ORIGINAL ARTICLE

Clinical Laboratory and Echocardiographic Profile of Rheumatic Fever in Children

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Abstract

This prospective study includes clinical, laboratory and echocardiographic evaluation of 50 cases of rheumatic fever admitted to the Department of Pediatrics, Govt. Medical College, Jammu over a period of 34 months from October 1994 to July 1997. All cases were diagnosed according to modified Jones criteria. Rheumatic fever formed 0.18% of total hospital admissions with majority of children in age group of 5-15 years. Male preponderance was observed. Carditis was the most common manifestation seen in 70% followed by arthritis, subcutaneous nodules and chorea in 60%, 10% and 8% respectively. Erythema marginatum was not seen in any case.

Key Words

Rheumatic, Childhood

Introduction

Rheumatic fever is a sequel of an immunological disorder initiated by group A beta haemolytic streptococcus (1). 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 countries. The commonest age group involved is 5-15 years (2). Genetic predisposition and overcrowding associated with low socio-economic status are some of the predisposing factors (3). 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. Markowitz *et al* has reported a changing pattern of the disease (4). 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. 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. The present study was contemplated with an objective to have an insight into clinical profile of the disease as observed in the pediatrics services of the SMGS Hospital, Govt. Medical College, Jammu.

Material and Methods

The present study was conducted on patients of acute rheumatic fever admitted to the Department of Pediatrics, SMGS Hospital, Govt. Medical College, Jammu over a period of 34 months from October 1994 to July 1997. About 50 patients suspected to have rheumatic fever were enrolled for the study. Besides detailed history, a general physical and systemic examination along with the progress record during the hospital study was carried out as per pre-structured performa. Patients were

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diagnosed using modified Jones criteria, which included major and minor criteria (1) as shown in Table No. 1.

Regarding laboratory profile, the hemoglobin less than 10 gm/dl was taken as significant. The total leucocyte count (TLC) > 10,000/mm3, ESR above 20 mm / hour were considered abnormal. X-ray chest and ECG were done to see for cardiomegaly, congestive heart failure, and rhythm and PR interval respectively. ASO titers more than 200 international units were considered to be abnormal and it revealed an recent evidence of streptococcal infection (4). Echocardiography (2D M-mode Colour Doppler) was carried out in 36 patients.

Results

The incidence and age and sex wise distribution of cases of rheumatic fever in hospitalised children and during the study period are depicted in table no. 2 & 3 respectively. The male, female ratio was 2.5:1. The major and minor criteria are shown in table no. 4. The final diagnosis in 50 cases of rheumatic fever is depicted in Table No. 5.

Table No. 1

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 & 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 included in study (3).

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

Table 2 Incidence Of Rheumatic Fever In Hospitalised Children

Period	Total Admissions	Rheumatic Fever	Percentage
OCT 94 - DEC 94	2211	7	0.31%
JAN 95 - DEC 95	9652	25	0.25%
JAN 96 - DEC 96	10862	12	0.11%
JAN 97 - JULY 97	4840	6	0.12%
TOTAL	27565	50	0.18%

Table 3 Age and Sex Wise Distribution of Cases.

Age group (in years)	Sex		Total	Percentage
	Male	Female		
0-5	1	0	1	2%
5-10	15	6	21	42%
10-15	20	8	28	56%
TOTAL	36	14	50	100

Table 4 Major and Minor Criteria

		No. of cases	Percentage
Major Criteria	Carditis	38	76
	Arthritis.	30	60
	S/C Nodules	5	10
	Chorea.	4	8
	Ery. Marginatum	--	--
Minor Criteria Clinical	Arthralgia	16	32
	Fever	42	84
	Previous H/O	--	--
	Rheumatic fever,	9	18
	RHD.	--	--
Lab.	Raised ESR	31	62
	+ve C.R. Protein.	43	86
	Leucocytosis.	16	32
	Prolonged PR interval	29	58

Table 5 Final Diagnosis in 50 cases of Rheumatic Fever.

