Multimodality Imaging in the Evaluation and Management of a Right Atrial Mass

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INTRODUCTION

Primary cardiac tumors are extremely rare,[1,2] and diagnosis and management can often be complex and challenging. While both benign and malignant masses can have substantial hemodynamic and arrhythmic consequences, accurate definition of a cardiac tumor is crucial in the management and treatment planning, as well as a predictor of prognosis.[3]

CASE PRESENTATION

We describe a 64-year-old female patient following the incidental discovery of a right atrial (RA) mass on computed tomography (CT). The patient initially presented with shortness of breath along with a cough with sputum production for 1 week. Medical history was significant for chronic obstructive pulmonary disease, type 2 diabetes mellitus, hypertension, dyslipidemia, and obesity. Computed tomography pulmonary angiography showed multifocal airspace opacities, and the patient was diagnosed with multifocal pneumonia and treated with doxycycline and cefdinir. Computed tomography pulmonary angiography also revealed a 6.0 cm mass in the right atrium (RA), in addition to biatrial enlargement (Figure 1A, Video 1).

Transthoracic echocardiography (TTE) was performed the day after discharge at an outside institution to further assess the mass (Figure 2, Video 2). Left atrial and RA volumes were 36.0 mL/m^2 and 28.0 mL/m^2 (indexed), suggestive of mild enlargement. A large RA mass was observed, measuring 6.2 cm × 4.4 cm and prolapsing into the tricuspid valve causing flow obstruction, without tricuspid regurgitation or right ventricular (RV) outflow tract obstruction. Right atrial pressure was also noted to be high (estimated 15 mm Hg). Initial evaluation was suggestive of cardiac myxoma, and follow-up was scheduled for further investigations and planning for surgical intervention, after optimization of pulmonary function.

The patient underwent coronary angiography 2 months later as part of presurgical workup, on which the tumor was found to have a well-defined blood supply from the auricular branch from the circumflex artery penetrating the atrial septum and the tumor (Video 3).

Follow-up CT scan performed 3 months after the initial presentation for preoperative pulmonary evaluation showed resolution of previously seen bilateral patchy lung opacities and unchanged appearance of the 6 cm RA mass, with apparition of a new small pericardial effusion (Figure 1B, Video 4). It additionally showed an unchanged previously seen 1.6 cm adrenal mass and an indeterminate partially seen 2.5 cm left renal lesion.

In light of the presence of pericardial effusion and increased vascularity, cardiovascular magnetic resonance (CMR) with multiple

VIDEO HIGHLIGHTS

Video 1: Baseline CT angiography (soft tissue window, axial stack view) showing a large atrial mass.
Video 2: Two-dimensional TTE, RV-focused apical 4-chamber view, demonstrates a large, heterogenous RA mass protruding into the right ventricle during diastole.
Video 3: Follow-up CT scan (3 months later) showing that very little changed in the appearance of the 6.0 cm RA mass and interval new small pericardial effusion.
Video 4: Left coronary angiography demonstrates blood supply to the RA mass from a large branch that arises from the circumflex coronary artery.
Video 5: Cardiovascular magnetic resonance balanced steady-state free precession sequence axial view through the base of the heart demonstrates the large, heterogenous RA mass occupying the bulk of the dilated RA cavity.
Video 6: Cardiovascular magnetic resonance balanced steady-state free precession sequence oblique sagittal RA-RV view demonstrates the large, heterogenous RA mass occupying the bulk of the dilated RA cavity.
Video 7: Cardiovascular magnetic resonance T2-weighted sequence axial stack view demonstrates the large, bright RA mass with high signal intensity relative to the adjacent myocardium.
Video 8: First-pass perfusion imaging (axial view, full cardiac cycle).
Video 9: Two-dimensional transesophageal echocardiography, midesophageal long-axis view (0°) obtained preoperatively, demonstrates the large, heterogenous, partially mobile atrial myxoma that obstructs the RV inflow during diastole.

