

Giant Metastatic Intracardiac Malignant Fibrous Tumor Presenting with Supraventricular Tachycardia: A Case Report

Supraventriküler Taşikardi ile Prezante Olan Dev Metastatik İntrakardiyak Malın Fibröz Tümör: Olgu Raporu

ABSTRACT

Solitary fibrous tumors (SFTs) are rare neoplasms originating from fibroblastic mesenchymal cells. This case report details a 43-year-old female patient with cardiac SFT, a rarity in the medical literature, who presented with palpitations and a known malignancy. Supraventricular tachycardia was diagnosed in the emergency department, and a subsequent echocardiography revealed a giant mass in the left atrium after sinus rhythm restoration. The mass, causing symptomatic severe mitral stenosis, necessitated surgical intervention. Pathological examination of the surgical specimens confirmed the diagnosis of SFT. Cardiac masses are extremely rare in clinical practice, and the literature on SFTs is primarily limited to case reports. More research is needed due to the scarcity of information on managing this condition.

Keywords: Cardiac mass, cardio-oncology, solitary fibrous tumor

ÖZET

Soliter fibröz tümörler (SFT), fibroblastik mezenkimal hücrelerden kaynaklanan nadir neoplazmlardır. Bu vaka sunumunda, literatürde nadir rastlanan kardiyak SFT'li bir hasta sunulmuştur. Bilinen maligniteye sahip 43 yaşında kadın hasta çarpıntı şikayeti ile acil servise başvurdu. EKG'sinde supraventriküler taşikardi izlenen hastada, sinüs ritmi sağlandıktan sonra uygulanan transtorasik ekokardiyografide sürpriz bir şekilde sol atriyum içerisinde dev bir kitle tespit edildi. Hasta, kitlenin semptomatik ciddi mitral darlığa neden olması nedeniyle opere edildi. Cerrahi örneğin patolojik çalışması sonucunda literatürde oldukça nadir rastlanılan SFT tanısı koyuldu. Literatürde SFT üzerine olan bilgiler vaka raporları ile sınırlıdır. Güncel literatür, kardiyak kitle yönetimi üzerine eksik kalmakta olup bu konu üzerinde ileri çalışmalara ihtiyaç duyulmaktadır.

Anahtar Kelimeler: Kardiyak kitle, kardiyo-onkoloji, soliter fibröz tümör

Cardiac masses are typically diagnosed incidentally during evaluations for non-cardiac clinical scenarios.^{1,2} The lack of definitive recommendations complicates their treatment and management.

Solitary fibrous tumors (SFTs) are rare neoplasms derived from fibroblastic mesenchymal cells.³ Initially described as pleural-based, extrapleural locations are now diagnosed more frequently than pleural ones.⁴ However, cardiac involvement is exceptionally rare, with few reports documented in the literature.³⁻⁶

This case report discusses the diagnosis and treatment of a patient diagnosed with malignant cardiac SFT after presenting with palpitations.

Case Report

A 43-year-old female patient, who was under palliative therapy for known metastatic anaplastic thyroid cancer and a malignant mesenchymal tumor of the right hip, presented to the emergency department with palpitations.

Upon physical examination, she exhibited a heart rate of 180 bpm, a respiratory rate of 18/min, a body temperature of 36.6°C, and an oxygen saturation of 90%. Bilateral

CASE REPORT OLGU SUNUMU

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Figure 1. Transthoracic echocardiography parasternal long axis view displays a large mass almost completely filling the left atrium and obstructing the mitral valve.

