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Right ventricle myxoma with massive pericardial effusion: a case report



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ABSTRACT

Introduction: Right ventricular myxomas (RVM) are exceedingly rare primary cardiac tumors, comprising less than 5% of all cardiac myxomas. The challenge of establishing diagnosis and proper management might improve survival in such patients.

Case illustration: A 53-year-old female presented with a primary complaint of shortness of breath. Further work-up diagnostics were done. Echocardiography findings suggestive of large pericardial effusion and echogenic material at the apical right ventricle (RV) with size 4.8x2 cm with normal left and right heart contraction. At first the echogenic material was suspected as RV aneurism with impending rupture. Due to high-risk profile patients with cardiac tamponade, open heart surgery was done and revealed solid tumor located at the apical RV. After that patient underwent a biopsy and excision of the tumor with ventriculectomy and reconstruction. A pathological exam of tumor reveals a spindle cell tumor with giant cell rich feature and finally decided as cardiac myxoma.

Conclusion: Right ventricular myxomas are exceptionally rare cardiac tumors that pose significant diagnostic challenges due to their atypical location and potential to mimic other life-threatening conditions, such as ventricular aneurysms.

Keywords: myxoma, right ventricle, excision.

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INTRODUCTION

Right ventricular myxomas (RVM) are exceedingly rare primary cardiac tumors, comprising less than 5% of all cardiac myxomas. These benign neoplasms, while most commonly originating in the left atrium, can occasionally present in the right ventricle, posing significant diagnostic challenges due to their atypical location and presentation. Clinical manifestations of myxomas are highly variable, ranging from asymptomatic findings to life-threatening conditions such as obstructive cardiac failure, embolic phenomena, or cardiac tamponade.¹

This case report discusses a rare presentation of RVM, initially misdiagnosed as an impending rupture of a right ventricular apical aneurysm, complicated by massive pericardial effusion. The patient presented with signs of pericardial tamponade, necessitating urgent intervention. Preoperative imaging suggested a diagnosis of an RV apical aneurysm with impending rupture, highlighting the potential for misdiagnosis

in the context of overlapping clinical and imaging findings.

Intraoperatively, a well-defined mass consistent with myxoma was discovered at the RV apex, contradicting the preoperative diagnosis. Histopathological examination confirmed the diagnosis of myxoma, emphasizing the critical importance of surgical exploration in cases with ambiguous diagnostic imaging.¹

Supporting evidence from existing literature highlights that cardiac myxomas, although benign, can have devastating clinical consequences if left untreated. Tumors in atypical locations, such as the right ventricle, are particularly prone to misdiagnosis due to their rarity and non-specific clinical features. For example, large myxomas may cause pericardial effusion, mechanical obstruction, or embolization, all of which can present similarly to other cardiac conditions, such as aneurysms or thrombi.^{2,3}

This report aims to contribute to the understanding of RVM by detailing its unique clinical course, diagnostic challenges, and intraoperative findings.

The case underscores the necessity of a multidisciplinary approach involving cardiologists, cardiothoracic surgeons, radiologists, and pathologists to achieve accurate diagnosis and optimal management. Furthermore, it highlights the role of intraoperative decision-making and histopathological evaluation in resolving diagnostic.⁴

CASE PRESENTATION

A 53-year-old female presented with a primary complaint of shortness of breath. Her symptoms had been persistent for a week prior to hospital admission, exacerbated by physical activity and alleviated by rest. The dyspnea was occasionally accompanied by chest pain and palpitations, alongside nausea and vomiting. Additionally, she reported a productive cough with blood-streaked sputum but had no prior history of tuberculosis or pulmonary evaluations.

Her past medical history was significant for kidney stone surgery in 2020 and fibroadenoma excision in 2007,

with no history of hypertension, diabetes mellitus, smoking, or other major systemic illnesses.

The patient was referred from a regional hospital with a provisional diagnosis of an impending rupture of an apical right ventricular (RV) aneurysm complicated by massive pericardial effusion. Imaging and laboratory findings supported this assessment. A chest X-ray demonstrated cardiomegaly with a cardio-thoracic ratio of 69%, prominent pulmonary segments, and signs of vascular congestion and infiltrates (Figure 1). Echocardiography revealed a massive pericardial effusion with a suspected tumor or thrombus measuring 4.8 x 2 cm located at the apical RV. The RV exhibited good systolic function with a tricuspid annular plane systolic excursion (TAPSE) of 20 mm. The left ventricular (LV) ejection fraction (EF) was preserved at 56%, with global normal kinetic function and no valvular abnormalities apart from mild tricuspid regurgitation (Figure 2).

