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Research Article

CLINICAL MANIFESTATIONS OF JUVENILE IDIOPATHIC ARTHRITIS IN TAMILNADU

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ABSTRACT

Objective: The objective was to study the functional disability, and clinical manifestations in children diagnosed with juvenile idiopathic arthritis (JIA) in Bhavani town, Tamil Nadu. JIA is a mixed group of disorders among various populations with various clinical manifestations. We have carried out a study herein the data presented.

Methods: Patient's diagnosis of arthritis with an age group of 10-15 years presenting to tertiary care hospital, Bhavani town, Tamil Nadu for a period of 4 months in the year of 2013-2014. A retrospective study have been conducted and reclassified as JIA based on the International League of Association for Rheumatology (ILAR) JIA diagnostic criteria.

Results: Totally 45 patients with arthritis at age of 10-15 years were evaluated during a period of 6 months at the hospital. These patients were further analyzed by ILAR JIA criteria. Average age of disease onset among 15 patients (33.33%) were 6 years with an average age at first visit to the hospital being 7 years and with a male to female ratio of 2:4. Polyarticular rheumatoid factor (RF) negative JIA, at 44.5% was the most frequent type of chronic arthritis encountered. 4.5% polyarticular RF was positive and JIA. 17.7% diagnoses with psoriatic arthritis among the population.

Conclusion: Polyarticular RF negative which was the subtype in JIA was found to be high at Bhavani town, Tamil Nadu. There is a need to elucidate environmental factors of JIA in this region.

Keywords: Juvenile idiopathic arthritis, Bhavani town, Clinical manifestations, International League of Association for Rheumatology, Rheumatoid factor.

INTRODUCTION

Juvenile rheumatoid arthritis (JRA) is the most prevalent rheumatic diagnosis among the children. This term represents a group of diseases that have in common chronic idiopathic inflammation of one or more joints [1]. Substantial evidence points to an autoimmune pathogenesis, including the observed immunohistopathology, the presence of autoantibodies, and the presence of T-cell clonality primarily seen in the joints [2].

Juvenile idiopathic arthritis (JIA) is a generic term for arthritis with unknown etiology and with the onset occurring before the age of 16 years [3]. In 1995, a new classification criteria for JIA, the so-called International League of Association for Rheumatology (ILAR) criteria were proposed [4]. As JIA is a disease with serious functional implications, a descriptive study would not be complete without reviewing both the functional and clinical parameters of the disease [5].

There are five major types of JRA

- Pauciarticular JRA - Affects four or fewer joints
- Polyarticular JRA - Affects five or more joints
- Systemic onset JRA (also called still's disease) - Affects the entire body; least common type of JRA
- Enthesitis associated arthritis-occurs with inflammation of the tendon at the bone
- Psoriatic arthritis-associated with a skin disease called psoriasis.

Risk factors

There are no clear risk factors for JRA. However, in general:

- Pauciarticular JRA
  - The first subtype typically affects girls under the age of 7 years
  - The other subtype typically affects boys over the age of 8 years.
- Polyarticular JRA usually affects girls more often than boys
- Systemic onset JRA affects boys and girls equally
- For enthesitis associated arthritis, risk factors include a family history of:
  - Anterior uveitis with eye pain
  - Inflammatory back arthritis
  - Inflammatory bowel disease.
- Psoriatic arthritis risk factor:
  - Arthritis and a positive family history of psoriasis in a first-degree relative.

Symptoms

The major symptoms of JRA are as follows:

- Joint stiffness, especially in the morning or after periods of rest pain, swelling, tenderness, or weakness in the joints
- Fever
- Weight loss
- Fatigue or irritability
- Eye inflammation
- Swollen lymph nodes
- Growth problems, such as:
  - Growth in affected joints may be too fast or too slow, causing one leg or arm to be longer than the other
  - Joints grow unevenly, off to one side
  - Overall growth may be slowed [6,7].

Background

Juvenile arthritis was a heterogeneous group of disorder seen in children of <15 years of age group, but its cause was unknown. It is a disorder that was based on genetical characterization [8-18].

Difficulties in classifying the type of JIA due to late diagnosis, irregular treatment, and limited diagnosis procedures.

If the JIA was not treated, then it may lead to fibromyalgia in elder children, and it is quite difficult to differentiate JIA and leukemia. In such cases, a bone marrow aspirates to be checked and tested. There is no certain confirmatory test to distinguish JIA. Antinuclear antibody (ANA) test can be of value when clinical findings strongly suggest JRA, as the test is positive in 60-80% of JRA patients, particularly those with pauciarticular disease. ANAs have a high prevalence (65-85%) in children with pauci JRA and chronic uveitis. test for identifying which children are at increased risk for developing chronic anterior uveitis. On the other hand, the rheumatoid factor (RF) test, often used to corroborate a diagnosis of adult rheumatoid arthritis, is positive in only 15-20% of children with JRA. It is more frequently seen in children with later onset polyarthritis and is associated with a more aggressive disease course. Between 40% and 50% of JRA patients have disease that persists into adulthood, and more than 30% have significant functional limitations after 10 or more years of follow-up. It is estimated that blindness occurs in as many as 15-30% of cases.

