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ORIGINAL ARTICLE

# Atypical Articular Presentations in Indian Children with Rheumatic Fever

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**Background** The objective of the study was to describe the clinical characteristics of atypical articular presentations during the initial outbreak and recurrence in patients with acute rheumatic fever (ARF) in the paediatric age group.

**Methods** This was a retrospective, observational study conducted between January 2012 and December 2014 on all suspected cases of acute rheumatic fever (ARF) fulfilling either WHO 2004 or Australian guidelines with atypical articular manifestations ie, presence of at least one of the following features: duration of symptoms more than three weeks; monoarthritis/arthralgia; involvement of small joints of hand and feet and/or cervical spine and/or hip joint; and, not responding to salicylates in one week.

**Results** 'Atypical' pattern was present in 63% (39/62) of patients with articular manifestations, of which arthralgia was a common manifestation (57%). Polyarticular afflictions were predominately non-migratory (additive) in both atypical (74%; 29/39) and typical (82%; 18/23) groups. Monoarticular (33%) affliction of the joints constituted a significant disease manifestation. Time from onset to diagnosis was >3 weeks in 79% of patients while small joints involvement and axial joint involvement occurred in half of the cases (51%). Inadequate response to NSAIDs was found in three (7%) cases.

**Conclusion** Atypical manifestations in ARF may well be mistaken for a connective tissue disorder, post streptococcal reactive arthritis and septic arthritis. Physicians should be made aware of these features to prevent diagnostic dilemma, and to effect institution of appropriate management including penicillin prophylaxis

**Keywords** Rheumatic fever • Atypical arthritis • Arthralgia • Jones criteria

## Introduction

Acute rheumatic fever (ARF) is a late, inflammatory, non-suppurative complication of group A beta-haemolytic streptococcal infection of the throat. It affects the heart, joints and the central nervous system, in genetically predisposed children and adolescents [1].

The lack of specific laboratory tests makes the diagnosis of ARF quite problematic. Jones criteria first introduced in 1944, requires the fulfilment of certain ''major and minor'' manifestations and essential criteria to diagnose ARF. The traditional teaching espouses arthritis of ARF as migratory polyarthritis, involving large joints of the lower limb, with its onset around two to three weeks after a streptococcal infection of the throat and typically the intense pain responds well to the use of salicylates. Without treatment, the signs of inflammation normally last from two to five days in each joint but usually not more than three weeks. Arthritis typically heals without sequelae [2].

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In some patients joint involvement may be mono rather than polyarticular, additive rather than migratory, involving small joints of the hands and feet, unusually the axial skeleton, and persisting for weeks if not treated [3,4].

Jones criteria did not address the difficulties in establishing the diagnosis of ARF, especially in the presence of these 'atypical' articular manifestations or where arthritis is the only clinical manifestation of the disease and the other important and more specific criteria (carditis, erythema marginatum) are not present, which may lead to inappropriate under-diagnosis.

The studies in the literature on this form of presentation of arthritis after streptococcal infection are relatively scarce. The objective of the present study is to describe the clinical characteristics of atypical articular presentations during the initial outbreak and recurrence in patients with ARF in the paediatric age group.

## Materials and Methods

We retrospectively analysed medical records of all the first episodes of ARF and recurrence cases in the paediatric age group (5–18 yrs) admitted between January 2012 to December 2014 to the Division of Paediatric Cardiology, Dr RML Hospital New Delhi, fulfilling either Jones criteria, ACC/AHA1992, WHO 2004, or Australian guidelines 2012. Of all the cases, those cases with articular manifestations were singled out and sorted into atypical and typical articular presentations. Children with congenital heart disease, other acquired heart diseases like cardiomyopathies, juvenile rheumatoid arthritis and other connective tissue disorders were excluded. A case was recruited only once in the study either reporting to the hospital as first episode or as recurrence.

