Juvenile Idiopathic Arthritis

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This study attempts to determine the clinical manifestations, severity and immunological features of JIA and its influence on growth and associated cardiac involvement in children below 16 years of age. This is a cross sectional study in a tertiary referral center on 50 consecutive children below the age of 16 years. Each patient was thoroughly examined and scored on Juvenile Arthritis Functional Assessment Scale. Relevant blood tests, cardiac and ophthalmic evaluation was done. Growth patterns were noted. There was an overall equal sex ratio, though there was a male preponderance in the systemic and oligoarthritis groups. Disturbance of growth frequently occurred in children suffering from JIA. Cardiac involvement should be looked for in cases of JIA. Significant number of cases of PSRA is diagnosed in children presenting with chronic arthritis.

Keywords: *JIA, PSRA, Uveitis.*

JUVENILE idiopathic arthritis (JIA) is the most common cause of chronic arthritis and represents up to 65% of arthritic disease in children(1). Various epidemiological studies of juvenile chronic arthritis (JIA) report divergent results owing to differences in diagnostic criteria, patient retrieval, and study designs(2-5).

It should be noted that a spectrum of reactive post-streptococcal diseases may also become manifest secondary to beta hemolytic streptococcal infection(6) in this age group, such as acute rheumatic fever (ARF) and poststroptococcal reactive arthritis (PSRA).

In adult rheumatoid arthritis cardiac involvements are found in 40% to 70% patients with rheumatoid arthritis(7). Valvular heart disease is less reported in juvenile idiopathic arthritis. The aortic valve is most commonly involved(8). Some authors have recommended cardiac follow-up in all patients of JIA(8).

Data of juvenile arthritis is not available from eastern India. This study is an attempt towards this, using a clinic-based approach.

Subjects and Methods

Patients were enrolled from the Rheumatology clinic at a tertiary referral center in eastern India from November 2003 to October 2004 and classified as per International League Against Rheumatism(9) criteria.

Inclusion criteria

Patients under the age of 16 years who present with complaints of joint pain, swelling and/or stiffness for a period of at least 6 weeks(9).

Exclusion criteria

Patients whose complaints are due to an obvious local cause, who are suffering from non rheumatoid disorders but presenting with joint related symptoms (neoplastic diseases,

sickle cell anemia, hemophilia), and patients of rheumatic fever.

PSRA may be classified as "Other arthritis" under that ILAR classification. These patients fit the inclusion criteria; having arthritis of more than six weeks.

After detailed history patients were put to thorough musculoskeletal, eye and cardiac examina-tions, and DAS28(11) (Disease Activity Score) and Juvenile Arthritis Functional Assessment Scale (JAFAS)(10) were noted.

Besides routine investigations, serum Rheumatoid Factor (RF) by latex agglutination and Antinuclear Factor by Immuno-fluorescence method was measured. Antistreptolysin O titers at diagnosis by latex agglutination were measured. Echocardiography with Doppler study and slit lamp examination of eyes were done in all patients.

Standard step down therapy was offered to all patients of juvenile idiopathic arthritis, along with physical therapy. Patients of PSRA(14) received penicillin prophylaxis.

Results

Of 50 patients, diagnosed as Juvenile Idiopathic Arthritis by the ILAR classification, 22% (11 patients) presented with systemic onset disease, 18%(9) with PSRA, 28%(14) with oligoarthritis, 10% (5) with RF positive polyarthritis, 20%(10) with RF negative polyarthritis and 2%(1) with enthesitis associated arthritis. There is a female preponderance in the polyarthritis group; both RF positive (all 5 patients were female) and RF negative (8 of 10 female; 80%). The oligoarthritis group had a male preponderance (10 of 14 male; 71.4%), as the group of systemic onset patients (7 of 11 male; 63.3%). Earliest disease onset was seen in male patients in systemic onset disease (7 years). Overall, the boys were affected earlier in systemic disease, PSRA, and RF negative arthritis, while in oligoarticular disease; the mean age of onset was similar.

All patients of systemic onset disease had fever, ranging from moderate to high grade, mostly of the classical quotidian variety. Low-grade fever was present in almost all subgroups. Three patients each of PSRA, RF negative arthritis, and oligoarthritis had no fever.

The mean American Rheumatology Association (ARA) functional class at onset was highest in the systemic onset disease with males having the more severe disease (3.6 for males, 2.8 for females). The patients of polyarthritis subtype and oligoarthritis had similar ARA scores at onset (2.0-3.0). The PSRA patients tended to have less severe disease.

The total joint scores varied according to the age of onset, and the type of disease. They were highest in systemic and polyarticular subtypes.

Sixty six per cent patients of JIA were below the 3rd percentile of height for age (CDC 2000 standard). This was distributed throughout the subtypes, commonest with oligoarthritis and systemic disease. However, no control was used.

