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Case Report

Rheumatic Chorea Without Cardiac Involvement in a 10-Year-Old Girl from a Remote Area: Diagnostic and Management Challenges

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Abstract

Rheumatic Chorea, also known as Sydenham Chorea, is a major neurological manifestation of acute rheumatic fever, an autoimmune complication following infection with Group A β-hemolytic streptococcus (GABHS). It remains common in low- and middle-income countries, where limited healthcare access may delay diagnosis and increase the risk of complications, particulary cardiac. We report a case of a 10-year-old girl from a remote area who presented with involuntary movements, joint pain, and a history of throat infection one month prior to symptom onset. Laboratory tests showed elevated erythrocyte sedimentation rate (ESR) and a positive Anti-streptolysin O (ASO) titer, indicating a recent streptococcal infection. Cardiac examination, including the electrocardiography (ECG), revealed no abnormalities. However, echocardiography, the gold standard for detecting subclinical carditis, was not performed due to lack of available resources. The patient was treated with haloperidol and trihexyphenidyl for chorea, penicillin G benzathine for eradication and prophylaxis of streotococcal infection, and aspirin for its anti-inflammatory effects. Clinical improvement was noted within one month of therapy. This case met the 2015 revised Jones criteria for moderate-risk populations, with major criteria including Sydenham chorea and polyarthralgia, and evidence of recent streptococcal infection (positive ASO). Absence of cardiac involvement may reflect early recognition and treatment, although echocardiography is required to exclude subclinical carditis. Symptomatic therapy and long-term antibiotic prophylaxis are crucial for preventing complications. Rheumatic Chorea can occur without cardiac involvement. Particularly in resource-limited remote areas, early diagnosis, adherence to Jones criteria, and secondary prophylaxis are essential to prevent long-term sequelae.

Keywords

Sydenham Chorea, Rheumatic Fever, Children, Without Cardiac Involvement, Case Report

1. Introduction

Sydenham chorea (SC) is a delayed neurological manifestation of infection with group A β-hemolytic streptococcus (GABHS) and is a major component of the diagnostic criteria for rheumatic fever (RF). It is the most common autoimmune chorea in children [1, 2].

The incidence is decreasing in high-income countries but remains a concern in specific population subgroups and developing countries across Latin America, Africa, and Asia [3].

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Sydenham chorea presents in 10-25% of rheumatic fever cases and is typically generalized, although it may be asymmetrical or unilateral in 20% of cases. The usual age of onset is 5-15 years, with a higher prevalence in females [2, 4, 5]. Failure to recognize the disease increases the risk of recurrent acute rheumatic fever (ARF) as well as rheumatic heart disease. Although carditis is reported in most cases, isolated rheumatic disease without cardiac involvement is rarely documented. Early treatment can prevent complications and often results in full recovery.

We present a case of Rheumatic Chorea (Sydenham Chorea) without cardiac involvement in a remote area, underscoring the diagnostic and management challenges in resource-limited settings.

2. Case Presentation

A 10-year-old girl was brought to the emergency room with complaints of involuntary movements of the extremities and face, along with clumsy movements that disappeared during sleep. The symptoms had been present for two weeks prior to admission. The patient also complained of pain and stiffness in both knees and elbows. There was a history of throat infection more than one month prior, for which she was treated with paracetamol and amoxicillin. Before seeking hospital care, the patient had pursued traditional remedies, including consultations with shamans and alternative therapies, without clinical improvement.

On physical examination, the patient was alert and conscious. Her temperature was 37.0 ℃, respiratory rate 24 breaths per minute, and heart rate 109 beats per minute. Growth parameters were within normal limits. Head and neck examinations were unremarkable. Cardiovascular examination revealed normal heart sounds with no murmurs or rubs. Pulmonary and abdominal examinations were within normal limits. There were no skin abnormalities. Local examination of both elbows and knees revealed tenderness over the medial and lateral aspects, with no evidence of rash or joint effusion.

Neurological examination revealed an alert, oriented child with no cranial nerve deficits. Choreiform movements were observed in the face and limbs. Muscle tone and motor strength were normal.

Complete blood count: Hemoglobin 12.9, Leukocytes 8300, Hematocrit 40.6, Plate. Estimated sedimentation rate (ESR) was elevated (42 mm/h in the first hour and 60 mm/h in the second hour). Anti-streptolysin O (ASO) titer was positive (Table 1). Electrocardiogram showed normal sinus rhythm (Figure 1). Echocardiographic examination was not performed due to limited diagnostic resources and lack of specialist availability in the remote area.

Based on the history, clinical presentation, laboratory findings, and electrocardiography (ECG), the patient was diagnosed with acute rheumatic fever, manifestation with sydenham chorea and polyarthralgia, without cardiac involvement.

