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Cardiomyopathy with Acute Rheumatic Fever in Children: A Case Report

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ABSTRACT

Cardiomyopathy is defined as a disease of the myocardium that can cause cardiac dysfunction. The complication of this condition usually can manifest in heart failure, arrhythmia, and even sudden death. This study will describe a rare case of cardiomyopathy with acute rheumatic fever in children. In this study, a girl aged 11 years and 2 months with no history of heart disease was admitted to the hospital with congestive heart failure due to cardiomyopathy and acute rheumatic fever. Acute rheumatic fever is diagnosed based on the presence of carditis, high erythrocyte sedimentation rate, and positive antistreptolysin titer-O (ASTO). Cardiomyopathy is diagnosed by echocardiography. The prognosis of this patient is poor due to poor left ventricular function. In conclusion, patients with clinical features of ARF/RHD should be promptly treated and referred for definite diagnosis and long-term management to limit the extent of heart damage.

1. Introduction

Cardiomyopathy is defined as a myocardium disease that can cause cardiac dysfunction. The complication of this condition usually can manifest in heart failure, arrhythmia, and even sudden death.[1] Based on anatomic and functional features, cardiomyopathies are classified by World Health Organization (WHO) into three types; they are hypertrophic, dilated, and restrictive cardiomyopathies. Cardiomyopathies in children are a heterogeneous group of rare disorders characterized by mechanical and electrical abnormalities of the heart muscle. The overall annual incidence of childhood cardiomyopathies is estimated at about 1 per 100,000 children and is significantly higher during the first 2 years of life. Dilated cardiomyopathies account for approximately half of the cases. Hypertrophic cardiomyopathies form the second largest group, followed by the less common left ventricular non-compaction and restrictive phenotypes. Infectious, metabolic, genetic, and syndromic conditions account for the majority of cases.[2-4]

Rheumatic heart disease is an acquired heart disease that is common in children.[5] Rheumatic heart disease cause heart valve abnormality that persists due to a history of acute rheumatic fever previously. This disease mainly affects the mitral valve (75%) and aortic (25%), rarely involves the tricuspid valve, and

never invades the pulmonary valve. The highest incidence of rheumatic heart disease in children is between 5-15 years of age. Rheumatic heart disease (RHD) is caused by the *Streptococcus*-β-*hemolyticus* type A bacteria, which can cause acute rheumatic fever (ARF) and may lead to complications such as congestive heart failure. Approximately 39% of patients with acute rheumatic fever can develop abnormalities in the heart ranging from valvular insufficiency, heart failure, pericarditis, and even death. With chronic rheumatic heart disease, patients may develop valvular stenosis, varying degrees of regurgitation, atrial dilatation, arrhythmias, and ventricular dysfunction.[6] This study will describe a rare case of cardiomyopathy with acute rheumatic fever in children.

2. Case Presentation

A girl, aged 11 years and 2 months-old, was hospitalized for the first time in the pediatric ward of Balimed Karangasem Hospital with the chief complaint of shortness of breath 3 days before admission to the hospital. The child also complained of palpitation 3 days before admission, exercise intolerance for the past 1 week before the symptoms worsened, breathlessness during rest and sleep on two pillows, history of fever 2 weeks before admission, but the temperature was not high during examination in hospital. The fever was usually intermittent, and no shivering. Exercise intolerance since 1 week before admission to the hospital, the child said that she was difficulty breathing after running and also could not play with her classmates in school. The patient looks pale and easily fatigued, and palpitation and worsening shortness of breath since 3 days before admission. There was a history of sore throat. There was a history of joint pain, with no uncontrolled movement and no pink rings on the trunk and inner surfaces of the limbs. No history of cyanosis since birth and excessive sweat after feeding and exercise. Bowel movement was normal. The patient had never suffered from a disease like this before. There is no other family member who suffered from heart disease, lung disease, or another metabolic disease.

The patient was the second child in the family member, spontaneous vaginal delivery assisted by a midwife, a term, with a birth weight of 3500 grams and birth length of 50 cm and vigorous. She was discharged home on the second day of life without a medical problem. Primary immunization was completed according to the public health program. The history of growth and development as standard. Before being hospitalized, she just started to play and learn in junior high school. The patient and her family live in their relatives' home in a rural area, a permanent house with good hygiene and sanitation.

Based on physical examination, the general condition was severely ill. The patient slept in a sitting position and leaned on two pillows. Vital signs were blood pressure 100/50 mmHg, heart rate 120 times per minute, respiratory rate 30 times per minute, body temperature 36^{o}C, oxygen saturation 96%, and body weight 34 kg with no edema. Jugular venous pressure (JVP) 5+3 cm H_20. The chest was symmetric; there was an epigastric retraction, heart: apex was not inspected. The apex was palpable at one finger lateral of left midclavicular line, intercostal space V, heart border: above: intercostal space II, dextra: sternalis dextra line, sinistra: 1 finger lateral left midclavicular line, heart sound: regular rhythm. There was systolic murmur grade 4/6 at the mitral valve, tricuspid, and aortic valve radiating to the axilla, lung: vesicular, rales +/+, no wheezing.