The hemodynamic assessment showed a stable sinus rhythm on electrocardiogram, but the patient had hypotension with a non-invasive blood pressure of 87/60 mmHg and a heart rate of 113 bpm. Respiratory parameters were within normal limits on supplemental nasal oxygen at 3 liters per minute. The patient demonstrated no signs of pulmonary hypertension.

The laboratory results showed moderate anemia, with hemoglobin levels suggesting severe depletion. Inflammatory markers were significantly elevated, with a procalcitonin level of 1.46 ng/mL and a C-reactive protein (CRP) of 200 mg/L, indicating a possible underlying infection or inflammatory process. Renal function tests revealed acute kidney injury (AKI) with a blood urea nitrogen level of 57 mg/dL and creatinine of 2.33 mg/dL. Other notable laboratory findings included hypoalbuminemia (2.8 g/dL) and mildly elevated uric acid levels (8.3 mg/dL). Coagulation profiles were within normal ranges, with a prothrombin time of 11.7 seconds and an international normalized ratio (INR) of 1.09.

These findings were consistent with the diagnosis of massive pericardial effusion and the potential for mechanical complications, including tamponade.



Figure 1. CXR: Cardiomegaly (Cor to thorax ratio 69%), pulmonary congestion, and infiltrates.

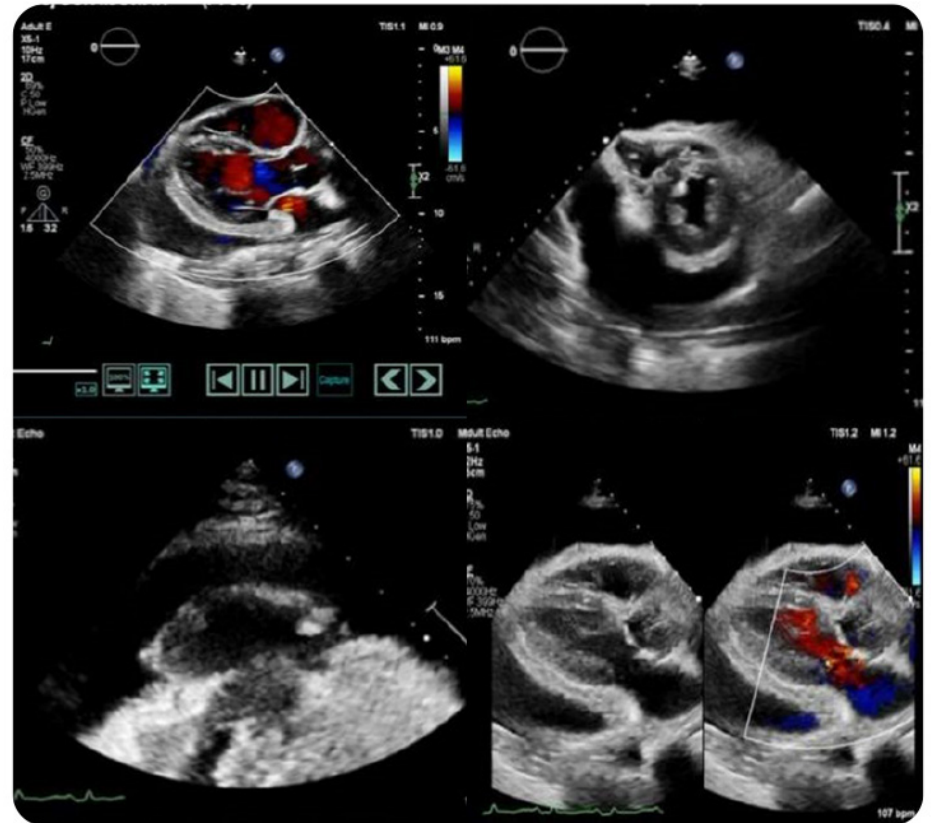


Figure 2. Echocardiography: Large pericardial effusion, 4.8 x 2 cm RV mass, preserved LV function (EF 56%), mild TR.

