**A Rare Case of Atrial Myxoma Mimicking Right-Sided Heart Failure**

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ABSTRACT

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| **Background**: Myxomas are rare, with an incidence of 0.0017% in the general population. The majority (75–80% of cases) of myxomas are located in the left atrium, and 18% of cases are located in the right atrium with diverse clinical manifestations.  **Presentation of Case**: A 30-year-old woman presented with a chief complaint of dyspnea and near syncope on effort, and a history of ascites and bilateral lower extremity edema. Transthoracic echocardiography (TTE) disclosed a substantial mass in the RA, indicative of myxoma, causing right ventricular (RV) inflow obstruction with a mean gradient of 18 mmHg. The patient was undergoing urgent open-heart surgery with resection of an 8.5 x 6.5 cm multilobulated mass. The residual septal defect was rectified with direct closure. Tricuspid valve coaptation testing was satisfactory. The diagnosis of myxoma was validated through a histological examination. The post operative recovery was unremarkable, and the patient was discharged on the sixth day following surgery.  **Discussion and Conclusion:** RA myxoma often goes undetected due to its low incidence rate and its character of mimicking right heart failure presentation. Postponement of surgical operation may result in irreversible consequences therefore surgical resection should be executed promptly following diagnosis. |

***Keywords:*** *intracardiac neoplasms, RA myxoma, right heart failure, edema*

1. INTRODUCTION

Intracardiac neoplasms represent 0.2% of all human malignancies.(Arruda et al., 2008) These masses are classified as primary (benign or malignant) and metastatic (malignant). Metastatic neoplasms occur 20-40 times more frequently than initial neoplasms. Primary cardiac neoplasms are uncommon. About 75% of primary neoplasms are benign, including myxoma, which in common population has an occurrence of 0.0017%. Myxoma is an authentic neoplasm originating from multipotent mesenchymal cells of the sub-endocardium histologically.(Arruda et al., 2008; De et al., 2008; Hermans et al., 2003; Jang et al., 2010)

Myxoma is located in the left atrium in 75-80% of cases and typically manifests with indications of mitral valve pathology or thrombo-embolic occurrences. They may occur in atypical locations within the right atrium (RA) in 18% of cases.(Diaz et al., 2011) The other assessment primarily distinguishes between thrombus and vegetation.(Jang et al., 2010) Thrombus and vegetation didn’t adhere to interatrial septum (IAS).

Due to its low incidence, the literature regarding signs, symptoms, and diagnoses is also very limited. Although its benign nature, myxoma has varied signs and symptoms that make it difficult to establish an initial intracardiac neoplasm diagnosis.(De et al., 2008) The symptoms of myxoma may remain asymptomatic or advance to thromboembolic events, potentially resulting in sudden death.(De et al., 2008; Hermans et al., 2003; Murayama et al., 2001) The symptoms are contingent upon the size, location, and mobility of the myxoma.

The conventional triad in patients with myxoma consists of flow obstruction, thromboembolic manifestation, and constitutional symptoms.(De et al., 2008; Hermans et al., 2003; Oliveira et al., 2010) RA myxoma can obstruct the tricuspid valve, which results in right heart failure signs and symptoms such as elevated jugular venous pressure, peripheral edema especially in lower legs, ascites, hepatic congestion, hypotension, shock condition, and syncope.(Oliveira et al., 2010)

If the diagnosis of myxoma is confirmed, surgical resection of the myxoma should be performed immediately due to the risk of flow obstruction and thromboembolism.(Arruda et al., 2008; Guhathakurta & Riordan, 2000) Surgical treatment generally results in good outcomes for patients and recurrence rates in cases of myxoma resection are very rare. This report details an uncommon case of RA myxoma, emphasizing the challenges in the awareness of this neoplasm due to its atypical location and size.

2. PRESENTATION OF CASE:

A 30-year-old woman came to the referring hospital with a primary complaint of ascites and bilateral lower leg edema. The patient had a history of dyspnea and near syncope during moderate-heavy activity for the past 6 months, which had worsened two months ago. There was no history of hypertension, diabetes, chronic disease, or malignancy. The patient also had no family history of malignancy. Physical examination was consistent with signs of right heart failure such as increased jugular venous pressure (JVP), ascites, and bilateral leg edema.

The patient was then treated with right heart failure management. During treatment, the patient underwent transthoracic echocardiography (TTE), and a suspected RA myxoma was found. The patient’s clinical condition then improved and was discharged on the 6th day of treatment with a plan to be referred to our hospital per polyclinic.

