

## Case Report

# Atypical arthritis in a toddler: beware, save the heart

Sharani Boopathy\*, Kanimozhi Thandapani, Arulkumaran Arunagirinathan

Department of Pediatrics, Sri ManakulaVinayagar Medical College and Hospital, Puducherry, India

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**\*Correspondence:**

Dr. Sharani Boopathy,

E-mail: [sharanboopathy@gmail.com](mailto:sharanboopathy@gmail.com)

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### ABSTRACT

Acute rheumatic fever still poses a dreadful threat to pediatric morbidity and mortality. A 2 year old toddler presented to us with non migratory polyarticular joint pain and swelling. General physical and systemic examination was normal. Local examination revealed swelling and tenderness of multiple small and large joints. Further evaluation fulfilled Modified Jones criteria and she was diagnosed to have acute rheumatic fever. Other close differentials were simultaneously ruled out. Child was started on naproxen because of aspirin toxicity. There was significant response for therapy and complete resolution of joint involvement was noted on follow up. We report this case to emphasize the fact that rheumatic fever might have bizarre clinical presentation and may also affect infants and toddlers deviating the typical age of onset. There is a need for earliest possible initiation of adequate management and follow up to prevent permanent cardiac complications. This warrants high index of suspicion even in uncommon age group.

**Keywords:** Atypical arthritis, Children, Rheumatic fever

### INTRODUCTION

Acute rheumatic fever is an autoimmune, multiorgan inflammatory disease that occurs as a result of Group A  $\beta$ -hemolytic streptococcal pharyngitis.<sup>1</sup> About 0.3 - 3% of those who had GAS infection develop acute rheumatic fever, based on genetic predisposition and virulence of the infecting strain.<sup>2</sup> Common primary sites of affliction include heart, joints and central nervous system, of which most important sequel of rheumatic fever is the rheumatic heart disease (RHD) because of its significant residual morbidity and mortality.<sup>3</sup>

Rheumatic fever in children younger than 5 years is extremely uncommon and had widely varied presentations compared to older children. Younger ones were noted to acquire more severe carditis often associated with congestive cardiac failure and those who were followed up had high frequency of recurrence unlike school going children.<sup>4,5</sup> Miserably its

heterogeneous presentation in preschool children provides a greater difficulty in suspicion leading to under diagnosis and progression to rheumatic heart disease, which is preventable. Here we report a 2 year old female toddler who had initial attack of acute rheumatic fever with isolated atypical articular manifestation.

### CASE REPORT

A 2 year old female toddler hailing from Villupuram, Tamil Nadu was hospitalized with complaints of fever on and off for one week, multiple joint pain and swelling for 4 days with difficulty in ambulation for 1 day. Initially she developed left ankle swelling and pain, followed by left knee, with subsequent additive involvement of right ankle and right knee (Figure 1). Eventually involvement of all metacarpophalangeal joints of both hands, all metatarsophalangeal joints of both feet with proximal and distal interphalangeal joints of all four limbs was also noted (Figure 2).



**Figure 1: Bilateral knee joint swelling with resolving bilateral ankle swelling.**



**Figure 2: Additive involvement of all metacarpophalangeal joints of both hands, all metatarsophalangeal joints of both feet with proximal and distal interphalangeal joints of all four limbs.**

There was no history of breathing difficulty, skin lesions, involuntary movements, prolonged fever or any other local or systemic symptoms. Child received acetaminophen, antitussive and an incomplete course of oral amoxicillin for symptoms of upper respiratory tract infection one week prior to the presenting illness. She did not have any past medical illness or significant contact history. She was the first born female child to non-consanguineous parents with no antenatal complications, smooth perinatal transition and uneventful postnatal period. There was no relevant medical history among family members. They belonged to lower middle class socioeconomic group based on modified Kuppusamy classification and resides in a pucca house with adequate ventilation and no overcrowding.

On examination she was non toxic, afebrile, alert, oriented and had moderate wasting. Admission vitals were axillary temperature of 98.4°F, respiratory rate 24 per minute, heart rate 102 per minute, blood pressure

92/56 mmHg measured in right upper arm and 100% oxygen saturation at room air. She had pallor. Ear, nose, throat examination was normal. Local examination revealed swelling and tenderness of left knee joint, both ankle joints with minimal terminally restricted range of movements. She also had swelling of all metatarsal, metacarpal and interphalangeal joints. Systemic examination was normal.

Complete hemogram showed anemia, leucocytosis with neutrophilic predominance. Peripheral smear was normal. Erythrocyte sedimentation rate at half an hour was 100 and 150 at one hour. C-reactive protein was 4.8mg/dl. Electrocardiogram showed prolonged PR interval (0.16sec) for that age. Anti-streptolysin O titre was elevated (400IU/ml). Chest roentgenogram and echocardiogram were normal. Urinalysis was normal. Ophthalmic examination had no abnormality. Evaluation fulfilled one major and two minor Revised Jones criteria of high risk population with supportive evidence of streptococcal infection favoring a diagnosis of acute rheumatic fever (Table 1).<sup>6</sup>

**Table 1: Modified Jones criteria (2015).**

Moderate to high risk population*	
<b>Major criteria</b>	
•	Carditis (clinical or subclinical)
•	Arthritis <ul style="list-style-type: none"> <li>• Monoarthritis or polyarthritis</li> <li>• Polyarthralgia</li> </ul>
•	Chorea
•	Erythema marginatum
•	Subcutaneous nodules
<b>Minor criteria</b>	
•	Monoarthralgia
•	Hyperpyrexia ( $\geq 38.0^{\circ}\text{C}$ )
•	ESR $\geq 30$ mm/h and/or CRP $\geq 3.0$ mg/dl
•	Prolonged PR interval (after taking into account the differences related to age; if there is no carditis as a major criterion)

Initial attack: 2 major / 1 major and 2 minor, + evidence of recent GAS infection.

