# Not all cardiac tumors are myxomas and not all benign tumors are benign: two case reports

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## ABSTRACT

**Background:** Primary cardiac tumors originating in the heart are much less frequent than metastasis from other organs. The majority of cardiac tumors are of benign nature and only 25% are estimated to be malignant. Common symptoms of cardiac tumors include systemic embolization, congestive heart failure, and arrhythmias.

**Case report:** We present two cases of cardiac tumors initially causing heart failure symptoms and valvular disease in relatively young patients. These tumors were initially diagnosed as myxomas, but post-op histological studies revealed the presence of angiosarcoma and hamartoma. It is important to differentiate and have a clear diagnosis because some malignant and invading tumors may require the use of adjuvant chemotherapy agents.

**Conclusion:** It should not be taken for granted that cardiac tumors are of benign nature even if initial evaluation is consistent with myxomas. Early resection and pathological study of primary cardiac tumors are important in identifying the tumor type and targeting management.

**Keywords**: Cardiac tumors, myxoma, cardiac angiosarcoma, cardiac hamartoma, case reports.

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## Background

Cardiac tumors are extremely uncommon and are diagnosed incidentally in the vast majority of cases. Metastasis to the heart is 30 times more common than primary cardiac tumors. Of the primary cardiac tumors, only 25% are estimated to be malignant [1]. Myxoma is the most common benign primary tumor, while angiosarcoma is the most common malignant tumor and is estimated to be 75% of malignant cardiac tumors; hamartomas are very infrequent [2,3]]. Surgical removal is the primary treatment modality associated with the most favorable outcomes [4]. Symptoms vary by location and size rather than the nature of the tumor. They may include systemic embolization, congestive heart failure, arrhythmias, and constitutional symptoms [1,4]. Left-sided sarcomas are generally less infiltrative and present with obstructive and severe heart failure symptoms. Right-sided sarcomas are generally malignant and infiltrating; they do not present with congestive symptoms until advanced disease stages [1].

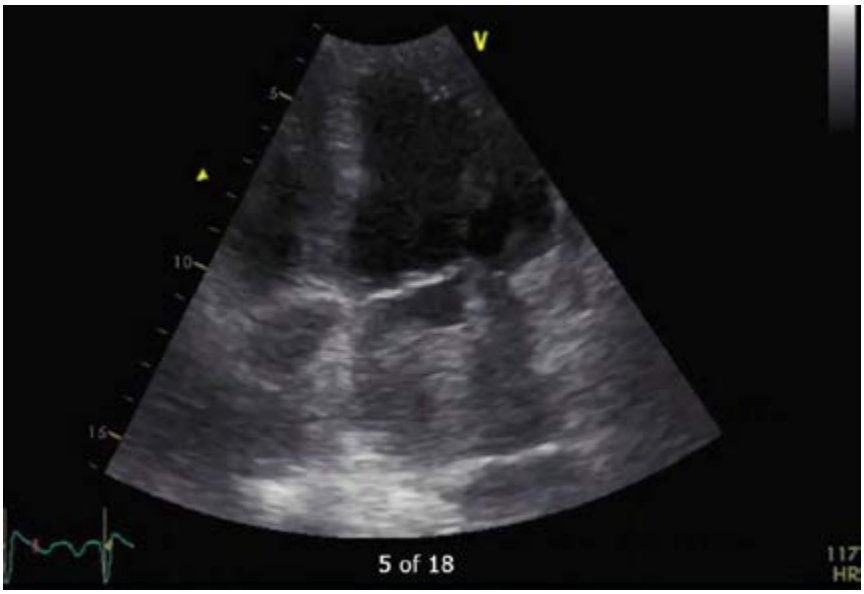
## Learning Objective

We present two case reports that highlight the importance of early detection, resection, and pathological diagnosis of cardiac tumors. Malignant cardiac tumors can lead to the invasion and destruction of cardiac structures mimicking valvular lesion or heart failure. It is important to determine the nature of the cardiac tumor in order to better direct management and improve patient prognosis.

### *Case presentation 1*

A 55-year-old female patient known to have hypertension and a history of cerebrovascular accidents presented for exacerbating heart failure symptoms of 2 months duration. On physical examination, the patient appeared alert, awake, and oriented. Cardiopulmonary examination revealed scattered crackles in bilateral lower lung lobes; no accompanying murmurs were identified. For further evaluation, a transthoracic echocardiogram was performed, which showed a large left atrial mass with features compatible with myxoma, protruding into the left ventricle causing functional mitral stenosis and severe mitral regurgitation (Video 1). Estimated Ejection Fraction (EF) at time of diagnosis was 35%.

