



Case Report

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Recurrent Rheumatic Fever

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Abstract

Rheumatic fever is a delayed autoimmune response following group A streptococcal infection, which may progress to carditis and lifelong rheumatic heart disease. Although recurrent rheumatic fever in adults is uncommon and the recurrence rate declines with age and with the interval from the initial episode, we describe a patient who has had two episodes of rheumatic fever, one in childhood and one in adult life.

Keywords: Rheumatic fever, Carditis, Rheumatic heart disease, Jones criteria.

INTRODUCTION

Rheumatic fever is a prevalent healthcare problem in the developing countries. It is estimated that there are at present about 15 million cases of rheumatic heart disease (RHD) worldwide, causing 233,000 deaths annually [1]. India contributes to 25%–50% of the global burden of Rheumatic Heart Disease [2]. A study from Orissa shows that Rheumatic heart disease still accounts for 45% of cardiac admissions in tertiary care hospitals [3]. The incidence of Recurrent Rheumatic fever has been reported between 15 and 34% in India [4]. Similarly, a population survey done in the year 2000 in northern India reported a high prevalence of Rheumatic heart disease (6.4/1000) [5]. According to a population survey in 2011, there are expected to be more than 3.6 million patients of RHD, almost 44,000 patients are added every year, and expected mortality is 1.5%–3.3% per year [6]. Repeated episodes of Rheumatic Fever are an important factor for worsening of Rheumatic Heart Disease and is associated with increased frequency of carditis [7,8]. Prior carditis and younger age have also been associated with higher rates of Recurrence [9].

CASE REPORT

A 28-year-old male, with a history of rheumatic heart disease presented to the Hospital with three days history of fever and polyarthralgia non migratory type, involving major joints (both shoulder joints, Elbow, Knee and Ankle). He had a past medical history of one episode of rheumatic fever at 5 years of age. During that episode he had Fever, Breathlessness and arthralgia following a sore throat and was diagnosed as acute rheumatic fever and was advised Oral Penicillin prophylaxis but patient was non-compliant with his medication. At the age of 20, the patient was diagnosed to have Moderate Mitral Stenosis and Moderate Mitral Regurgitation in Atrial Fibrillation and underwent Mitral Valve Repair. Since then patient is on Oral Anticoagulation and Penicillin prophylaxis.

On physical examination, the patient was febrile, Pulse rate was 84 beats per minute, Irregularly irregular, Normal Volume, no specific character, all peripheral pulses felt. Blood Pressure was 130/80 mmHg, Right arm in supine position, Equal in both limbs. No Peripheral signs of Aortic regurgitations or Infective Endocarditis or Rheumatic fever were present. Cardiovascular examination revealed Apex beat in fifth intercostal space and 1.5cm lateral to mid clavicular line and A crescendo decrescendo Ejection systolic murmur of grade IV heard in Aortic area, An Early diastolic murmur in Erb's area both heard best with the diaphragm of the stethoscope in sitting and leaning forward position and a rough rumbling low pitched Mid diastolic murmur in Mitral area heard with bell of the stethoscope in left lateral position. Other Systemic Examination were Normal.

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Initial laboratory findings included a Hemoglobin 13.2 g/dL, White cell count 10700 cells/cu mm and Platelet count of 4.60 lakh /cu mm. Renal and Liver parameters were within normal Limit, Viral Markers were Negative, Blood and Urine Cultures were Negative. Acute Phase Reactants were Elevated [C-reactive protein (CRP) was 9.6 mg/dL and Erythrocyte sedimentation rate (ESR) was 112 mm/first hour] and Antistreptolysin O titre (ASO titre) was Elevated 859 IU/ml (Normal upto 116 IU/ml).

Chest X ray: Straightening of Left Heart border with double density sign and Increased Cardinal angle. Suggestive of Left Atrium Enlargement and Left Ventricular Enlargement.



Figure 1: Chest X Ray

Echocardiogram:

- Hugely dilated Left Atrium and Left Ventricle
- Mitral Ring insitu
- AML thickened and PML Calcified with Restricted Mobility
- Gradient Across Mitral Valve (PG – 17MMHG, MG – 10MMHG)
- Mitral Valve Area by PHT – 1.7CMSQ
- Mild Mitral Stenosis with Mild to Moderate Mitral Regurgitation
- Gradient Across Aortic Valve (PG – 46MMHG, MG – 27MMHG)
- Mild Aortic stenosis with Moderate Aortic Regurgitation (AR by PHT – 376 MS)
- Mild to Moderate Tricuspid Regurgitation with Mild Pulmonary artery Hypertension (EPAP- 44mmHg)
- Paradoxical Septal Motion and Adequate Left Ventricular Systolic Function (Ejection Fracture 52%)
- IVC 1.7CM Partially Collapsing

Diagnostic criteria (Jones’s and WHO criteria) for rheumatic fever [10] is explained in table 1.

Diagnostic Criteria for Recurrent Rheumatic Fever with established rheumatic heart disease includes two minor manifestations plus evidence of a preceding group A streptococcal infection.

Diagnostic Criteria for Recurrent Rheumatic Fever without established rheumatic heart disease includes two major or 1 major and 2 minor manifestations plus evidence of a preceding group A streptococcal infection.



