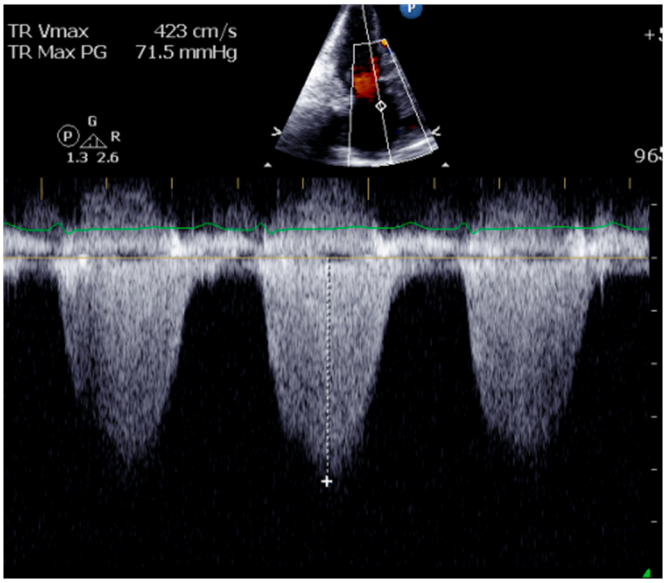

Fig. 3. Severe pulmonary hypertension instead of Severe TR.

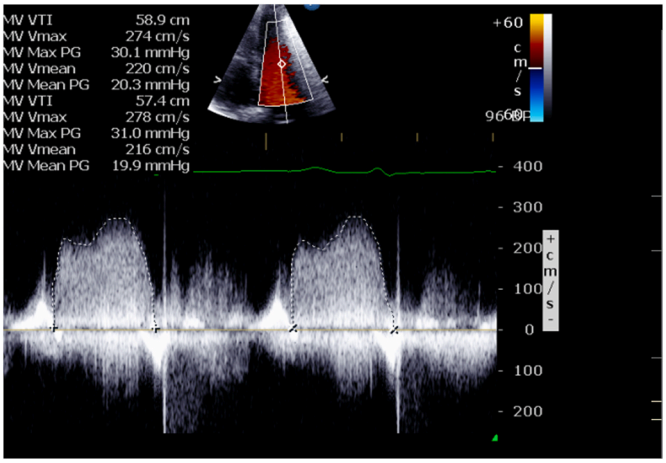

Fig. 4. Severe mitral stenosis instead of Severe pulmonary hypertension.

partially adherent to the inter-atrial septum and contained multiple highly mobile intracavitary components. The mass dynamically prolapsed through the mitral valve into the left ventricle resulting in obstruction of mitral valve flow. Post contrast tissue characterization demonstrated extensive lack of contrast uptake within the central aspect of the mass with slight peripheral enhancement. Morphology and tissue properties were suggestive of minimally vascular neoplasm and less likely thrombus.

Normal left ventricular size and systolic function (LVEF = 59%), moderate to severely dilated right ventricle with moderately reduced systolic function (RVEF = 32%), severely dilated right atrium, moderate pericardial effusion, and moderate right and small left pleural effusions were also noted.

(Figs. 5 and 6) Based on the above findings, the patient underwent surgical resection of the cardiac mass.

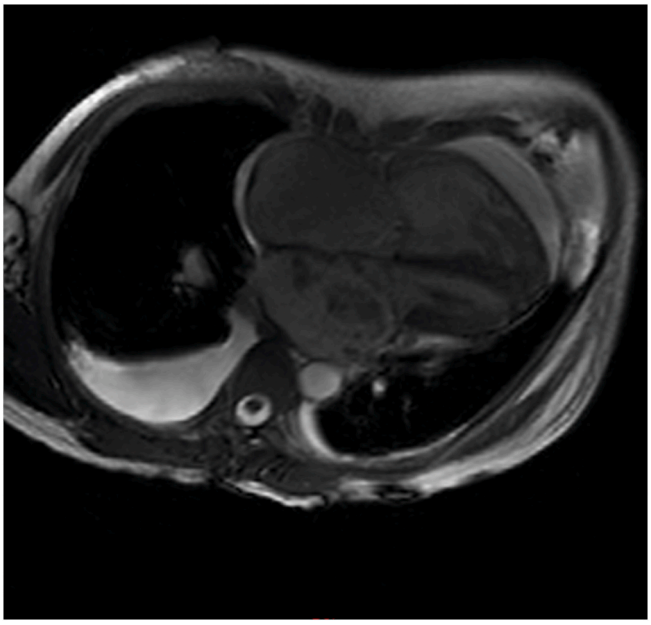

Fig. 5. 4 chamber view MRI.

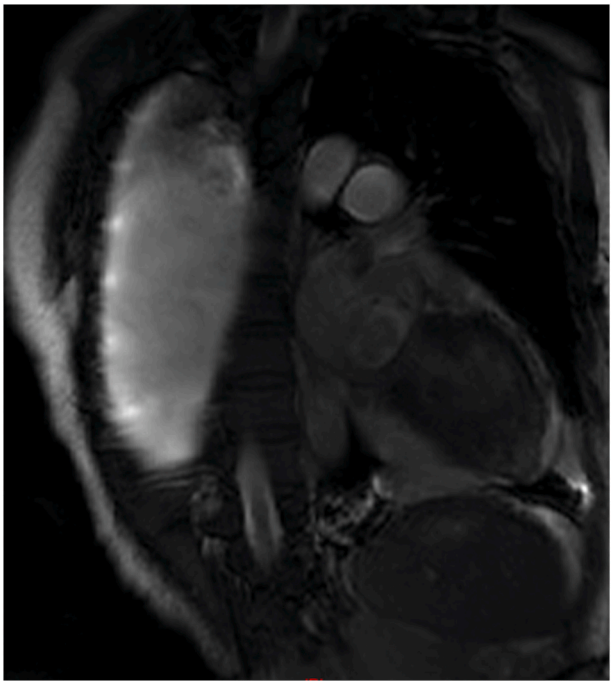

Fig. 6. 2 chamber view MRI.