Final Diagnosis	Number of cases	Percentage
ARTHRITIS ONLY.	8	16%
CARDITIS ONLY.	13	26%
CARDITIS + ARTHRITIS.	20	40%
CARDITIS + S.C. NODULES.	3	6%
CARDITIS + ARTHRITIS + S.C. NODULES.	2	4%
CHOREA	4	8%

Discussion

Rheumatic fever formed 0.18% of total hospital admissions. The incidence is quite low as compared to other studies where it used to be 0.8% to 1.05% (5-7). Incidence of rheumatic fever among hospitalised children have decreased over years. This is because of increasing awareness among people. Antibiotics have a great role to play in this regard. Standard of living has improved over years and there is an easy access to medical facilities. Also an important reason of low incidence of rheumatic fever in hospital admission is possibility of easy access to private clinics. Even patients from poor socio-economic group prefer to go to a private clinic than visiting a government hospital. Therefore it is not possible to say with certainty that incidence of rheumatic fever has decreased. The maximum number of cases in our study was in age group of 5-15 years (98%) as and noted in other studies (7-9). Only one case was admitted with age less than 5 years. Liability to rheumatism is far greater in the early than in later years, although no age is altogether exempt from its attack.The

male, female ratio observed to our study was 2.5:1, while Mayer et al and Robinson et al have recorded equal incidence of rheumatic fever in both sexes (9-10). Increased occurrence of rheumatic fever in males is perhaps related to greater concern to ailing males in the developing countries.

Carditis and arthritis were commonest major manifestations seen in 76% and 60%. Subcutaneous nodules were seen in five patients (10%) and chorea in 4 patients (8%). None of the patients had erythema marginatum. Most common manifestation was carditis. The manifestations were comparable with other studies. Some other studies show arthritis as commonest picture, others show carditis as commonest (10-12). According to Ghai arthritis is more common than carditis seen in 70-75% and 50-60% respectively. He has described subcutaneous nodules in 3-5% and chorea in 10% of patients (2). Nelson has similar observations of carditis in 40-80% and chorea in 10% (3). Reports from West indicate a similar pattern as noted in our study. Massell et al, have reported carditis and subcutaneous nodules in 59% and 12% respectively (9).

Polyarthritis is the most confusing of the major criteria and probably leads to more diagnostic errors than any of the other manifestations. The arthritis of acute rheumatic fever is exquisitely tender. It is not uncommon for children with this form of arthritis to allow even bed sheets or clothing to cover an affected joints. The joints are red, warm and swollen. It affected mostly elbows, knees, ankles and wrists. In many patients with early arthritis of rheumatic fever because of treatment with anti-inflammatory drugs the classical migratory polyarthritis does not develop thus confusing the diagnosis. In our series there were at least three such patients who had joint pains in one of major joints but had disappeared after taking some anti-inflammatory drugs outside. But we could diagnose these patients because all three at admission had developed carditis.

In majority of our patients with arthritis two or more joints were affected. These were definite objective findings, pointing towards involvement of joints. Further more nearly all the patients with joint pain due to acute rheumatic activity had fever.

About 30 patients in our study group had arthritis and out of them 22 patients (73%) were associated with carditis whereas in 2 patients it was associated with subcutaneous nodules. In none of the patients did arthritis occur simultaneously with chorea. Joint involvement was polyarticular in all. The joints involved in order of frequency were knee (80%), ankle (60%), wrist (25%), elbow (10%) and hip (6%). Metacarpophalyngeal joint involvement was noted in two patients and there was one instance each where arthritis affected the spine, and temporo-mandibular joint.

Carditis was noted in 76% of our patients. Carditis occurred as an isolated lesion in 13 cases (20%) and was associated with arthritis in 22 cases (44%) and with subcutaneous nodules in 5 cases (10%). It was not associated with chorea in any case. Out of 38 patients of carditis, all had significant murmur. About 76.3% had muffled heart sounds. Pericarditis, cardiomegaly and congestive cardiac failure were seen in 7.8%, 23.5% and 18.8% respectively.

Out of 38 cases who had significant murmurs, 30 (79%) had apical systolic murmur. Carry coombs murmur, which is typical of acute rheumatic fever, was found in 10 cases (31%), whereas two patients (6.2%) had a basal diastolic murmur. The most common and generally the earliest clinical signs of carditis in acute rheumatic fever are significant murmurs. Although rheumatic myocarditis may play a role in the production of murmurs, the presence of significant murmurs usually point to involvement of the valves. The early lesions are always those of regurgitation (insufficiency) and the valves that are practically always involved are the mitral and or aortic valves. In our study 93% had involvement of mitral valves, 24% had involvement aortic valve and only one case (3%) had involvement of tricuspid valve as seen in echocardiography. None of the patients had pulmonary valve involvement. Mitral valve in association with aortic and tricuspid valve was involved in 16% and 3% respectively. Similar observations have been noted by Massell et al (9). They have reported significant murmurs in 51%. Out of these 71% had mitral regurgitation alone, 22% had both mitral as well as aortic regurgitation and 7% had aortic regurgitation alone.

