

## Case Report

# Chorea as the first presenting sign of rheumatic fever in a 12-year-old female child

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## ABSTRACT

Rheumatic chorea (RC\*) is a movement disorder seen in young children and adolescents with a recent history of incompletely treated group A beta-hemolytic streptococcal (GABHS) pharyngitis. Although, it rarely presents as the first manifestation of the rheumatic fever, physicians should be aware of the disease, so that early diagnosis and prompt treatment may lead to elimination of the pathogen and prevent further disease progression. We present a case of a 12-year-old female child who presented with only RC as the first clinical sign.

**Keywords:** Group a beta hemolytic streptococcus, Rheumatic chorea, Rheumatic fever

## INTRODUCTION

Rheumatic chorea (RC) is a debilitating neurological presentation of rheumatic fever (RF). RC is a rare isolated and initial presentation of RF; however, with rheumatic carditis it becomes fairly common.<sup>1</sup> The burden of RF/ RHD has been described in detail by Carapetis and colleagues.<sup>2,3</sup> Excluding developed economies, the global burden of RHD in the 5 to 14 years old children was estimated to be 0.8 - 5.7/1000 with a median of 1.3/1000. Population based surveys for prevalence are very few and scattered.

In a study in rural Haryana prevalence of RHD was found to be 2.2/1000 in 5 to 30-year-old subjects.<sup>4</sup> Mathur in a study of the urban population of Agra found RHD in 1.8/1000 in the same age group.<sup>5</sup> Berry studied the urban population of Chandigarh and found RHD in 1.23/1000 male and 2.07/1000 in the female population of all age groups.<sup>6</sup>

A recent Indian Council of Medical Research (ICMR) study (between 2000 and 2010) in 10 different, mostly urban, locations of the country found the prevalence to

range from 0.2 to 1.1/1000 for RHD and 0.0007 to 0.2 /1000 for RF. The data were based on registration of all cases in one million population by approaching hospitals, private practitioners and extensive advertising for establishing a registry of all known cases.

In India population based study indicates prevalence of RHD to be about 2/1000 population, however survey conducted in school children in the age group of 5-16 years by ICMR gives overall prevalence of 6/1000 (range 1.8 to 11/1000).<sup>7</sup> In various study in India including (Roy, Padmavati, Arora, Sanya, Agrawal, Vaishnava) 5 to 21% patients having chorea as a major manifestation in rheumatic fever.<sup>8-12</sup>

Follow up of patients with pure chorea without RHD or of patients who have had RF but no clinical evidence of carditis indicates that RHD can develop over a period of time. Bland in a 20 year follow up of patients with isolated chorea found 23 per cent patients without clinical carditis to develop RHD predominantly mitral valve obstruction (MS).<sup>13</sup> Similarly, Aron et al, in a 30-year follow up of 50 patients of pure chorea ended up with RHD, predominantly MS, in 34 per cent patients.<sup>14</sup>

Chorea alone has been identified in 0.6% cases of RF in Nepal and along with carditis its incidence increased to 2.3%.<sup>15</sup> In Pakistan, 16% children with rheumatic carditis developed chorea in the first presentation and another 4% in recurrent attacks.<sup>16</sup> RF occurs as the autoimmune response of the body to group A beta-hemolytic streptococcal (GABHS) infections. These infections are more common in school-going children and arthritis and carditis remain the common presentation in this age group. RC remains the most common cause of any choreiform movement in children. Failure to recognize chorea as a presenting sign of acute RF and subsequent management predisposes the child to recurrent attacks of RF and also rheumatic heart disease. We present a case of chorea as the first presentation of RF in a 13-year-old child.

## CASE REPORT

A 12-year-old female child was brought by her mother to the outpatient department (OPD) of a tertiary care hospital, in Vadodara, Gujarat, with a three-day history of sudden onset of restless abnormal movements of the body. The abnormal movements began unilaterally in the upper limbs then involved ipsilateral lower limb (on the same day). Her mother reported she has difficulty performing her daily activities such as bathing, picking up utensils, and eating since then. The mother also reported that there were no abnormal, involuntary movements during sleep. She had a history of sore throat and fever four months back, which resolved spontaneously and had not taken treatment.

Detailed history including origin, duration, progress of complaint, past history, family history, development history taken. General examination and detailed systemic examination had been done. The general physical examination was unremarkable. She was alert and oriented to place and time. While lying, she was anxious and restless with involuntary movements. Her vitals were taken which showed, heart rate 88/min, respiratory rate 18/min, afebrile, and blood pressure 108/70 mmHg. She was mildly anemic.

Pertinent examination findings included unclear speech, difficulty in walking. There were irregular contractions and relaxation of muscles with pronation of forearm on outstretched hand above her head. so "Milkmaid's grip." and 'pronator sign' were present.

Her laboratory investigations showed hemoglobin (Hb) levels: 12.2 mg/dL (normal range: 12.0-15.5), hematocrit (HCT) level: 37.5% (normal range: 37%-48%), total leukocyte count (TLC): 7600 IU/L (normal range: 130,000-400,000), erythrocyte sedimentation rate (ESR): 15 mm 1st hour (normal: <20), anti-streptolysin O (ASO titer): 284 (normal: <200 units), electrocardiogram (ECG) was normal (PR interval <0.15 sec) and echocardiogram was suggestive of mild mitral regurgitation so rheumatic carditis without congestive cardiac failure was present.

