

A Ticking Time Bomb in the Heart: A Case Report of Cardiac Myxoma

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Abstract

Background: Cardiac myxoma is the most common primary cardiac tumor and is frequently found in the left atrium. Symptoms and signs are most often non-specific, making diagnosis difficult. We present a case report of an atrial cardiac myxoma with an atypical presentation

Case Illustration: A 55-year-old man presented to our outpatient clinic with recurrent and intermittent chest discomfort that had lasted for months. No other cardiac symptoms such as dyspnea or palpitations were noted. The patient's medical history was positive for hypertension and smoking. Thorough chest pain evaluations were conducted, there were no ST-T changes on electrocardiogram. However, Transthoracic Echocardiography (TTE) revealed an incidental finding of a 2.1 x 2.6 cm mass, located in the Right Atrium (RA), with the stalk attached to the interatrial septum. Subsequently, patient was consulted to cardiothoracic surgeon and underwent successful tumor resection via median sternotomy. The tumor was confirmed histopathologically.

Conclusions: The nonspecific symptoms of cardiac myxomas pose a diagnostic conundrum to clinicians. TTE plays a pivotal role in diagnosis, and surgical removal of the mass should be undertaken as soon as possible. Early detection and intervention remain imperative to rid the risk of this cardiac pathology.

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Introduction

Cardiac myxomas are one of the most common primary neoplasms of the heart, accounting for 50-70% of all primary cardiac tumors.¹ It originates from the limbus fossa ovalis of the Interatrial Septum (IAS), although it may also originate from other places, such as the posterior atrial wall, the anterior atrial wall, and the atrial appendage.² Symptoms are typically a triad of obstruction (e.g., syncope), embolization, or constitutional symptoms (e.g., fever, weight loss); however, in one-fifth of the cases, the patient is asymptomatic, leading to potential diagnostic delay.¹ Echocardiography is the gold standard for the diagnosis of cardiac myxoma, with reported sensitivity of 97% and specificity of 50%.³ The objective of this case report is to highlight the diagnostic challenges associated with Right Atrium (RA) myxoma, specifically when presenting with isolated, atypical chest pain. We emphasize the critical role of echocardiography in the early detection of masses.

Case Illustration

A 55-year-old man presented to our outpatient clinic with recurrent and intermittent chest discomfort, described as sharp, non-radiating, that had lasted for months. No other cardiac symptoms such as dyspnea or palpitations. Constitutional symptoms such as fever, weight loss, or myalgia were also not present. Physical exam revealed no remarkable findings. Transthoracic Echocardiography (TTE) was subsequently performed, which revealed normal left ventricular systolic function. No regional wall motion abnormalities were observed, and the valves were normal. However, there was a pedunculated mass in the RA, approximately 2.1 x 2.6 cm, with the stalk attached to the IAS (Figure 1). The electrocardiogram showed a normal sinus rhythm without ST-T changes, and the cardiac catheterization showed non-significant coronary artery disease. Following a thorough discussion during the case conference involving both cardiology and cardiac surgery, the consensus was that surgery was imperative.

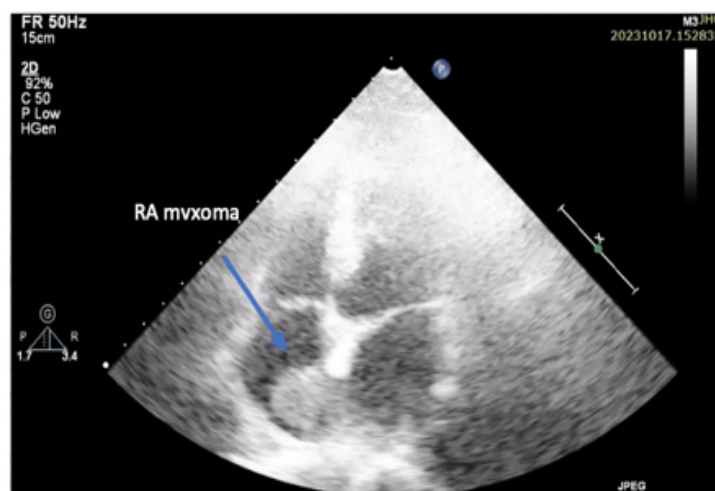


Figure 1. Transthoracic echocardiography of right atrial myxoma.

