CASE REPORT

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Recurrent acute rheumatic fever: a case report

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ABSTRACT

Introduction: It is estimated that about 3% of people untreated for group A streptococcal infection will develop rheumatic fever. In most case, an appropriate treatment with antibiotics will prevent acute rheumatic fever. However, not all case of acute rheumatic fever showed an apparent clinical presentation. Furthermore, some symptomatic patients did not seek medical treatment. These caused rheumatic fever and rheumatic heart disease still prevalent, especially in the developing country. Proper management, according to the latest guideline should be prompted in those individuals to halt the progression of cardiac damage. This article describes one such case.

Case Description: A 15-year-old boy with a chief complaint of breathlessness during activity and improved with rest and multiple joint pain. He had a history of recurrent upper respiratory infection, which was not treated with antibiotics. On physical examination, the blood pressure was 110/70 mmHg and heart rate 110 bpm. On cardiac examination, he had holosystolic, and mid-diastolic murmur heard best at the apex. Laboratories test found WBC 15.420/µL, ASTO 400 IU/ml and CRP 48 mg/dL. Chest x-ray showed cardiothoracic ratio of 59% and echocardiography showed left atrial enlargement and left ventricular hypertrophy (ejection fraction 66%), mild mitral stenosis (MVA 1,6 cm^2, mean MVG 13 mmHg), and severe mitral regurgitation. The patient was then diagnosed with recurrent acute rheumatic fever and treated with erythromycin 500 mg q.i.d and aspirin 500 mg q.i.d.

Conclusion: Adequate management of acute rheumatic fever during and after the acute episode aimed to reduce the recurrence, prevent cardiac deterioration and expected to improve quality of life.

Keywords: Recurrent, Rheumatic Heart Disease, Carditis

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INTRODUCTION

Acute rheumatic fever is a delayed autoimmune response after group A streptococcal infection of the pharynx, which can develop into carditis and rheumatic heart disease for life. This disease usually responds well to medical management if started early.[1] Rheumatic heart disease refers to long-term heart damage caused by one severe episode or several recurring episodes of acute rheumatic fever. RHD is a significant cause of morbidity and mortality throughout the world, especially in areas that lack resources. At present acute rheumatic fever and rheumatic heart disease predominantly affect people in low and middle-income countries, as well as in native populations in rich countries where group A hemolytic streptococcal infections may be left untreated which allows for the development of sequelae after the infection cease.[2]

Research from Lawrence and colleagues' states that the highest recurrence rate was in the first year after the first episode of acute rheumatic fever and decreases to 0 in 10 years after the first episode. The recurrence rate in the first year after diagnosis is 4.5%, and the 5-year recurrence rate is 12.5%. Recurrence is more frequent in younger patients and decreases in older age at initial presentation (by 7% per year of age).[1,3]

CASE DESCRIPTION

A 15-year-old boy came to the cardiac center with major complaint breathlessness during activity in the school which decreased with rest and recurrent upper respiratory infection, which was not properly treated. The patient also complains of polyarthralgia. History of trauma, convulsions or body movements that cannot be controlled, redness of the skin and swelling in the joints denied by the patient. On physical examination, the blood pressure was 110/70 mmHg, heart rate 110 bpm, respiratory rate 20 times per minutes, the body temperature of 37°C. On cardiac examination, the patient had holosystolic, and mid-diastolic murmur heard best at the apical area. Other physical exams were within normal limits.

On ECG examination (Figure 1), sinus rhythm is obtained. The pulse rate is about 110 times per minute regular, with normal axis, P mitral wave with a duration > 0.10 seconds and notched in lead

