

# Recurrent cardiac rhabdomyosarcoma with multiple metastases: A case report

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## Abstract

Rhabdomyosarcoma, which accounts for 20% of all malignant tumors of the heart, is an aggressive tumor originating in the ventricular wall. These tumors are the second most common malignant primary tumor of the heart after angiosarcoma. Despite treatment options, such as surgical resection, radiotherapy, and chemotherapy, recurrence is common and mortality is high. Among these patients, survival with surgical resection is around six months to one year. In this case report, we discuss a patient who presented with recurrent rhabdomyosarcoma with distant metastasis after surgical intervention. A 56-year-old male patient who underwent left atrial mass excision and mitral valve replacement ten months prior was admitted with recurrent metastatic rhabdomyosarcoma.

**Keywords:** Rhabdomyosarcoma, Cardiac, Recurrent tumor

## Introduction

Primary cardiac tumors are rare and have been reported at between 0.001% and 0.28% in autopsy series [1]. Of these primary tumors, 25% are malignant [2,3]. Rhabdomyosarcoma accounts for 20% of these malignant tumors [4,5]. In fact, rhabdomyosarcoma is very rare in adults compared to children [6]. Rhabdomyosarcoma, which often originates in the ventricular wall, spreads aggressively and has a high mortality rate [5]. In this case report, we will present a patient who was operated on for an intracardiac tumor with recurrent metastatic rhabdomyosarcoma detected ten months later.

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### Informed Consent

The authors stated that the written consent was obtained from the patient presented with images in the study.

### Conflict of Interest

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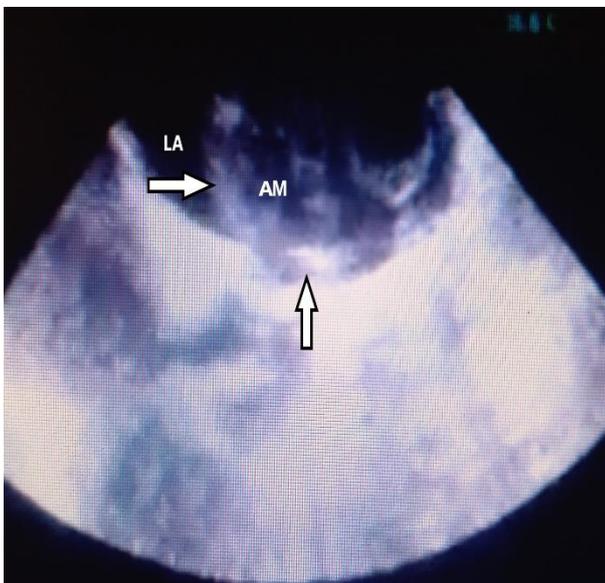
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## Case presentation

A 56-year-old male patient who had undergone left atrial mass excision and mitral valve replacement ten months previously at another center presented to the cardiology outpatient clinic with dyspnea, malaise, and fatigue. The international normalized ratio (INR) test showed an effective INR value. Other laboratory results revealed no pathology except a hemoglobin value of 10.6 gr/dl and a C-reactive protein (CRP) value of 163 mg/lt. The patient's transthoracic echocardiography (TTE) revealed a mitral valve gradient of 25/19 mmHg and a suspicious mass surrounding the left atrium. The patient was initially diagnosed as having possible left atrial mass and mechanical valve dysfunction. He was scheduled for transesophageal echocardiography (TEE) for a definitive diagnosis. The TEE showed a mass completely covering the left atrium and a raised gradient across the mechanical valve (Figure 1).