Preoperative transesophageal echocardiography revealed a mass approximately 2.8 x 2.4 cm, which was filling the RA with the stalk on the atrial septum near the sinus venosus (Figure 2). No obstruction of RA-Right Ventricle (RV) flow due to the mass was appreciated. Surgical excision was performed using a median sternotomy approach. An incision was made in the RA, revealing a pedunculated mass arising from the interatrial septum. The mass and a portion of the IAS were resected, followed by

closure of the IAS defect with a pericardial patch. The specimen was sent for pathology evaluation, and a myxoid growth with oval, bland nuclei and eosinophilic cytoplasm was observed. Pathologic examination further revealed a perivascular ring of numerous myxomatous cells surrounding small intratumoral vessels (Figure 3). No malignant features were observed. The patient's postoperative course was uneventful, and they were discharged on the fifth day.

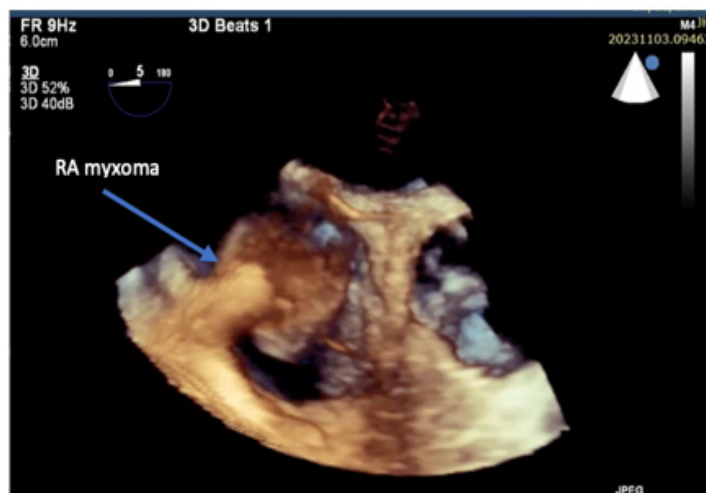


Figure 2. Transesophageal echocardiography 3D reconstruction of right atrial myxoma.

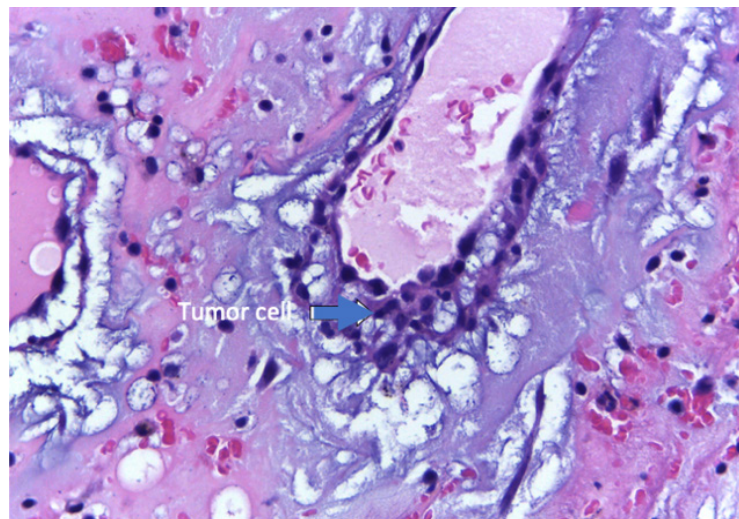


Figure 3. Histopathological examination with hematoxylin eosin smear demonstrating perivascular ring with numerous myxomatous cells.

Discussion

Cardiac myxomas have an uncertain origin. The prevailing hypothesis proposes that it originates from multipotent cardiac stem cells. Although familial inheritance is possible, the majority of cases occur sporadically, approximately 90% of cases.⁴ Location of myxomas varies; one study reported that the majority (75.7%) originate in the left atrium, 16.8% in the RA, 7.5% in biatrial or either ventricle. The majority of myxomas that arise in the RA were attached to the fossa (61.1%), tricuspid valve (22.2%), atrial appendage (5.6%), coronary sinus (5.6%), and inferior vena cava (5.6%).⁵ All age groups could be affected, although the majority of patients are between 30 and 50 years of age.⁶

