International Journal of Contemporary Pediatrics
Patel BG. Int J Contemp Pediatr. 2025 Feb;12(2):341-342
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20250109

Case Report

Rheumatic chorea as the first presenting sign in a 12-year-old male child

Bhargav G. Patel*

Department of Pediatrics, Sadvichar Parivar Godhra, PT Mirani Hospital, Godhra, Gujarat, India

Received: 24 December 2024
Accepted: 21 January 2025

***Correspondence:**
Dr. Bhargav G. Patel,
E-mail: dr bhargavpatel381@gmail.com

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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-haemolytic streptococcal (GABHS) pharyngitis. Although, it rarely presents as the first manifestation of the disease, 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 years old male child who presented with only RC as the first clinical sign.

Keywords: Case report, Group a beta-hemolytic streptococcus infection, 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.[1] however, with rheumatic carditis it becomes fairly common.[2]

Chorea alone has been identified in 0.6% cases of RF in Nepal and along with carditis its incidence increased to 2.3%.[3] In India there is no evidence of presentation of rheumatic fever as chorea alone. 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.[4] We present a case of chorea as the first presentation of RF in a 12 years old child male child.

CASE REPORT

A 12 years old male child was brought by his parents to the outpatient department (OPD) of a P.T. Mirani trust hospital, Godhra, Gujarat, India with a fifteen-days history of sudden onset of restless abnormal movements of the body. The abnormal movements first began unilaterally in the right upper limbs then involved the trunk (on the same day) and finally right lower limb were involved (on day 2). His parents reported he has difficulty performing his daily activities such as bathing, picking up utensils and eating since then. The parents also reported that there were no abnormal, involuntary movements during sleep. He had a history of sore throat and fever a few weeks back, which resolved spontaneously.

The general physical examination was unremarkable. He was alert and oriented to place and time. While lying, he was anxious and restless with involuntary movements. His vitals were taken which showed, heart rate 102/min, respiratory rate 24/min, afebrile and blood pressure 106/80 mmHg. Other examination findings included unclear speech, difficulty in walking. There were

irregular contractions and relaxation of muscles showing pronation of forearm on outstretched hand above his head called as "Milkmaid's sign". His laboratory investigations showed hemoglobin (Hb) levels 12.4 mg/dl (normal range 12.0-15.5), hematocrit (HCT) level 39.2% (normal range 37%-48%), total leukocyte count (TLC): 6750 (normal range: 4000-1000/cu mm), erythrocyte sedimentation rate (ESR): 22 mm 1st hour (normal:<15), antistreptolysin O (ASO titer): 429.85 (normal-<200 IU/ml), electrocardiogram (ECG) and echocardiogram were with normal PR interval of 0.12 s thus, rheumatic carditis was ruled out.

The patient was managed on OPD basis. Strict bed rest for 10 days along with oral amoxicillin-clavulanic acid and phenobarbitone in maintenance dose were advised. As he is not having any sign of arthralgia or arthritis, aspirin was not started. Injection Benz penicillin 1.2 MU every three weeks (till the age of 21) was also advised. At his first follow-up visit two weeks later, child's condition had significantly improved. The patient has since resumed his normal daily routine activities with resumption of his school and continues to receive injection Benz penicillin every three weeks.

DISCUSSION

Rheumatic chorea or Sydenham chorea (SC) is a neurological disorder of childhood triggered as a result of autoimmune response to GABHS infections. SC is categorized as a major criterion for acute RF diagnosis. As per modified Jones criteria, clinical appreciation of choreiform movements is sufficient to diagnose RF in the presence of acute infective history.[5-6] Females develop choreiform movements more commonly than males.[3] 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.[6]

To the best of our knowledge, this is the first report of pure RC in a child from Gujarat, India. Other pediatric reports have been in presence of rheumatic carditis as discussed earlier, however, in our case, cardiac complications were rules out on echocardiogram.[2] There have been other case reports of pediatric RF-both pure as well as with carditis-from other regions including Nepal (13 years old female), Saudi Arabia (5 years old male), India (18 years old female) and the United States (7 years old female).[4,5,7,8] In a four-year retrospective analysis from Nepal, 672 cases of pediatric acute RF were identified.[3] 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.[3] In an Israeli study with 24 SC patients, there were twice as many female children as male. Their ages ranged from 4 to 13 years. Familial chorea was appreciated in 8.3%, 21% had pure isolated chorea and 42% developed recurrent choreic episodes in the subsequent years.[9]

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.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Patel BG. Rheumatic chorea as the first presenting sign in a 12-year-old male child. Int J Contemp Pediatr 2025;12:341-2.