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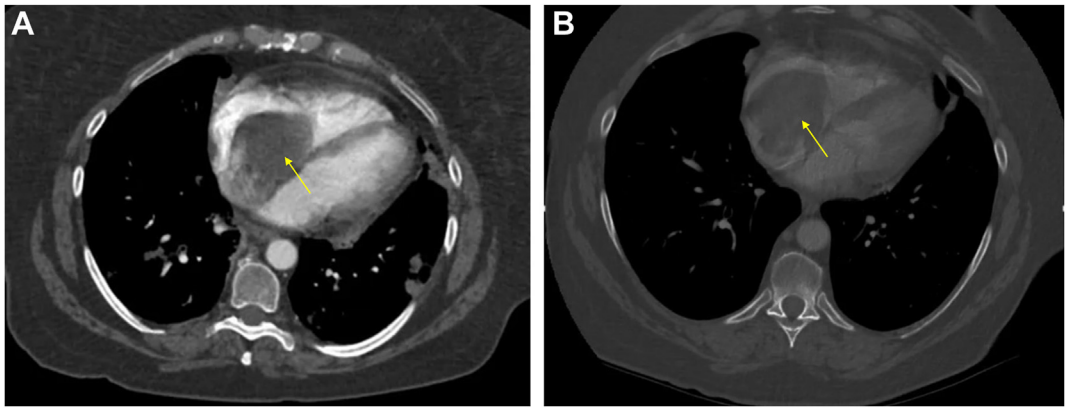

Figure 1 Chest CT (axial view, soft tissue window), at baseline **(A)** and 3 months later **(B)** showing very little change in the appearance of the 6.0 cm RA mass (*arrow*) and interval new small pericardial effusion.

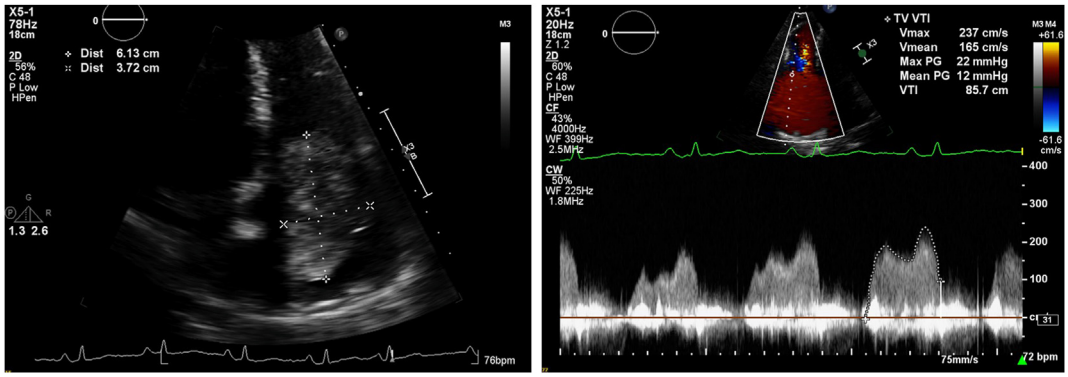

Figure 2 Two-dimensional TTE showing a 6.0 cm RA mass prolapsing into the right ventricle on apical 2-chamber view (*left*), as well as tricuspid flow obstruction with a mean gradient of 10 mm Hg on color-flow Doppler (*right*).

sequences was obtained shortly after at an outside institution. The latter showed a large heterogeneous mass measuring 4.7 × 5.0 cm occupying almost the complete RA and prolapsing through the tricuspid valve in diastole (Videos 5 and 6). The mass extended into the lower superior vena cava, without extension into the inferior vena cava, left atrium, or pulmonary veins (Figure 3). A stalk attached to the interatrial septum was also visualized (Figure 4). On T1-weighted images, the mass was isointense with focal areas of hyperintensity suggestive of hemorrhage or tissue breakdown (Figure 5), further visible as hypointensities on a T2 star map. It also appeared hyperintense on T2-weighted sequences, suggesting high water content (Figure 3, Video 7). First-pass perfusion images demonstrated patchy enhancement demonstrating the vascular nature of the tumor (Video 8). Additional delayed enhancement imaging showed florid heterogenous enhancement of the mass (Figure 6). Pericardial thickness was 3.0 mm, along with a small to medium sized circumferential pericardial effusion with echogenic material, without tamponade. Mild mediastinal lymphadenopathy was also seen. While most of the previous features were consistent with atrial myxoma,[4] the amount of gadolinium uptake and heterogeneous enhancement during first pass of gadolinium, as well as the presence of pericardial effusion, were considered slightly unusual for a myxoma, suggesting a possible malignant component.[3,5] Based on the outside report, the initial working diagnosis was angiosarcoma. Lymphoma, fibroma, hemangioma, or lipoma was deemed unlikely. However, repeat reading of the CMR at our institution and further discussion after combined assessment with other imaging findings suggested that the most likely diagnosis was a myxoma.