She was managed with oral haloperidol 0.5 mg twice daily, trihexyphenidyl 1 mg once daily, and intramuscular penicillin G benzathine 1.2 million units every 21 days. She was also given aspirin 500 mg four times daily, followed by tapering off of the dosage.

Education regarding the need for echocardiographic evaluation at a better-equipped facility has been provided; however, referral has not yet been approved by the patient's family, primarily due to the necessity of sea travel to reach the facility and ongoing financial limitations.

Subsequent monitoring was carried out at the Pediatric Clinic of Maren Regional Hospital. Electrocardiography (ECG) was performed at each follow-up visit. Penicillin G Benzathine injections were administered every 21 days as secondary prophylaxis and are planned to be continued until five years following the last episode or until the patient reaches 18 years of age, while awaiting the family's consent for referral for further evaluation, particularly echocardiographic examination.

Choreiform movements gradually improved within one month of initiating treatment.

Table 1. *Laboratory findings in 10-year-old girl with Rheumatic Chorea (Sydenham's chorea) without Heart Involvement.*

Test	Result	Reference range
Hemoglobin (g/dl)	12,9	12-16
Hematocrit (%)	40,6	38-47
Mean Corpuscular Volume (fl)	87	80-100
White blood cells (cmm)	8.300	4.000-10.000
Neutrophils (%)	57	50-70
Lymphocytes (%)	36	20-40
Platelet (cmm)	240.000	150.000-450.000
Estimated sedimentation rate (mm/h)	42 mm/I hour 60 mm/II hour	0-20

Test	Result	Reference range
Anti-streptolysin O	(+) Positive	Negative

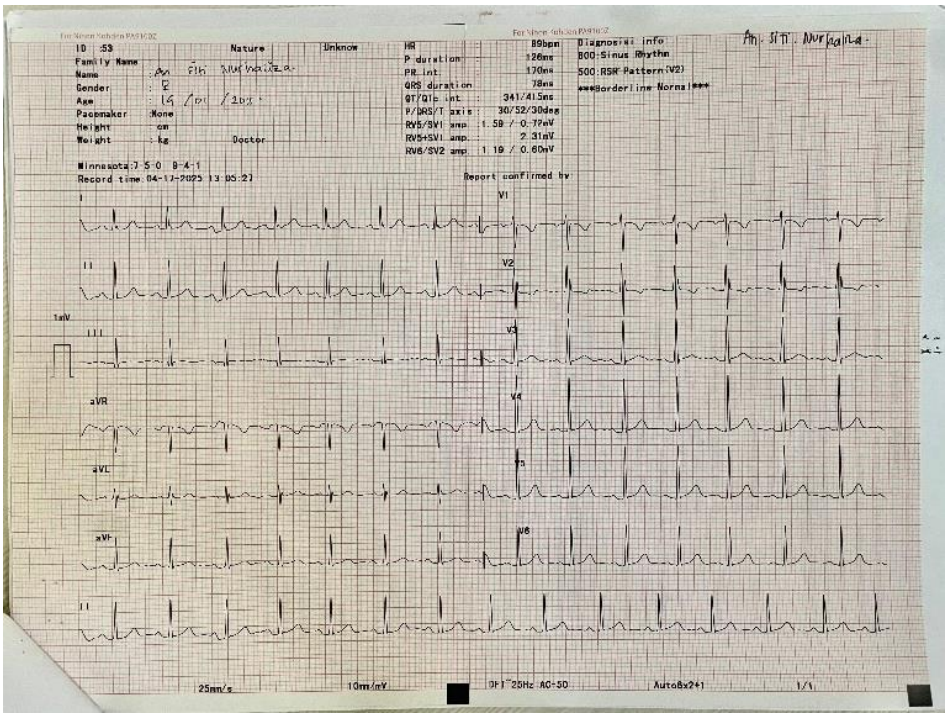

Figure 1. *Electrocardiogram of 10-year-old girl with Rheumatic Chorea (Sydenham's chorea) without Heart Involvement.*

3. Discussion

Rheumatic Chorea (Sydenham chorea) may be the sole or major manifestation of Rheumatic Fever (RF). The Jones criteria remain the cornerstone for diagnosis, with the 2015 revision including echocardiography to detect subclinical carditis [6]. Sydenham chorea (SC) is an antibody-mediated autoimmune disorder caused by the interplay of genetic and environmental factors, with a high prevalence in low- and middle-income countries [1, 7]. SC generally manifests 4 to 8 weeks following an episode of streptococcal pharyngitis, however delayed onset extending to several month after initial infection has been reported [8], earlier onset may occur. In this case, the patient had a history of sore throat about one month prior, treated with acetaminophen and amoxicillin. This suggests a possible recent streptococcal infection as a trigger. The patient's age (10 years) and female gender fall within the typical demographic for SC (5-15 years) [5].