The diagnosis of cardiac myxoma is often challenging. Although the textbook clinical presentation is classically a triad of obstructive cardiac symptoms, embolic phenomena, and constitutional symptoms, some patients might remain asymptomatic. The location of the myxomas may play a role in the different presentations of patients. For example, unlike Left Atrial (LA) myxomas, which often present with systemic embolization (Stroke) or pulmonary congestion mimicking mitral valve disease, RA myxomas are often asymptomatic until they grow large enough to obstruct the tricuspid valve and cause right heart failure. Physical exams may reveal a diastolic or systolic murmur or “tumor plop”.⁷ In our case reports, chest discomfort was the only symptom

reported by the patient. Our case report describes a patient with chest discomfort as the sole presenting symptom. This presentation contrasts with previously published cases of similarly sized RA myxomas, which commonly report findings such as heart failure or a systolic murmur on physical exam.⁸ One study of 61 patients reported that the incidence of chest pain at presentation for RA myxoma was 4.9% (3 patients), and that the tumor was an incidental finding in 6% of cases.⁹

Our patient initially presented with chest discomfort, for which our initial differential diagnosis was chronic coronary syndrome. However, echocardiography revealed the patient had a cardiac myxoma. Transthoracic echocardiography has a reported sensitivity of 95% for cardiac myxomas, which approaches 100% with TEE. The modality is essential of determining the tumor's location within the heart and its point of attachment. When evaluating a cardiac mass, examiners should consider differential diagnoses such as thrombus and other types of tumors (e.g., angiosarcoma). A thrombus is typically associated with conditions such as heart failure with low ejection fraction or atrial fibrillation. Key differentiating features on the echo are the morphology and mobility. A thrombus often has a layered, laminated, and irregular appearance with a heterogeneous texture, whereas a myxoma is a well-defined oval mass. Thrombi are often sessile, while myxomas are almost always pedunculated, attached by a stalk. Malignant tumors like angiosarcomas often invade the myocardium and extend into the pericardium, but confirmation of tissue specimen is required as a confirmatory test for the definite diagnosis of cardiac myxomas.¹⁰

According to the World Health Organization (WHO), the diagnosis of cardiac myxoma is dependent on the presence of cells with ovoid nuclei, eosinophilic cytoplasm, and indistinct cell borders, termed myxoma cells. The presence of an aneuploid cell population needs to be evaluated, as it indicates the presence of neoplasia. Our pathology findings in this case are in line with the WHO description. We found numerous myxomatous cells surrounding the intratumor small vessels, and there were no aneuploid cells, indicating this is a benign cardiac myxoma.¹¹

Surgical excision of the cardiac myxoma is the treatment of choice for this condition. This is necessary to reduce the risk of tumor embolization. Surgery is classically performed by median sternotomy; the root of the stalk and the full thickness of the adjacent IAS are excised, and

the corresponding defect is closed appropriately. Excellent outcomes of both survival and tumor recurrence have been reported after surgical resection.² In a series of 403 patients, over of 4.5-year follow-up, the mortality rate was 5.2%. Another study reported that 10-, 20-, and 30-year survival rates were 77%, 52%, and 34%, respectively.¹² The recurrence rate of cardiac myxoma ranges between 3-5.2%, with ventricular myxoma having a higher recurrence.¹²⁻¹³ In our case, the patient has shown echocardiographic evidence of recurrences during two years of follow-up. Although no official guidelines exist, our institution's protocols include echocardiograms before discharge, 6 months post-surgery, and annually thereafter to monitor for recurrence. Family screening may be considered if the myxomas present with features suggestive of Carney Complex, though none were present in this patient.

Conclusion

Cardiac myxoma is a rare tumor with low prevalence in the general population. Its symptoms are often non-specific, and its diagnosis requires a high degree of suspicion. Echocardiography is a crucial modality for diagnosis. Surgical resection should be promptly undertaken once the diagnosis has been made.

List of Abbreviations

IAS	Interatrial Septum
RA	Right Atrium
RV	Right Ventricle
WHO	World Health Organization
TTE	Transthoracic Echocardiography
TEE	Transesophageal Echocardiography

Ethical Clearance

Not applicable.

Publication Approval

All authors consent to the publication of this manuscript.

Author Contributions

RS: conception and design, drafting article, revising article; TMHP: conception and design, drafting article, revising article, final approval of version publish; ID: drafting article, revising article, final approval of version publish; GRF: drafting article, revising article, final approval of version publish;

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