The surgical approach was through a vertical sternotomy. After the installation of a classical bypass between




***Video 1**. Large left atrial mass protruding into the left ventricle causing functional mitral stenosis and severe mitral regurgitation. Mass was found to be an Angiosarcoma on histopathology.*

*URL: https://www.youtube.com/watch?v=YHWGSqdnC3U&ab_channel=EuropeanJournalofMedicalCaseReports*

the aorta and a double venous cannulation, a right atriotomy was performed and the left atrium was reached through a resection of 2.5 cm at the foramen ovale.

The tumor of 8 cm × 4 cm was adherent to the inferior right pulmonary vein and to the posterior wall of the left atrium but not to the septum. The tumor was passing through the mitral valve, with invasion of the valve, leading to a high-grade mitral stenosis. The patient was hypervolemic as confirmed by echocardiographic measures of pulmonary artery pressure reaching 100 mmHg.

A complete resection of the tumor was performed, and no residual tissue was seen macroscopically (Figure 1). Severe mitral regurgitation was discovered due to a prolapse of the P3 leaflet and a restriction of the anterior leaflet. The mitral valve was replaced by a biological St Jude #29 prosthesis and the atrial septum was closed by a Dacron patch. Re-airing and injection of a hot bloodshot into the aorta was performed, and going off pump was easy and without need for inotropes. Cytological studies confirmed the presence of an angiosarcoma and not a myxoma.

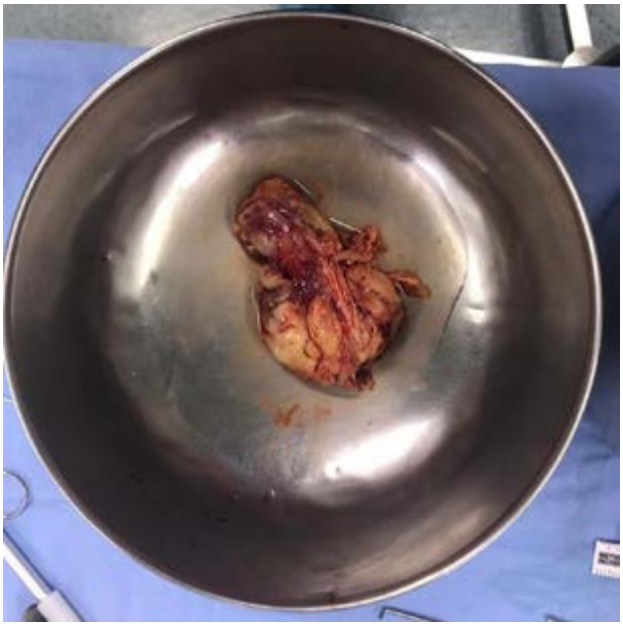


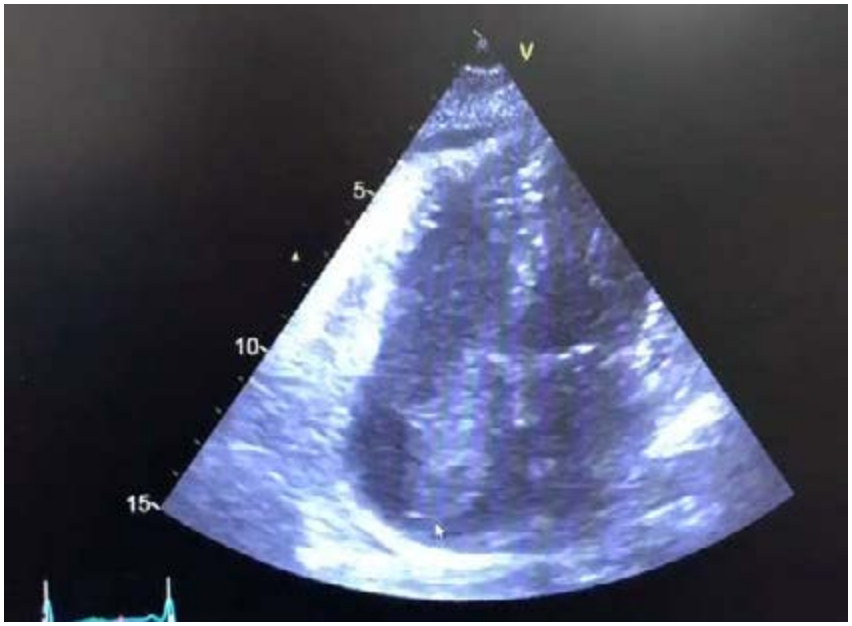
***Figure 1.** Complete resection of angiosarcoma tumor in Patient 1. Tumor was protruding through the mitral valve and occluding it, but without any adherences to the valve or annulus.*

