Isolated Cardiac Sarcoidosis Presenting with Acute Ruptured Chordae

Jonathan Powell*, Emanuel Ebin

Internal Medicine Resident Physician, Florida Atlantic University
*Corresponding author: powellj@health.fau.edu

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Abstract Isolated cardiac sarcoidosis is a rare but life threatening sub classification of sarcoidosis. The symptomology can range from incidental asymptomatic disease to a life threatening disorder causing sudden cardiac death [3]. Conduction disorders are a well documented manifestation of cardiac sarcoid, however a lesser studied yet life threatening problem includes granulomatous involvement of the myocardium leading to heart failure and cord rupture. Early diagnosis and initiation of appropriate treatment are paramount in preventing destruction of myocardial tissue and improving the survival rate of this unique patient population. We present a case of an otherwise healthy young adult male that presented with acute decompensated heart failure with complete bi-valve failure requiring urgent cardiac surgery.

Keywords: cardiac sarcoidosis acute ruptured chordae acute decompensated congestive heart failure


1. Case Presentation

Mr. TM is a 31-year-old male without significant past medical history who does not take any medications or supplements that presented to the emergency department with a four month history of progressively worsening dyspnea. Symptoms began with inability to exercise secondary to easy fatigability dating back more than a year with gradual progression of dyspnea with lesser exertion. Upon presentation to our ED the patient had been unable to perform daily activities without dyspnea and fatigue. Associated symptoms included a persistent dry cough, orthopnea, paroxysmal nocturnal dyspnea, palpitations, extremity edema and weight gain. He denied fever, nausea and chest pain.

Mr. TM lives at home with his fiancé and their six-month-old son. He has an 8-pack year history of smoking and quit in his late 20s. He drinks two to three beers a week and denied illicit drug use. His family history is significant for a grandfather who died of sudden cardiac death in his 40’s.

Examination of the patient revealed a well-nourished young male, lying in bed, in moderate distress. Vitals as follows: Blood pressure 125/94, pulse 106, respiratory rate 23, and O2 saturation 99% on 2 L nasal cannula. Cardiac exam was significant for tachycardia, a normal S1 and S2, with an irregular rhythm. A 5/6 high pitched holosystolic murmur best auscultated at the apex with axillary radiation was appreciated. There was no JVD. No cervical lymphadenopathy appreciated. No carotid bruits appreciated bilaterally. A brisk carotid upstroke was noted. His extremities were not edematous. Strong and equal pedal pulses were appreciated. No cyanosis nor clubbing of the distal extremities noted.

Admission basic laboratory including cardiac enzymes were within normal limits as was his urine drug screen.

His ECG is significant for left atrial enlargement and tachycardia. Right axis deviation. Prolonged QTc interval and non-specific ST-T wave abnormalities.
2-D echocardiogram demonstrated: The left ventricle is moderately to severely dilated. The left ventricle is normal in wall thickness. Visually estimated ejection fraction is 60% (overestimated secondary to severe mitral regurgitation). There was no aortic insufficiency. A flail posterior mitral valve leaflet with an incompetent chordal structure was seen. Severe anteriorly directed eccentric mitral regurgitation is present back to the pulmonary veins. Mildly thickened tricuspid valve leaflets with normal excursion and mild tricuspid regurgitation was present. Right ventricular systolic pressure is estimated to be 53 mmHg. The aortic root is normal in size. The IVC is mildly dilated. A thin interatrial septum is present without evidence of shunting.

CT chest with contrast imaging (above): cardiomegaly with enlarged hilar lymph nodes bilaterally in the hilum of the lungs. Chest X-Ray (below) emphasizing cardiomegaly

Cardiac cath revealed: normal coronary arteries and anatomy. Severe mitral regurgitation with a large V wave, and moderate pulmonary hypertension which is reversible to IV cardene infusion

The patient then proceeded to open bileaflet mitral valve repair, tricuspid valve repair and left atrial appendage ligation. The operative course was uneventful, intraoperative TEE demonstrated an estimated LVEF of approximately 20% with functioning valves without evidence of leaks. Intraoperatively, two mediastinal lymph nodes were excised and sent for pathologic examination as was the posterior leaflet of the mitral valve. The results are as follows:

Part 1. Anterior mediastinal lymph node, excisional biopsy: 
Noncaseating granulomatous inflammation
Special stains for acid fast bacilli and fungal organisms negative.