Laboratory findings revealed hemoglobin 11.8 g/dL, leukocyte 6560/mm^3, hematocrit 39.9%, thrombocyte 299.000/mm^3, and erythrocyte sediment rate 45 mm in the first hour. MCV: 89 fl, MCH: 26.3 PQ, MCHC: 29,5%. Troponin I <0.01 ng/mL. Chest x-ray examination presented cardiomegaly with pulmonary congestion (Figure 1).

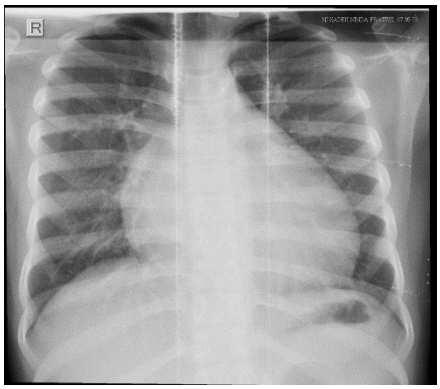

Figure 1. Chest X-ray imaging.

The diagnosis of this patient was congestive heart failure and suspected acute rheumatic fever. Treatments from the pediatric department were oxygenation 2 liters per minute (lpm), bed rest, and ceftriaxone 1 gr twice daily intravenous. Further examination revealed as follows; electrocardiogram result was sinus rhythm, heart rate 103 bpm, without prolonged interval PR wave, no arrhythmia, and left ventricular hypertrophy sign. Antistreptolysin O (ASTO) reveals positive. Impression was suitable with acute rheumatic fever and treated with benzathine penicillin 1.200.000 unit intramuscular and methylprednisolone 20 mg intravenous. Echocardiography results presented cardiomyopathy dilatation with moderate mitral regurgitation (MR), moderate tricuspid regurgitation (TR), mild pulmonary regurgitation (PR), mild aortic regurgitation (AR), global hypokinetic, poor left and right ventricle function (Figure 3).

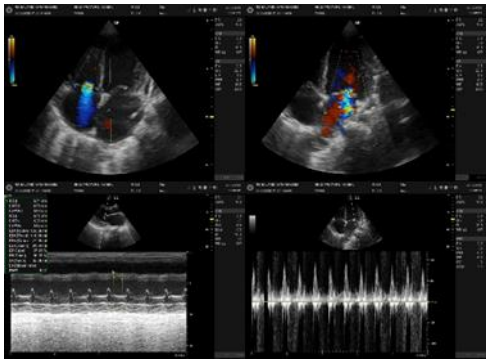

Figure 2. Echocardiography result.

Based on further examination, the patient was diagnosed with congestive heart failure et causa cardiomyopathy/functional class III with acute rheumatic fever. Additional management for this patient was NaCl 0.9% drip 8 guttae/minute, furosemide 40 mg OD IO, spironolactone 25 mg OD IO, digoxin 0,25 mg OD IO, captopril 6,25 mg IO, and bisoprolol 1,25 mg OD IO. Education for management of this case was strict bed rest, providing age-ill-appropriate activities, and early diet advice, if overweight and in failure. Monitoring for vital signs and blood pressure 4 times daily. If the pulse rate were more than 100 bpm, apical heart rate was found, and liquid and electrolyte balance were signs of digoxin intoxication. Electrolyte evaluation on the third day of inpatient revealed sodium 138,7 mmol/L, potassium

4,65 mmol/L, calcium 9,99 mg/dL, and chloride 98,39 mmol/L.

The patient was discharged with several education related to her disease. Parents were encouraged to limit the patient's activity for three months later, visit for heart control every 3 weeks, take long-standing prophylaxis benzathine penicillin, and do dental care and antibiotic prophylaxis to treat sore throat and skin sores. The most important of education is awareness of parents about the prognosis of dilated cardiomyopathy and the condition of heart contractility.

3. Discussion

Rheumatic heart disease is still the most common cause of mitral valve stenosis and valve replacement in adults in the United States.[6] The presence of valve malfunctions can lead to pump failure either by pressure overload (obstruction in the outflow of the pump chamber, such as aortic valve stenosis or pulmonary stenosis) or with volume overload, which indicates an increase in blood volume to the left ventricle so that the end product of valve malfunction due to rheumatic heart disease is congestive heart failure.[5,6] This study presents a case of 11 years and 2 months old girl with unremarkable cardiac history hospitalized for congestive heart failure due to cardiomyopathy with acute rheumatic fever. The diagnosis of congestive heart failure was made based on a history of shortness of breath 3 days before admission, exercise intolerance for the past 1 week before the symptoms worsened, and breathlessness during rest and sleep on two pillows.

Cardiomyopathy is an abnormality in cardiac muscle function that can cause ventricle dysfunction.[7,8] It can appear along a spectrum of no symptoms, subtle symptoms, or, in the more severe cases, congestive heart failure (CHF), which occurs when the heart is unable to pump blood well enough to meet the body tissue's needs for oxygen and nutrients. In this case, the cardiomyopathy symptoms appear as congestive heart failure. Congestive heart failure is the combination of both clinical forms of left and right heart failure.[7,8] In this case, the patient has the symptoms of congestive heart failure due to cardiomyopathy, which are dyspnea on effort characterized by exercise intolerance, orthopnea, which is the patient sleeping on two pillows, jugular venous congestion, and heart enlargement.