The patient underwent surgery for complete excision of the mass (Figure 7 and Video 9), 4 months after initial discovery (Figure 8), with subsequent tricuspid valvuloplasty for severe tricuspid regurgitation due to annular dilation. The mass was well circumscribed, measuring 6.4 × 5.4 × 4.6 cm with a sessile base of transected myocardium and endocardium measuring 4.0 × 1.5 cm. Serial sectioning revealed yellow and gelatinous cut surfaces with areas of consolidated hemorrhage measuring up to 3 cm, which complemented CMR findings of hemorrhage and heterogenous uptake.

Microscopic examination showed a heavily myxoid stroma populating the abundant canaliculi lined by lepidic cells, as well as rich neovascularization of areas of interstitial hemorrhage with hemosiderin-laden macrophages. The tumor had a sessile base with sheets of lymphocytes at the interface between the tumor and endocardium and mild hypertrophy of cardiac myocytes underneath the endocardium. Movat stain showed alternance of fibrous tissue and myxomatous tissue forming the stroma throughout the tumor. There was no evidence of Gandy-Gamna bodies or glandular differentiation.

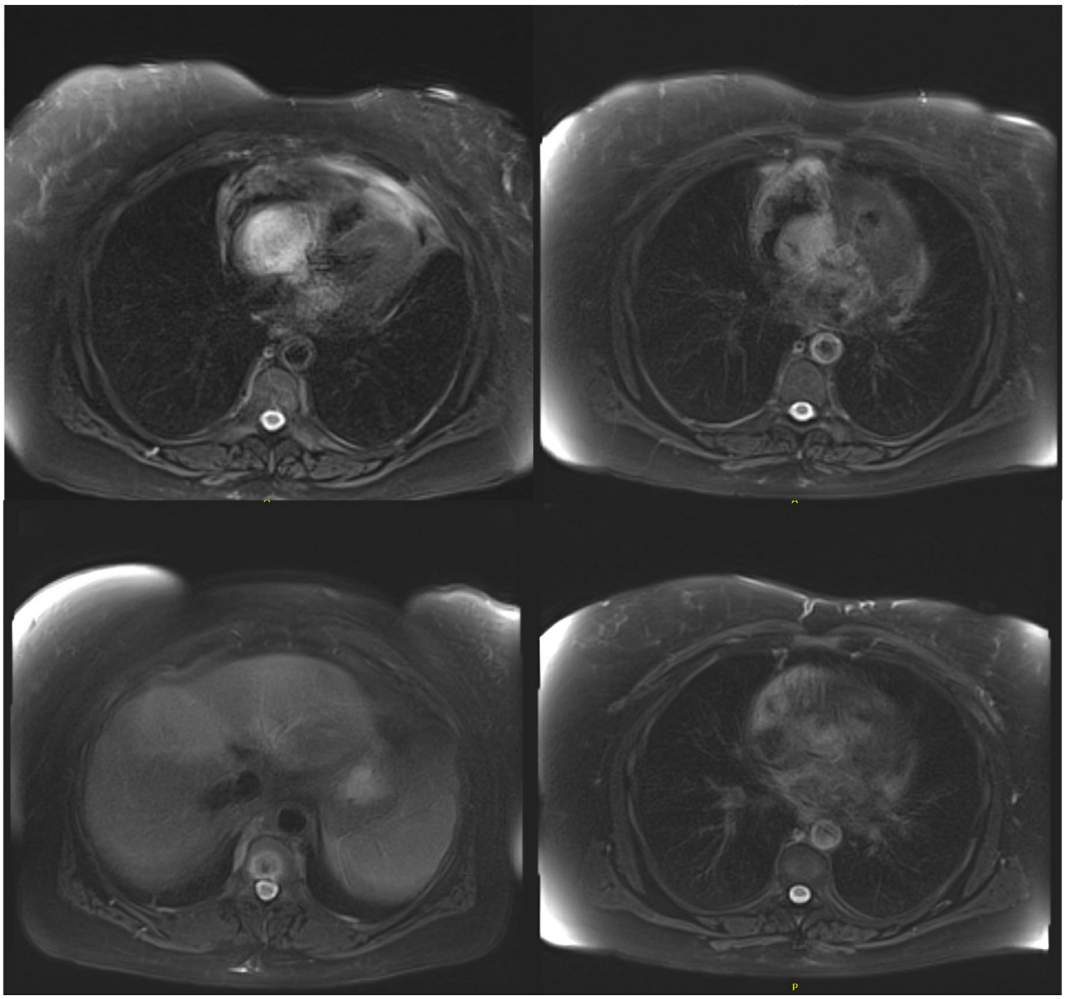

Figure 3 Cardiovascular magnetic resonance, three-plane T2-weighted images. The tumor is bright, suggesting high water content, and demonstrates some extension into the lower superior vena cava, without extension into the inferior vena cava, left atrium, or pulmonary vein. *3C*, Three-chamber; *4C*, four-chamber; *SA*, sagittal.

