

Cardiac tamponade as a manifestation of extrapulmonary tuberculosis in β thalassemia major patient

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Abstract. Cardiac tamponade is a medical emergency condition. Rapid diagnosis and determination of the etiology with epidemiologic consideration may lead to earlier treatment and improved survival. Occasionally, the etiology may be clearly related to a recognized underlying disease, but the possibility of unrelated etiologies should be considered. Pericarditis tuberculosis, a rare manifestation of extrapulmonary tuberculosis in a non-HIV patient, has to be deliberate as one of the etiology, especially in the endemic area. Here, we report a case of 28 years old male with β thalassemia major presented with excessive exertion breathlessness progressing to orthopnea. Sign of cardiac tamponade was identified from echocardiography which showed large pericardial effusion with swinging heart and right atrial systolic collapse. Pericardiocentesis was performed immediately, drained 870 ml of hemorrhagic fluid from inserted pigtail. The patient was treated with the anti-tuberculosis regimen and oral corticosteroid after real-time polymerase chain reaction of *Mycobacterium tuberculosis* positivity in pericardial fluid. MRI T2 confirmed no haemosiderosis in patient's heart. After treatment, the patient responded well and showed clinical improvement.

1. Introduction

The term Extra Pulmonary Tuberculosis (EPTB) has been used to describe an isolated occurrence of tuberculosis (TB) at body sites other than the lung.[1] In immunocompetent adults, EPTB constituted about 15 to 20% of all TB cases, where cardiovascular involvement is a relatively uncommon manifestation. A recently published literature suggests that, in the developed country, TB is a relatively rare cause of pericardial diseases in HIV negative, immunocompetent persons and accounts for only 2% of cardiac tamponade.[2] Here we report a case of β thalassemia major patient with large hemorrhagic pericardial effusion presenting cardiac tamponade as EPTB.

2. Case report

A 28 years old male patient with β -thalassemia major who got a regular blood transfusion, admitted to an emergency room of Cipto Mangunkusumo National General Hospital with a chief complaint of breathlessness, which was insidious onset, progressive, even at rest, more in laying down. He reported chest pain, weight loss, night sweats, but no cough. He also complains of swelling in both legs since a



week before admitted to hospital. He had no history of a chronic cough, pulmonary TB and intense contact with TB patient.

At the time of admission, the general condition of the patient was poor; he was restless, tachypnoea, hypotension with pulses paradoxes and tachycardia. On systemic examination, inferior palpebral conjunctiva was anemic, there was a dull note on the right and left side thorax with reduced air entry suggestive of bilateral pleural effusion along with raised jugular venous pressure, muffled heart sound secondary to large pericardial effusion, tender hepatosplenomegaly, and pedal edema. Laboratory findings showed anemia with hemoglobin level 8.48 g/dl. Viral serology for hepatitis and human immunodeficiency virus (HIV) were negative. Chest x-ray showed marked cardiomegaly with bilateral pleural effusion (Figure 1). ECG features were sinus tachycardia with electrical alternans pattern (Figure 2). Urgent 2D-ECHO was done immediately, showed swinging heart, the diastolic collapse of the right ventricle, the systolic collapse of atrium along with large pericardial fluid (4 cm of thickness) which was suggestive of cardiac tamponade. Consent was obtained immediately, and pericardiocentesis with echocardiography guiding was done with pigtail catheter in situ, 870 ml of hemorrhagic fluid was drained. The patient responded well to the pericardial fluid drainage with subsidence of the dyspnea and can lay down after that.

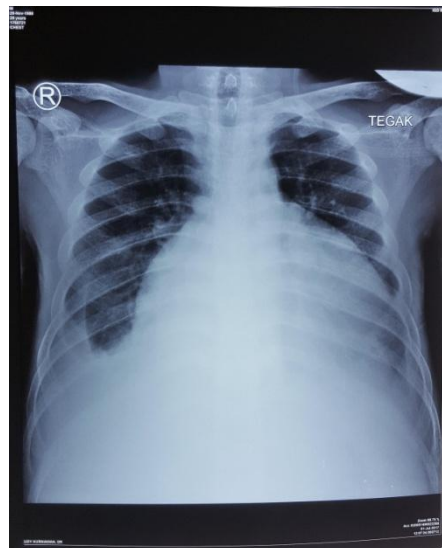


Figure 1. Chest X-Ray on admission showing marker cardiomegaly with bilateral pleural effusion.

The pericardial fluid was sent for analysis, cytology, microbiology investigation, real-time Polymerase Chain Reaction (PCR) for TB and Adenosine Deaminase (ADA) value. Analysis of pericardial fluid confirmed hemorrhagic which contained 2000/ μ L of segment leucocyte, 1300/ μ L of lymphocytes, 2034U/L of lactate dehydrogenase (LDH) and 5.1 g/dl of total protein. There was no malignant cell found from cytology of pericardial fluid. Microbiology yielded a negative result. Gram staining, acid-fast bacilli, and low level of ADA (15.7 IU/L) were not supporting for tubercular origin. However, pericardial fluid was positive for real-time PCR TB. Contrast-enhanced computed tomography thorax was performed after pericardiocentesis which shows left pleural effusion with compressive atelectasis inferior left lobe of the lung, cardiomegaly with pericardial effusion and catheter in the pericardial cavity. No nodule or infiltrate seen in lungs, no lymphadenopathy seen in trachea, mediastinum and hilar.