The patient's risk factors included dyslipidemia but no family history of cardiovascular diseases. The clinical team deemed her case high-risk and initiated preparations for urgent surgical repair of the RV apical aneurysm. Preoperative preparation included blood transfusion protocols, with the arrangement of 1000 mL of packed red blood cells, fresh frozen plasma, and platelet concentrates. Other preoperative interventions involved fasting, site preparation, and administration of prophylactic antibiotics and medications to optimize surgical outcomes.

The multidisciplinary team, comprising cardiothoracic surgeons, anesthesiologists, and cardiologists, planned an operative strategy to address the aneurysm and associated complications. The goals included preventing rupture, relieving the pericardial effusion, and evaluating the suspected tumor or thrombus.

During surgery, the patient underwent a biopsy and excision of a tumor located at the right ventricular (RV) apex, followed by ventriculectomy and reconstruction of the RV-free wall using Teflon material. Intraoperative findings revealed a firm, rough-surfaced tumor measuring 5 × 4 × 4 cm with well-defined borders, extending through the epicardium to a transmural location. The subsequent pathological examination of the mass revealed a spindle cell tumor with giant cell-rich features, which appears to originate from bone or soft tissue. Given the histological characteristics, the lesion is most likely a myxoma. A massive pericardial effusion of 450 mL serosanguinous fluid was also identified. Cardiopulmonary bypass (CPB) was employed for 42 minutes, with an aortic cross-clamp time of 25 minutes. Postoperatively, the patient was transferred to the ICU with stable hemodynamics (blood pressure: 86/50 mmHg, heart rate: 122 bpm, central venous pressure: 7 mmHg) and supported by vasopressors, including adrenaline and vasopressin. A brief episode of supraventricular tachycardia (SVT) occurred during bleeding control, which was resolved using synchronized cardioversion at 150 J. These measures ensured the patient's stability following the procedure.