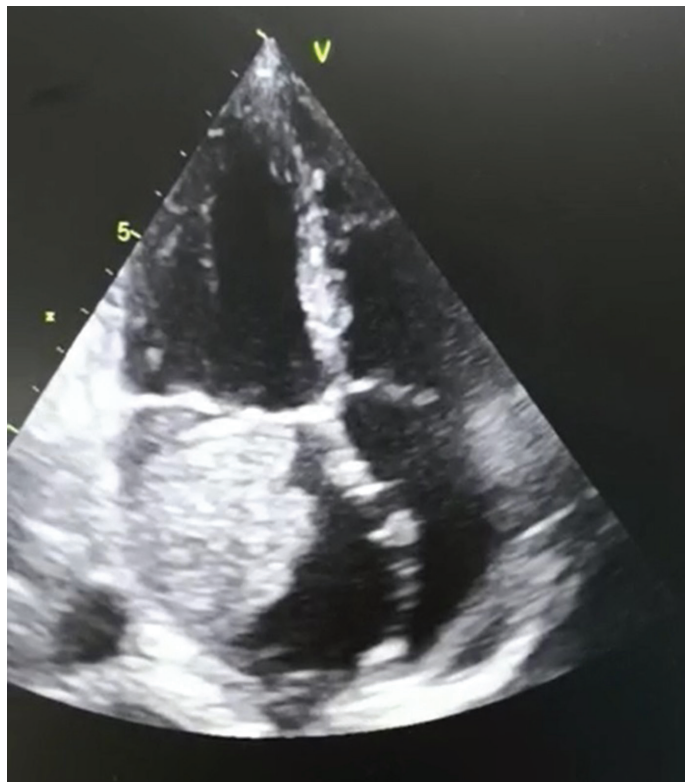


Figure 2. Apical 4-chamber view of the mass on transthoracic echocardiography.

pretibial edema was noted, more prominent on the right lower extremity. Auscultation of the lungs and heart revealed bilateral basilar crackles and a 3/6 early diastolic murmur at the apex.

Electrocardiography indicated regular tachycardia at 180 bpm with a narrow QRS complex. After the administration of a 12 mg followed by a 6 mg dose of adenosine, sinus rhythm was restored. Laboratory tests showed hemoglobin of 7.2 g/dL, a D-dimer level of 19,050 µg/L FE, with electrolytes, thyroid-stimulating hormone (TSH), and troponin levels within normal ranges. The patient's anemia was compatible with anemia of chronic disease. Transthoracic echocardiography (TTE) showed a left ventricular ejection fraction (LVEF) of 60% and enlarged left atrial dimensions (60 x 48 mm on the parasternal long axis). A significant finding was a giant mass in the left atrium, measuring 53 x 41 mm (Figures 1, 2), which almost completely obliterated the left atrium and caused severe mitral stenosis (Figure 3). This mass was also visible on contrast-enhanced computed tomography of the thorax (Figure 4A), where it narrowed to the left main pulmonary artery due to its mass effect (Figure 4B). Additionally, the lung parenchyma showed consolidations in the form of ground-glass areas compatible with

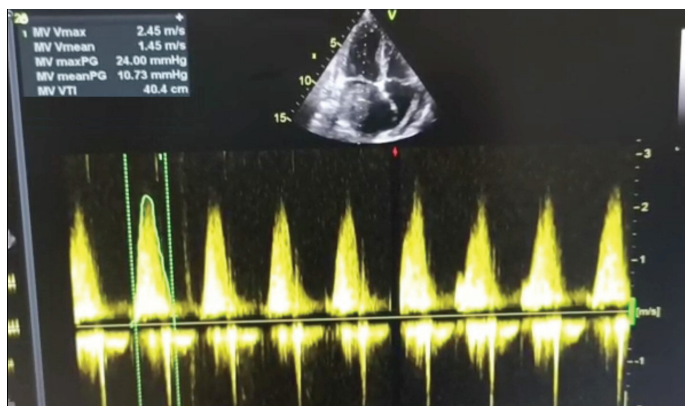


Figure 3. Doppler echocardiography shows severe mitral stenosis.

congestion and metastatic pulmonary nodules. No pulmonary thromboembolism was detected. Doppler ultrasonography of the right lower extremity, which was notably larger in diameter than the left, revealed deep vein thrombosis.

Due to the left atrial mass causing severe obstruction of the mitral valve, the patient was admitted to the cardiovascular surgery intensive care unit, and surgery was performed.