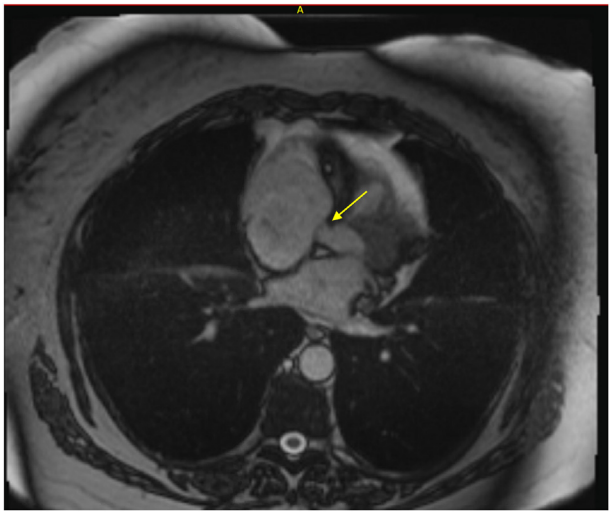

Figure 4 Cardiovascular magnetic resonance, true fast imaging with balanced steady-state free precession sequence (axial view). A stalk is visualized attached to the interatrial septum (*arrow*).

All previous findings were compatible with the diagnosis of cardiac myxoma.

The patient experienced no major complications and was discharged a week after surgery. Follow-up echocardiogram at 2 weeks showed a left ventricular ejection fraction of 70%, mild RV dysfunction (RV systolic tissue Doppler velocity 7.0 cm/sec, tricuspid annular displacement 1.7 cm), which resolved over the following months, and mild tricuspid regurgitation.

The patient was readmitted 1 week later for acute bilateral pulmonary embolism and treated with anticoagulation, with subsequent resolution of their condition. No tumor recurrence or cardiac complications occurred after 2 years. The renal mass was resected 2.5 years later and was found to be a grade 2 clear-cell renal carcinoma without invasion or metastasis.

DISCUSSION

This case highlights the importance of careful multimodality imaging in the diagnosis and treatment of cardiac tumors. Transthoracic echocardiography is often the initial approach, allowing for an initial look at the mass, and in particular its repercussions on hemodynamics.[3]

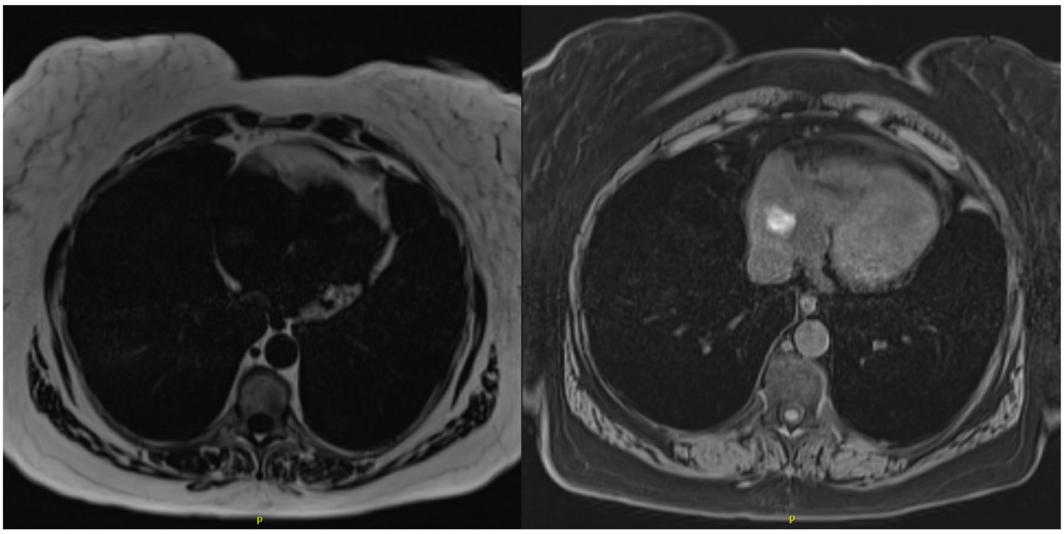

Figure 5 Cardiovascular magnetic resonance, T1-weighted sequences (axial view) showing the mass as isointense with extensive areas of hemorrhage/breakdown within the mass, accounting for focal areas of hyperintensity.

