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Pattern of Juvenile Rheumatoid Arthritis Seen in 91 Patients, Presenting to An Urban Rheumatology Clinic in Pakistan

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ABSTRACT

Objective: To determine various types of Juvenile rheumatoid arthritis (JRA) seen in patients visiting our rheumatology clinic. **Materials and methods**: A retrospective review of case records of 91 patients ages <= 16 yrs, who satisfied American College of Rheumatology (ACR) criteria for Juvenile rheumatoid arthritis was done. These patients visited rheumatology clinic from April 2002 to Jun 2004. **Results:** Total patients were 91. Females were 49(54%) and males were 42(46%). There was an overall female predominance except for pauciarticular type where males were more than females 24(60%) vs. 16(40%) p value<0.05. Mean age of onset was 10.7 yrs± 04. Polyarticular sub-type was the commonest pattern seen in 51.6% of cases while 44% of the patients had pauciarticular disease. Systemic JRA was seen in 4.4%. Rheumatoid factor was positive in 48.4% cases with Polyarticular disease and in this sub-group of seropositive polyarticular patients 88% were females. **Conclusion:** Pattern of JRA in this case series was quite different in comparison to data generated from western countries. Mean age of onset was much higher than quoted in literature. Polyarticular sub-type was the commonest pattern seen in our patients, where as pauciarticular is more prevalent in the west. Rheumatoid factor was positive in 48% of the patients with polyarticular disease in contrast to 5-10 % seen in western data. As sero positive polyarticular JRA is associated with more aggressive disease and disability so these patients have to be identified earlier and need aggressive treatment with disease modifying agents.

Key words: Arthritis, Juvenile, Pakistan.

INTRODUCTION

Juvenile rheumatoid arthritis (JRA) is the commonest form of childhood arthritis occurring in children younger than 16 years. Etiology of JRA is un-known and it has got different clinical sub-types. The prevalence of JRA has been estimated to be between 57 and 113 per 100,000 children younger than 16 years in United States[1,2]. JRA affects approximately the same number of children as juvenile diabetes and at least ten times as many as hemophilia and acute lymphocytic leukemia[3].

JRA has been extensively studied in the Western population, but to our knowledge there is no local data about the disease profile.

Different Classification systems have been developed so for and are still being evolved[4]. American College of Rheumatology (ACR) classification is the most widely accepted classification. According to this classification JRA has been divided into three different types polyarticular, pauciarticular and systemic.

These three sub-types demonstrate unique clinical presentations, immunogenetic associations and clinical course. These sub-types can be further classified. International League against Rheumatism (ILAR) and European League against Rheumatism (EULAR) classifications are being used widely. However for the next several years all classification systems will continue to appear in the literature. We used the ACR classification to categorize our patients

METHODS

This was a retrospective study conducted in the rheumatology clinics of Fatima Memorial Hospital and National Hospital Lahore, Pakistan. Case records of all patients coming to us with diagnosis of JRA from April 2002 to June 2004 were reviewed. The attending rheumatologist used following criteria to establish the diagnosis of JRA

1. Onset of disease less than 16 years of age
2. Evidence of arthritis in one or more joints for at least 6 weeks.
3. Exclusion of other possible causes of arthritis.

Patients were assigned JRA sub-type based on following criteria[5].

a) Polyarticular JRA
These had evidence of arthritis in 5 or more joints without systemic features. They were further divided into two sub-sets depending upon their Rheumatoid factor status.

b) Pauciarticular JRA
Those having evidence of arthritis in four or fewer joints and without systemic features.

c) Systemic JRA
Those having arthritis along with daily intermittent fever up to 103°f or more with or without rash or other organ involvement.

Data of all patients was recorded on a flow sheet. Following variables were included:-

1) Age of onset
2) Sex
3) JRA sub-type
4) RA-factor status in polyarticular sub-type

Chi square test was used to find out the level of significance for the observed difference between the two sexes, within the same JRA subtype.