The patient was initially managed with IV penicillin, and oral carbamazepine. After 48 hrs of hospital admission, she was started with oral aspirin along with proton pump inhibitor. the patient was discharged on oral antibiotics, aspirin 60 mg/kg/day in a 4 divided dose for twelve weeks. Oral carbamazepine was also advised for 30 days. Injection Benzathine penicillin 1.2 mil IU every three weeks (till the age of 21) was also advised. At her first follow-up visit three weeks later, child's condition had significantly improved. The patient has since resumed her normal daily routine activities with resumption of her school and continues to receive injection Benz penicillin every three weeks.

## DISCUSSION

Rheumatic fever caused by antecedent infection by group A streptococcus upper pharyngitis infections by several mechanisms. Including cytotoxin theory, immunologic theory.

Certain serotypes of group A streptococcus (M type 1,3,5,6,18,29) are more frequently isolated from patients with acute rheumatic fever than other serotypes.

Cytotoxic theory suggests that a streptococcal toxin is involved in the pathogenesis of acute rheumatic fever and rheumatic heart disease.

An immune mediated pathogenesis for rheumatic fever explained by antigenicity of several group A streptococcus cellular and extracellular epitopes and their immunologic cross reactivity with cardiac antigenic epitopes. in rheumatic chorea, antibodies against N-acetyl-B-D-glucosamine epitope of streptococcal group A carbohydrate target intracellular B-tubulin and extracellular lysoganglioside GM1 in human caudate-putamen preparations. These antibodies are capable of directing calcium/calmodulin-dependent protein kinase 2 activation, which may cause the neurologic manifestation of Sydenham chorea by increasing dopamine release into synapse.

Rheumatic chorea or Sydenham chorea (SC) is a neurological disorder of childhood triggered as a result of autoimmune response to Group-A beta hemolytic streptococci infections. SC is categorized as a major criterion for acute RF diagnosis. It is characterized by sudden, brief, nonrhythmic, nonrepetitive twitching of limbs with facial grimacing. There is a history of sore throat several weeks before onset of symptoms which include, involuntary movements, slurred speech, hypotonia, and difficulty holding objects, writing, eating, and dressing. As per modified Jones criteria, clinical appreciation of choreiform movements is sufficient to diagnose RF in the presence of acute infective history. The criteria for the diagnosis enunciated by Dr T. Duckett Jones' have been modified, revised and updated by the American Heart Association (AHA).<sup>17</sup> The diagnostic criteria consist of major manifestations carditis, arthritis,

subcutaneous nodules, erythema marginatum and chorea. Rheumatic carditis resulting in a more or less permanent damage to the heart is the main virulent manifestation of RF. The minor manifestations consist of fever, arthralgia, elevated sedimentation rate, C-reactive protein (CRP) and prolonged PR interval in the electrocardiogram. Presence of two major or one major and two minor manifestations with an evidence for recent GAS infection (essential criterion) indicate acute RF. Evidence for recent GAS infection can be in the form of a positive throat culture, elevated anti-streptococcal antibodies or presence of features for recent scarlet fever, rare in our country.

The components of major, minor and essential criteria for the diagnosis remain more or less as before in AHA guidelines.<sup>17</sup> The updated criteria emphasize the value of indolent carditis and chorea to be accepted as evidence of RF and have removed previous RF or presence of RHD as a minor manifestation to simplify the diagnosis of first attack of RF. In the presence of previous history of RF/RHD one major or more than one minor criterion is acceptable for the diagnosis of recurrent RF. Additionally echocardiogram-based diagnosis of carditis has been questioned in the absence of clinical findings to indicate cardiac involvement. The clinical manifestations of RF, except for minor differences in frequency, is the same all over the world. In our country erythema marginatum is not recognized possibly because of the darker skin complexion. Females develop choreiform movements more commonly than males.<sup>8-12</sup>

Identification of rheumatic fever: RHD can occur only after a patient has had RF. Evaluation of data indicates that about 65 per cent patients get clinically recognizable RHD following RF. In the global estimate a conservative figure of 60 per cent carditis has been used for calculating the burden of RHD.<sup>2</sup> This suggests that at least 40 per cent patients who have had RF could be potentially patients of subclinical carditis. On the basis of Utah study, 27 per cent patients had subclinical carditis.<sup>18</sup> Hence, the actual estimated burden could be much more than the actual burden. There have been other case reports of pediatric RF-both pure as well as with carditis-from other regions including Nepal (13-year-old female), Saudi Arabia (5-year-old male), India (18-year-old female), and the United States (7-year-old female).<sup>15,19-21</sup>

In a four-year retrospective analysis from Nepal, 672 cases of pediatric acute RF were identified. There were more females than males (55% vs. 45%). The incidence of SC was 3.8%. SC was much more common in females than males (77% vs. 23%) and 73% lied between the age of 10 and 16 years. Only 0.6% children had isolated chorea, 0.8% had chorea with arthritis, and 2.3% had chorea with carditis.<sup>22</sup>

## CONCLUSION

Pure isolated chorea as the first presentation of rheumatic fever cannot be underestimated in older children. RC

should be among the top differential diagnoses in children presenting with movement disorders in developing countries. Keen observation of the progression of erratic movements helps in suspecting chorea. With strong clinical suspicion unnecessary neurological investigations can be avoided. Recognition of RC and its timely management is crucial in preventing rheumatic heart disease.

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