Sydenham chorea pathogenesis is a Group A β-hemolytic streptococcus (GABHS) induced autoimmune process with multiple mechanisms, but most likely poly-reactive antibodies against streptococci, via the process of molecular mimicry, recognize neuronal extracellular surface and/or intracellular antigens. This process is believed to disrupt signaling within cortical-basal ganglia-thalamo-cortical (CBGTC) circuits. Within CBGTC pathways, Chorea is generally associated with several neurotransmitters, including excessive dopaminergic transmission, a lack of gamma aminobutyric acid (GABA), or altered acetylcholine activity [7]. The patients may have Tic-like movements and vocalizations along with neurobehavioral characteristics, such as anxiety, irritability, attention deficit hyperactivity disorder, and obsessive-compulsive behaviors, that can occasionally be disruptive [1, 7, 9]. The symptoms, including clumsy movements and facial twitches, are typical of this condition, and they often disappear during sleep, as seen in our cases.

Patients suspected of having SC should have several blood tests to help rule out other causes of chorea. These tests should include checks for acute phase proteins, anti-streptococcal antibodies, blood cell count serum electrolytes, glucose level, renal, hepatic and thyroid function, ceruloplasmin, antinuclear antibodies, antiphospholipid and anticardiolipin antibodies, rheumatoid factor are also needed [7]. Due to limited laboratory facilities in the remote area, only a complete blood count, include erythrocyte sedimentation rate (ESR) and Anti-streptolysin O (ASO) titer were performed in our case.

Early diagnosis of SC is important since patients will require a cardiac workup, particularly echocardiography [1]. Cardiac involvement, particularly mitral valve dysfunction, is

common associated with Sydenham chorea. Although holosystolic murmurs are detected in only a minority of patients, echocardiographic findings consistent with rheumatic carditis are present in up to 63% (60%-70%) of cases, indicating high prevalence of subclinical cardiac involvement [7]. However, our case demonstrates the heterogeneity of the disease. Absence of cardiac involvement may be attributed to early recognition and treatment of streptococcal infection or to host immune variability. Remote areas pose additional challenges, diagnosis of ARF is based on the 2015 revised Jones Criteria, which are adapted depending on the population's risk level [10]. The patient lives in a remote area, where rheumatic fever remains more common, placing her in a moderate-risk population. According to the revised criteria for moderate-risk populations, a diagnosis of ARF requires two major criteria, or one major and two minor criteria and evidence of a recent streptococcal infection.

This patient met the two major criteria (Sydenham chorea and Polyarthralgia). She also had a positive ASO titer, indicating a recent group A streptococcal infection. Minor criteria present include elevated ESR, although these were not required since two major criteria were already met. Carditis, another possible major criteria, could not be evaluated fully due to the lack of echocardiography. Physical examination showed no murmurs, and ECG was normal, but subclinical carditis cannot be ruled out without imaging. This case highlights the importance of applying the revised Jones Criteria in moderate-risk areas, especially where diagnostic resources are limited. Recognizing the signs early is essential to start treatment and prevent long-term complications like rheumatic heart disease. We advised the patient and her family to seek further evaluation, particularly echocardiographic assessment, at a facility with more adequate resources. Due to financial constraints and limited access to such facilities, the patient's family has not yet given consent for the referral.

The therapeutic goals in SC involve treating and preventing additional GABHS Infections, symptomatic control of the chorea and decisions regarding the need for immune modulation. Apart from the long-term use of penicillin to reduce the risk of recurrent carditis and persisting valvular damage. The most commonly used regimens include either a single dose of intramuscular (IM) penicillin G benzathine 1.2 million units or a 10-day oral course of penicillin VK 500mg twice daily. Secondary prophylaxis with penicillin G benzathine 1.2 million units administered IM, every 3-4 weeks is recommended, and the duration of treatment depends on how much time has passed since the ARF episode, patient age. Carditis may indicate greater inflammatory activity and more severe disease; a recent report similarly identified arthritis as a risk factor for longer chorea duration. Patients who are allergic to penicillin should receive oral macrolides, like erythromycin PO 250mg twice daily, for the same number of years they would receive penicillin [7, 11].

In remote areas, diagnosing and treating acute rheumatic fever may be challenging due to limited healthcare resources. However, the first priority is to prevent further streptococcal infections. Our patient should receive antibiotic treatment with penicillin G benzathine as secondary prophylaxis to reduce the risk of recurrent infections, which can cause long-term complications like heart valve damage and planned for five years after the last attack or until the age of 18, whichever is longer, while awaiting the family's consent for referral for further evaluation, particularly echocardiographic examination.