RESULTS

Total patients were 91. Females were 49 (54%) and males were 42 (46%) p value>0.05. There was an overall female predominance except for pauciarticular type where males were more than females 24(60%) vs. 16 (40%) p value<0.05. Mean age of onset was 10.7±04 yrs. Polyarticular sub-type was the commonest pattern seen in 51.6% of cases while 44% of the patients had pauciarticular disease. Systemic JRA was seen in 4 (4.4%). Rheumatoid factor was positive in 48.4% cases with Polyarticular disease and in this sub-group of seropositive polyarticular patients 88% were females (Table 1).

DISCUSSION

In this study the main focus was on identifying the profile of JRA in terms of age of onset, sex of patient and JRA sub-type. Both age and sex are very important predictors of long-term outcome[6]. Female sex and early age at onset both are associated with poor functional outcome. Similarly outcome also depends on JRA sub-type. Polyarticular and systemic JRA are associated with a worse outcome in terms of functional impairment, delayed growth and disability[7].

Mean age of onset in this case series was 10.76 years. This was much higher than quoted in different western studies conducted on Caucasian population. Anderson et al from Sweden have reported a peak age of onset around 4years[8]. Dracut et al from Greece showed that there were two peak ages of onset, one between 2 years to 5 years and other between 9 years to 12 years[9]. But the studies conducted on blacks showed age of onset around 11.8 years[10]. This is closer to our finding of 10.76 years.

Another important finding in this study was regarding the sex distribution of patients. Inspite of overall female predominance, it was observed that males were more prevalent in pauciarticular disease sub-type. A study from Singapore parallels our finding of male predominance in pauciarticular sub-type[11]. Similar results have been reported in studies involving black and South African Indian with JRA[12].

Regarding JRA sub-type, a polyarticular pattern was the commonest pattern seen. In most of the Western countries including North-America, a pauciarticular pattern is more prevalent. Khuffash et

al.[13] from Kuwait conducted a retrospective study

Table 1: Distribution of Patients According to JRA Sub-type and Sex (n = 91)

JRA Sub-type	Total Number		Male		Female		P value
	No.	%	No.	%	No.	%	
Polyarticular	47	51.6	16	35.7	31	64.3	P<0.01
Pauciarticular	40	44.4	24	60.0	16	40.0	P<0.05
System	4	4.4	2	50.0	2	50.0	
Total	91		42		49	100.0	

like ours. They reported polyarticular pattern in (42%) of the cases and systemic in 2.9 of cases. Rests were pauciarticular[13]. Surprisingly systemic disease was hardly seen in our study. In a population based study from Sweden the prevalence of systemic JRA was only (6.6%) of total cases[14]. In a referral based pilot study in Costo Rica not even a single case of systemic JRA was reported[15]. Both these results are similar to our. Aggarval et al.[16] from India conducted a retrospective study on 214 patients. They found polyarticular disease in (43.4%) and pauciarticular in (35.5%). However, systemic disease was seen in (21%) of cases which is much higher than our finding[16]. In another study from India polyarticular type was the most common, followed by pauciarticular and systemic types[17].

Positive Rheumatoid factor status in polyarticular disease is associated with prolonged disease activity, joint erosions, deformities and poor functional outcome[18]. In this study Rheumatoid factor was positive in a large proportion of patients (48.4%) with polyarticular disease. 88% of those who had positive RA factor in this subset were females. Western literature quotes 5-10% patients with polyarticular disease having positive rheumatoid factor. The prevalence of polyarticular disease with sero-positivity had been shown to be very high in Black, Indian & South African children[12]. As most of our patients came to us, after six months of onset, so their disease pattern had already been established. As early onset pauciarticular JRA may extend later during course of the illness, into polyarticular disease this might be one reason why polyarticular disease was more commonly seen in this study[19].

Finally, divergent results seen might be because of inconsistencies in classification systems, racial, ethnic & environmental differences.

CONCLUSION

The pattern of JRA was quite different in various respects in this case series as compared with data from the Western Countries. The age of onset is much higher. There was an overall female predominance; however, males were significantly more in pauciarticular sub-type. Polyarticular disease was more frequent than other JRA sub-types. Systemic JRA was the least common. Rheumatoid factor was positive in almost half of the patients with polyarticular disease and in this subset (88%) were females. These are the patients who require aggressive treatment to prevent disability and improve functional outcome.

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