

Rare Recurrence of Sydenham Chorea in an Adult: A Case Report

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Background: Sydenham chorea is thought to be an autoimmune condition that usually develops following a group A beta-hemolytic streptococcal infection. The onset of Sydenham chorea in adults is rare and most of the adult cases usually are secondary to recurrence following childhood illness. Risk factors for chorea recurrence include irregular antibiotic prophylactic use, failure to reach remission within 6 months, and symptom persistence for longer than a year.

Case Presentation: A 27-year-old young adult Ethiopian female patient with chronic rheumatic valvular heart disease for the last 8 years experienced repetitive uncontrollable movements of her extremities and torso for three years prior to her current visit. Physical examination was significant for holosystolic murmur at the apical area radiating to the left axilla and choreiform movements apparent on all limbs and trunk. Investigations were significant for mildly raised ESR, echocardiography findings of thickened mitral valve leaflets and severe mitral regurgitation. She was successfully treated with valproic acid and the frequency of penicillin injection was made every 3 weeks with no recurrence for the first 3 months follow-up period.

Conclusion: We believe that this is the first case report of adult onset recurrent Sydenham chorea (SC) from a resource-limited setting. Though Sydenham chorea and its recurrence is rare in adults, it should be considered in adults after ruling out other competing differential diagnoses. Because of the lack of evidence on treatment of such rare cases, individualized mode of therapy is advised. Valproic acid is preferred for symptomatic treatment and more frequent benzathine penicillin G injections, for example every three weeks, may help in the prevention of recurrence of Sydenham chorea.

Keywords: Sydenham chorea, rheumatic fever, recurrence

Background

Sydenham chorea (SC) is thought to be an autoimmune condition that usually develops following a group A beta-hemolytic streptococcal infection. The immune system antibodies that are produced in response to the streptococcal bacterial infection, presumably have a cross-reaction with the basal ganglia cells, which play crucial roles in regulating motor activity. The onset of Sydenham chorea in adults is rare and most of the adult cases usually are secondary to recurrence following childhood illness.¹ Females are twice more affected than males.² SC ranges in severity from mild to severe and has an impact on a variety of daily tasks, including writing, walking, eating, and getting dressed.³

The reason for recurrence of Sydenham chorea is not yet clearly known. Recurrent chorea and rheumatic fever do not share the same underlying etiology and recurrent chorea is not always brought on by rheumatic fever. Following a single attack of Sydenham chorea, the basal ganglia may sustain persistent subclinical injury or a basic abnormality leading to recurrence.⁴ Risk factors for chorea recurrence include irregular antibiotic prophylactic use, failure to reach remission within 6 months, and symptom persistence for longer than a year.⁵

Case Presentation

A 27-year-old young adult Ethiopian female patient who is known to have chronic rheumatic valvular heart disease for the past 8 years started to experience repetitive uncontrollable movements of her extremities and trunk since 3 years ago

and which were occurring once to three times per month. The abnormal body movements usually lasted up to a week duration and got worse while she tried to sit or walk and improved with rest. She was consistently taking intramuscular injection of benzathine penicillin G 1.2 million units every month for the past 8 years. She did not have previous history of behavioral and emotional lability. She does not remember childhood history of rheumatic fever or movement disorder. She did not use oral or injectable hormonal agents. There was no family history of rheumatic fever, social stress, psychiatric disease, or tic disorders. Upon physical examination, temperature was 36.7°C, heartbeat was 90 beats per minute, and blood pressure was 100/70 mmHg. There was holosystolic murmur at the apical area radiating to the left axilla. Choreiform movements were apparent on all limbs and trunk.

Laboratory investigations revealed moderate leukopenia (but with a normal repeat complete blood count after 3 days), normal liver enzymes, normal renal function tests, erythrocyte sedimentation rate of 37 mm/hour, negative qualitative C-reactive protein, weakly positive anti-streptolysin O antibody, negative qualitative antinuclear antibody, negative urine human chorionic gonadotrophin and normal level of thyroid stimulating hormone. Magnetic resonance imaging of the brain showed left parietal periventricular tiny white matter non-specific lesion. Rheumatic heart involvement was confirmed by echocardiography which showed thickening of mitral valve leaflets with severe mitral regurgitation (Figure 1).

The present case was diagnosed as recurrent Sydenham chorea based on clinical evidence and she was prescribed valproic acid 500 mg PO/day for 2 weeks and benzathine penicillin G injection was made every 3 weeks. She was re-evaluated after 2 weeks and she had significant improvement with good control of motor activity and reduced involuntary movements of her extremities and trunk. She was then followed every 3 weeks for the subsequent 3 months and she did not have any abnormal body movement. She was finally referred to an advanced cardiac center for possible mitral valve replacement.