The operation began with a median sternotomy and left atriotomy. The mass was excised from the left atrium using blunt finger dissection (Figure 5). The mitral valve was examined for regurgitation; no regurgitation was detected, so no repair or replacement of the mitral valve was necessary. Left atriotomy

ABBREVIATIONS

CD34	Cluster of Differentiation 34
CMRI	Cardiac magnetic resonance imaging
IGF2	Insulin-like growth factor II
LVEF	Left ventricular ejection fraction
STAT6	Signal Transducer and Activator of Transcription 6
TSH	Thyroid-stimulating hormone
TTE	Transthoracic echocardiography

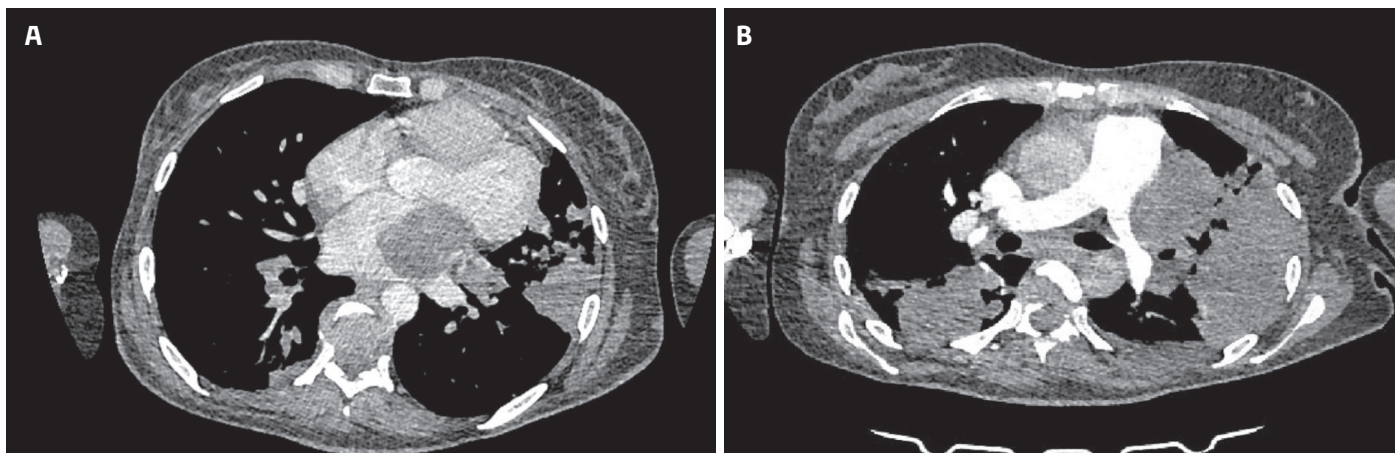


Figure 4. Thorax CT shows a large left atrial mass (A) and left main pulmonary artery was narrowed due to mass effect (B).



Figure 5. Macroscopic view of the surgically excised mass.

was sutured primarily. There were no significant hemorrhages or additional complications during the surgery. The patient was admitted to the intensive care unit for postoperative treatment and care.

Patient was extubated on the second postoperative day. TTE performed postoperatively showed no residual mass and was free of any surgery-related complications. Mitral valve function was normal, with no obstruction or regurgitation noted on echocardiography. Postoperative period was uneventful, and the patient was discharged on the seventh day. Discharge medications included metoprolol 50 mg orally (po) once a day, cefuroxime 500 mg po twice a day, furosemide 40 mg po every two days, and enoxaparin sodium 60 mg subcutaneously twice a day.

Histopathological examination of the surgical specimen revealed a tumor consisting of cells with round-oval nuclei, eosinophilic cytoplasm, nucleolar prominence, and local intranuclear inclusions. With Cluster of Differentiation 34 (CD34), B-cell

lymphoma 2 (Bcl-2), and Signal Transducer and Activator of Transcription 6 (STAT6) positivity, the case was diagnosed as "malignant solitary fibrous tumor." It was noted that the left atrial mass had similar morphological features to the mass removed from the right hip, which was diagnosed as undifferentiated sarcoma.