A classical maculopapular rash, waxing with the onset of high fever, was seen in 2 patients with systemic onset disease. Two patients of the same subtype had a vasculitic rash. Palpable purpura was present in 1 patient of oligoarthritis and 1 patient of RhF negative polyarthritis.

One patient each in oligoarticular, systemic arthritis and enthesitis associated arthritis groups had uveitis by slit lamp examination (6%). None of them was symptomatic.

Rheumatoid factor was positive in 16% of

all patients, including 1 patient of PSRA It was not correlated with severity of disease.

ANF was positive in low titer (1:40) in 1 patient of oligoarticular disease, 2 of patients of RF positive polyarthritis and 1 patient of PSRA (8%). It was not correlated with severity of disease or presence of uveitis.

In echocardiography, cusp thickening was noted in 2 patients of PSRA, 1 patient of RF positive arthritis and 1 patient of RF negative polyarthritis. Mild mitral regurgitation (grade 1) was noted in 1 patient of systemic onset disease, 1 patient of oligoarthritis, 1 patient of Rh. Factor negative polyarthritis and 2 patients of PSRA. None of the patients had cardiovascular symptoms.

Discussion

In this study, 22% (11 patients) presented with systemic onset disease, 18% (9) with PSRA, 28% (14) with oligoarthritis, 10% (5) with RF positive polyarthritis, 20% (10) with RF negative polyarthritis and 2% (1) with enthesitis associated arthritis. PSRA constituted a large number of cases in this study. In a population-based survey in Germany, 294 of 457 cases had a para/postinfectious etiology(13). There has been no major population or clinic based estimate of PSRA among the patients of juvenile arthritis; this study suggests that it may form a significant category of patients.

There was an overall equal sex ratio. There was a female preponderance in the polyarthritis group, especially in rheumatoid factor positive polyarthritis, where all five patients were female. There was a male preponderance in the systemic and oligoarthritis groups. This is consistent with the prevalence patterns from previous Indian studies.

In an Indian study(4), the mean age of onset was earliest in systemic arthritis (5.2 years), followed by oligoarticular (6.8 years) and polyarticular (7.2 years) subtypes. The overall onset of disease in our study tended to peak in the later age groups. The mean age of onset was lowest in male patients of systemic arthritis-7 years. Patients with oligoarticular subtype had a later onset at the age of 10 to 11 years. Male patients were affected earlier in systemic disease, PSRA, and RF negative arthritis, while in oligoarticular disease; the mean age of onset was similar.

In this study, there was a significant reduction in height-for-age in all subtypes of the disease. Sixtysix per cent of patients were below the third percentile for height-for-age in patients up to 16 years of age. Twentytwo per cent of the population below 12 years is below the third percentile of height-for-age in India.

In 16% of patients, RF was positive; 2 patients of systemic arthritis, 1 patient of PSRA and all 5 patients with polyarthritis arthritis. The RF was present in 15% of polyarticular JIA, 7 and 9% of patients with pauciarticular and systemic subtypes respectively in a study in Delhi(4), 9% in the population-based study in Sweden(12).

One of major differences between Indian and Western studies of juvenile arthritis has been the incidence of ANF positivity. In studies done in Delhi(4) and Chennai(17), 3 of 66 patients and 5 of 44 patients were found to be ANA positive respectively. In comparison, 30% in Sweden(12), and 40% patients in Puerto Rico(15) were found to be ANA positive. In this study, ANA was positive in low titer in 3 patients.

Both ANA and RF were not correlated with severity of disease or presence of uveitis.

The presence of uveitis has also been found to be rare in the Indian population. In

Key Messages

- PSRA should be differentiated from other Juvenile idiopathic arthritis cases, because management and prognosis differs.
- Disturbance of growth frequently occurs in children suffering from juvenile idiopathic arthritis.
- Cardiac involvement should be searched for in cases of JIA.

our study, one patient each in oligoarticular, systemic arthritis and enthesitis associated arthritis groups had uveitis by slit lamp examination (6%). However, 12 out of 50 (24%) patients had complaints of redness of eyes in the beginning of the illness especially among the patients with systemic onset.

Heart disease is a rare complication of juvenile rheumatoid arthritis (JIA). None of the patients had any cardiovascular symptoms apart from occasional palpitations or breathlessness unrelated to any organic cardiac disease. On echocardiography, cusp thickening was noted in 4 patients (2 of PSRA). Mild mitral regurgitation (grade 1) was noted in 5 patients (2 of PSRA). The findings were not diagnostic of rheumatic carditis in any case. In the absence of a control group, these findings cannot be taken as significant.

Contributors: AG provided the concept and design for the study; and along with BG, edited the manuscript. DS and BG were involved in literature search, data acquisition, manuscript preparation. Statistical analysis was performed by DS and SH. SH contributed to studies and data acquisition. All authors stand guarantor to the study.

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