

Resurgence of Acute Rheumatic Fever and Rheumatic Heart Disease? A Case Series from Tertiary Care Centre of Upper Assam

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I. Introduction

Acute Rheumatic fever (RF) is a multisystem inflammatory disorder presenting with self limited migratory arthritis. It may be also accompanied or followed by carditis, and less frequently by chorea and skin involvement. It is related to post-infectious autoimmune mechanisms driven to group A streptococcus antigens. Jones criteria are the main diagnosis guidelines, by combining major and minor signs at presentation. There is no single biologic marker, and the disease course is characterized by relapse after re-exposure to streptococcal antigens.¹⁻⁶ It is estimated that carditis may occur in up to 60% of the cases.^{5,6} Rheumatic valvular disease may be caused by a single severe attack, but it is often related to recurrent attacks. However, relapsing risks should be considered even for RF mild forms.

Epidemiology

According to WHO,⁷ at least 15.6 million people have RHD, 300 000 of about 0.5 million individuals who acquire ARF every year go on to develop RHD, and 233 000 deaths annually are directly attributable to ARF or RHD. However, these estimates are based on conservative assumptions, so the true disease burden is likely to be substantially higher. Furthermore, the overall quality of epidemiological data from developing countries is poor, particularly with respect to research documenting the incidence of ARF.⁷ The incidence of rheumatic fever in developed countries is in the range of 0.2 to 0.5 per 1000 but that for developing countries is variable from place to place reaching as high as 20 per 1000 children living in Johannesburg, South Africa. The prevalence of RHD increases with age, peaking in adults aged 25–34 years, reflecting ARF activity in previous decades.⁸ In young patients, mitral valve regurgitation is the predominant cardiac lesion, but mitral stenosis becomes progressively more common with increasing age.⁹ This trend, whereby the incidence of ARF peaks in childhood and adolescence, but the prevalence of RHD peaks in adulthood, has been documented in studies done in the USA, in Aboriginal Australians, in India, and in Burma, and is likely to be seen in all populations with high rates of RHD.^{10,11,12} In many populations, ARF and RHD are more common in females than males.⁴⁰ Whether this trend is a result of innate susceptibility, increased exposure to group A streptococcus because of greater involvement of women in child rearing, or reduced access to preventive medical care for girls and women is unclear.

II. Aims and Objectives

We have recently diagnosed quite a few number of cases of Acute rheumatic fever over a short period of six months. We report them in this case series.

Case 1

Dipika dao a 18 yr. old female patient presented with dyspnoea on exertion for 1 year, palpitations for 2 months. There is no history of swelling of lower limbs, no tremors, no chorea, no skin manifestations. There is h/o pulmonary kochs 6 months back, h/o severe excruciating joint pain and fever is present when she was 11 years old which subsided after taking pain killers without leaving any joint deformity. On examination the BP is 100/60, pulse is 112/min and normal in rhythm and there is decreased air entry in the left lower chest, abdominal and CNS systems are normal, CVS examination revealed the apex is located in Lt. 5thics 2 cm lateral to MCL a mid diastolic murmur in apex with opening snap is heard, a mid-diastolic murmur is also heard in the right Lower sternal edge which is increased on inspiration. On investigating Hb is 10.1 gm/dl, ESR & CRP are within normal and rest of the reports are within normal range. Echocardiography showed severe MS, mild AR, severe TR and severe PAH.

Case 2

Pinky Keot a 17 year old female patient presented with fever and multiple joint pains associated with swelling and redness for 7 days. It started in the rt. Knee and then few days later the lt. knee and then the elbows. The pain and redness of the knee joints subsided by the time the elbows was painful, there is h/o sore throat 1 month back which subsided on medications, there is no h/o chorea, no skin manifestations. There is similar type of joint pains 4 years back. On examination Cardiovascular system revealed a soft diastolic murmur of changing character which is best heard in the apex. Neurological and respiratory systems were normal. On investigation the ESR is raised and ASO titer is >800 rest of the parameters are within normal limit. Echocardiography revealed no abnormality.

Case 3

Dipamoni Das a 13 year old female presented with fever and multiple joints pain for 15 days. The joint pain started in the lt. ankle and lt. knee joint and after few days it involved the rt. Knee and then the hips and both the elbows. There is no h/o chest pain and palpitations abnormal body movements and skin manifestations. On examination the joints are warm and tender and there is severe limitation of movements. Respiratory and nervous system were normal. Cardiovascular examination revealed a soft pansystolic murmur best heard in the apex radiating to the axilla. On investigations ESR is 140 and total count is 17,000; ASO titer is >400, rest of the blood parameters are within normal limit. Echocardiography showed moderate MR mild AR and AML slightly thickened.