Discussion

Cardiac masses are rare in clinical practice and are mostly diagnosed incidentally.^{1,2} Due to their rarity, there is relatively less experience managing them compared to other cardiac diseases, and due to the malignant nature of these masses, they may cause mortality.

Primary cardiac tumors are extremely rare, while metastatic tumors are more common and frequently originate from the lung, breast, esophagus, and lymphomas.^{1,2} Metastatic cardiac tumors have a worse prognosis than primary cardiac tumors.¹ They can result from direct tumor invasion, hematogenous spread, or extension of the growing tumor into the right atrium via the inferior vena cava.² It is well established that metastatic cardiac tumors are more common than primary cardiac tumors.¹ Autopsy studies reveal that metastatic cardiac involvement is more prevalent in cancer patients than those diagnosed in clinical practice. In studies conducted on patients who died of cancer, cardiac involvement was reported in 8-12% of cases.² It has been found that metastases are predominantly located in the pericardium and often remain asymptomatic.

Although the clinical characteristic of cardiac masses are mostly asymptomatic, conditions such as pericardial tamponade, arrhythmias, embolic events, obstruction, heart failure, and shock can occur.^{1,2} Symptoms vary according to size, location, relationship with cardiac and extracardiac structures, embolic nature, and the cytokine production patterns of the masses.²

Echocardiography is useful in diagnosing cardiac tumors in most cases because it can be performed at the bedside, is cost-effective, and has a sensitivity and specificity of up to 90% in detecting tumors.⁷ It can also reveal the dynamic relationship of the mass with cardiac and paracardiac structures.⁷ However, it does not provide tissue characterization. Since pre-surgical

biopsy for intracardiac tumors is a risky procedure, cardiac magnetic resonance imaging (CMRI) is essential for determining the treatment approach by enabling tissue characterization.⁷

SFTs, rare neoplasms originating from fibroblastic mesenchymal cells, can occur in all anatomical regions such as the pleura, superficial and deep soft tissues, bone, abdominal cavity, and visceral organs. Data on cardiac involvement are primarily limited to case reports. The tumor may manifest signs depending on its location and size but is mostly asymptomatic. Rarely, it can cause Doege-Potter Syndrome, a paraneoplastic syndrome characterized by hypoglycemia due to secretion of insulin-like growth factor II (IGF2).⁸ In the limited case reports available in the literature, pericardial and epicardial tumors appear predominantly benign. Surgical resection is recommended as the best treatment method, although chemotherapy may be beneficial in managing unresectable, symptomatic cases.⁹

In the case presented here, our patient had known metastatic cancer but no prior cardiac disease or symptoms, and no abnormalities were detected on echocardiography before admission. However, during a hospital visit prompted by palpitations, a surprising mass was found in the left atrium causing severe obstruction. CMRI was unavailable at our hospital due to the radiologist's limited experience with this imaging technique. The mass was successfully removed through surgery, and the patient was discharged complication-free after postoperative follow-up. A follow-up for surgical care and echocardiographic evaluation was scheduled one week post-discharge. Unfortunately, when she failed to attend, her relatives informed us that she had died suddenly of respiratory distress seven days after discharge, and the exact cause of death remains unclear. It is speculated that pulmonary embolism from deep vein thrombosis, late postoperative complications, or her primary malignancy could have been contributing factors.

Conclusion

Cardiac masses are exceedingly rare in clinical practice, with most data on SFTs limited to individual case reports. The size, location, and behavior of these tumors can vary, potentially remaining asymptomatic or manifesting diverse cardiac symptoms.

It is crucial to consider that the detection of asymptomatic minimal pericardial effusion, which can progress insidiously, in patients with known cancer may indicate the onset of cardiac involvement. Consequently, closer echocardiographic follow-up should be performed, and other cardiac imaging modalities should be consulted if there is any doubt.

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Video: <https://youtu.be/22RicLFJOWY>

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