The patient came to our hospital polyclinic in a stable hemodynamic state with GCS E4M6V5, blood pressure 96/55 (MAP 68) mmHg, heart rate 88 bpm, and a respiratory rate of 18-20 tpm. No abnormalities were found in the blood laboratory examination. A chest x-ray revealed a clear lung with ra dilatation. TTE showed a mass suspected RA myxoma, which was fixed to the fossa ovalis of the IAS measuring 7.7 cm x 5.3 cm, which caused RV inflow obstruction with a mean gradient of 18 mmHg.

The urgent cardiology team discussion was held with the conclusion of the discussion: the patient was undergoing emergency surgery for the RA myxoma. After obtaining informed consent, our patient was performed open heart surgery under general anaesthesia. The atriotomy procedure was performed to access the RA. The large neoplasm was seen affixed to the margin of the fossa ovalis and then was excised together with the IAS to which the neoplasm was connected. The residual septal defect has been repaired with a direct closure approach. The tricuspid valve leaflet coaptation was satisfied and remained without surgical intervention. Transesophageal echocardiography (TEE) revealed modest tricuspid valve regurgitation during the weaning process from cardiopulmonary bypass. The length of the cardiopulmonary bypass was 64 minutes.

Macroscopically, the neoplasm presented as a 8.5 x 6.5 cm lobulated mass with an irregular surface, featuring polypoid regions of elastic consistency and vulnerable tissue. Microscopy revealed a profusion of myxoid stroma accompanied by stellate cells. Necrosis, mitotic activity, atypia, and pleomorphism were absent. The histological analysis indicated a benign myxoma. The postoperative recovery went well, and the patient was discharged on the 6th postoperative day.

3. discussion

Primary cardiac neoplasms are infrequent, with an estimated frequency ranging from 0.0017-0.19%, constituting less than 5% of all cardiac malignancies.(Azevedo et al., 2010) Myxoma is the most common primary cardiac neoplasm. The RA is an atypical site, accounting for 15% to 20% of myxoma occurrences.(Diaz et al., 2011) A low prevalence of RA myxoma has been documented for decades. About 70% of the individuals afflicted are women(Jang et al., 2010; Manfroi et al., 2001) Primarily between the ages of 30 and 60,(Guhathakurta & Riordan, 2000; Ojji et al., 2008) In accordance with our patient, of the 30-year-old woman presented in this report.

The clinical manifestations of myxomas depend on the size, mobility, and location of the neoplasm, the clinical presentation of RA myxoma differs from LA myxoma.(Oliveira et al., 2010) The RA myxoma can be asymptomatic(Yuce et al., 2007), or manifest also through three symptom categories: (1) hemodynamic impairments due to obstruction of blood flow (2) neoplasm embolization or pulmonary embolism, and (3) constitutional symptoms.(Kirklin & Blackstone, 2012; Klaus, 2024) If the neoplasm is large, it can entirely block the valve orifice, resulting right heart failure sign and symptoms. The severe blockage of tricuspid valve potentially resulting in cardiogenic shock and even sudden death.(Modi et al., 2010)

Dyspnea is the most prevalent sign, occurring in 80% of patients, with instances of right heart failure also documented. Dyspnea on exertion was established in this case. Physical examination findings consistent with right heart failure, including elevated jugular venous pressure (JVP), ascites, and bilateral lower extremity edema. Echocardiographic evaluation revealed a mean gradient of 18 mmHg across the tricuspid valve, indicative of right ventricular (RV) inflow obstruction.

Familial cardiac myxomas are characterized by involvement in sites other than the left atrium, including biatrial involvement and ventricular involvement, and by a high recurrence rate. Familial cardiac myxomas may present as part of a syndrome (Carney's complex, LAMB, or NAME syndrome) or as nonsyndromic familial cardiac myxomas. Carney's complex, as described by J.A. Carney, is characterized by the association of cutaneous pigmentation, fibromyxoid tumors of the skin, myxomas of the heart, endocrine overactivity, and autosomal dominant inheritance.

The RA myxoma typically originates in the fossa ovalis or the base of the IAS (STOLF et al., 2000) which aligns with our case. A recent study documented 19 years of surgical management for primary myxoma and reported that seven (17%) of the 41 patients originated from the RA. The average maximal size of the RA myxoma was 5.1 ± 1.8 cm.(Samanidis et al., 2011) Our case represents one of the largest RA myxomas documented in the literature (7.7 cm x 5.3 cm).

Echocardiography is the most effective diagnostic technique for identifying and evaluating the amount of myxoma and finding reoccurrence, with a sensitivity of up to 100%. Despite that, TTE may not be enough to find cancers less than 5 mm wide, indicating a TEE is needed when an impression of a minuscule neoplasm exists.(Manfroi et al., 2001) Transthoracic echocardiography suggested the presence of an RA myxoma, which was later confirmed through histological analysis. Echocardiography is the preferred method to evaluate cardiac masses; however, MRI and CT scans offer insights into tissue features and provide a comprehensive view of cardiac and paracardiac morphology.(Oliveira et al., 2010) CT scan was not performed on this case considering that an immediate cardiac CT scan could not be performed, and TTE showed that the myxoma had caused RV inflow obstruction with a mean gradient of 18 mm, which could at any time cause shock condition, total obstruction, or even mortality.