The patient's demographic data, clinical history, examination and laboratory reports including erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), into streptolysin O (ASO) titre, anti DNAseB, electrocardiography, echocardiography were collected from the medical records.

Arthralgia was defined as involvement of the joint without swelling whereas arthritis was defined as joint swelling with restriction of movement [1,2]. Monoarthritis meant involvement of a single joint with polyarthritis having several joints involvement.

The atypical pattern of joint involvement involved the presence of at least one of the following features: duration of symptoms more than three weeks, monoarthritis/monoarthralgia, involvement of small joints of hand and feet and/or cervical spine and/or hip joint and not responding to salicylates in one week [5].

Statistical analysis was conducted using SPSS 17.0 (Chicago, IL, USA). Results are expressed as mean ± SD, numbers and percentages. Categorical variables were analysed using either the chi square or Fisher's exact test. For all statistical tests, a p value less than 0.05 was considered significant.

## Results

103 cases of suspected ARF and recurrence were analysed; 54 as first episode of ARF and 49 with recurrence. The male: female ratio was 1.9:1 with a mean age of 11 ± 2.5 years (range 7–18 years). On applying either of the Jones criteria, 71.8% (74) had clinical carditis, 14.6% (19) had subclinical carditis, 60% (62) had joint manifestations, 9.7% (10) had chorea, and 0.9% (1) had subcutaneous nodules while none had erythema marginatum.

Of the 62 patients with articular manifestations, 40 were first episode and 22 with recurrence. Mean age of these patients was 11.1 ± 2.2 years with a male to female ratio of 2:1. Arthralgia (62%, 39/62) was more common than arthritis (37%, 23/62). The atypical pattern of articular manifestations was observed in a considerable number of cases: (63%, 39/62), with again arthralgia being the predominant presentation (arthralgia 57%, 22/39; arthritis 43%, 17/39). Most of the patients presenting with arthritis (23/62) had atypical arthritis (74%, n = 17/23).

In the atypical cases 71% (28) had clinical carditis, 23% (9) subclinical carditis, 2% (1) chorea, and 2% (1) had subcutaneous nodules. A similar distribution of major manifestations was observed in typical arthritis. Fever was present in 73% and 59% of cases with typical and atypical arthritis respectively (Table 1). Ninety-four per cent of patients were regularly on secondary prophylaxis.

Among these 39 cases of atypical joint affliction, isolated arthritis was found in 11 (28%) patients, arthritis/arthralgia associated with carditis was found in 28 cases (71%); arthritis associated with carditis and chorea in 1 case; and arthritis, carditis, and subcutaneous nodules in 1 case.

Of the cases with atypical joint afflictions, involvement of small joints and/or axial skeleton occurred in 51% (20/39), duration of joint symptoms was greater than three weeks in 31 (79%) and with inadequate response to NSAIDs in 3 (7%). Monoarticular involvement constituted one third of the atypical cases (13/39, 33%) of which monoarthritis constituted 13% (5/39) while monoarthralgia was observed in a fifth of cases (8/39, 20%). Monoarthritis was the predominant affliction in the first episode while monoarthralgia was equally observed in the first episode and recurrence.

Polyarticular involvement constituted a less common presentation in the atypical variant (66%) as compared to the typical group (100%) due to the overt presence of monoarticular afflictions in the atypical group. Of the polyarticular affliction, arthralgia was more common than arthritis in both typical group (arthralgia 74%, arthritis 26%) and the atypical variant (arthralgia 54%, arthritis 46%). Polyarticular afflictions were predominately non-migratory (additive) in both atypical (74%; 29/39) and typical (82%; 18/23) cases and in the first episode (75%; 30/40) and recurrence (77%; 17/22) respectively (Table 2).

The majority (84%, n = 22) of the patients had 2–5 joints affected while a minority had more than 6 joints afflicted (two had 6–10 joints while the other two had >11 joints). The most common joint involvement was knee followed by ankle joint.

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