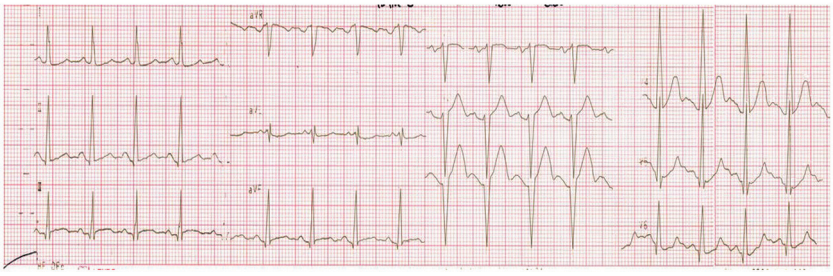

Figure 1. Electrocardiography

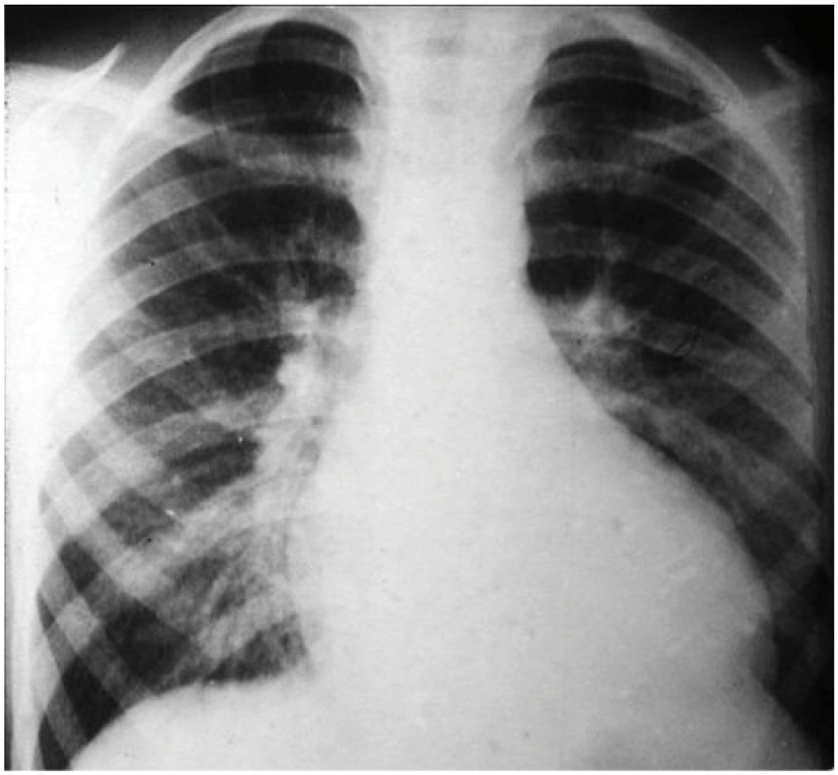

Figure 2. Chest X-ray

II and biphasic in V1 with depth > 1 mm, interval PR 0, 14 seconds, the QRS complex is narrow, duration 0.08 seconds with R wave height at V5 25 mm and in S wave at V1 17 mm, ST-segment and T-wave were normal. The overall result concludes a sinus tachycardia with left atrial enlargement (LAE) and left ventricular hypertrophy (LVH).

In laboratory tests, the hemoglobin levels were 13.3 g/dL, hematocrit 43.1%, erythrocytes 5.61 million/µL, leukocytes 15,420/µL, platelets 327,000 / µL, SGOT 24 U/L, SGPT 14 U/L, urea 23 mg/dL, creatinine 0.8 mg/dL, blood sugar at 63 mg/dL, sodium 134 mEq/L, potassium 3.4 mEq/L, and chloride 95 mEq/L. The ASTO examination showed 400 IU/ml and CRP 48.00 mg/dL. On chest radiograph examination (Figure 2), the impression of heart's waist disappeared with Cardiothoracic Ratio (CTR) of 59%.

The patient had an echocardiography, and the results were LA and LV dilatation, normal global Left Ventricular systolic function with EF 66%, global normokinetics, Mitral Stenosis (MS) mild (MVA 1.6 cm2, mean MVG 13 mmHg), vegetation (-), Mitral Regurgitation (MR) severe likely resulted from Rheumatic Heart Disease (RHD), trivial Aortic Regurgitation, Trivial Tricuspid Regurgitation, mild Pulmonary Hypertension, normal Right Ventricular contractility, Inferior Vena Cava (IVC) diameter 0.9 cm, collectability> 50% with Right Atrial Pressure (RAP)/Central Venous Pressure (CVP) 3-5 mmHg.

Based on the history, physical examination, and supporting examination, the patient is diagnosed as recurrent acute rheumatic fever. Patients have treated with paracetamol 500 mg every 8 hours if fever, erythromycin 500 mg q.i.d and aspirin 500 mg q.i.d.