The recommended treatment for myxomas is surgical excision.(Arruda et al., 2008; Guhathakurta & Riordan, 2000) Thorough excision of the neoplasm and its implantation base, ensuring an adequate margin, is crucial for curing the symptoms, averting reoccurrence, and further surgical interventions, which may subject the patient to additional difficulties like hemorrhage and requirements for blood products. Myxoma is also often excised through extensive excision of the pedicle or attachment to avert recurrence. The myxoma was attached to the IAS, doing an excision with an extensive margin potentially causing residual atrial septal defect. The surgical procedure involved myxoma excision and direct closure for IAS repair.

The reoccurrence of sporadic neoplasms is minimal, ranging from 1% to 3%. Seven mortality from operation varies from 0% to 3% across several studies.(Murayama et al., 2001; Oliveira et al., 2010) The postoperative survival rate is increased.(Guhathakurta & Riordan, 2000) in this case, our patient improved, and no events occurred during treatment.

4. Conclusion

Although an uncommon site for a substantial myxoma, the RA myxoma must consistently be included in the diagnosis of a right-sided cardiac neoplasm, particularly when it also exhibits manifestations of right heart failure with an indeterminate cause. The cardiologists and surgeons must promptly diagnose and give further treatments to patients with these malignancies to enhance prognosis.

Consent

All authors declare that ‘written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

**DISCLAIMER (ARTIFICIAL INTELLIGENCE)**

Author(s) hereby declares that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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Abbreviations

TTE : Transthoracic Echocardiography

TEE : Transesophageal Echocardiography

RV : Right Ventricular

RA : Right Atrium

LA : Left Atrium

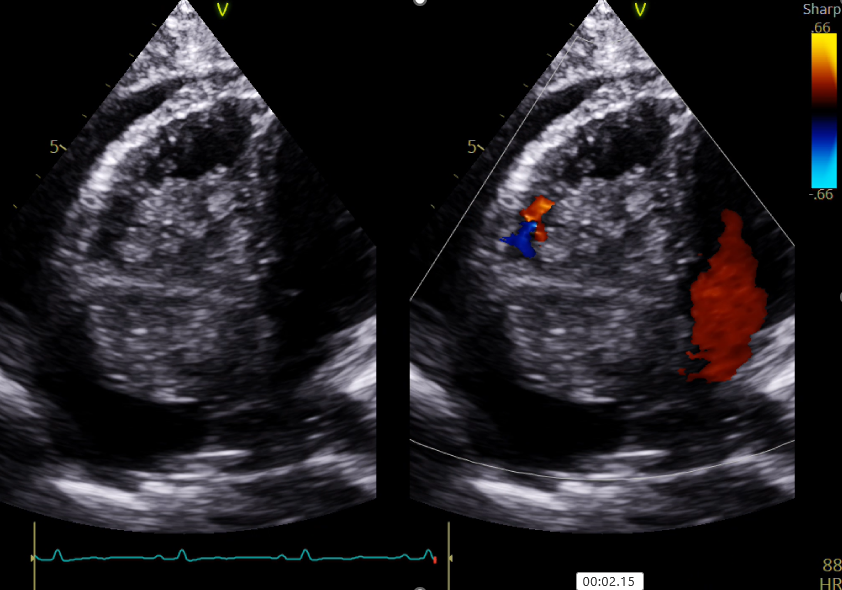
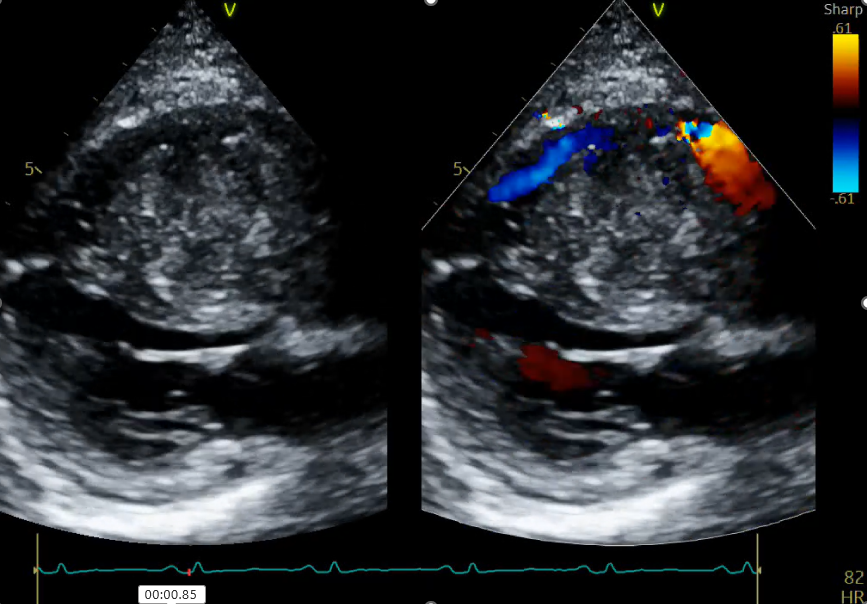
JVP : Jugular Venous Pressure

