

A Rare Cause Of Acute Respiratory Failure And Hypoxia: Huge Left Atrial Sarcoma

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Abstract: A 29 year-old male, not known to have any medical illness. He was admitted with fever and shortness of breath for 4 days not improving with medical therapy. He was diagnosed with acute respiratory failure and was admitted to a local hospital, intubated for 10 days and treated as pneumonia. He was transferred to our hospital and after reviewing his condition, other diagnoses such as pulmonary embolism (PE) were considered. CT-scan for PE showed large left atrial (LA) mass. Transesophageal Echo (TEE) showed a large LA mass obstructing the mitral valve. He underwent emergency surgical excision of the mass via median sternotomy. His respiratory failure and hypoxia improved immediately post-operatively and he was extubated the next day. Large LA mass could mimic acute respiratory failure clinically and should be considered in the differential diagnosis.

Keyword: acute respiratory failure, cardiac sarcoma, left atrial mass, left atrial tumor, sarcoma.

INTRODUCTION

Primary cardiac sarcomas are extremely rare. About 25% of primary cardiac tumors are malignant, and of those 75% are sarcomas (1). Presenting symptoms of primary cardiac sarcomas depends on the location and the size of the tumor. Left heart sarcomas are usually broad based and large in size. This may result in obstruction of mitral valve diastolic inflow which may present clinically with severe pulmonary edema and acute respiratory failure (2,3). Complete surgical resection is the standard treatment. We present here a 29-year-old male who presented with acute respiratory failure and was intubated for 10 days with no improvement. Full workup showed left atrial mass obstructing the mitral valve diastolic inflow. The mass was excised surgically via median sternotomy and histopathology showed sarcoma. Echocardiography is often the initial diagnostic modality of choice to rule out cardiac causes of acute respiratory failure and should be considered early in the management of such rare condition.

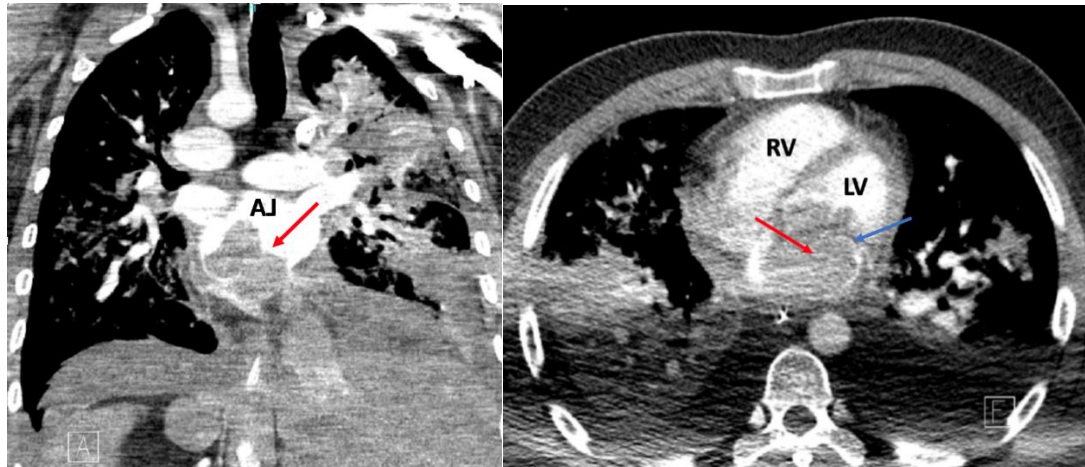
Case Presentation

A 29-year-old male presented from home to a local hospital with 4 days history of fever and shortness of breath with no abnormal past medical history. He was treated with oral antibiotics for suspected pneumonia, but his symptoms did not improve after 10 days of treatment. He required endotracheal intubation at the local hospital and transferred to our intensive care unit (ICU) with respiratory failure. On arrival to our ICU he was sedated, intubated and hemodynamically stable. His physical examination was notable for bibasal crepitations, and a grade 2/6 mid-diastolic heart murmur. His ventilator settings were: pressure control ventilation–volume guaranteed (PCV-VG), positive end-expiratory pressure (PEEP) of 12, tidal volume 7 ml/kg, and 70% fio₂.

His arterial blood gas (ABG) showed PH 7.45, PaO₂ 60 mmHg, PaCo₂ 40 mmHg, HCo₃ 24 mEq/L. His Chest-Xray (CXR) showed bilateral pulmonary infiltrates and his laboratory tests were unremarkable. Blood and

sputum cultures were negative. His electrocardiogram showed sinus rhythm of 90 beats/minute, without any significant abnormalities.