### *Case presentation 2*

A 52-year-old female patient with a past medical history of deep venous thrombosis presented for exacerbating heart failure symptoms of 3 months duration. On physical examination, the patient was awake, alert, and oriented. Cardiopulmonary examination revealed no murmurs. 1+ bilateral lower limb pitting edema was present with jugular venous distention 1 cm above the sternal angle. A transthoracic echocardiogram was performed to assess the origin of dyspnea and it showed a massive right atrial tumor adherent to the inferior vena cava, protruding into the right ventricle (Video 2). On echocardiography, EF was estimated to be 32%.

An urgent surgery was planned and performed by a vertical sternotomy. After the installation of a classical bypass between the aorta and a venous cannulation of the superior vena cava, and a femoral right vein cannulation, a right atriotomy was performed.

The tumor of 15 cm × 5 cm was adherent to the inferior vena cava and not to the septum. The tumor was protruding through the tricuspid valve and occluding it, but without any adherences to the valve or the annulus.




***Video 2.*** *Massive right atrial tumor adherent to the inferior vena cava, protruding into the right ventricle. Mass was found to be a Hamartoma on histopathology.*

*URL: https://www.youtube.com/watch?v=gTbSTLbIZ74&ab_channel=EuropeanJournalofMedicalCaseReports*

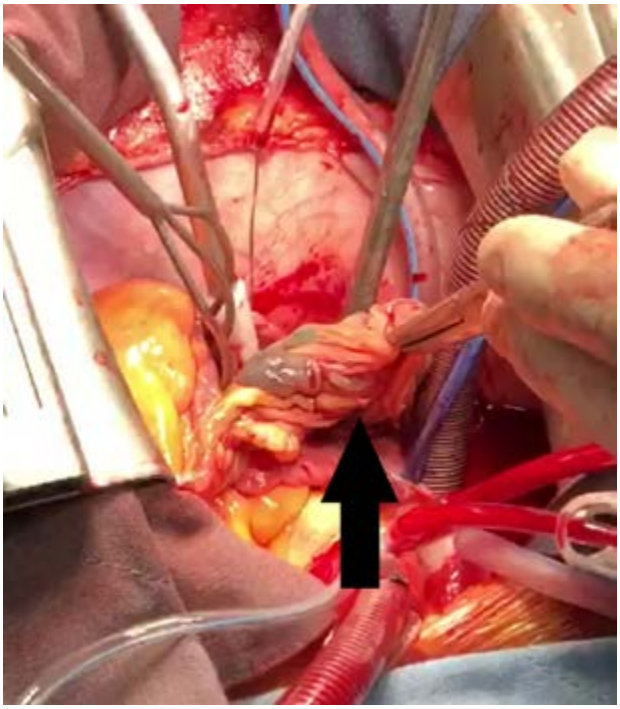


***Figure 2.*** *Complete resection of hamartoma performed in Patient 2. Tumor was protruding through the tricuspid valve and occluding it, but without any adherences to the valve or annulus.*

A complete resection of the tumor was performed and no residual tissue was seen macroscopically (Figure 2). Re-airing and injection of a hot bloodshot into the aorta was performed, and going off pump was easy and without use of inotropes. Cytological studies confirmed the presence of a hamartoma and not a myxoma.

## Discussion

Cardiac tumors are extremely rare, accounting for 0.03% of overall tumors, and can be divided into primary tumors and metastatic tumors. It is well established that advanced tumors can lead to development of arrhythmias, heart failure symptoms, valvular heart diseases, and systemic embolization [1,5]. In left-sided heart tumors, emboli can travel to the brain or peripheral organs, whereas emboli from the right-sided tumors can reach the lungs. In order to detect a tumor, the main diagnostic modality is echocardiography, which helps to differentiate between the different types of tumors. The gold standard test to confirm the diagnosis is to resect a tumor and take a biopsy [6].