Child was started on Aspirin at 50mg/kg/day for 12 weeks but was switched over to Naproxen (20mg/kg/day) in view of drug intolerance. Secondary prophylaxis with Benzathine penicillin 0.6million units every 4 weeks till 21 years of age was advised. On follow up, her symptoms resolved and arthritis subsided with no residual deformity.

## DISCUSSION

Acute rheumatic fever still continues to be more prevalent and has huge impact on pediatric morbidity and mortality in developing countries till date though there is evident decline in incidence among developed countries.<sup>7-</sup>

<sup>9</sup> Poor socioeconomic status, undernutrition, overcrowded homes, are proven to be most common risk factors for the same.<sup>10,11</sup> The overall prevalence of acute rheumatic fever in India varies from 0.5 to 11/1000 children of age group 5 to 14 years.<sup>12</sup>

Shah et al observed that in India, children get afflicted even at earlier age compared to developed countries.<sup>13</sup> Tani et al reported 5% (27 children) affected were younger than 5 years out of 541 cases in a 15 year retrospective analysis.<sup>4</sup> Canter et al observed 609 rheumatic fever attacks, out of which 16 (2.6%) had initial attack in the first 5 years of life, with a mean age of 4.6.<sup>14</sup> Chockalingam A et al in a retrospective study at Chennai found that 6.8% of the rheumatic fever admissions were aged less than 5 years.<sup>15</sup>

Clinical profile of acute rheumatic fever in children younger than 5 years was analysed by several researchers.<sup>16</sup> Majeed HA et al studied prospectively in 53 children seen over a period of seven years. Majority had arthritis (81%) and Carditis was seen in 42%, with a high incidence of pericarditis (6%) and congestive heart failure (15%) and a mortality of 2%.<sup>17</sup> Tani et al observed isolated arthritis to be commonest presentation (41%) followed by isolated carditis (30%) which was more severe than in older children, erythema marginatum and chorea were observed only in 11% children.<sup>4</sup> Canter et al noted isolated carditis and isolated arthritis in 32.5% and 48.1% respectively while nil presented with chorea.<sup>14</sup> In a prospective analysis by Chockalingam A et al, 75%, 50% and 4% had arthritis, carditis and chorea occurred respectively.<sup>15</sup> In all of their observations arthritis was the predominant feature with most of them being atypical as in our case.

Pileggi and Ferriani et al described rheumatic arthritis to be atypical if at least one of the following characteristics were present - Longer duration than 3 weeks, involvement of small joints and/or cervical spine and/or hip joints, presence of monoarthritis/arthritis, and unsatisfactory response to salicylates in one week.<sup>18</sup> Bhutia et al in a retrospective study conducted at PGIMER found 63% of children had atypical arthritis. Also, polyarticular afflictions were predominately non-migratory (additive) in both atypical (74%) and typical (82%) cases.<sup>19</sup> In a study by Robazzi et al atypical pattern was observed in (70.9%) with predominant pattern being oligoarthritis (71%) followed by involvement of small joints and/or axial skeleton in 38.7%.<sup>20</sup> Banna et al observed atypical arthritis in 86.36 % of ARF patients associated with oligoarthritis, monoarthritis, polyadditive arthritis and small joint affliction in 43.18%, 27.27%, 15.90% and 9% respectively.<sup>21</sup> Our child had non migratory polyadditive arthritis of large joints (bilateral knee and ankle) with involvement of small joints (MCP, MTP,IP).

Atypical arthritis is a confounding factor making diagnostic and therapeutic delay which may be dreadful

because of its risk of progression to rheumatic heart disease. This spotlights the need for high index of suspicion despite erratic presentations even in an uncommon age group. Adding to this, there is no definitive investigation available for GAS pharyngitis / acute rheumatic fever. Hence clinching clinical diagnosis supported by laboratory evidence at the earliest manifestation combating both over and under diagnosis is mandatory to prevent or limit both acute and chronic complications.

## CONCLUSION

Rheumatic fever still continues to be a major health problem in India. Awareness on need for appropriate management of streptococcal pharyngitis among public coupled with improved hygiene and socioeconomic conditions forms the base for successful primary prophylaxis. Further studies on pathogenic mechanisms that link Group A Streptococcal exposure to RHD are to be done which has hampered GAS vaccine development efforts. Rheumatologists and pediatricians should acknowledge rarer variants of rheumatic fever facilitating early initiation of management and they also have a pivotal role in educating necessity of compliance and importance of secondary prophylaxis. It is an immediate priority globally to prevent acute rheumatic fever and rheumatic heart disease by establishing comprehensive and sustainable surveillance systems for the disease.

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