Case 4

Dalimi Jamo a 38 yr. female presented with fever, chest pain, pain in the multiple joints for 7 days. The joint pain is associated with swelling and limitation of movements. There is no swelling of the lower limbs no respiratory distress. There is similar type of joint pain 1 year back which got subsided on taking pain killers. There is no history of TB no significant past history noted. On examination the joints are tender but not hot and there is crepitations in the lt. lower zone of the lung. Cardiological examination revealed a soft pansystolic murmur best heard in the apex. Nervous system is normal. On investigating the ESR is 62 and ASO titer is > 400 rest of blood parameters are within normal limit. Echocardiography revealed mild pericardial effusion with no tamponade mild MR mod TR and mod PAH were noted.

Case 5

Malati Murah a 16 yr. female presented with breathlessness and easy fatigability for the last 2 years with history of multiple joint pain 2 years back. There is history of PND no h/o palpitation or chest pain is there. No fever and joints pain is present. On examination the BP is 100/60. Cardiological examination revealed a loud S1, mid diastolic rough and rumbling murmur in the apex, neurological and respiratory system were normal. On investigating ESR and ASO titer are within normal limit. Echocardiography revealed severe MS, mod TR and enlarged LA.

Case 6

Papu gogoi a 13 year male presented with fever and multiple joint pains for 2 months, the pain started in the rt. Ankle joint and then it progressed to the lt. Knee and within 2 days the rt knee joint and lt. Ankle also got involved, the pain was associated with swelling and marked restriction of the movements. The patient had palpitations which was sudden in onset however there is no history of chest pain or breathlessness, no skin manifestations and no abnormal body movements. On examination the joints are swollen and tender, cardiological examination revealed an early diastolic murmur best heard in the 3rd lt. Sternal area. Nervous system and respiratory system were normal. On investigation ESR is 130 ASO titre is 443 IU/ml, CRP and TC was raised, rest of the blood parameters are within normal limit. Echocardiography revealed moderate AR.

Case 7

Mohendra sonowal a 20 yr. male presented with multiple joints pain for 1 month. It started in the left knee joint followed by the left elbow and then right elbow and right knee joint. It was associated with fever on and off of same duration. There is no history of chest pain, palpitations and breathlessness, no skin manifestations and abnormal jerky movements of the body. On examination the joints are swollen and tender. Cardiological examination reveals no abnormality, respiratory and nervous system were normal. On investigation the ESR is 75 and ASO titre is >400, rest of the blood parameters are within normal limit. Echocardiography shows normal valves and chamber.

Case 8

Saher Ahmed a 20 year male presented with multiple joint pain for 2 months and fever for 1 and ½ months . It was associated with chest pain and breathlessness. On examination the joints are swollen and tender and there is marked limitation of movements. There is no history of skin manifestations and abnormal jerky movements of the body. Cardiological examination revealed a pansystolic murmur in the apex and an early diastolic murmur in the Lt 3rd intercostals space. Respiratory and neurological examination are within normal limit. On investigations ESR is 20, CRP is >2.4 mg /dl and ASO titre is >1600 IU/ ml , rest of the blood parameters are within normal limit. Echocardiography revealed mild MR, mild AR and enlarged left atrium, ecg reveals 1st degree AV block.

Case 9

Lakhi gogoi a 13 year male presented with multiple joint pains for 15 days, it started in the rt knee joint, ankle joint and then the Lt knee joint . It was associated with fever .There is h/o sore throat in the past. There is no h/o chest pain, no palpitations ,no skin manifestations and abnormal jerky movements of the body. On examination the joints are swollen and tender . cardiological examination revealed a soft pansystolic murmur in the apex. Respiratory and neurological examination are within normal limit. On investigation the ASO titer is 227.4, ESR 110 ,CRP 4.47. Echocardiography shows trivial MR and trivial AR.

Case 10

Kamal Gohain, 13 year old male from sivsagar presented with migratory poly arthritis, fever and chest pain for one week. His involvement started with left knee followed by left ankle followed by right knee. He was found to have a pan systolic murmur radiating towards left axilla. Pulmonary component of second heart sound was prominent. ECG showed prolonged PR interval. Echocardiography showed anterior and posterior mitral valve thickened, severe mitral regurgitation, moderate tricuspid regurgitation. Ejection fraction was preserved at 69%.

