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Recurrent Acute Rheumatic Fever with Severe Rheumatic Mitral Stenosis in 11-years-old Patient: A Case Report

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Abstract

Background: Acute Rheumatic Fever (ARF) was an immune-mediated complication of Group A Streptococcus (GAS) infection that could progress to Rheumatic Heart Disease (RHD) through repeated or untreated episodes. While RHD typically developed over years, yet some children in endemic settings can develop severe multivalvular disease rapidly, likely due to unrecognized/subclinical ARF and inadequate secondary prophylaxis. This case adds to the literature by illustrating severe rheumatic mitral stenosis at a young age with clinical features suggestive of recurrent ARF despite no documented prior ARF, emphasizing rapid progression could occur in endemic settings.
Case report: An 11-year-old male presented with exertional dyspnea and intermittent joint pain without swelling or redness. There was no previously documented ARF episode. Serology showed positive anti-streptolysin O (ASO) supporting recent streptococcal exposure. Echocardiography demonstrated severe mitral stenosis, moderate mitral regurgitation, moderate aortic regurgitation, moderate aortic stenosis, and severe tricuspid regurgitation with high probability of pulmonary hypertension. Diagnosis of recurrent ARF with severe RHD was established using the modified Jones criteria, supported by echocardiographic evidence of multivalvular involvement. Initial management was adjusted for penicillin allergy and included azithromycin, corticosteroids, beta-blockers, diuretics, and nutritional rehabilitation, followed by erythromycin for secondary prophylaxis.
Conclusion: This case highlighted the possibility of rapid progression to severe RHD in children due to subclinical ARF. Early diagnosis, routine echocardiography, strict adherence to secondary prophylaxis, and patient education were vital to prevent long-term complications, including heart failure and surgical interventions.

Keywords: *acute rheumatic fever, rheumatic heart disease, mitral stenosis*

Introduction

Acute Rheumatic Fever (ARF) is a systemic inflammatory disease characterized by involvement of the heart, joints, nervous system, and skin. This condition results from an abnormal immune response to infection by Group A Streptococcus (GAS), primarily following tonsillopharyngitis. ARF can affect all age groups but most commonly occurs in children. If inadequately treated or recurrent, ARF may progress to Rheumatic Heart Disease (RHD). RHD remains one of the leading preventable causes of cardiovascular mortality in developing countries.[1,2]

Approximately 470,000 new cases of ARF are reported globally each year, with a higher disease burden in developing countries due to the high incidence of untreated GAS infections. The global incidence of ARF ranges from 8 to 51 cases per 100,000 population. Crowded living conditions and low socioeconomic status are directly

associated with increased incidence of ARF. The Global Burden of Disease estimates 33 million cases of RHD with 275,000 deaths annually and over 9 million disabilities.[3,4]

The exact prevalence of ARF in Indonesia remains unclear, but published studies estimate RHD prevalence in children approximately 0.3 to 0.8 per 1,000 school-aged children. The incidence of ARF in Indonesia continues to pose a significant public health challenge, particularly among populations with low socioeconomic status. During the period of 1973–1977, the Internal Medicine Department at Dr. M. Djamil Central General Hospital reported the highest prevalence of ARF/RHD among patients aged 10 to 40 years as an etiology of heart disease requiring hospitalization, with a mortality rate of 12.4%. In contrast, during the period 2009 to 2012, 54 patients diagnosed with RHD were admitted to the inpatient ward of the Internal Medicine Department at Dr. M. Djamil Hospital, with the highest prevalence observed in the 11 to 20 years age group and a mortality rate of 9.26%.[5,6]

One in five ARF patients experiences recurrence within 10 years. Recurrences and severe initial episodes increase the risk of RHD, with over half developing RHD within 10 years of the first ARF episode. Factors influencing recurrence include age at first attack, presence of RHD, intervals between attacks, family history, social and educational factors, local streptococcal infection risk, and treatment adherence. Chronic inflammation in RHD leads to valve leakage or stenosis. Progressive valve deformity leads to chronic adult manifestations and death. However, progression from ARF to RHD takes years. Meanwhile, in this case discusses recurrent ARF in an 11-year-old boy with severe valvular disease, highlighting the importance of secondary prevention and medical intervention to prevent serious complications.[7,8]