Chorea was observed in patients forming 8% of total rheumatic fever admissions. In all cases it was present as alone without being associated with carditis or arthritis. Out of 4 cases, 3 were females and one was male thus favouring the observations made by previous studies that chorea is more common in females. The usual history was a gradual onset over a week or two of fumbling with pencils, knives and the spilling of drinks. All had asymmetrical, jerky, involuntary, purposeless, incordinate non-repetitive movements accompanied by difficulties of speech. In 2 cases of 4, Jones criteria were not fulfilled in the sense that both these cases had no evidence of recent streptococcal infection as well as these cases had no other minor or major criteria. But still these 2 cases were taken as acute rheumatic fever as suggested by WHO, that chorea alone without evidence of streptococcal

infection can be taken as diagnostic of acute rheumatic fever. The incidence of chorea had been reported to be between 3 to 19% in various other studies (9-13).

We found subcutaneous nodules in 10% i.e. 5 cases. In all five they were associated with carditis, whereas in two cases they were associated with arthritis. Similar observations have been made by Massell et al (9). Sites of location of subcutaneous nodules were extensor surfaces of wrist, elbows and ankles. We looked for them over spine also but could not find any. All these nodules were noted on clinical examination and were not noticed by the patients or their attendants.

No case of erythema marginatum was seen in present study. It is similar to the study of Benakappa et al (12). Massell et al, from Boston have reported 7.1% cases (9). So the incidence of erythema marginatum is very low here as well as in India. Erythema marginatum is a evanescent, pink or faintly red, circinate, non itching rash which occurs on various parts of body. One possible reason of its low incidence could be dark skin of patients in this part, which makes it difficult to detect.

ESR was found to be raised in 62% cases in the present study (once chorea was excluded) an incidence similar to that of Saxena et al, (14). This is low as compared to observations made by Benkappa et al, who could demonstrate raised ESR in 100%(12).

C. reactive protein, another acute phase reactant, was positive in 86% of our patients. CRP is neither specific nor does it indicate accurately the degree of severity. It is an sensitive indicator of inflammation. Absence of CRP is strongly against the diagnosis of acute rheumatic fever (2). Presence of CRP however is not diagnostic since it becomes positive in many respiratory infections.

Leucocytosis, another acute phase reactant was present in 32% cases. The count of more than 10,000/ cubic mm were taken as significant. Wilson's figures suggest that the greatest degree of leukcocytosis occurs with the acute carditis while joints pains show a lower range (5). It is important to remember that the leukocytosis count will remain elevated if the patient is being treated with steroids. Under these circumstances it therefore loses its value as an index of activity.

29 cases (58%) in our study group had prolongation of PR interval. All 29 cases had carditis. There were 9 cases of carditis in whom PR interval was normal.

ASO titers were raised in 96% patients in our study group. ASO titers of more than 200 I.U were taken as significant. Only two patients (4%) had insignificant ASO titers of less than 200 I.U. Both these patients had chorea in which increased ASO titers were not a must for diagnosis. 36% patients had ASO. titers above 400 IU. Massell et al in their series found significant ASO titers in 82%(9) Robinson et al reports it to be 89%(8). ASO titers while of considerable value for diagnosis show no clear relationship to imflammatory activity of the disease. It is very important to have serial estimation of ASO titers. Rising titers are of more significance for recent evidence.

Echocardiography is useful in evaluating patients suspected of having rheumatic fever(3). Echocardiogram of 36 patients revealed that 72.2% had valvulitis, 5.5% had pericardial effusion. Mitral, aortic, tricuspid regurgitation were recorded in 66.6%, 13.8% and 2.7% respectively.

The incidence of rheumatic fever in hospitalised children is low and carditis and arthritis are the most common manifestations. Though some studies in recent past have pointed an altered clinical profile over the last few years, our study has not shown any convincing evidence of such change.

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