A Trial of weaning from ventilator was attempted and failed due to tachypnea and hypoxemia. Hence, other causes of respiratory failure were considered such as pulmonary embolism (PE). A CT-scan of the chest was done to rule out PE. It was negative for PE, however, it showed a mass in the left atrium (Fig-1).



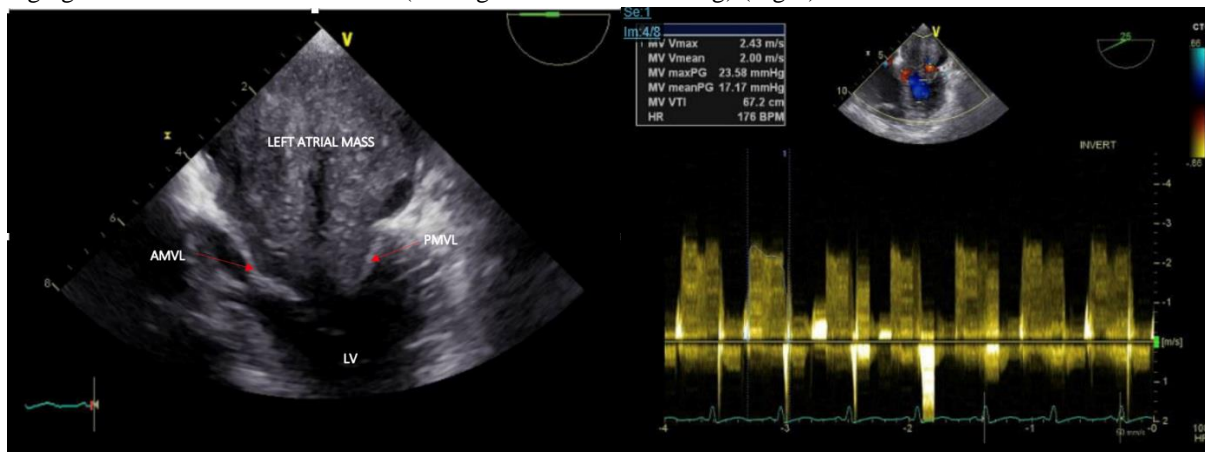
A. Coronal view: showing the large left atrial mass (red arrow).

B. Axial view: showing large mass in the left atrium (red arrow) prolapsing through the mitral valve (blue arrow).

Fig.1. Ct-chest showing left atrial mass

LV: left ventricle, RV: right ventricle, LA: left atrium, MV: mitral valve

The CT-PE did not show any coronary artery calcification requiring further testing such as coronary angiogram. The patient subsequently underwent Transesophageal echocardiogram (TEE) which showed huge LA mass prolapsing into the mitral valve and obstructing the left ventricle (LV) inflow. The mass was generating a high gradient across the mitral valve (mean gradient was 17 mmHg) (Fig-2).



A. 4-chamber view showing a large hyperechogenic mass within the left atrium obstructing the left ventricle inflow through the mitral valve

B. Continuous wave doppler showing a significantly high gradient across the mitral valve indicative of severe mitral stenosis secondary to the obstructive large left atrial mass.

Fig.2. Trans-esophageal Echo (TEE) showing

AMVL: Anterior mitral valve leaflet

PMVL: Posterior mitral valve leaflet
LV: left ventricle

This was the cause of such severe respiratory failure and hypoxia secondary to pulmonary edema and heart failure related to high left atrial pressure causing severe mitral stenosis. The patient underwent emergency median sternotomy and excision of large left atrial tumor (Fig-3).