Discussion and Conclusions

Patient characteristics, associated clinical features and treatment outcomes of Sydenham chorea in the present case report are quite different from those of the available case reports (Table 1). In contrast to a prior case series of children with SC, where the time between the initial episode and the recurrence ranged from 3 months to 10 years,⁴ the recurrence in the present case was once to three times per month. Adult patients may experience neuropsychiatric consequences of Sydenham chorea such as chorea gravidarum, obsessive-compulsive disorder, and schizophrenia.⁵ In contrast to a case report of an adult male patient who had psychiatric manifestations including emotional incontinence and abnormal behavior accompanying chorea,⁶ there was no associated psychiatric symptom in this case report.



Figure 1 Transthoracic echocardiography from a patient with rheumatic valvular heart disease and recurrent Sydenham chorea in parasternal long axis view. Mitral valve leaflets are thickened (arrows).

Abbreviations: LA, left atrium; LV, left ventricle; Ao, aorta.

Table I Case Reports of Sydenham Chorea in Adolescents and Adults

Source of Data	Age (Years)	Sex	Duration of Chorea	Additional Clinical Features	Recurrence of Chorea	Treatment Response
Present case report	27	Female	3 years	Rheumatic valvular lesion and raised ESR	Very frequent recurrences (in weeks)	Responded to sodium valproate
S. Kin et al ⁶	56	Male	3 years	Arthritis, psychiatric symptoms, fever, increased ESR and elevated CRP	Unknown	Unknown
S. Bouchal et al ¹¹	16	Female	2 years	Recurrent angina	Occasional recurrences (in years)	Responded to sodium valproate
J. Sie et al ¹²	18	Female	3 days	Rheumatic valvular lesion, fever and raised CRP	Unknown	Responded to diazepam and haloperidol

Abbreviations: ESR, erythrocyte sedimentation rate; CRP, c-reactive protein.

The risk of recurrence of SC in the present case may be explained by the absence of remission in the first 6 months of onset of SC and its extension beyond a year. This is supported by findings of a study done by Gurkas et al, which found that failure to achieve remission within 6 months and prolongation of symptoms for more than 1 year were important risk factors for recurrence of SC.⁷

Our patient had recurrence of SC despite taking monthly benzathine penicillin G, unlike the findings of a study done in children with SC, which showed a significant reduction of recurrence for those who were given prophylactic long-acting penicillin G compared with those who were not given this.⁸

In the previous case reports, where most patients were children, those developing recurrent or non-recurrent SC were oftentimes treated effectively with valproic acid and neuroleptic agents.^{4,9} Despite the paucity of data on how to treat recurrent or non-recurrent SC in adults, symptomatic treatment of the present case with valproic acid was successful. There was no recurrence while she was followed for three months, which might be because of the more frequent administration of benzathine penicillin G injection.

A study done by Dean et al reviewed available case reports and case series and finally recommended the initial use of valproic acid or carbamazepine instead of neuroleptics for symptomatic treatment of Sydenham chorea. Neuroleptics are not preferred because of their adverse effects and lack of evidence to show their superior efficacy.¹⁰ A similar response to sodium valproate was seen in a case report of a 16-year-old female patient with recurrent Sydenham chorea after failure of haloperidol.¹¹ Another case report has shown that an 18-year-old female patient with Sydenham chorea responded to diazepam and haloperidol.¹²

In conclusion, we believe that this is the first case report of adult onset recurrent Sydenham chorea from a resource-limited setting with frequent episodes of choreiform movements in the background of chronic rheumatic valvular disease. Though SC and its recurrence is a rare condition in adults, it should be considered in the appropriate clinical context after ruling out other competing differential diagnoses. Because of the lack of evidence on treatment of such rare cases, individualized mode of therapy is advised. For symptomatic treatment, we favor valproic acid instead of haloperidol to make sure patients are treated with an effective and well-tolerated drug. In settings with a burden of group A streptococcal infection, we advise more frequent benzathine penicillin G injections, which might help to curb the recurrence of Sydenham chorea.

Limitations of the Case Report

The first limitation is the short follow-up period of the patient, which is not adequate to trace for any recurrence of Sydenham chorea. The second limitation is the absence of adequate similar case reports to compare and contrast with the current case report.

Ethical Clearance

Ethical clearance including publication of this patient's case details was obtained from Yekatit 12 Hospital Medical College Review Board.

Consent for Publication

The patient gave a written informed consent for the publication of her case details including the history, physical findings, laboratory reports, and the echocardiography image.

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Disclosure

We don't have conflicts of interest in this work.

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