Cardiomyopathy, in this case, is characterized by a decrease in ventricular contractile function due to dilatation of the cardiac chambers. The echocardiography reveals cardiomyopathy dilatation with moderate MR, moderate TR, mild PR, mild AR, global hypokinetic, poor LV, and RV function. We suspected the etiology of cardiomyopathy in this patient might be caused by the infection of bacteria and make inflammation of the myocardium. Inflammatory cardiomyopathy (ICM) is defined as inflammation of the heart muscle associated with impaired function of the myocardium. To diagnose inflammation, results from an endomyocardial biopsy are still the reference standard. Another modality, like cardiac magnetic resonance, is also a reliable diagnostic technique for cardiomyopathies because it offers important information on cardiac morphology and function, disease mechanism, treatment guidance, and prognosis evaluation.[3,4]

The cause of heart failure, in this case, is probably also related to acute rheumatic fever. This diagnosis was made based on other related signs and symptoms that were present in the patient. Based on the clinical and supporting diagnostic examination, the patient had a history of fever 2 weeks before shortness of breath appeared, but on our examination, the temperature was not high, with intermittent fever with no shivering. There was a history of sore throat and joint pain. Antistreptolysin O reveals positive. Echocardiography reveals cardiomyopathy dilatation with moderate MR, moderate TR, mild PR, mild AR, global hypokinetic, poor LV, and RV function.

Acute rheumatic fever (ARF) is diagnosed based on the guidelines for the diagnosis of ARF, which fulfill one major, and two minor manifestations plus evidence of a preceding group A streptococcus (GAS) infection. This patient presented carditis diagnosed by echocardiography finding with moderate MR and mild AR. A positive antistreptolysin O reveals evidence of a preceding group A *Streptococcus* (GAS) infection. Based on the diagnosis according to the criteria of the World Health Organization (WHO) 2002-2003 for diagnosis of rheumatic fever and rheumatic heart disease (based on revised Jones criteria), this patient included a condition of first attack or a recurrent episode where found 1 major criteria and 2 minor criteria with the evidence previous infection of *Streptococcus β hemolyticus* group A.[9-11]

ARF is an autoimmune consequence of GAS pharyngitis mediated by molecular mimicry. ARF usually affects school-age children, with preceding infection of group A streptococcus (GAS) that manifests in multiple symptoms. Risk factors of streptococcal infection include poverty, malnutrition, overcrowding, poor housing, and shortage of healthcare resources, underlining the high prevalence in developing countries. All manifestations of RF resolve completely after, except for cardiac valvular lesion, which is the hallmark of RHD. Post-rheumatic valvulopathies remain the main cause of heart failure among children and young adults. The type of cardiomyopathy caused by ARF is inflammatory cardiomyopathy. Beta-haemolytic streptococci is a causative agent of rheumatic myocarditis involving valvular and perimyocardium and is a major cause of paediatric hospitalizations and cardiac deaths globally, with the disproportional incidence in developing countries. Autoimmune response to group A streptococcal antigens during pharyngitis leading to cross-reaction of antibodies and immunocompetent cells with antigens in the myocardium is believed to be the primary pathogenic mechanism in rheumatic myocarditis.[3,12,13]

The management of heart failure depends on the etiology, hemodynamics, clinical symptoms, and severity of heart failure. The treatment includes 5 components the form of handling in general, treating the underlying disease, preventing further damage to the heart, and controlling the degree of CHF. Treatment is generally based on symptoms in the form of restriction of fluid intake due to most of the fluid will be absorbed by the body and increasing the amount of fluid in the body, thereby making the heart works harder. Administering diuretics is necessary to remove existing fluids from the body until the blood flow back to the heart (preload) decreases. In this case, furosemide was used as a diuretic. Although furosemide has a role in reducing cardiac workload, furosemide as well has debilitating side effects on potassium levels in the blood. Therefore, the administration of potassium-sparing diuretics such as spironolactone is also provided in the treatment. Digoxin was given in this case for positive inotropic effect and with bisoprolol to achieve optimal heart rate for the patient.[11,13,14]

The prognosis is based on the heart's pumping ability to compensate and improvement of clinical symptoms after treatment. To see the anatomical abnormalities of the heart then, the patient is examined by echocardiography.15 Prognosis of this patient remains worse because of poor LV-RV function (EF 30,32%, TAPSE 1.25 cm), so the ad functionam prognosis quo is dubia ad malam. But, clinically, in this patient, there is improvement so that the prognosis quo ad vitam is dubia ad bonam. Based on its social function, patients with heart failure have limitations in strenuous activity, so the prognosis quo ad sanationam is dubia ad malam.

4. Conclusion

Patients with clinical features of ARF/RHD should be promptly treated and referred for definite diagnosis and long-term management to limit the extent of heart damage.

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