The pleural fluid drainage was performed, resulted in 700 ml of hemorrhagic fluid. Analysis of pleural fluid was done and showed exudative. Acid-fast bacilli stain and culture of microorganism from pleural fluid yielded negative findings. There was no malignant cell found in the pericardial and pleural fluid.

Since the patient was having β thalassemia major, Magnetic Resonance Imaging (MRI) T2* was performed to see iron cardiomyopathy in patient's heart. Normal cardiac iron loading, 20.95 ms was found. While liver iron loading was 1.96 ms and pancreas iron loading was 8.32 ms which indicating moderate haemosiderosis in patient's liver and pancreas.

Pigtail catheter remains placed for 14 days with total drainage of 1191 ml of hemorrhagic pericardial fluid. Patients were treated with first-line oral anti TB regimen consist of rifampicin, isoniazid, pyrazinamide, and ethambutol plus oral corticosteroid. He also got a blood transfusion with deferiprone and deferoxamine as iron chelating agents. After days, the patient showed clinically improvement and follow-up echocardiographic findings showed normal cardiac function without residual pericardial effusion when he discharged.

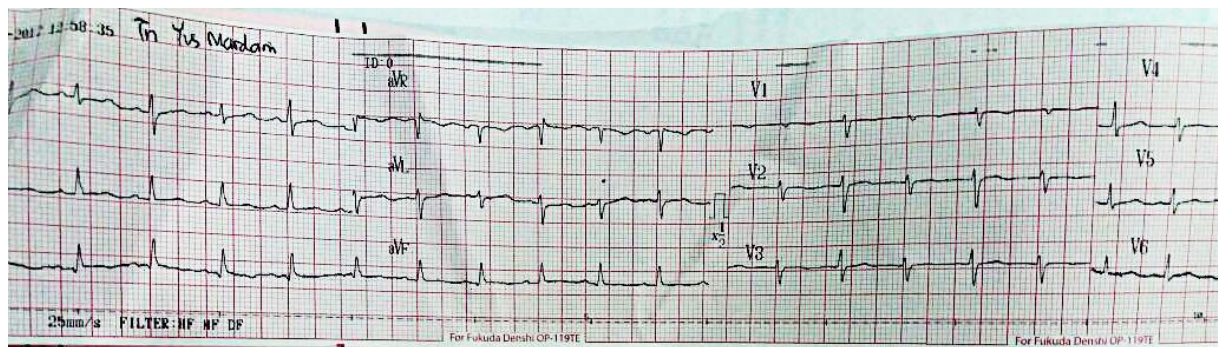


Figure 2. Electrical alternans patterns on ECG.

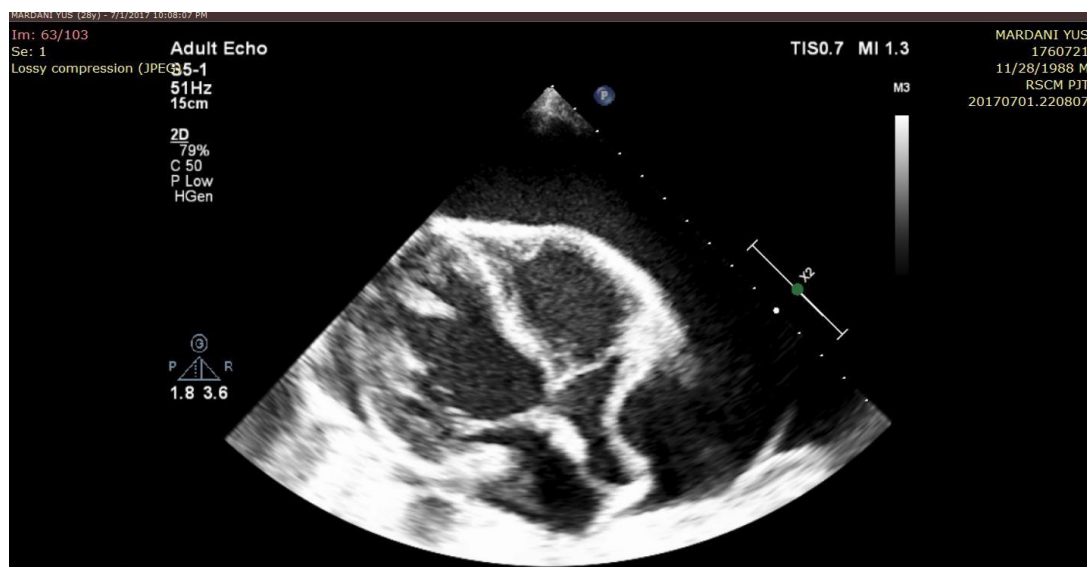


Figure 3. Transthoracic 2-dimensional echocardiography showed swinging heart with large pericardial effusion and right atrial systolic collapse.