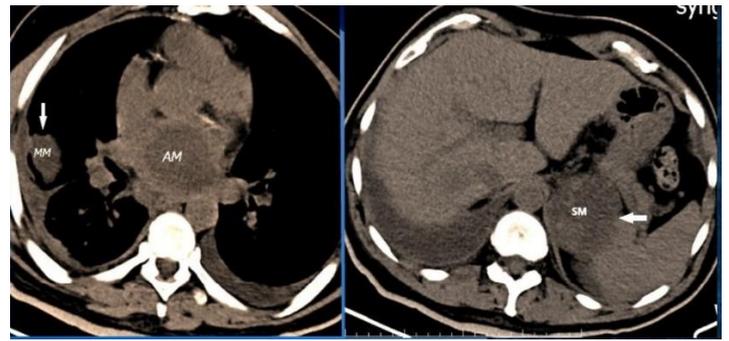
Figure 1: Transesophageal echocardiography revealed a mass filling the left atrium (LA: left atrium, AM: atrial mass)



Given that the patient had symptoms of decompensated heart failure, emergency surgical preparation was made. Preoperative computed tomography (CT) was performed for possible metastases. The CT examination showed multiple mediastinal lymph nodes, with the largest one measuring 13 mm, a mass appearance of 7 x 4 cm in the lower lobe of the right lung, a right paracardiac mass of 42 x 30 mm and a mass of 7 cm in the left adrenal gland, as well as a hypodense lesion with a diameter of 58 mm in the left atrium (Figure 2). The patient's data from the hospital where he had been previously been operated on showed that the postoperative histopathological result of the intracardiac mass excised was rhabdomyosarcoma. An interview with the patient and his relatives revealed that he was called to the hospital for radiotherapy and chemotherapy planning after the diagnosis, but the treatment was not continued, because he did not want to receive chemotherapy. After a council discussion with the cardiology, cardiovascular surgery, and oncology departments, the patient was considered to have a possible metastatic rhabdomyosarcoma and it was decided that this patient's condition was inoperable. The patient, who was

followed up in the intensive care unit, died on the fifth day after diagnosis due to multiorgan failure secondary to cardiac failure.

Figure 2: Computed tomography showing recurrence of the mass lesion in the left atrium, right lung, and surrenal gland (MM: metastatic mass in the lung, AM: left atrial mass, SM: surrenal metastatic mass)



## Discussion

Rhabdomyosarcoma, which was first described by Raycoff in 1937, is a malignant tumor arising from embryonic mesenchymal cells that can differentiate into skeletal muscle [1]. The literature shows that cardiac sites involved include the left atrium (55%), left ventricle (15.7%), right ventricle (15.7%), and right atrium (13%) [3].

This condition can be diagnosed by TTE, TEE, CT and magnetic resonance imaging [5]. The prognosis is often poor, with 75% of patients having distant metastases at the time of diagnosis. The spread of metastases occurs via lymphatic and hematogenous routes. Symptoms develop due to intracardiac obstruction by the mass. Patients often present with heart failure, dyspnea, mitral/tricuspid stenosis, arrhythmia, inferior/superior vena cava obstruction, restrictive cardiomyopathy, tamponade, and sudden death [1]. They may present with tumor-induced cerebral, pulmonary and peripheral embolism. Among these patients, survival with surgical resection is around six months to one year [3]. Although complete surgical resection is the most important prognostic factor, systemic chemotherapy and local radiotherapy for residual tumors are both important for possible survival [1]. However, there is no consensus on radiotherapy and adjuvant chemotherapy due to the poor prognosis [5]. Heart transplantation has also been attempted in cases where these treatments were ineffective and metastasis was excluded [7]. In their series of four patients, Uberfuhr et al. [7] reported a mean survival of 18 months, while Li et al. [8] reported a mean survival of 16 months after transplantation in a literature analysis of 46 patients.

In our case, a review of the patient's past surgical notes showed that the tumor was located in the left atrium and had invaded the mitral valve. During the surgical intervention, the tumor was removed, the mitral valve was completely resected and mechanical valve replacement was performed to avoid residual tumor tissue. Although the surgical intervention was successful, we do not know whether there was a residual focus or microscopic metastasis during the surgery. Failure to administer postoperative chemotherapy and radiotherapy may have caused his early recurrence within ten months. Notably, surgical intervention would have been a controversial decision in terms of prolonging survival if no metastasis had been detected in the patient.

## Conclusion

Intracardiac rhabdomyosarcoma is a malignancy with an aggressive course and high mortality despite all available treatment modalities, such as surgical resection, chemotherapy, and radiotherapy. Continuation of chemotherapy and radiotherapy after surgical intervention is important in terms of prolonging the survival of such patients.

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