Part 2. Mediastinal lymph node, excisional biopsy:
Noncaseating granulomatous inflammation
Special stains for acid fast bacilli and fungal organisms negative

Part 3. Mitral valve leaflet (P2 segment of P3), excision: 
Degenerative changes of cardiac valve.
No vegetations identified; noncaseating granulomatous inflammation identified

Mr. TM’s post-operative course was complicated by low EF, approximately 20%. 2-D complete ECHO performed 3 days postoperatively demonstrated: a dilated left ventricle with severely reduced left ventricular contractility. There was severe global hypokinesis with regional variation. The right ventricle was also enlarged however contractility appeared preserved. His valves were functioning appropriately without evidence of leakage.

The patient was given a Life Vest for his low EF and high risk of sudden cardiac death. By the date of discharge the patient was walking without assistance and tolerating a regular diet. His incisions were clean, dry, intact and healing well. The patient was discharged home on post operative day 7.

The patient has followed up regularly with his PCP, cardiologist and CT surgeon as an outpatient. He has reported significant improvement in his dyspnea and fatigability. His exercise tolerance has been increasing, he has denied any palpitations, chest pain, chest pressure, syncope or pre-syncope. He also states that his PND and orthopnea are resolving. He does not have any complaints of lower extremity edema. He is tolerating cardiac rehab well.

2. Discussion

The clinical presentation of cardiac sarcoidosis can range from an asymptomatic course to sudden cardiac death [3]. Cardiac sarcoidosis has three histologic phases: edema, granulomatous infiltration, and fibrosis with eventual scarring. The typical sites of involvement are the LV free wall, basal septum, right ventricle, and the atrial wall presumably because of the relatively increased myocardial mass present in these areas [1]. Currently there are no ideal non-invasive means of diagnosing cardiac sarcoidosis. The gold standard is still endomyocardial tissue biopsy demonstrating the presence of non-caseating granulomatous disease however even that has a sensitivity of 20-30% as the myocardium is not uniformly affected [10]. Cardiac MRI has 100% sensitivity and 78% specificity for cardiac sarcoidosis [5]. Sarcoidotic lesions are detected by late gadolinium enhancement and are localized predominantly subepicardially [1,4,5,6,10]. Differentiating between active inflammatory lesions vs scar tissue is a difficult task to perform as both the scarred and actively inflamed tissue demonstrate a slowed rate of
gadolinium uptake. It is imperative to control active inflammation to prevent further myocardial degradation [11]. Utilization of 18-fluorodeoxyglucose (FDG PET) allows for the differentiation of hypermetabolic (active inflammatory) tissue from fibrosed scarred myocardium. FDG PET scan has an 85% sensitivity and 95% specificity for cardiac sarcoidosis [7,8,9]. A simultaneous use of PET/MRI allows the detection of distribution of the inflammation, fibrotic changes, and the overall disease activity in cardiac sarcoidosis [9]. The mainstay of treatment for cardiac sarcoidosis is corticosteroids [19]. The appropriate dose is not yet determined however current guidelines recommend initial therapy with 1mg/kg/day tapering down to 10 mg/day [19]. The goal of therapy is to minimize active inflammation to prevent further cardiac damage. Early initiation of steroid treatment has been shown to decrease recurrence of ventricular tachycardias and in some cases patients have had resolution of AV nodal blocking as the inflammation was quelled before scarring took place [11,13].

The overall prognosis for patients with cardiac sarcoidosis is not well established. Several reports demonstrate a five-year survival rate exceeding 70% [1,3]. According to Yazaki et al., patients treated with corticosteroids with a left ventricular ejection fraction >50% had a ten year survival rate of 89% compared with 27% for the same patients with LVEF <50%. Independent predictors of mortality were NYHA functional class, left ventricular end-diastolic diameter, and the detection of sustained ventricular tachycardia [3]. The leading cause of death in these patients is sudden cardiac death caused by ventricular tachycardia, conduction block or congestive heart failure. Additional therapeutic modalities including permanent pacemaker implantation for conduction disturbance or cardioverter-defibrillator implantation in case of ventricular arrhythmias, are indicated given the high propensity of this population to suffer from sudden cardiac death. Patients with refractory heart failure unresponsive to medical therapy should be considered for heart transplantation [15]. According to Zaidi et al., patients undergoing orthotopic heart transplantation due to cardiac sarcoidosis had better short and intermediate term survival than the majority of heart transplant recipients [15]. Therefore, the diagnosis of sarcoidosis should not disqualify potential candidates for cardiac transplantation.

3. Conclusion

Isolated cardiac sarcoidosis is a rare but life threatening disease. Unfortunately the diagnosis typically is not made until significant myocardial destruction has already occurred. Early diagnosis and initiation of appropriate treatment are paramount in preventing further degradation of myocardial tissue and improving the survival rate of this unique patient population.

References