The dopamine antagonists (i.e. antipsychotics) and anticonvulsants are used to manage chorea symptoms, as they help regulate on the autoimmune-elicited basal ganglia dysfunction marked by excessive dopaminergic activation and reduced activation of the basal ganglia inhibitory pathways. However, haloperidol, as the first-generation antipsychotic, is often linked to severe side effects, such as hypertonia or parkinsonism. One possibility is that some patients treated with haloperidol had such a susceptibility due to baseline differences (eg, worse disease severity or lower resource health care settings) [4, 7, 11]. Medication-induced parkinsonism is managed by discontinuing or reducing the dosage of the causative medication, switching to another medication less likely to cause parkinsonian symptoms, or using antiparkinsonian medications, including amantadine, anticholinergic medications, L-dopa, and selegiline. Trihexyphenidyl, an antimuscarinic agent, is commonly co-administered with haloperidol. It has been in clinical usage for decades [12]. In our case, the patient was treated with a combination of Haloperidol and Trihexyphenidyl, along with additional therapy to prevent recurrent infections. The patient's condition improved significantly, within one month of treatment initiation.

In cases where symptoms don't improve with symptomatic therapy, more advanced options like intravenous immunoglobulin (IVIg) or steroids may be considered. These therapies have shown some success in improving patient outcomes and may be especially useful in severe or persistent cases [7, 9]. Our patient was not administered IVIg due to the favorable outcomes observed with initial therapy, as well as the unavailability of IVIg at our healthcare facility located in a remote area.

Aspirin was given to our patient to manage arthralgia symptoms and may potentially lower the risk of arthritis through its antioxidant properties and [13]. The previously recommended aspirin dose of 80-100 mg/kg/day in divided dose has been revised to 50-60 mg/kg/day due to its potential toxicities, including gastrointestinal irritation and tinnitus. Although up-titration may be necessary based on clinical response. The dosage is gradually tapered as symptoms improve and typically continued for 1-2 weeks following symptom resolution [14].

Clinical resolution in 80% of patients occurs within 2 years, but some continue to have persistent condition. Episodes of relapse are typically linked to new streptococcal infections, hormonal exposure, and pregnancy [2]. Our patient demon-

strated gradual improvement in choreiform movements within one month of initiating treatment. Continued follow-up in the pediatric polyclinic is recommended to ensure appropriate long-term management and to implement preventive measures aimed at reducing the risk of future streptococcal infections and subsequent recurrences. This recommendation is supported by recent data showing that 76.7% of patients achieved complete resolution of chorea within a median duration of two months (1-3 months); however, 43.5% of these patients later experienced relapse. These findings underscore the importance of sustained clinical monitoring and long-term care in patients with Rheumatic chorea [15].

4. Conclusions

Rheumatic chorea (Sydenham chorea) represents a significant clinical manifestation of rheumatic fever, necessitating prompt diagnosis and management to avert complications such as rheumatic heart disease. In resource-limited and remote settings, establishing a diagnosis can be particularly challenging, especially in the absence of evident or clinically apparent cardiac involvement. This case underscores the importance of thorough clinical assessment, appropriate laboratory testing, and strict application of the Jones criteria to ensure diagnostic accuracy. Despite these diagnostic difficulties, effective treatment—comprising antibiotic therapy for secondary prophylaxis and symptomatic management of chorea—can result in favorable clinical outcomes. Moreover, early identification and prevention of recurrent streptococcal infections are vital to mitigating long-term complications in such environments. The case further highlights persistent barriers to optimal care, including limited public awareness regarding the urgency of timely medical intervention in remote communities. Addressing these challenges requires enhanced health education, improved access to specialized healthcare services, and consistent long-term follow-up to prevent the progression to rheumatic heart disease and reduce the overall disease burden in underserved populations.

Abbreviations

SC	Sydenham Chorea
RF	Rheumatic Fever
ARF	Acute Rheumatic Fever
ECG	Electrocardiogram
ASO	Anti-Streptolysin O
ESR	Erythocyte Sedimentation Rate
GABHS	Group A β-Hemolytic Streptococcus
CBGTC	Cortical-Basal Ganglia-Thalamo-Cortical
GABA	Gamma Aminobutyric Acid
IM	Intramuscular
IVIg	Intravenous Immunoglobulin

Declaration

This case report was conducted in compliance with ethical guidelines, and the patient's guardians provided informed consent for treatment and participation in this report. The authors declare no conflicts of interest. We extend our gratitude to the healthcare providers at our hospital for their dedication and care in managing this case. The report highlights the importance of timely diagnosis and treatment, particularly in resource-limited settings where access to diagnostic tools like echocardiography may be restricted.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this case report.

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