Criteria for polyarticular JRA

- Persistent arthritis in one or more joints for at least 6 weeks
- Arthritis in five or more joints over the first 6 months of disease
- Exclusion of other forms of arthritis including systemic JRA
- Uveitis (uncommon)- 5% of patients.

Polymorphic markers in JRA

- Immunoglobulin A deficiency
- Complement deficiency
- α1-antitrypsin
- Amyloid P component
- Interleukin 5-1a promoter
- Tumor necrosis factor a/b
- T cell receptor Vb6.1 null gene
- Interleukin-6 promoter
- Interleukin-10
- Chromosome [19,20].

METHODS

The record of patients who were diagnosis with JIA was collected. Patients who diagnosis at primary age that is., before 15 years of age or children suffering from joint pains continuously for a period of 4-6 months getting treatment in a tertiary care hospital, Bhavani town during the period of 2013-2014. The data's were collected from both the out-patient and in-patient departments for a period of 4 months from 2013 to 2014. The patient was diagnosed prospectively using the ILAR classification. European Union League of Association or rheumatology system was used to classify the patient into onset sub-types as follows:

1. Pauciarticular onset in which four or fewer joints involved
2. Polyarticular onset in which five or more joints affected
3. Systemic onset in which arthritis associated with spiking fever for at least 2 weeks with or without a typical rash.

This retrospective study of cases involves reclassifying the every patient with ILAR criteria and compiling respective clinical data of each patient. All the patient data such as clinical, hematological, immunological, radiological data's were collected. Classifying the patient into sub-type was based on the presence and absence of the RF and history of articular diseases. Thus, patients were categorized as systemic arthritis, oligoarthritis, polyarthritis (RF negative), polyarthritis (RF positive), psoriatic arthritis, enthesitis related arthritis. Oligoarthritis cannot be differentiated because it needs continuous observation which was not possible with the retrospective study, and some are due to lack of follow-up. Patients with clinical manifestations of acute rheumatic fever, septic arthritis, systemic lupus erythematous, dermatomyositis, cancer, human immune deficiency virus type-1 were taken and included in the study. The study was conducted with the approval of research Ethical Committee [21].

RESULTS

Prevalence of JIA

In total 45 patients with chronic arthritis onset at the age of 1-15 years were seen at Bhavani town hospital undergoing a treatment from 3 years onwards shown in Table 1. Of these 45 patients, 15 were male and 30 were female. The average age of the disease onset was 6.5 years. Age of first visit to the hospital was 10.7 years. In the total 50 patient record some were having incomplete data, and few did not accept to expose their data due to the inconvenience. Polyarticular RF negative JA at 20 (44.5%) was more frequent type. Polyarticular RF positive was 2 (4.5%), psorotic arthritis 8 (17.7%). Polyarticular disease with the limb involvement was similar and in those with the symmetrical arthritis, mainly wrists part was getting affected. The pattern of joint involvement was asymmetric with oligoarticular disease and symmetric in polyarticular and systemic onset diseases. Predominant distribution of the RF shows high in the females than the males. The mostly seen in subtype of the JIA was polyarticular RF (-), psoriatic arthritis, and then the polyarticular RF (+).

JIA clinical manifestations

As most of the pediatrics will be having the joint injuries due to falls, sprains it is not possible to assess the JIA that is rare in children's. It is helpful to remember that joint pain alone in case of absence of swelling, stiffness, tenderness, which was never sufficient to diagnosis the JIA. If the joint pain extends for more than 6 weeks that the child have JIA, which can be further confirmed by conducting tests such as laboratory tests and the ANA test.

Common features are shown in Table 2 are fever, chills, conjunctivitis, increase in erythrocyte sedimentation rate, anemia. Most of the cases the hemoglobin level has been dropped to below 50%. Fever persists for a period of at-least for a period of 2 weeks for few patients, intermittent for few, evening raise in temperature for few patients were noticed. The patients were hospitalized for an average duration of 1-2 weeks. The elevated ESR level indicates the severity of the disease and also the presence of infection. However, in the most of the records the ESR level was not available only 10-15 records have the information about the ESR

Table 1: Details regarding JIA patients presenting to tertiary care hospital Bhavani town, Tamil Nadu

JIA type	Number of patients (%)	Male (%)	Female (%)	Age at onset	Age at diagnosis
Total	45 (100)	15 (33.3)	30 (66.6)	01-15	4-17
Polyarticular RF (-) JIA	20 (44.5)	08 (17.8)	12 (26.6)	01-11	2-13
Polyarticular RF (+) JIA	10 (22.2)	02 (4.4)	08 (17.8)	01-12	7-18
Psoriatic arthritis JIA	15 (33.3)	05 (11.1)	10 (22.2)	01-14	3-10

