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Case Report
Heart Threat

The Hidden Heart Threat: Subclinical RHD Mimicking Tic Disorder

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Rheumatic fever (RF) is a post-infectious inflammatory disease that occurs after a Group A streptococcal (GAS) pharyngitis, which affects multiple systems [1], [2]. It primarily affects children aged 5 to 15 years [2], [3].

Sydenham's chorea (SC), also known as Saint Vitus' Dance, manifests as rapid, uncoordinated movements mainly impacting the face and hands due to the destruction of cells in the corpus striatum of the basal ganglia [6], [7]. The prevalence of chorea in patients with acute rheumatic fever (ARF) is 5-36% as reported by WHO and can occasionally be the sole manifestation of acute rheumatic fever (ARF) [6], [7]. It is included as a major ARF criterion per the Jones criteria, with a higher prevalence in females and children under 18 [7], [8].

In some cases of acute rheumatic fever (ARF), cardiac involvement may not be observed, this condition is known as subclinical carditis with a prevalence of 14-35% [8], [9]. Carditis is the only manifestation of acute rheumatic fever (ARF) that may result in permanent disability [10], [11]. Subclinical carditis can be identified in patients during their first ARF episode using ECHO, even if they present solely with isolated rheumatic chorea or migratory polyarthritis [10], [12]. Studies have shown that the mitral valve is the most common valve involved in RHD followed by the aortic valve [2], [8]. Most RHD patients with mitral valve involvement have shown improvement in valve symptoms with regular secondary prophylactic antibiotics over 5–10 years [10].

Keywords: Rheumatic chorea, subclinical carditis, Sydenham's chorea

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Introduction

Rheumatic fever (RF) is a post-infectious inflammatory disease that occurs after a Group A streptococcal (GAS) pharyngitis, which affects multiple systems [1], [2]. It primarily affects children aged 5 to 15 years [2], [3].

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Treatments for Sydenham's chorea (SC) include acute and long-term penicillin therapy, symptomatic medications like anticonvulsants (valproate, carbamazepine, phenobarbitone) and neuroleptics (pimozide, haloperidol, risperidone), and immunotherapy for resistant cases [12], [13]. Medications should be tapered once the patient's symptoms improve. Literature suggests that while most cases respond well to symptomatic treatment, immunotherapy is typically reserved for cases unresponsive to standard approaches [13].

We highlight a case where subtle valvular regurgitation was detected only by echocardiography, initially misinterpreted as a tic disorder.

Case report

A 10-year-old boy presented to paediatric OPD with complaints of involuntary movements of the upper and lower limb, predominantly on left side for 10 days along with involuntary movements of face. The symptoms were insidious in onset, gradually progressive and disappeared during sleep. He also exhibited facial grimacing, random tongue movements, and slurring of speech. Notably, he had joint pain five months ago, a body rash sparing head and neck one and a half months ago, and a fever 10 days ago. The child had no history of similar complaints in past. There was no history of behavioural or neuropsychiatric disorders in family members. The birth history was uneventful and child is immunised to date with an average scholastic performance & no developmental delays.

On examination, he was vitally stable and afebrile. Heart examination revealed normal heart sounds with no audible murmurs. Skin examination was unremarkable. Neurologically, the patient was conscious, alert, and oriented to time, person, and place, displaying cooperation and intelligence. On head-to-toe examination, choreiform movements were noted in the face and upper and lower extremities. The cerebellar signs were negative, cranial nerve function was intact and sensory examination was normal. The motor system assessment showed normal muscle tone and strength globally. Brief, nonrhythmic, nonrepetitive involuntary movements were observed in the left upper limb. He also showed facial twitching and darting movements of the tongue on protrusion.

Differential diagnosis:

- Tic disorder
- Dystonia
- Rheumatic chorea

Investigations:

Routine laboratory studies were normal. The erythrocyte sedimentation rate (ESR) was 50 mm/h and C- reactive protein (CRP) was negative. The anti-streptolysin O titre (ASLO) was elevated with 1:1600IU/ml dilution. Thyroid function tests were normal and ECG was within normal limits. An ECHO was done and found to have moderate MR with AML prolapse and posteriorly directed MR jet, which along with the given history was suggestive of Rheumatic heart disease (RHD).

The case was diagnosed as acquired subclinical RHD with chorea.

Intervention:

The patient was started on Tab. Penicillin V 250mg BD, Tab. Phenobarbitone 30mg TDS and Aspirin 75mg 8 tablets QID.

The Tab. Penicillin was given as a long-term prophylaxis for at least 10 years.

Follow-up:

At follow-up visits, the patient showed a reduction in involuntary movements, allowing for the gradual tapering of Tab. Phenobarbitone. Lifelong penicillin prophylaxis was advised to prevent recurrence and further complications.

Discussion

Rheumatic heart disease (RHD) is a complication of Acute rheumatic fever (ARF) and it remains a major health concern as it contributes to significant cardiovascular disease burden and mortality, especially in developing countries due to factors such as overcrowding, poor nutrition, and limited access to healthcare [2], [4]. India ranks among the top two countries that contribute to the highest estimated numbers of RHD-related deaths [1], [5].

This case underscores the necessity of recognizing Sydenham's chorea (SC) as an indicator of acute rheumatic fever (ARF), especially in children who present with recent choreiform movements. Despite SC being a well-established major criterion for ARF within the Jones criteria, it can often be misdiagnosed as other movement disorders, such as tic disorders or dystonia, particularly when clinical signs are subtle. Subclinical RHD refers to the detection of structural or functional valvular damage via echocardiography, despite the absence of a heart murmur [2], [9]. Identifying subclinical RHD early allows for timely intervention and the commencement of secondary prophylaxis, crucial for preventing recurrent ARF episodes and limiting long-term cardiac damage.

Research indicates that consistent secondary prophylaxis with penicillin can greatly reduce the risk of disease progression and recurrence, contributing to better outcomes, particularly for patients with early-stage mitral valve involvement [10], [12].

In this case, the introduction of prophylactic penicillin, combined with symptomatic treatment using phenobarbitone and aspirin, led to significant clinical improvement and a reduction in the patient's involuntary movements.

Conclusion

Sydenham chorea should be considered in young children with choreiform movements, as it may be the first sign of acute rheumatic fever [6], [12]. This case highlights the essential role of echocardiography in detecting subclinical carditis associated with Sydenham's chorea.

Early identification and symptom management, along with long-term prophylaxis, are key to preventing recurrent ARF and mitigating the risk of early, severe rheumatic heart disease which can improve patient outcomes and minimize the risk of permanent cardiac damage.

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