DISCUSSION

Risk factors for acute rheumatic fever and rheumatic heart disease include age, gender, and various environmental factors. In terms of age, ARF mostly affects children aged 5-14 years, and some cases of ARF can affect even younger children. Recurrent episodes generally affect older children and can occur in young adults. Rheumatic heart disease often has cumulative damage, the peak prevalence of RHD occurs in the twenties and thirties of individuals, although the burden of RHD in children and adolescents remains large.[4] In these patients, the age, gender and environmental factors were risk factors for being affected acute rheumatic fever. Although in the context of gender, women had a higher likelihood of acute rheumatic fever.[3]

There is no single gold standard test to diagnose rheumatic fever. The diagnosis of acute or recurrent rheumatic fever depends on a set of clinical criteria. The most well-known criterion is the Jones criteria, which has undergone the fifth revision in 2015 that extends its application to low and high-risk populations (Table 1).[2,3]

The revision also includes subclinical carditis outcomes as the main criteria, which are diagnosed through echocardiographic evaluation. The diagnosis of acute rheumatic fever based on Jones criteria requires two major criteria or one major and two minors, along with evidence of previous streptococcal infection. Chorea and chronic onset late carditis remain exceptions to these requirements, and each is considered sufficient evidence. Apart from that, there are WHO criteria for diagnosing rheumatic fever and rheumatic heart disease based on the revised Jones criteria (Table 2).[2–4]

Tabel 1. Revision of Jones criteria[5]

A. All patients with evidence of previous group A streptococcal infection	
Diagnosis: First acute rheumatic fever	Two major manifestations or one major plus two minor manifestations
Diagnosis: Recurrent acute rheumatic fever	Two major or one major and two minor or three minor
B. Major Criteria	
Low-risk population	**High-risk population**
Carditis: Clinical and/or subclinical	Carditis: Clinical and/or subclinical
Arthritis: -Only polyarthritis	Arthritis: - Monoarthritis or polyarthritis - Poliarthralgia
Sydenham Chorea	Sydenham Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
C. Minor Criteria	
Low-risk population	**High-risk population**
Polyarthralgia	Monoarthralgia
Fever (³ 38,5ºC)	Fever (³ 38,0ºC)
LED ³ 60 mm in the first hour and / or CRP ≥ 3.0 mg / dL	LED ³ 60 mm in the first hour and / or CRP ≥ 3.0 mg / dL
Prolongation of PR interval after calculating for age variability (except carditis is a major criterion)	Prolongation of PR interval after calculating for age variability (except carditis is a major criterion)

Table 2. WHO criteria for the diagnosis of rheumatic fever and rheumatic heart disease (based on revised Jones criteria).[4,6]

Diagnostic category	Criteria
Primary rheumatic fever	Two major* manifestations or one major and two minor** plus evidence of group A Streptococcal infection***
Recurrent rheumatic fever *without* rheumatic heart disease	Two major manifestations or one major and two minor plus evidence of preceding group A streptococcal infection
Recurrent rheumatic fever *with* rheumatic heart disease	Two minor manifestations plus evidence of preceding group A streptococcal infection
Rheumatic chorea Rheumatic carditis with insidious onset	Other major manifestations or evidence of group A Streptococcal infection is not required
Chronic rheumatic heart disease with valve abnormalities	Does not require criteria for the diagnosis of rheumatic heart disease
*Major manifestations	Carditis Polyarthritis Korea Erythema marginatum Subcutaneous nodules
**Minor manifestation	Clinical: fever, polyarthralgia Laboratory: acute-phase proteins (CRP, LED)
***Evidence of Streptococcal infection in the last 45 days	ECG: Prolonged P-R interval ↑ ASTO Throat swab culture (+) Group A streptococcal antigen test Recent Scarlet fever

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Jones criteria accentuate high specificity to avoid false-positive diagnoses which include history taking, physical examination and investigations. In this patient based on the patient history, we found fever, joint pain, a history of upper respiratory tract infection accompanied by dysphagia when the patients at the age of 5 years, this can cause rheumatic fever that develops into rheumatic heart disease.

On physical examination, the left heart margin has been dilated and was found approximately at the anterior axillary line. On auscultation, we found holosystolic and mid-diastolic murmurs in the apical region with suspicion of regurgitation and stenosis of the mitral valve. This finding point to the possibility of rheumatic carditis as it always associated with the murmur.