III. Discussion

The exact mechanism streptococcal throat infection result in rheumatic fever is not known. There is neither direct invasion nor toxin-mediated damage to the tissues involved. The most popularly accepted theory is immune mediated damage, where antibodies produced against streptococcal antigen cross reacts with host tissue (like cardiac muscles, synovial and brain tissues, among others) .The m-protein is an antigenically variable cell wall component in some of these bacteria. There are more than 80 subtypes but m-subtypes 1,3,5,6 and 18 are most often associated with acute rheumatic fever.

The main clinical features of ARF are outlined in the Jones Criteria, which were established in 1944 and then modified, revised twice,^{13,14} and updated (panel)¹⁵ by the American Heart Association. Every revision increased the specificity but decreased the sensitivity of the criteria,¹⁶ largely in response to the steadily declining incidence of ARF in developed countries. In regions of the world where ARF is endemic or epidemic, however, and where the risk associated with missed diagnoses—lack of provision of secondary prophylaxis to prevent recurrent ARF and worsening RHD—might outweigh the consequences of over-diagnosis, the 1992 Jones criteria might not now be sufficiently sensitive. As such, the 2002 03 WHO criteria⁸ which, among other things, specified less stringent requirements for the diagnosis of recurrent ARF in patients with established RHD should probably be adopted (panel). The Jones and the WHO criteria are only diagnostic guidelines, however, and should be adapted in certain circumstances, for example to increase sensitivity of diagnosis in populations at high risk of ARF.

Panel: Diagnosis of AR^{15,16}

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| Jones criteria (1992) ¹⁵ |
| Two major or one major and two minor manifestations must be present, plus evidence of antecedent group A streptococcus infection Chorea and indolent carditis do not require evidence of antecedent group A streptococcus infection Recurrent episode requires only one major or several minor manifestations, plus evidence of antecedent group A streptococcus infection |
| Major manifestations |
| Carditis |
| 1) Polyarthritis |
| 2) Chorea |
| 3) Erythema marginatum |
| 4) Subcutaneous nodules |
| Minor manifestations |
| 1) Arthralgia |
| 2) Fever |
| 3) Raised erythrocyte sedimentation rate or C-reactive protein concentrations |

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| 4) Prolonged PR interval on electrocardiogram |
| Evidence of antecedent group A streptococcus infection |
| 1) Positive throat culture or rapid antigen test for group A streptococcus |
| 2) Raised or rising streptococcal antibody titre |
| WHO criteria (2002–03) ¹⁶ |
| Chorea and indolent carditis do not require evidence of antecedent group A streptococcus infection |
| First episode |
| As per Jones criteria ¹⁵ |
| Recurrent episode |
| In a patient without established RHD: as per first episode |
| In a patient with established RHD: requires two minor manifestations, plus evidence of antecedent group A streptococcus infection. Evidence of antecedent group A streptococcus infection as per Jones criteria, but with addition of recent scarlet fever |

Rheumatic Fever is a public health concern due to carditis and heart damage, which may be aggravated by late diagnosis and poor penicillin prophylaxis adherence. A systematic evaluation of patients care is important to define further strategy for disease control and treatment. Arthritis was the most frequent major sign, followed by carditis in the present case series. Arthritis had migratory or additive pattern in most of our cases, affecting predominantly large joints as described originally in the 1950's. The migratory pattern predominated in lower limbs. Presentation with symmetric small joints, neck, and hip arthritis involvement, as well as longer duration was not observed. Small joints, cervical spine involvement and longer arthritis duration need a differential diagnosis with post-streptococcal reactive arthritis, but in the pediatric age range this feature is still a matter of controversy, where it possibly represents an atypical RF profile. Concomitant arthritis and carditis was seen in most of our cases, carditis the second most frequent major sign, and mitral valve was the most frequent valve to get involved.

None of the patients reported of Chorea, Subcutaneous nodules, Erythema marginatum An important clue for diagnosis ascertainment is high or rising titres of antistreptolysin-O. In our series almost all had high antistreptolysin-O titres.

IV. Conclusion

ARF prevalence has fallen in many developed countries, however it is still a challenge for doctors who work in developing countries. In the present study we have tried to present clinical aspects of ARF and RHD patients presenting in the Department of Medicine. In spite of the limitations of small case series study, the study reinforce the need for early diagnosis and long term surveillance, in order to get better disease control to prevent heart damage.

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