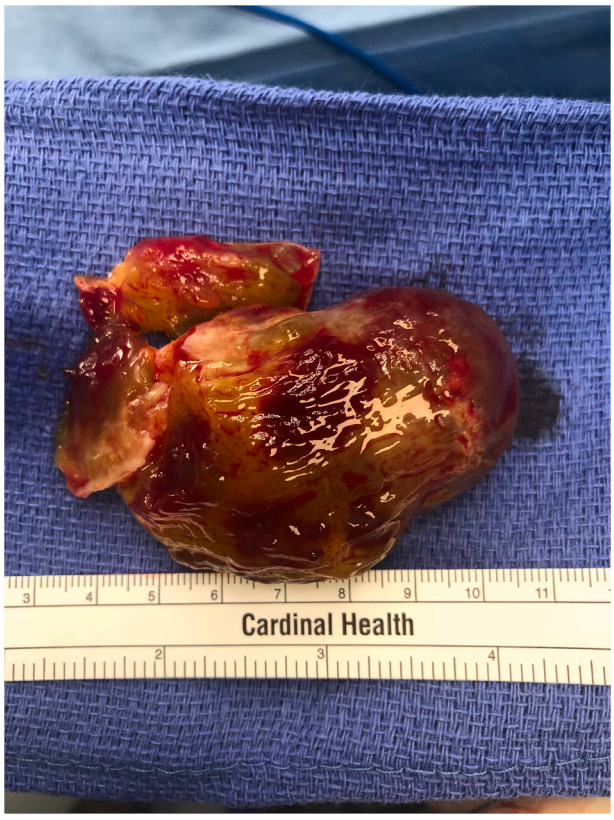

Fig. 7. Excised surgical specimen of the tumor.

(Fig. 7). Pathology confirmed a left atrial myxoma. She was extubated the following day after surgery. In a few days repeat echo showed improvement in RV size and function. Septum was midline without any significant flattening in systole or diastole.

Patient was discharged home in a stable and well-compensated state 5 days postoperatively.

2. Discussion

WHO Group 2 pulmonary hypertension (PH) includes PH due to left heart disease [1]. In this extensive group of left heart diseases, valvular heart disease such as mitral valve obstruction by a tumor such as a myxoma can cause pulmonary hypertension with right heart failure. As the valve narrows, the resting diastolic mitral valve gradient increases and subsequently left atrial pressure increases [2].

Pulmonary hypertension can develop as a result of many causes, including but not limited to retrograde flow from the left atria, interstitial edema, constriction of the pulmonary arterioles, or hyperplasia/hypertrophy of the pulmonary vasculature. Right ventricular dilation and tricuspid regurgitation can occur as a result of increased pulmonary arterial pressure which is evidenced by elevated JVP, third spacing of fluid, and hepatic vascular congestion.

Myxoma is the most common primary cardiac tumor, accounting for 30–35% of cases. They can manifest with obstructive, embolic, and constitutional symptoms [3]. When this patient initially presented with symptoms of right heart failure and an echocardiogram showing a giant tumor causing severe mitral stenosis with severe right ventricular dysfunction and severe pulmonary hypertension, the question always remained whether severe pulmonary hypertension was due to functional mitral stenosis or whether pulmonary thromboembolism was also playing a role. Other imaging modalities such as CT and cardiac MRI can be of immense help in arriving at a more definitive diagnosis. As is illustrated in our case report, MRI was better able to elucidate right heart failure due to obstructive mitral valve disease. Patient underwent successful surgical resection of the myxoma. Subsequently she had a decrease in her pulmonary artery pressure with improvement in RV systolic function.

Prior case reports have been published regarding the rare incidence of pulmonary hypertension caused by left atrial myxoma [4]. Although pulmonary hypertension secondary to mitral valve obstruction is not uncommon in left atrial myxoma, severe right ventricular systolic dysfunction and tricuspid regurgitation are less expected outcomes.

In a study performed at the All India Institute of Medical Sciences, a tertiary cardiac care center in Northern India, it was shown that out of 93,500 echocardiograms performed over a period of 11 years, only 70 cases of cardiac myxoma were found [5]. Only five of these 70 patients had moderate to severely elevated RV pressures with tricuspid regurgitation and only three of them had associated pulmonary hypertension as was seen in our patient. In a broader retrospective review of 112 patients with atrial myxoma over 40 years in a French Hospital symptoms of right heart failure such as abdominal bloating or peripheral edema were not reported and nor were pulmonary artery pressures as affected [6].

The purpose of this case report is to share with our physician colleagues the dilemma that we face when we see such rare cases and how multimodality imaging with echocardiogram, MRI, and CT can help us in arriving at the proper diagnosis.

Our case report also highlights the important role of cardiac surgery in reversing the course of severe pulmonary hypertension due to a cardiac mass.

Declaration of competing interest

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2021.101422.

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