Methods

This report presents an 11-year-old boy presented to the emergency department at Dr. M. Djamil General Hospital with complaints of intermittent shortness of breath lasting for two months. The dyspnea was primarily exertional, with no history of orthopnea or paroxysmal nocturnal dyspnea disrupting sleep. The patient also experienced intermittent pain in both knees over the past month, without any swelling or redness. He had frequent episodes of fever recurring over the past year with afebrile intervals in between. No involuntary movements were reported. Skin rash was absent. The patient denied sore throat at presentation but had a history of 2-3 self-limited sore throat episodes per year. There was no prior history of similar complaints, nor joint pain or migratory arthritis. There was no history of cardiac disease. There was no report of cyanosis at birth, and the parents denied any feeding difficulties during infancy.

On physical examination, the patient appeared moderately ill with stable vital sign, blood pressure was 97/58 mmHg, pulse rate 71 beats per minute, respiratory rate 22 breaths per minute and temperature 36.2°C. Body weight was 21 kg and height 130 cm, indicating the patient was undernourished (weight-for-height 77%). The throat examination showed tonsils graded T2-T2 with no signs of redness, and the pharynx appeared normal without inflammation. Cardiac auscultation revealed a low-pitched mid-diastolic murmur grade 2/4 with punctum maximum at the apex, accompanied by a high-pitched systolic ejection murmur grade 3/6, with punctum maximum at the upper right sternal border with radiation to the neck, and an early diastolic murmur grade 2/4, high-pitched, punctum maximum at the left lower sternal border. No peripheral edema, erythema marginatum, or subcutaneous nodules were observed. Knee joints showed no swelling or redness.

Electrocardiogram (ECG) showed sinus rhythm with no prolonged PR interval. Laboratory results showed leucocyte 7.520/mm3, ASO test was positive, while the CRP test was negative. The patient underwent TTE (Figure 1) revealing situs solitus, all PV to LA, LA, LV dilated, AV-VA concordance, normal relations of great arteries, ASD (-), VSD (-), PDA (-), good LV contractility EF 58%, good RV contractility (TAPSE 2,4 cm), mitral stenosis (MS) severe, mitral regurgitation (MR) moderate due to RHD, aortic stenosis (AS) moderate, aortic regurgitation (AR) moderate due to RHD, tricuspid regurgitation (TR) severe with high probability of PH, pulmonic regurgitation (PR) mild to moderate, and left arch. Based on the modified Jones criteria and supported by echocardiographic findings, the patient was diagnosed with recurrent acute rheumatic fever and severe RHD.

