

Sydenham's Chorea as Presentation of Rheumatic Heart Disease

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INTRODUCTION

Rheumatic fever is now considered an acute non-suppurative inflammatory disease that follows group A Streptococcal infection of the throat. It has remained the most common cause of acquired heart disease in underdeveloped countries, in children and young adults. It has continued to be a major public health problem worldwide, but particularly in developing countries.¹ Sydenham's chorea is a late manifestation of acute rheumatic fever and can occur several months after group A β -hemolytic Streptococcus infections. From 1920 to 1950, more than half of the patients with rheumatic fever had Sydenham's chorea. The incidence decreased to less than five percent in more recent studies.² Although the incidence of acute rheumatic fever and Sydenham's chorea has declined significantly in developed countries, they are still serious health concerns in developing countries. A 13 years old female child presenting with features of chorea and subsequently diagnosed to have rheumatic heart disease is reported.

ABSTRACT

Sydenham's chorea is the most common type of acquired chorea in childhood which is a major neurological manifestation of rheumatic fever. We describe a 13 years old girl who presented with weakness and purposeless involuntary movements of upper and lower limbs. The symptoms slightly affected the child's daily activities and had an unstable gait on walking which was aggravated during stress. Grade II ejection systolic murmur was noticed on cardiovascular examination. Echocardiography evaluation showed thickened aortic and mitral valve leaflets with mild to moderate degree of mitral regurgitation. Anti-streptolysin O titer was positive (≥ 200 IU/ml). CT scan of brain was normal. Subsequently child was diagnosed as Rheumatic heart disease with Sydenham's chorea and kept on regular Benzathine penicillin prophylaxis. Symptoms subsided spontaneously after 3 months without any further complications. Although decreasing, early diagnosis and management of Sydenham's chorea and Rheumatic heart disease are very crucial and should be considered with such presentation.

KEYWORDS

Carditis, penicillin prophylaxis, rheumatic fever, sydenham's chorea.

CASE-REPORT

A 13 years old female child from Falametar, Kavrepalanchowk presented in our pediatric outpatient clinic with weakness and involuntary movements of upper and lower limbs since last 3 months. Left sided extremities were affected more than her right side. The symptoms had slightly affected the child's daily activities and had an unstable gait on walking. Symptoms were aggravated by stress and did not persist during sleep. Parents don't recall the child suffering from any recent febrile illness or sustaining any major trauma. No history of any drug intake. No history of rashes or joint swelling or chest pain. There was no significant past medical history and no similar history in the family. Regarding her birth history she was a home delivery, and the perinatal period was uneventful. Child was fully immunized according to the EPI schedule.

On clinical examination child was afebrile, alert and oriented. Her vital signs were within normal limits. Her anthropometry measurements, growth and developments were appropriate for her age. Her higher mental function was normal and had no signs of cerebellar ataxia. She was

having bilateral involuntary, irregular and purposeless movements, mostly affecting the left sided extremities and also involving facial muscles with bilateral facial twitching. Child had bilateral flexed wrist joint with hyper-extended metacarpal joints. Strength and tone of the bilateral upper and lower extremities were found to have minimal weakness with hypo-tonia. Deep tendon reflexes were symmetrical and normal. Milkmaid's grip sign, darting tongue sign, pronator sign were positive and her hand writing were deteriorating. She had an unstable gait and frequently tried to put her left hand on her back in an attempt to prevent the involuntary movements. In cardiovascular examination 1st and 2nd heart sounds were heard normal and regular with a heart rate of 82/ min and blood pressure of 110/70 mmHg taken in the right arm. Grade II ejection systolic murmur was appreciated which was best heard in the apex, in the left 5th intercostal space medially to the mid clavicular line with no radiation. Other systemic examination was normal.

Sydenham's chorea was suspected, echocardiography was done and lab investigations were send for further evaluation. Echocardiography showed thickened aortic and mitral valve leaflets with mild to moderate degree of mitral regurgitation with ejection fraction of 61% and no pericardial effusion or vegetations were seen. Anti-streptolysin O titer was positive (≥ 200 IU/ml), ECG showed sinus rhythm with normal PR interval of 124 ms (upper limit for age and rate 160 ms), chest x-ray showed normal cardiac shadow with cardiothoracic ratio of 42%, CT scan of brain done to exclude other causes of chorea was normal, throat swab showed no growth, other lab investigations send were within normal limits including CRP < 5.0 mg/dl and ESR 18 mm/hr. In view of Rheumatic heart disease with Sydenham's chorea child was started on secondary antibiotic prophylaxis with Benzathine penicillin 1.2 million IU intramuscularly repeated every 3 weeks. The choreiform movements gradually subsided after 3 months spontaneously without any specific treatment and with no further complications. Child is on regular follow up for last 12 months on regular penicillin prophylaxis with no recurrence of symptoms.

DISCUSSION

There has been a decline in the incidence of rheumatic fever in developed countries, it is believed to be due to improvement in living standard, overall socio-economic improvement and the wide availability of penicillin.³ Rheumatic fever still continues to be a major public health problem in developing countries.