JIA: Juvenile idiopathic arthritis, RF: Rheumatoid factor

Table 2: Clinical manifestations in the various JIA subtypes presenting to tertiary care hospital Bhavani town, Tamil Nadu

Clinical	Polyarticular RF (-) JIA (N=20) (%)	Polyarticular RF (+) JIA (N=10) (%)	Psoriatic arthritis JIA (N=15) (%)	Total (%)
Fever	20 (100)	04 (40)	10 (66.6)	34 (75.5)
Conjunctivitis	03 (15)	00	06 (40)	09 (20)
Chills	20 (100)	04 (40)	10 (66.6)	34 (75.5)
Headache	10 (50)	03 (30)	00	13 (28.8)
HB	10.6	12.4	11.8	11
ESR	58±28	43±20.6	38±28	40±20

Hb: Hemoglobin, ESR: Erythrocyte sedimentation rate

level and the severity of the disease. The record that shows the ESR was only for those patients who were diagnosed at the age of 13-15 years. Some patients are having changes in nails and altered appetite.

Joints involved in the disease such as knee (50%), wrist (10%), elbow (30%), ankle (48%), fingers (58%), and toes (26%).

The hospital provided with non-steroidal anti-inflammatory drugs, disease modifying anti-rheumatic drugs such a methotrexate, leflunomide. Biologics such as etanercept, infliximab, adalimumab, abatacept, anakinraand tocilizumab. Corticosteroid such as prednisolone at low dose that was mainly and commonly used drug to treat JIA. However, in the clinical settings there are certain regimens that should be used up regularly without missing, whereas in the case of these patients the follow-up was missing, which leads to progression of the disease (Fig. 1).

The patient over this area seems to be more prone to the JIA disease, but exact reason was unknown. Record shows that the cause of the disease would be genetic, geographical reason, lifestyle management, and dietary habits. Most of the patients experienced the growth problems such as too fast growth or too slow growth. Reduced joint movement, swelling of both hands and feet, fever with chills that lasts for 1-2 weeks and the temperature within the range of 102°F and conjuctivitis.

DISCUSSION

In the past, the diversity of the disease, the lack of diagnostic tests and the differences in diagnostic criteria may have made it difficult to understand fundamental epidemiological comparisons such as incidence, prevalence and clinical manifestations. The current ILAR JIA classification is contributing to a more uniform nomenclature and thus improving comparative disease diagnosis and epidemiology across countries and ethnic populations. For this reason, we chose to adopt the ILAR classification criteria for this study. We realize that there is an inherent risk of error arising from a retrospective reclassification process for the patients during the period prior to the formulation and adoption of the ILAR criteria. However, we are confident of the validity of this process because of the meticulous, detailed prospective recordings and observed during this era.

About one-third of Caucasian children with juvenile arthritis that during the first 6 months affects less than four joints will continue to develop arthritis in more joints thereafter [22]. It is possible therefore that some children classified as polyarticular may in reality be "oligoarthritis extended."

The gender ratios in our study show a paucity of pre-school girls, compared to Caucasian studies where a ratio of about 3:2 is the norm. Social and cultural rather than biological reasons may lie behind this observation. It is highly likely that so-called "milder" cases of oligoarticular disease might never reach a tertiary care facility in many developing world settings. In the developed world children with oligoarthritis will usually be reviewed in a hospital setting and have access to diagnostic and therapeutic facilities. It is apparent that differences in prevalence of oligoarthritis JIA in India may simply be the result of a selection bias imposed by a dearth pediatric rheumatology services and expertise. In this context, it is of interest that in true community-based studies in the developing world the prevalence of oligo-articular disease matches or exceeds that of polyarticular disease [23-25].

Fig. 1: Right ankle joint deformity in polyarticular arthritis

The extra-articular features were as expected, apart from the poor ophthalmic outcomes in those with chronic uveitis. As rheumatology knowledge increases amongst doctors and other care providers in India, leading to the application of standard diagnostic and classification criteria, prevalent cases are likely to continue to resemble those reported elsewhere. Clinicians are working in parts of India where rheumatological services are non-existent or rudimentary face enormous challenges. Distinguishing common rheumatic fever from rarer JIA subtypes is one example, and unless clinicians are well-trained to recognize the distinctive features of the two conditions, they will have doubts about diagnosing JIA. In our setting, JIA may often be attributed to trauma or thought to be infective, and children with persistent or more severe joint symptoms may be subjected to arthrotomy and prolonged courses of antibiotics. Therefore, increasing awareness of JIA among clinicians should lead to improvements in reporting and adherence to the standard diagnostic criteria of the disease.

CONCLUSION

Almost all types of JIA among children in the age group of 1-15 years were identified in our Bhavani town, Tamil Nadu. Polyarticular RF negative disease was the most common presentation as per the study result. The females with polyarticular rheumatoid disease are underrepresented in this part that was predominant distribution of RF in females' shows higher levels in the males. Therefore by increasing the awareness among the people and clinicians in this area the JIA and its subtypes may be treated well completely and satisfactorily.

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