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**Case Report**

# Atypical recurrence of rheumatic chorea

**Gunjan Pankaj Kumar*, Ankita Singh, Swati Mulye**

Department of Pediatrics, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India

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***Correspondence:**
Dr. Gunjan Pankaj Kumar,
E-mail: drgunjankumar30@gmail.com, drgunjanpankajkumar30@gmail.com



## ABSTRACT

Syndenhams Chorea in acute rheumatic fever is reported to occur in 20-30% of patients. It is usually late onset, occurring upto 6 months after acute infection but may occasionally be present as presenting symptom of rheumatic fever. It is a self-limiting condition with spontaneous remission lasting from 1 week to 6 months. The risk of recurrence is present in 1st 1-2 years in about 20% of cases. Most of children (two thirds) with rheumatic fever are of school age (5-15 years of age). It is common in India and the incidence has not shown the declining trends seen in the developing countries. We report the clinical findings, investigations and the course of clinical development of a 14-year-old girl, who presented with Rheumatic chorea which recurred 3 years after the initial episode.

**Keywords:** Rheumatic fever, Chorea, relapse

## INTRODUCTION

Acute rheumatic fever is a multisystem inflammatory disease which occurs as delayed sequelae to group A streptococcal pharyngitis. The important clinical manifestations are migratory polyarthritis, carditis, chorea, subcutaneous nodules and erythema marginatum occurring in various combinations. Sydenham's chorea, once considered as a self-limiting condition, is now felt to need more aggressive treatment because it can cause great functional impairment to a patient.[1,2]

This is a self-limiting condition & spontaneous remission can occur within 1 week to 3 to 6 months. The risk of recurrence is present in 1st 1-2 years in about 20% of cases.

We report the clinical findings, investigations and the course of clinical development of a 14-year-old girl, who presented with Rheumatic chorea which recurred 3 years after the initial episode.

## CASE REPORT

The patient was a 14 year old girl presented to us with history of involuntary movements of her body since 15days. The involuntary movements involved her limbs, fingers, toes & facial muscles. The involuntary movements were severe enough to impair her daily activities such as deterioration in her handwriting skills and increasing difficulties with speaking, walking, and daily activities such as brushing, writing. The involuntary movements disappeared during sleep and became prominent when the girl felt anxious. Liability of mood was also noted during this period of time.

There was a significant past history with similar involuntary movements 3 years back for which the child was admitted at some local hospital & was treated symptomatically and took treatment regularly for 6-8 months.

There was no history of any fever, sore throat, breathlessness, chest pain, dyspnea, arthralgia, jaundice,

photosensitivity, recurrent mouth ulcers. There was no history of any long term drug intake.

Physical examination showed that she was afebrile, alert and oriented. She had dysarthria but no dysphonia. There was chorea involving all her four limbs with writhing movements of her fingers. Blessing sign was present with jack in the box tongue & dinner fork hand. The muscle tone was decreased & deep tendon reflexes & muscle power were all symmetrical & normal. No cerebellar ataxia was demonstrated. Due to the choreic movements, she failed the finger-to-nose test and heel-to-shin test. There was also dysdiadochokinesia. Cardiovascular examination her heart rate was regular at 82 per minute. Apex was not displaced. Heart sounds were normal with a soft systolic murmur. Chest was clear. Abdomen was soft with no organomegaly.

Blood test showed normal electrolytes, calcium and complete blood picture. Anti-streptolysin O-titre (ASOT) was 200 IU. The Erythrocyte Sedimentation Rate (ESR) was 28 mm/hour. Anti-nuclear antibody was negative. Electrocardiogram was normal with no prolonged PR interval. No ST change was observed. 2D Echo was done which was suggestive of rheumatic heart disease with mild mitral regurgitation. A diagnosis of Sydenham's chorea was made based on the presence of abnormal 2D Echo & chorea.

The child was started on benzathine penicillin injection with sodium valproate & haloperidol. The Choreic movements gradually reduced and disappeared within 2 weeks of starting treatment. Follow up period of 4 months was uneventful.

## DISCUSSION

The guidelines for the diagnosis of acute rheumatic fever are based on the 1992 updated Jones criteria. Chorea consists of rapid, involuntary movements observed on physical examination which do not have a cause which can be demonstrated by laboratory tests. It is worsened by stress and anxiety and subsides during sleep.[3] Sydenham's chorea, first described in 1686 by Thomas Sydenham, is generally used to signify a diagnosis of a form of rheumatic fever. It is a cardinal feature of rheumatic fever and is sufficient alone to make the diagnosis.

Sydenham's chorea is the most common cause of acquired chorea in children.[4] Sydenham's chorea is a major late clinical manifestation in acute rheumatic fever. Migratory polyarthritis, carditis, subcutaneous nodules and erythema marginatum are the other major clinical manifestations according to Modified Jones criteria. There is often a long latent period between group A beta-haemolytic streptococcal infection and the onset of chorea, making it an uncommon initial presentation in acute rheumatic fever.

There are other diseases that cause chorea in childhood. The differential diagnosis can be Sydenham's chorea, lupus-associated chorea and drug-induced chorea.[5] Sometimes, chorea induced by another disease is misdiagnosed as Sydenham's chorea. Other common causes of chorea include atypical seizures, brain tumor, cerebrovascular accident, collagen vascular disorders such as systemic lupus erythematosus and Behcet's disease, drug intoxication, endocrine disorders such as hyperthyroidism and hypoglycemia, hereditary disorders such as Huntington's chorea and Wilson's disease, pregnancy and viral encephalitis.[5-7]

For a child with chorea, the initial examination includes chest X-ray, echocardiography, electrocardiography, anti-streptococcal antibody studies, erythrocyte sedimentation rate and C-reactive protein. If the initial examination does not support a diagnosis of Sydenham's chorea, further approaches include evaluation of thyroid function, antinuclear antibody titer, antiphospholipid antibodies, copper metabolism, and neuroimaging studies. Several drugs are used in chorea with a variable degree of efficacy; haloperidol, valproic acid, tetrabenazine, chlorpromazine, barbiturates and corticosteroids.

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