Sarcomas can be seen in all age groups (mostly among patients in their 30s-50s). They are mainly located in the right-sided cavities and can affect the pericardium as well. In rare cases, the tumor can invade the atrioventricular valves, leading to valve stenosis and regurgitation [7]. It has been noted that left-sided tumors are less infiltrative, have better overall survival, and have a later onset of metastasis as compared to right-sided ones [8].

Among sarcomas, angiosarcomas are the most common subtype (representing 40% of the cases), characterized by aggressive local growth and early spread [6]. Echocardiography is the best initial test for diagnosis [6].

Hamartomas is a benign primary tumor that is extremely rare (approximately 25 cases have been reported in the literature). The most common origin is in the ventricular free wall but can rarely originate in the atria [9]. Patients having this tumor are mostly asymptomatic; however, as the tumor grows, it becomes obstructive and can rarely progress to sudden cardiac death [10].

All types of cardiac tumors can grow rapidly, leading to the invasion of heart structures, damaging valves. This can happen in both malignant and benign tumors. The most essential part of the management is to avoid such complications by performing early diagnosis and surgery [6]. The tumors in our two cases induced the development of heart failure symptoms in patients that were previously healthy. In contrast to previously reported cardiac tumors, our patients did not have systemic symptoms or symptoms of metastasis at the time of presentation. The rapid development of severe heart failure was the only, but significant, unusual sign that led to extensive workup. Surgical intervention has led to the complete removal of the tumor and reversal of symptoms. In fact, repeat echocardiography a few weeks postsurgery in both patients led to the resolution of heart failure signs, drop in pulmonary artery pressure and improvement of valvular function.

This article gives additional insight on the presentation of cardiac tumors and the misleading nature of their presentation. It should be noted that in our cases, both tumors were initially mistaken to be myxomas of benign nature by the initial diagnostic tools. The decision of surgery was hastened by the severe heart failure symptoms in both patients. Histological studies revealed angiosarcoma (malignant type) and a hamartoma (benign but a very rare cardiac tumor) [3,6]. It is important to differentiate and have a clear diagnosis because some malignant and invading tumors may require the use of adjuvant chemotherapy agents. Thus, it is highly important to perform surgery and pathological studies as soon as possible on all cardiac tumors, even if they present like myxomas.

A transthoracic echocardiography performed 2 days post-op in case presentation 1 showed complete resolution of the mass, excellent functioning of the prosthesis, and a decrease in pulmonary pressure from 100 to 45 mmHg. Her symptoms improved and she resumed normal ambulation. Cytology confirmed the presence of angiosarcoma and not a myxoma.

A transthoracic echocardiography performed 2 days post-op in case presentation 2 demonstrated complete resolution of the mass, her symptoms improved, and she resumed normal ambulation. Cytology confirmed the presence of a hamartoma and not a myxoma.

### What is new?

This case highlights the importance of early detection, resection, and pathological diagnosis of cardiac tumors. It is important to determine the nature of the cardiac tumor and not assume the benign nature in order to better direct management and improve patient prognosis.

## Conclusion

Benign and malignant cardiac tumors have the ability to produce debilitating symptoms mimicking heart failure and valvular disease. Early surgical resection is the main modality of treatment with favorable outcomes for benign and malignant tumors. Surgery has been shown to lead to a complete reversal of heart failure symptoms in the two cases discussed.

### Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

### Funding

None.

### Consent for publication

Written and informed consent was taken from the patients to publish this case report.

### Ethical approval

Approval and consent of the ethics committee institutional review board was received for the publication of this article.

### Data availability statement

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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## Summary of the case

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| 1 | **Patient (gender, age)** | 1) Female, 55 year old<br>2) Female, 52 year old |
| 2 | **Final Diagnosis** | 1) Cardiac angiosarcoma<br>2) Cardiac hamartoma |
| 3 | **Symptoms** | 1) Exacerbation of heart failure<br>2) Exacerbation of heart failure |
| 4 | **Medications** | |
| 5 | **Clinical Procedure** | 1) Resection of tumor<br>2) Resection of tumor |
| 6 | **Specialty** | 1) Cardiology, Cardiothoracic surgery<br>2) Cardiology, Cardiothoracic surgery |