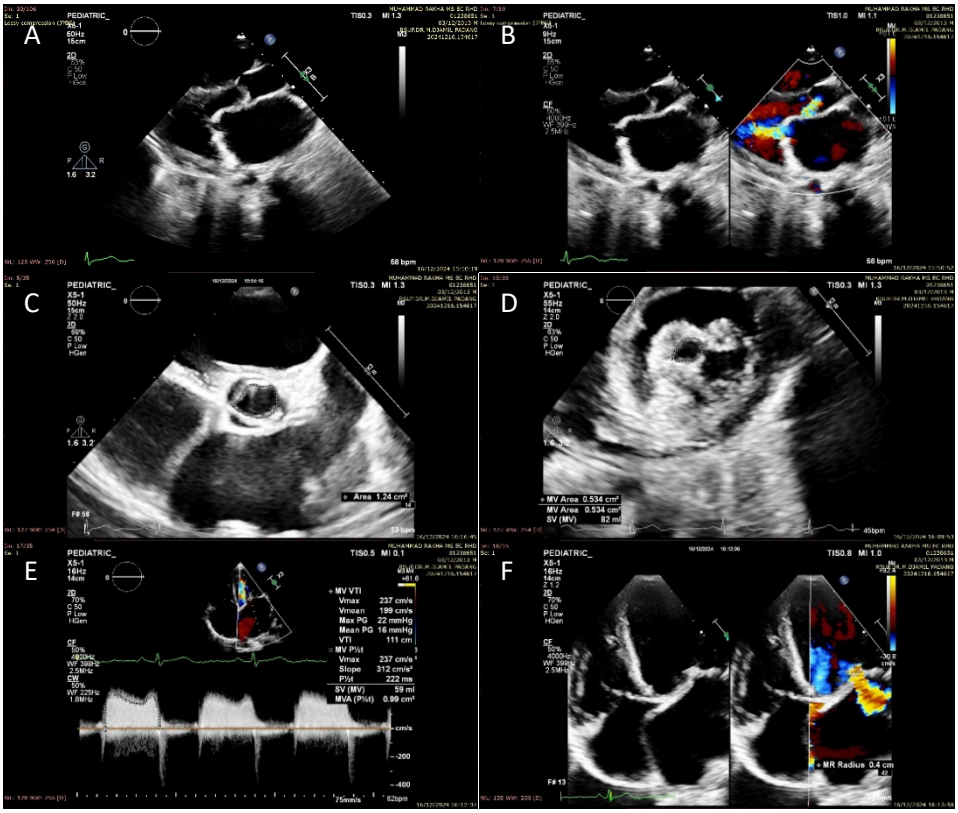

Figure 1. Echocardiography at Dr. M. Djamil General Hospital, (a) parasternal long axis showed thickening valves with doming AML and restricted PML; (b) parasternal long axis view with colour; (c) short axis view showed AVA planimetry 1,24 cm^2; (d) short axis view showed MVA planimetry 0,5 cm^2; (e) MVA PHT 0,6 cm2, mean gradient 15 mmHg; (f) four chamber view with showed MR VCD 0,4 cm

Results

The patient was diagnosed with definite recurrent ARF, severe MS, moderate MR Ross classification class II due to RHD, moderate AS, moderate AR Ross classification class II due to RHD, with undernourished. The initial treatment included antibiotic. A skin test was performed before administering a 600,000-unit injection of benzathine penicillin; however, the patient developed an allergic reaction, requiring modification of the antibiotic regimen. Subsequently, the patient was treated azithromycin 250 mg once daily for 5 days, spironolactone 25 mg once daily, bisoprolol 2.5 mg once daily, prednisone 20 mg twice daily, and paracetamol 500 mg three times daily. Nutritional support was provided with a 1600 kcal diet.

By the fifth day of hospitalization, the patient no longer experienced joint pain or fever, and vital signs remained stable within normal ranges. The patient was then discharged with instructions to adhere strictly to the prescribed medications, continue total bed rest for 2 to 4 weeks at home, and attend regular follow-up visits. Ongoing medications included bisoprolol 2.5 mg once daily, spironolactone 25 mg once daily, prednisone 20 mg twice daily, along with secondary prophylaxis using erythromycin 250 mg twice daily.

Discussion

The 2020 Australian Guideline for the Prevention, Diagnosis, and Management of ARF and RHD categorizes ARF diagnosis into seven classifications: (1) Definitive ARF (confirmed); (2) Probable ARF (highly suspected); (3) Possible ARF (uncertain); (4) Definitive Recurrent ARF (confirmed); (5) Probable Recurrent ARF; (6) Possible ARF; and (7) No ARF. Clinical manifestations are divided into major and minor criteria. Major manifestations include carditis, polyarthritis or aseptic monoarthritis, polyarthralgia, Sydenham's chorea, erythema marginatum, and subcutaneous nodules. Minor criteria vary by risk groups. In high-risk populations, minor clinical criteria include fever ≥38.0°C and monoarthralgia; laboratory criteria include elevated acute phase reactants (erythrocyte

sedimentation rate [ESR] ≥30 mm/hr or CRP ≥30 mg/L) and a prolonged PR interval on ECG. For low-risk populations, minor clinical criteria consist of fever ≥38.5°C and polyarthralgia; laboratory criteria include elevated acute phase reactants (ESR ≥60 mm/hr or CRP ≥30 mg/L) and prolonged PR interval on ECG.[7]

The patient was diagnosed with definite recurrent ARF, confirmed by the presence of two major manifestations (carditis and polyarthralgia) along with evidence of GAS infection, a positive ASO test. Echocardiography findings revealed severe rheumatic involvement of mitral and aortic valves. The diagnosis met established criteria for definitive recurrent ARF.