3. Discussion

Pericardial involvement in TB may result in acute pericarditis, chronic pericardial effusion, cardiac tamponade or pericardial constriction and has a mortality rate of 17-40 % at six months after diagnosis.[1] The clinical manifestations of TB pericarditis as EPTB are wide-ranging and varied. While a cough, chest pain and dyspnea are common, non-specific constitutional symptoms, encompassing fatigue, fevers, night sweats and weight loss, may also emerge. Patients may present subacutely with the development of constrictive pericarditis, or as described in this case acutely with

pericardial fluid accumulation leading to cardiac tamponade.[2] Indonesia, together with India, China, Nigeria, Pakistan and South Africa, is one of the six leading countries accounted for 60% of the new cases of TB.[3] It is reasonable when a patient presents with pericardial effusion in Indonesia, tuberculosis should be ruled out first as the etiology.

Definitive diagnosis of tuberculous etiology is challenging. From the analysis of pericardial effusion, TB pericardial effusions are typically exudative and characterized by high protein content and increased leukocyte count, with a predominance of lymphocytes and monocytes. Approximately 80% of cases of TB pericarditis are bloodstained effusions. In endemic areas, TB has known as the frequent causes of hemorrhagic pericardial effusion. Acid-fast bacilli are infrequently observed (6% sensitivity) and the yield is increased by culture (25 -75%). The determination of interferon gamma (sensitivity of 92% and specificity of 100%) is better than the elevation of ADA levels (sensitivity of 87% and specificity of 89%). Although the specificity of polymerase chain reaction is high, its sensitivity (32%) is low for the diagnosis according to published studies. If necessary, a pericardial biopsy should be carried out for culture and pathology study.[4, 5, 6]

Cardiac tamponade, slow or rapid compression of the heart due to the pericardial accumulation of fluid, pus, blood, clots or gas as a result of inflammation, trauma, rupture of the heart or aortic dissection, is a life-threatening condition resulting in a reduced ventricular filling and subsequent hemodynamic compromise. The magnitude of clinical and hemodynamic abnormalities in cardiac tamponade depends on the rate of accumulation and amount of pericardial contents, the distensibility of the pericardium and the filling pressures and compliance of the cardiac chambers. This condition is a medical emergency, the complications of which include pulmonary edema, shock, and death.[7,8] Because of its emergency nature, prompt diagnosis and rapid determination of the etiology must be done immediately and will guide to better management for life-saving.

Echocardiography is the single most useful diagnostic tool to identify pericardial effusion, estimate its size, location and degree of hemodynamic impact, and potential complications such as constrictive pericarditis. Also, echocardiography is also used to guide pericardiocentesis with excellent safety and efficacy. Signs of tamponade can be identified by echocardiography are swinging of the heart, early diastolic collapse of the right ventricle, late diastolic collapse of the right atrium, abnormal ventricular septal motion, inspiratory decrease and expiratory increase in pulmonary vein diastolic forward flow, exaggerated respiratory variability (>25%) in mitral inflow velocity, respiratory variation in ventricular chamber size, aortic outflow velocity (echocardiographic pulses paradoxus) and inferior vena cava plethora.[7]

However, this patient also has β thalassemia major. Cardiac involvement represents a notable complication of β thalassemia major (TM) and results in increased mortality and morbidity rates. Historical series show a broad range of cardiac complications including pericarditis, myocarditis, heart failure, and arrhythmia. In the era of deferoxamine iron chelation, the clinical manifestation cardiac disease in TM has changed, pericarditis and myocarditis are now rare. Heart failure is the most common cause of death in TM and primarily results from cardiac iron accumulation, accounting for 70% of cases, which also can present as heart failure. Myocardial iron deposition can be quantified reproducibly with myocardial T2*, a relaxation parameter that arises principally from local magnetic field inhomogeneities that increase with iron deposition. In TM, chronic anemia also leads to background symptomatology such as dyspnea, which can mask the clinical diagnosis of cardiac dysfunction.[9]

In this case, from clinical and PCR TB, cardiac tamponade with large hemorrhagic pericardial effusion was confirmed due to TB. The treatment of cardiac tamponade mainly involves drainage of the pericardial fluid, preferably by needle pericardiocentesis, with the use of echocardiographic or fluoroscopic guidance, and should be done without delay in unstable patients. Alternatively, drainage can be performed by a surgical approach. Treatment of EPTB itself does not differ from pulmonary TB treatment regimens. Treatment schemes include a two month period on rifampicin, isoniazid, pyrazinamide, and ethambutol followed by a four-month period on rifampicin and isoniazid.[1,7]

4. Conclusion

Cardiac tamponade is a life-threatening condition thus need to be diagnosed and managed immediately. As a developing country, Indonesia is an endemic area for TB. Although cardiac manifestation of EPTB is rare, TB has to be included in top consideration as an etiology of pericardial effusion with cardiac tamponade in Indonesia.

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