IAS : Interatrial Septum

MRI : Magnetic Resonance Imaging

CT Scan`: Computed Tomography Scan

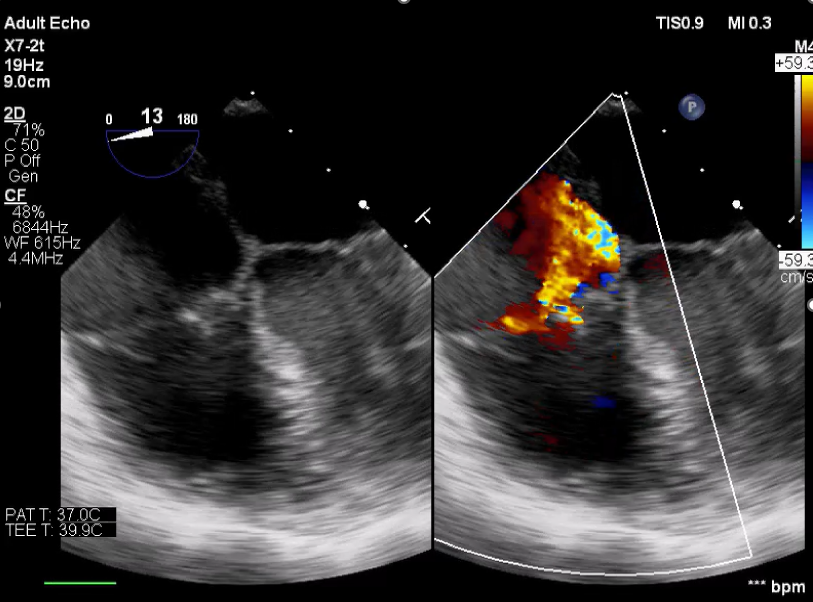
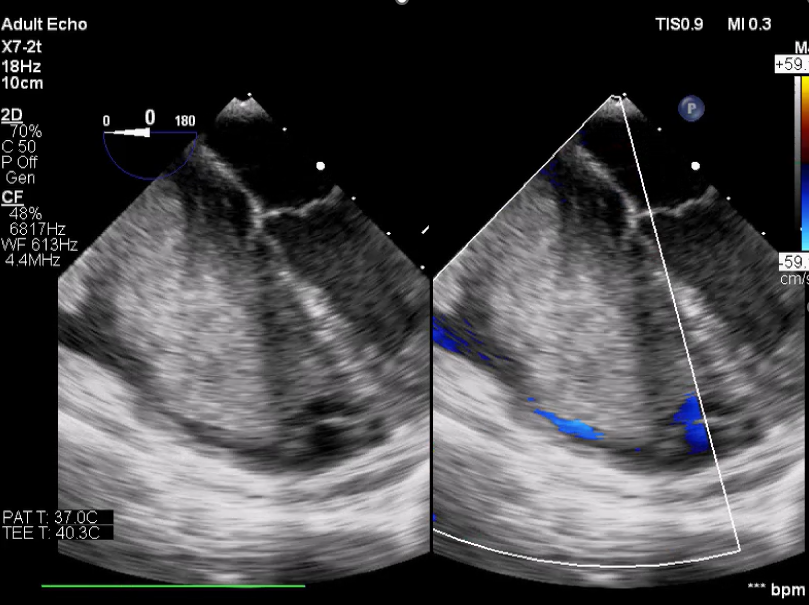
**Figure 1**. TTE showed a large RA myxoma on both PLAX view (A) and apical 4-chamber view (B) with RV inflow obstruction with a mean gradient of 18 mmHg. TTE: transthoracic echocardiography; RA: right atrium; PLAX: parasternal long axis; RV: right ventricular.



**A**

**B**

**Figure 2**. (A) TEE pre-surgery showed a large RA myxoma with RV inflow obstruction, (B) TEE post-surgery showed complete myxoma resection, complete IAS repair, and tricuspid regurgitation. TEE: transesophageal echocardiography; RA: right atrium; RV: right ventricular; IAS: interatrial septum.



**A**

**B**

**Figure 3**. Large RA myxoma after complete resection surgery. RA: right atrium.

A piece of food on a white surface

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