The patient's clinical picture included exertional dyspnea, intermittent bilateral knee pain without swelling or redness, and a history of recurrent fever episodes over the past year, though no sore throat was reported at admission. This absence likely reflects subclinical or undiagnosed initial streptococcal infections leading to mild or silent carditis, which progressed to severe valvular damage. Malnutrition is noted as a compounding factor that may impair immune defenses and facilitate disease progression.[9, 10]

The underlying pathophysiology involves molecular mimicry where antibodies and T-cells directed against streptococcal antigens cross-react with cardiac valve tissue, initiating inflammation and fibrosis. The inflammatory cascade in ARF exerts both structural and functional effects on various cardiac valve components, leading to acute inflammatory damage and ultimately RHD. These changes include dilation of the valve annuli and elongation of the chordae tendineae. Together, these alterations result in inadequate coaptation of the valve leaflets, causing regurgitation. Mitral regurgitation (MR) is the most common valvular abnormality observed in the early stages of rheumatic heart disease. Consequently, children under 10 years of age frequently present with isolated MR. During the healing phase of rheumatic valvulitis, progressive thickening of the mitral valve leaflets and commissural fusion may occur. These structural changes can restrict leaflet mobility and reduce the mitral valve orifice area, potentially leading to mitral stenosis over time. In addition, the rheumatic process may involve the subvalvular apparatus, resulting in fusion and shortening of the chordae tendineae and papillary muscles. [7, 11, 12]

Rheumatic mitral stenosis typically starts with an asymptomatic latent phase after the first episode of rheumatic fever, lasting on average 16 ± 5 years. Over time, there is a gradual fusion of the commissures, which eventually leads to the development of clinical symptoms. This progression towards severe disability occurs over a span of approximately 9 ± 4 years. The clinical progression of rheumatic mitral stenosis (MS) varies significantly, marked by gradual disease advancement over several years. It is estimated that the mitral valve area decreases by approximately 0.1 to 0.3 cm^2 per year, depending on individual risk factors. Following ARF, rheumatic MS typically progresses slowly. However, in endemic countries, rheumatic MS can advance rapidly, often presenting with severe disease in young adults and even children. Moreover, stenotic lesions can also develop in children under 15 years of age, which may result from extensive and rapid valvular damage due to inadequate prophylaxis or high rates of recurrent infections that are not appropriately treated. These factors likely contribute to the rapid progression of severe mitral stenosis observed in this 11-year-old patient.[7, 11, 12]

Management in this patient included eradication of streptococcal infection with azithromycin, due to penicillin allergy, corticosteroid therapy to control inflammation, symptomatic treatment with beta-blockers and diuretics, and nutritional support. Secondary prophylaxis with erythromycin was instituted to prevent further GAS infections and ARF recurrences. The severity of valvular disease indicates that surgical intervention such as valve repair or replacement may become necessary in the future. Education on medication adherence, hygiene, and regular follow-up remains essential to reduce recurrence and complications.[7, 13, 14]

This case highlights the importance of early diagnosis, regular echocardiographic monitoring, strict adherence to secondary prophylaxis, and patient education to mitigate disease progression and prevent complications such as heart failure, arrhythmias, and thromboembolic events.

Conclusions

Rheumatic Heart Disease (RHD) is a heart disease resulting from residual symptoms of ARF, characterized by the development of heart valve defects. However, the progression to severe valvular abnormalities in RHD takes a

long time. In this case, we report an 11-year-old male patient with recurrent ARF who has already developed severe valvular damage. Although the patient had no prior history of ARF, severe valvular abnormalities were found, likely due to a subclinical episode of ARF that went undiagnosed. A history of missed GAS infections, even without obvious symptoms, can trigger an autoimmune response that leads to gradual valve damage. This process may occur asymptomatically or with mild symptoms but ultimately causes severe mitral valve stenosis over time. Management included bed rest, antibiotics for GAS eradication, analgesics, corticosteroids, betablocker and mineralocorticoid receptor antagonist. Patient education is crucial to prevent recurrence and further disease progression.

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Declaration concerning generative AI and AI-augmented technologies in the compositional process

In the course of preparing this paper, the authors utilized ChatGPT to enhance readability and linguistic quality. Subsequent to utilizing this tool/service, the writers assessed and amended the information as necessary and assume complete accountability for the publication's content.

Declarations of competing interest

The authors declare that there are no competing interests associated with this publication.

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