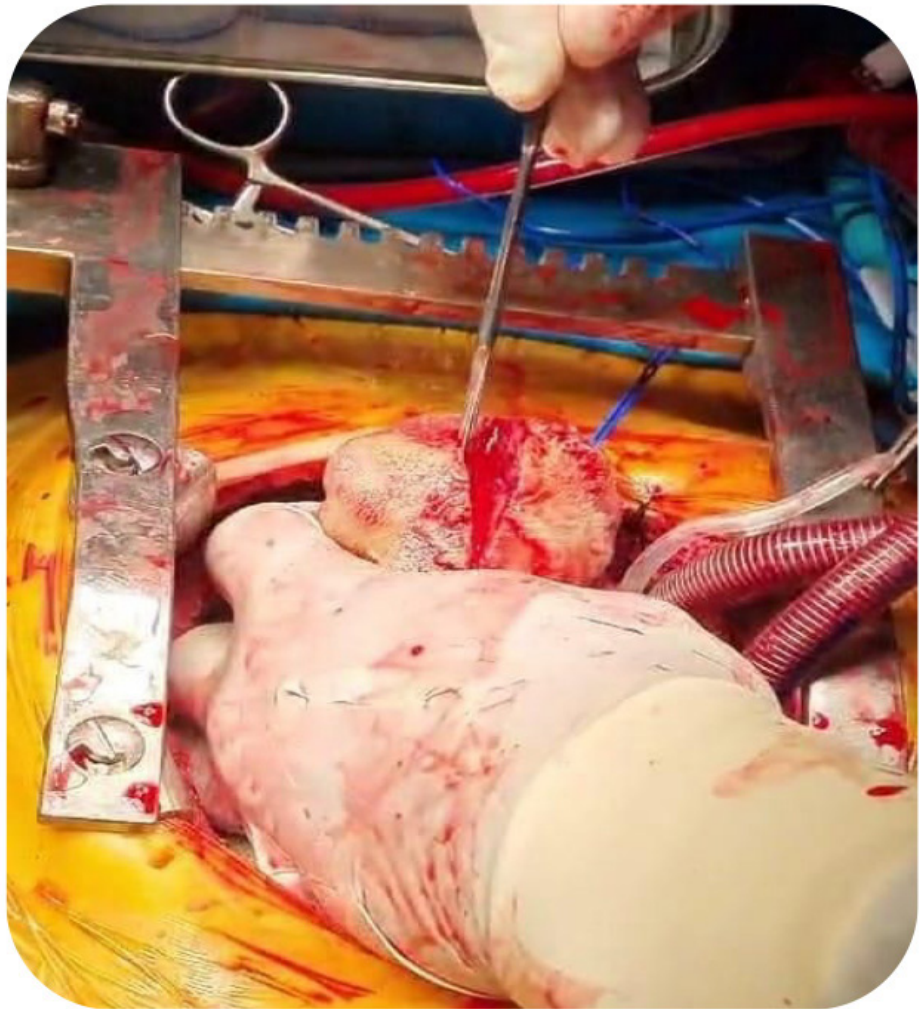


Figure 3. Intraoperative findings: myxoma

DISCUSSION

Right ventricular myxomas are exceedingly rare primary cardiac tumors, comprising less than 5% of all cardiac myxomas. These benign neoplasms, while most commonly originating in the left atrium, can occasionally present in the right ventricle, posing significant diagnostic challenges due to their atypical location and presentation. Clinical manifestations are highly variable, ranging from asymptomatic findings to life-threatening conditions such as obstructive cardiac failure, embolic phenomena, or cardiac.

The pathophysiology of RVM is complex and multifaceted, with symptoms primarily driven by tumor size, mobility, and location. Myxomas in the right ventricle can cause mechanical obstruction, leading to right-sided heart

failure, syncope, or severe dyspnea. Additionally, their potential for embolization and compression of adjacent structures further complicates the clinical presentation. Massive pericardial effusion, as observed in some cases, is thought to arise from tumor-induced irritation or venous obstruction, resulting in fluid accumulation in the pericardial space. This can obscure the underlying pathology and mimic other cardiac conditions such as aneurysms or thrombi.²

The diagnostic evaluation of cardiac myxomas relies heavily on imaging modalities. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are typically the first-line investigations, providing detailed visualization of intracardiac masses. Additional imaging studies, including cardiac computed

tomography (CT) and magnetic resonance imaging (MRI), can aid in differentiating myxomas from other cardiac pathologies and delineating their anatomical relationships. However, in cases where the tumor location or morphology is atypical, imaging findings may be misleading, as exemplified by masses mimicking ventricular aneurysms.³

The initial diagnosis of an impending rupture of a right ventricular apical aneurysm with massive pericardial effusion and suspected intracardiac tumor was based on clinical and imaging findings. Preoperative transthoracic echocardiography and subsequent imaging studies revealed a large pericardial effusion with signs suggestive of cardiac tamponade. Additionally, echocardiographic findings demonstrated a mass-like lesion at the apex of the right ventricle, which was interpreted as an aneurysmal dilatation with a high risk of rupture. These findings necessitated urgent surgical intervention to prevent catastrophic outcomes.

Intraoperatively, however, the diagnosis shifted, instead of an aneurysmal rupture, a well-encapsulated mass consistent with a tumor was discovered at the right ventricular apex. The tumor exhibited characteristics typical of myxomas, including a gelatinous texture and irregular surface, firmly attached to the endocardium. Histopathological examination postoperatively confirmed the tumor as a myxoma. This intraoperative discovery not only clarified the nature of the intracardiac pathology but also altered the course of management from aneurysmal repair to complete tumor.¹

The operative findings were critical in redirecting the management strategy from aneurysmal repair to tumor excision, confirming the diagnosis of a right ventricular apical myxoma. The surgical intervention involved careful excision of the mass to minimize damage to the surrounding myocardial tissue and avoid embolization. The removal was performed via a transventricular approach, ensuring complete resection of the tumor along with its attachment point. Intraoperative assessment revealed no evidence of rupture or other structural abnormalities associated with the initial suspicion of

aneurysm, further emphasizing the role of direct visualization in resolving diagnostic uncertainties.