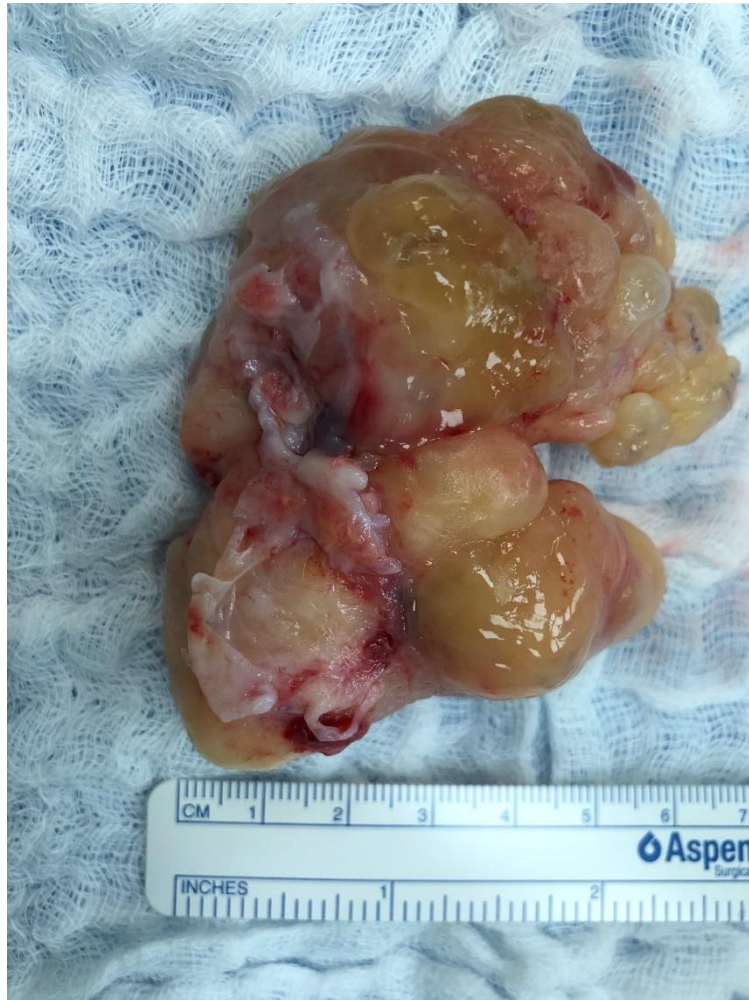


Fig.3. surgically removed large left atrial mass.

The mass was attached to the posterior wall of the left atrium. The mass was fully excised with good clear surgical margins. The mitral valve was not involved with the lesion and postresection TEE showed no significant mitral valve abnormality, a good left ventricular function, and no evidence of any remaining tumor.

Patient was extubated on post-operative day one and he stayed in the hospital for 30 days. He was transferred under oncology service for further workup and to rule out metastasis. His workup showed no metastasis and he was not given any adjuvant chemotherapy or radiotherapy.

Histopathology report confirmed the diagnosis of sarcoma. He was discharged home from oncology service after full recovery. Pre-discharge transthoracic echocardiogram (TTE) showed no recurrence of tumor in LA. The patient was seen 3 months in the clinic, and he was in good condition and recovering well. A follow-up TTE 3 months after surgery showed no recurrence and LA free of any masses. After treatment, patient returned to his country of origin and lost follow up.

DISCUSSION

Primary cardiac sarcomas are exceedingly rare with a reported prevalence of up to 0.0017% in an autopsy study. Majority of primary cardiac tumor are benign tumor. The malignant cardiac tumors are 25%, mostly sarcomas. Metastatic cardiac tumors are far more common (1,2).

Cardiac tumors may present with one or combination of the following: embolization, hemodynamic compromise interfering with cardiac function or cancer-like symptoms such as fever or fatigue or weight loss. Other potential presenting signs may be arrhythmias, syncope, and presyncope. Some cardiac tumors are found incidentally (3).

The delay in diagnosis of such rare condition is possible and high index of suspicion is required. Failure of improvement after full treatment of common causes of respiratory distress such as pneumonia, should alert clinician for rare causes such as intracardiac tumor (3,4,5).

TTE is simple, noninvasive, widely available as the first modality to consider in diagnosing cardiac tumors. It can identify the structural abnormalities within the heart chambers as well as the functional abnormalities including obstruction related to the tumor. TEE has more sensitivity and specificity compared with TTE in diagnosing cardiac tumors especially within the atria (6). However, echocardiogram sensitivity and specificity are limited in visualizing the right heart. Some features of the intracardiac masses on echocardiography may suggest malignancy. These include: broad-based mass, invasion of surrounding tissue, mass presence in more than a single chamber, poor definition of mass border, large size >5 cm, and presence of either pericardial or pleural effusion (6).

Cardiac MRI showed clear superiority in diagnosing cardiac masses when compared with echocardiography (7). Moreover, cardiac MRI can determine the tissue characterization of the tumor and assess for mediastinal and pericardial invasion (7). Cardiac CT is an excellent modality as well in diagnosing cardiac tumor. However, cardiac MRI has better resolution and tissue characterization when compared with cardiac CT (8).

Cardiac tumors are almost always managed by surgical excision (9,10). Because of the risk of embolization or hemodynamic compromise, left side lesions, should undergo resection. In the case of malignant cardiac tumors, surgical excision in combination with systemic chemotherapy remains the best available treatment (9,10,11).

CONCLUSION

Large left atrial mass could present with acute respiratory distress syndrome like picture. Delay in diagnosis is possible as these patients may present with respiratory failure symptoms and signs. Thus, differential diagnosis of such rare condition should be considered and a baseline TTE is highly recommended as a general assessment of the heart chambers and function.

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