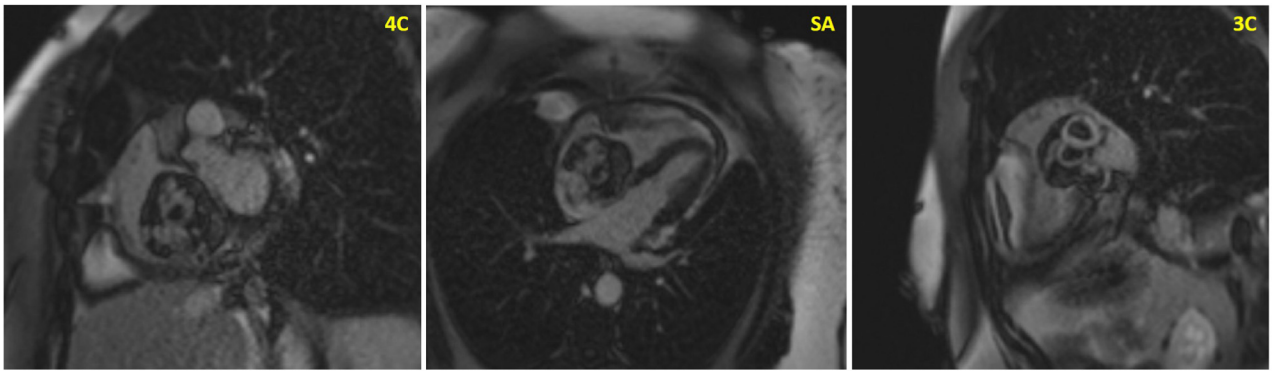

Figure 6 Three-plane CMR images showing heterogeneous late gadolinium enhancement on CMR.

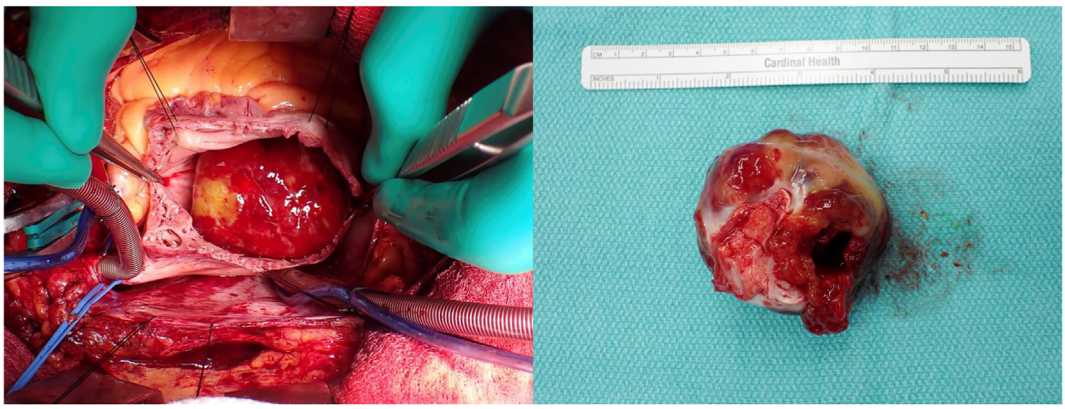

Figure 7 Intraoperative and postoperative images showing complete resection of the mass.