Figure 2: Echocardiogram showing deformed aortic and mitral valves

DIAGNOSIS

Based on Revised Jones Criteria and WHO guidelines for diagnosis of Recurrent Rheumatic Fever, Our patient had One Major (Carditis), Three Minor Criteria (Fever, Polyarthralgia and Elevated Acute Phase Reactant) and supporting evidence of a preceding streptococcal infection i.e., elevated Anti-streptolysin-O (ASO) titre, fulfilling the criteria for Diagnosis of Recurrent Rheumatic Fever in patient with established Rheumatic Heart Disease.

TREATMENT AND FOLLOWUP

After confirming the diagnosis of Recurrent rheumatic fever, patient was given one dose of Injection Benzathine Pencillin G 1.2 million units intramuscularly and was treated with Oral Prednisolone 1mg/kg body weight and high dose Salicylates (Aspirin) 600mg three times a day less than the advised dose (i.e, 50 to 60mg per kg body weight) as patient could not tolerate the full dose. Patient was discharged once he was stable and on follow up the patient was symptomatically better, CRP and ASO levels were found to be in decreasing trend and Oral Steroids was tapered gradually every two weeks.

DISCUSSION

We present a 28 year old male with recurrent rheumatic fever who presented with carditis, fever, arthralgia, elevated acute phase reactants (ESR, CRP) and a high ASO titre satisfying modified jones criteria and WHO guidelines for recurrent rheumatic fever. Very few proven cases of recurrent rheumatic fever have been reported worldwide.

Rheumatic fever is a delayed autoimmune response following group ‘A’ streptococcal infection, which may progress to carditis and lifelong rheumatic heart disease [11]. Rheumatic fever develops as a result of molecular mimicry. There are structural similarities between streptococcal antigens and human proteins which elicit an unwanted adaptive immune response against valvular interstitial cells. Early diagnosis and treatment favours good prognosis preventing complications like valvular heart diseases, infective endocarditis etc. [12]

According to a study conducted by Nassri Camara. et.al [1] young age (≤23 years), non-compliance to prophylactic antibiotics and heart failure were the strongest and independent risk factors for recurrence

of rheumatic fever. Our patient had history of non-compliance to medication and a prior rheumatic valvular heart disease. Recurrence of rheumatic fever in already existing rheumatic valvular heart disease further worsens the damage leading to bad outcome and increases the morbidity of the patient. Hence a strict compliance to the medication and regular follow up is mandatory

Secondary prevention of rheumatic fever with prophylactic antibiotics is highly recommended worldwide. Benzathine penicillin G with a dose of 1,200,000 units for adult patients is recommended single intramuscular injection every 3-4 weeks. Other regimens are oral penicillin V, sulfonamides, and erythromycin. Penicillin either given orally or intramuscularly is the preferred antibiotic as very less resistance has been reported across [13]. Duration of secondary prophylaxis is explained in table 2. The total duration of prophylactic treatment depends on the clinical presentation of the patient, severity of the disease, involvement of the valves.

Table 1: Diagnostic criteria for rheumatic fever

	Jones Criteria	World Health Organization Criteria
Major Criteria	<ul style="list-style-type: none"> • Carditis • Polyarthritits • Chorea • Erythema marginatum • Subcutaneous nodules 	<ul style="list-style-type: none"> • Carditis • Polyarthritits • Chorea • Erythema marginatum • Subcutaneous nodules
Minor Criteria	<ul style="list-style-type: none"> • Fever • Polyarthralgia • Elevated acute phase reactants (ESR OR CRP) 	<ul style="list-style-type: none"> • Fever • Polyarthralgia • Elevated acute phase reactants (ESR OR CRP) • Electrocardiogram (prolonged P-R interval)
Supporting Evidence of A Preceding Streptococcal Infection	<ul style="list-style-type: none"> • Electrocardiogram (prolonged P-R interval) • Elevated or rising antistreptolysin O or other streptococcal antibody titre • A positive throat culture, or • A rapid antigen test for group A streptococci or recent scarlet fever 	<ul style="list-style-type: none"> • Elevated or rising anti-streptolysin O or other streptococcal antibody • A positive throat culture, or • A rapid antigen test for group A streptococcus, or • Recent scarlet fevere

Table 2: Duration of secondary prophylaxis

Category of Patient	Duration of Prophylaxis
Rheumatic fever without carditis	For 5 years after the last attack or 21 years of age (whichever is longer)
Rheumatic fever with carditis but no residual valvular disease	For 10 years after the last attack, or 21 years of age (whichever is longer)
Rheumatic fever with persistent valvular disease, evident clinically or on echocardiography	For 10 years after the last attack, or 40 years of age (whichever is longer); sometimes lifelong prophylaxis

CONCLUSION

Patients with risk factors like young age, non-compliance and prior heart failure have high chances of recurrent rheumatic fever. And hence these group of population needs regular follow up, screening and drug compliance. Although infective endocarditis is the first diagnosis usually thought of in a patient with previous history of rheumatic heart disease with fever, recurrent rheumatic fever should be considered as a differential diagnosis.

Conflicts of Interest: None.

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