RESEARCH LETTER

Pitfalls in diagnosis of Acute Rheumatic Fever

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Acute Rheumatic fever (ARF) is a sequel of an immunological disorder initiated by group A beta hemolytic streptococcus (1-5). It continues to be a major problem in pediatric population and is one of the leading causes of heart disease in children in the developing and underdeveloped countries and accounts for about 600,000 cases of rheumatic heart disease (RHD) in India and 120,000 new cases of ARF annually(2) . The commonest age group involved is 5-15 years .Genetic predisposition and overcrowding associated with low socio-economic status are some of the predisposing factors .Rheumatic fever principally involves the heart, joints, central nervous system, skin and subcutaneous tissues. Although the name acute rheumatic fever emphasizes the involvement of the joints, rheumatic fever owes its importance to the involvement of the heart; as it leads to rheumatic heart disease because of scarring and deformity of the heart valves. With the advent of Penicillin one expected the incidence of rheumatic fever to go down but several other factors including increase in population, rapid urbanization, increasing schools, adverse environment conditions and increasing awareness account for its persistence in developing and undeveloped countries and for the resurgence in areas where it might have been extinct.(4-7) Some of the most characteristic manifestations have become less common and it has become more difficult to establish the diagnosis on clinical grounds. More patients are being seen who have arthritis as their only clinical finding. At times rheumatic fever is over diagnosed and also the immediate institution of anti-rheumatic drugs masks further development and may confuse the clinical profile leading to wrong label of rheumatic fever.

The Jones Criteria which have been used since 1944 for diagnosing this illness have undergone changes and revision many times .The most recent revisions being in1992 and 2003.(1-5)

Major Criteria	**Minor Criteria**
- Carditis. - Migratory polyarthritis. - Chorea. - Erythema marginatum. - Subcutaneous nodules.	**Clinical:** - Arthralgia - Fever. - Previous rheumatic fever, Rheumatic heart disease. **Laboratory:** - Raised acute phase reactants like ESR, CRP and leucocytosis. - Prolonged PR interval.
Essential Criteria:- Evidence of preceding streptococcal infection in form of: - Increased ASO titers. - Positive throat culture for group A streptococcus. - Recent scarlet fever.	
The presence of two major or one major and two minor criteria along with evidence of recent evidence of streptococcal infection were used for diagnosis.	
Few patients who did not fulfil Jones Criteria but were falling in following three categories were considered for diagnosis of theumatic fever. I. Chorea: if other causes have been excluded. II. Insidious or late onset carditis: with no other explanation. III. Rheumatic recurrence: in patients with documented rheumatic heart disease, the presence of one minor criterion with evidence of previous streptococcal infection	

In view of changing pattern of disease with its variable clinical profile and also many other causes like viral infections responsible for similar presentation, the diagnostic criteria which earlier had been held for years and being put to question now. But in absence of any full proof confirmation for the disease entity the support is still dependent on same clinical and laboratory criteria. It becomes relevant to have a review of the disease profile in view of the resurgence of rheumatic fever in certain parts of world and the reported changing pattern of clinical character of rheumatic fever. A study was contemplated with an objective to have an insight into

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clinical profile of the disease as observed in the pediatrics department of the SMGS Hospital, Govt. Medical College, Jammu (1994-97).

Carditis in ARF is always a pancarditis. However, a number of patients may present with the classical clinical findings of valvulitis at the initial phase of the illness. Hence it is now advisable to use Doppler echocardiography as a supplementary diagnostic tool whenever available to pick up sub clinical valvular lesions .Echocardiography also helps to identify the cause of congestive cardiac failure (CCF) in difficult situations such as in cases of established RHD, i.e. , whether CCF is due to recurrent rheumatic activity or due to development of infective endocarditis(7-9).The arthritis of ARF may not confirm to the traditional description of migrating polyarthritis of the big joints . Occasionally it may present as a monoarthritis or additive arthritis when clinical progression of the illness becomes important to establish a final diagnosis.Poststerptococcal reactive arthritis is occasionally difficult to distinguish from ARF. The classical clinical features are that of an additive rather than migratory arthritis, poor response to NSAIDS, persistence for more than two months, and may fulfill the Jones criteria with the presence of elevated acute phase reactants and positive anti-DNAase B. However one must remember that an arthritis lasting for more than 8-12 weeks virtually rules out ARF.The presence of chorea warrants a careful exclusion of other diseases such as systemic lupus erythematosus(SLE),Wilson's disease and intake of drugs such as phenytoin or oral contraceptives .Erythema marginatum is difficult to assess in dark skinned individuals and may be confused with rashes occurring with more common conditions such as sepsis, drug reactions or sometimes juvenile idiopathic arthritis (JIA).Subcutaneous nodules mostly occur with carditis and often be missed if not looked for actively.(2-4) The minor manifestations like fever and arthralgia are very non specific and may occur in many rheumatic conditions. Similarly ESR and C-reactive protein can be elevated in stressful conditions .The evidence of a previous streptococcal infection has been given special consideration in the diagnosis of ARF.However throat cultures are positive in only about 25% of cases and here too it is difficult to say whether the positivity is because of a real infection or a carrier state considering the fact that the prevalence of group A beta streptococcal sore throat is quite high in some populations. The antigen test is again specific but with very low sensitivity thus hampering its utility. The estimation of streptococcal antibodies such as anti sterptolysin O (ASO) and anti-DNAase B gives a reliable indicator of a previous GABS infection but there is an over reliance on ASO titers for the diagnosis of ARF.About 60% of the population may show an elevated ASO titers normally in developing countries such as ours. Hence one must remember that single raised ASO titers does not equate to ARF. Paired sera, i.e, a two fold increase or decrease in ASO titers done at an interval of 4-8 weeks gives a more meaningful interpretation. Similarly a negative ASO titer does not exclude the diagnosis of ARF.(3) Interpretation of ASO must be done in concurrence with other clinical features especially so because about 30% of systemic onset JIA have elevated ASO titers. Anti- DNAase B has good reproducibility but is not readily available to most of us even today.

Though a number of pitfalls in the diagnosis of ARF has been discussed , an over diagnosis at the initial phase of the illness is always better than missing the diagnosis altogether thereby delaying appropriate treatment at an early stage and prevention of serious cardiac morbidity in the long run.

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