Supporting investigation performed on the patient is electrocardiography, laboratory and echocardiography. Electrocardiography in these patients did not reveal any prolongation of the PR interval, only enlargement of the left atrium and hypertrophy of the left ventricle. Laboratory tests are in the form of antistreptolysin-O (ASTO) and acute-phase protein (C-reactive protein) tests with a yield of 400 IU/ml and 48 mg/dL, respectively. Echocardiography provides evidence of valve involvement and it can confirm the suspicion of valve regurgitation and exclude non-rheumatic causes of valve disease. This patient found severe mitral valve regurgitation and mild mitral valve stenosis.

In this patient based on Jones criteria, there were two major criteria, carditis and polyarthralgia. We also found the minor criteria, fever and CRP value ≥ 3.0 mg/dL. Based on the WHO criteria, we obtained two minor manifestations (fever and an increase in CRP) coupled with evidence of group A streptococcal infection (increased ASTO). Based on these, this patient can be diagnosed with recurrent acute rheumatic fever.

Therapy in rheumatic heart disease aims to eliminate infections from pathogenic bacteria, suppress the inflammatory process of the autoimmune response, prevent the re-occurrence of acute rheumatic fever and treat the symptoms related to cardiac complications. Patients are usually given antibiotic treatment for eradication of streptococcal infections, an anti-inflammatory which usually consists of aspirin and steroids. Aspirin is very effective in reducing all manifestations of acute rheumatic fever except chorea and usually produce a rapid response. The recommended dose is 50-60 mg/kg/day up to a maximum dose of 80-100 mg/kg/day in 4-5 divided doses. In this case, the patient was treated with aspirin 500 mg 4 times a day.[7]

Eradication of streptococcal infections using penicillin in the form of oral penicillin V (phenoxymethyl-penicillin) is given 250 mg 2 to 3 times a day for children weighing £ 27 kg, while children ³ 27 kg, adolescents, and adults given 500 mg 2 to 3 times per day. Amoxicillin is given 50 mg/kg per day (maximum 1 gram). The other option is intramuscular Benzathine penicillin G 600,000 U in patients weighing £ 27 kg and 1,200,000 U in patients weighing ³ 27 kg. In patients who are hypersensitivity to penicillin, they can be given the first-generation cephalosporin, cefadroxil or cefalexin. The macrolide group, such as erythromycin, clarithromycin, and azithromycin, should only be given to patients with hypersensitivity to beta-lactam antibiotics. In this case, erythromycin was given 500 mg 4 times per day. The duration of therapy for eradication of streptococcal infection is ten days.[8,9]

Secondary prevention of rheumatic fever is the continuous administration of specific antibiotics in patients with previous rheumatic fever attacks or with rheumatic heart disease. The aim is to prevent the colonization of group A beta-hemolytic streptococcal infection in the upper respiratory tract and the development of recurrent attacks of rheumatic fever. Benzathine penicillin G is given 600,000 U for children weighing £ 27 kg and 1,200,000 U for body weight ³ 27 kg intramuscularly every four weeks. Penicillin V 250 mg twice daily orally, Sulfadiazine 0.5 gram once daily for body weight £ 27 kg and 1 gram once daily for body weight ³ 27 kg orally. If the patients were allergic to penicillin and sulfadiazine, macrolide or azalide could be given. In this case, erythromycin 250 mg 2 times a day was planned.[9]

The duration of secondary prevention must be determined individually, depending on whether the patient has suffered from carditis and complications in the form of chronic valve disease. Secondary prevention must be given from 5 to 10 years from the recurrence of the last rheumatic fever or until the age of 21 years (whichever is longer). In cases of rheumatic fever with carditis leading to chronic heart disease, prevention must be given for 10 years or up to 40 years (whichever is longer).[10]

CONCLUSION

This case describes a patient with recurrent acute rheumatic fever who has been diagnosed based on history, physical examination and supporting investigations according to the latest guidelines. Management of rheumatic fever consists of adequate treatment during the acute rheumatic fever and the secondary prevention to reduce the risk of its recurrence that would cumulatively add

up the cardiac damage and is expected to improve the overall quality of life.

CONFLICT OF INTEREST

The authors declare that there are no conflict of interests or whatsoever regarding the writings of this study.

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AUTHOR CONTRIBUTION

All authors have contributed equally to all process in this research, including preparation, data gathering and analysis, drafting and approval for publication of this manuscript.

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