The diagnostic shift from an aneurysm to a tumor underscores the limitations of imaging modalities in accurately characterizing cardiac masses, particularly in atypical locations. While imaging techniques like echocardiography, cardiac CT, and MRI provide critical information about size, location, and morphology, distinguishing between structural anomalies such as aneurysms and tumors may remain challenging in some instances. This diagnostic complexity highlights the indispensable role of surgical exploration and histopathological confirmation in refining the diagnosis and guiding definitive treatment.³

The initial misdiagnosis of aneurysmal rupture could be attributed to the overlapping features of aneurysms and tumors on imaging. Both conditions can present with localized dilatations or masses, making differentiation particularly difficult in emergent scenarios. Moreover, the presence of massive pericardial effusion likely contributed to the diagnostic uncertainty by obscuring clear visualization of the underlying.^{2,4}

Histopathological analysis remains the definitive diagnostic tool for myxomas. These tumors are histologically characterized by stellate or spindle-shaped cells embedded in a myxoid stroma, often accompanied by areas of hemorrhage or calcification. Immunohistochemical staining may further aid in differentiating myxomas from other cardiac masses or thrombi. Despite their benign nature, myxomas can cause significant morbidity if left untreated, emphasizing the importance of prompt surgical intervention.^{1,4}

Surgical resection is the treatment of choice for cardiac myxomas and is typically curative. The surgical approach depends on the tumor's size, location, and attachment site, with right ventricular myxomas often requiring a transventricular or transatrial approach. Complete excision with adequate margins is essential to minimize the risk of recurrence. Intraoperative findings, such as tumor texture, mobility, and attachment, play a critical role in guiding surgical management. Long-term outcomes are generally favorable, although

regular follow-up is recommended to monitor for recurrence, particularly in familial or syndromic cases such as Carney complex.⁷

Literature on RVM highlights their diverse clinical presentations, which can complicate diagnosis and management. Massive pericardial effusion in the context of RVM is often misattributed to infectious or malignant processes, delaying definitive treatment. Pericardial effusion in these cases underscores the importance of considering myxomas in the differential diagnosis of unexplained effusions, particularly when imaging reveals intracardiac abnormalities.^{2,3}

A multidisciplinary approach is crucial in the management of rare cardiac tumors. Collaboration between cardiologists, cardiothoracic surgeons, radiologists, and pathologists ensures a comprehensive evaluation and optimal treatment outcomes. The integration of advanced imaging modalities with intraoperative findings and histopathological confirmation forms the cornerstone of accurate diagnosis and effective management.⁴

Emerging advancements in the diagnosis and management of cardiac myxomas aim to improve diagnostic accuracy and minimize recurrence. Novel imaging techniques, such as three-dimensional echocardiography and functional cardiac MRI, provide enhanced visualization and assessment of tumor hemodynamics. Furthermore, molecular and genetic studies have identified potential pathways involved in myxoma formation, offering insights into targeted therapies and risk stratification.^{3,5}

The rarity of RVM emphasizes the importance of comprehensive documentation and analysis of their clinical presentations, diagnostic challenges, and surgical outcomes. Insights gained from these cases contribute to the broader understanding of cardiac myxomas and underscore the need for ongoing research to refine diagnostic and therapeutic strategies. Future advancements in molecular diagnostics and imaging techniques hold promise for improving outcomes for patients with rare cardiac tumors.^{6,8}

CONCLUSION

Right ventricular myxomas are exceptionally rare cardiac tumors that pose significant diagnostic challenges due to their atypical location and potential to mimic other life-threatening conditions, such as ventricular aneurysms. The clinical presentation, often complicated by associated findings such as massive pericardial effusion, can obscure the underlying pathology and lead to diagnostic uncertainty. Intraoperative findings, as demonstrated in this case, play a pivotal role in clarifying the diagnosis and guiding appropriate management.

Surgical resection remains the gold standard for treating cardiac myxomas and provides excellent outcomes when performed with complete tumor excision. The shift in diagnosis from an impending rupture of a right ventricular aneurysm to a right ventricular apical myxoma during surgery underscores the critical importance of surgical exploration and histopathological confirmation. This highlights the limitations of preoperative imaging in accurately characterizing certain cardiac masses, particularly in rare locations. Early recognition, comprehensive evaluation, and timely intervention remain the cornerstones of successful management of rare cardiac tumors like right ventricular myxomas.

CONFLICT OF INTEREST

All author declares there is no conflict of interest.

FUNDING

None.

PUBLICATION ETHICS

The patient had received signed written informed consent regarding publication of medical data in scientific medical journal with confidentiality to personal information.

AUTHOR CONTRIBUTION

All authors had contributed to manuscript writing and agreed for the final version of manuscript for publication.

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