Advanced echocardiographic studies such as transesophageal echocardiography and ultrasound-enhancing agents may provide additional information. Typically, in high suspicion of cardiac myxoma without features suggestive of malignant process, surgical removal is the appropriate next step, without the need for additional evaluation. However, the presence of alarming features, including pericardial effusion and rich neovascularization, as in this case, usually prompts further investigations.[3,6] Several combinations of imaging techniques to assess cardiac neoplasms have been reported, including advanced echocardiographic assessment, CT, and coronary angiography.[7,8]

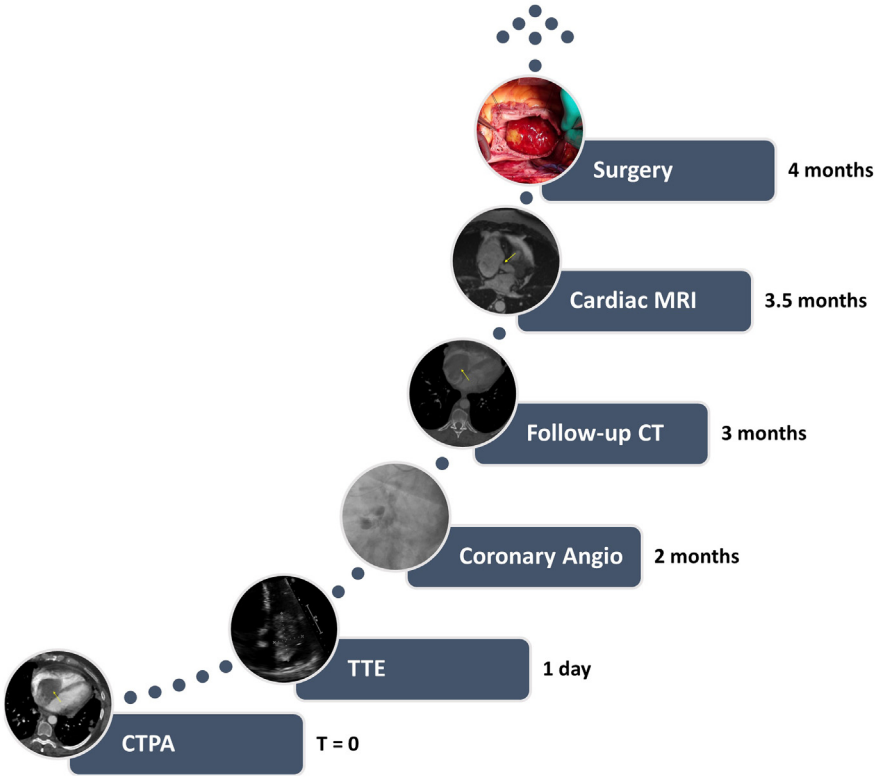

Figure 8 Clinical timeline from discovery of the mass to surgical resection. *CTPA*, Computed tomography pulmonary angiography; *MRI*, magnetic resonance imaging.

The use of coronary angiography remains under debate but has been used for preoperative assessment of tumor blood supply, which can affect surgical management.[9] Cardiovascular magnetic resonance is a robust cardiac imaging tool when assessing suspected tumors,[10] as it offers complete three-dimensional evaluation of the mass, allowing for more accurate tissue discrimination and morphological and anatomical characterization.[11] This helps identify the nature of the mass and differentiate it from thrombi, in addition to its role in surgical planning.[12] Computed tomography scan may be used when CMR is not available; in this case, it was performed due to concomitant pulmonary disease.

Atrial myxomas are the most frequent primary cardiac tumors, with only 15% to 20% arising in the RA.[6] Complete surgical resection allows for definitive diagnosis as well as prevention of embolic and hemodynamic complications and confers the best long-term outcome. In this case, the main differential diagnosis was angiosarcoma. These tumors are the most frequent malignant cardiac neoplasms in adults (about two-thirds) and most often occur in the RA. They are associated with worse prognosis and require more aggressive management, including radiation therapy and chemotherapy, although the effectiveness of these treatments remains controversial.[13,14] In contrast to myxomas, angiosarcomas rarely have interatrial septum origin and tend to be adherent to the wall and invade the pericardium and the atrioventricular groove, which was not the case here. Some features of the mass on CMR, such as the amount of gadolinium uptake, enhancement during the first pass of gadolinium, and presence of pericardial effusion, raised suspicion for malignancy. However, myxomas are known to have highly variable imaging characteristics on CMR, including heterogeneous composition due to areas of hemorrhage, owing to varied pathologic makeup.[11] The pericardial effusion was believed to be secondary to elevated RA pressure, as shown initially on echocardiography.[15]

CONCLUSION

Multimodality imaging offers incremental value in the evaluation of cardiac masses and is often encouraged for more accurate guidance in management and treatment planning. Nonetheless, this case suggests that one should keep in mind the variability of imaging findings and careful selection of additional imaging techniques in regard to different tumor types. Surgery followed by pathology remains the gold standard for definitive diagnosis and treatment.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.09.010.

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