The most commonly used criteria for the diagnosis of rheumatic fever include the modified Jones criteria (table 1). To increase specificity, these criteria have been revised four times - in 1955, 1965, 1984 and 1992. The latest revision of the Jones criteria suggests that the probability of acute rheumatic fever is high when there is evidence of a preceding streptococcal infection, usually measured by elevation of the antistreptolysin O titre together with two

Table 1. Guidelines for the Diagnosis of Initial Attack of Rheumatic Fever

Major Manifestations	Minor Manifestations
Carditis	Clinical Findings:
Polyarthritis	Arthralgia
Chorea	Fever
Erythema marginatum	Laboratory Findings:
Subcutaneous nodule	Elevated acute phase reactants (ESR,CRP)
	Prolonged PR interval
SUPPORTING EVIDENCE OF ANTECEDENT GROUP A STREPTOCOCCAL INFECTION	
Positive throat culture or rapid streptococcal antigen test	
Elevated or rising streptococcal antibody titer	

*If supported by evidence of preceding group A streptococcal infection, the presence of two major manifestations or of one major and two minor manifestations indicates a high probability of acute rheumatic fever. CRP: C-reactive protein; ESR: erythrocyte sedimentation rate.

major manifestations (arthritis, carditis, chorea, erythema marginatum and subcutaneous nodules); or one major and two minor manifestations (fever, arthralgia, high C-reactive protein (CRP) or elevated erythrocyte sedimentation rate (ESR) and a prolonged PR interval on electrocardiogram).⁴

Arthritis is the most common manifestation, present in 80% of patients. Carditis occurs in 40-75% of patients in the first 3 weeks of the illness. Erythema marginatum and subcutaneous nodules are rare, less than 10% of patients are affected. Sydenham's chorea is also a rare presentation, affecting less than 5% of patients.⁵ Rheumatic carditis is the most serious consequence of the disease process while migratory polyarthritis and the neurologic manifestation as Sydenham's chorea may present in conjunction with carditis.

Sydenham's chorea was first described in 1686 by Thomas Sydenham, it is a major criterion for the diagnosis of acute rheumatic fever and according to the modified Jones criteria, its presence alone is sufficient to make this diagnosis.² Sydenham's chorea, also known as St. Vitus dance, is a neuropsychiatric manifestation of rheumatic fever with an incidence varying from 5 to 35%.⁶ The incidence decreased to less than 5% in more recent studies.⁵ It usually occurs after a latency of several months and hence the lab evidence of prior streptococcal infection may be absent at the time of presentation.⁶ Although it is reported to occur in a quarter of patients with rheumatic fever and rheumatic heart disease in some regions, it may also be the presenting symptom.⁷ Cardiac involvement occurs in 42-70.5% of cases with chorea.² Sydenham's chorea is a post-streptococcal autoimmune disease and it is the most common cause of chorea during childhood, it manifests in children aged 5-15 years. The major neurological features of Sydenham's chorea are involuntary movements, which are exacerbated by stress and disappear during sleep. Classical presentation includes rapid, purposeless, involuntary movements that are irregular, non-stereotyped and can affect all limbs, face, and trunk. Sydenham's chorea is usually bilateral and female predominant, hemi-chorea has been reported in only 15-20% of patients.⁸ Psychological and psychiatric manifestations can precede the onset of

chorea, such as depression, anxiety, personality changes, emotional lability, obsessive compulsive disorder, tics and attention deficit disorder.⁹ Usually the patient has abnormal neurological signs with hypo-tonia and motor restlessness which can lead to coordination problems, gait disturbances, speech impairment and difficulty in writing. As a result, the activities of daily living can be severely disrupted. Generalized weakness and hypo-tonia can be so severe that the patient becomes bedridden. This special form of Sydenham's chorea is called "chorea paralytica". The diagnosis relies on a careful clinical history and laboratory assessment to rule out other causes of the symptoms.² The duration of Sydenham's chorea can last for 4-6 months, however, it can range from as little as one week up to over 2 years.⁹ Although most patients recover completely, some may have persistent choreiform movements especially when they are under stress.

Sydenham's chorea is associated with antibodies against group A β -hemolytic streptococci that cross react with either neuronal extracellular cell surface and/or intracellular (cytoplasmic or cytoskeletal) antigens.¹⁰ In most studies it is suggested that, there is an evidence of basal ganglia dysfunction which can be showed by morphologic and functional neuroimaging techniques. Basal ganglia dysfunction can also present with cognitive impairment, characterized mainly by executive dysfunction.¹¹ Sydenham's chorea is a self-limiting condition with a mean duration of 2-4 months. However, treatment is necessary for patients whose chorea is not mild. Antiepileptics, neuroleptics and phenothiazines have been reported to reduce the abnormal movements by affecting the dopaminergic or alpha-aminobutyric acid pathways. Intravenous immunoglobulin, plasma exchange and corticosteroids effectively reduce involuntary movements due to its autoimmune pathogenesis. However, because of the side effects of corticosteroids, their use is recommended only in chorea paralytica. In a previous study, recurrence was observed in approximately 30% of patients and was associated with discontinuation of the antibiotic prophylactic therapy or poor compliance

and perhaps with subclinical damage to the basal ganglia following the initial episode.²

Although increased antistreptolysin O titers exist in two thirds of cases, it is not helpful in the diagnosis of Sydenham's chorea. There are no typically defined MRI features in Sydenham's chorea. MRI may show varying degrees of signal hyperintensity on T2-weighted images in focal regions, such as the corpus striatum, caudate nucleus, putamen and multiple other areas. These abnormalities may be localized to the basal ganglia, but they are often not consistent with the patient's clinical signs, it is thought to develop as a result of vasculitis or inflammation.² CT abnormalities, including low attenuation in the corpus striatum without mass effect and hypodensity of the right caudate, have been reported less frequently. Abnormalities that are seen on imaging of the central nervous system usually improve when chorea resolves; however, some times the injury is permanent. Because of the great variability of MRI and CT findings, neuroimaging cannot be used to diagnose Sydenham's chorea; they can be used only to exclude other causes of chorea.¹²

CONCLUSION

Acute rheumatic fever still continues to be a major public health problem in our country. Sydenham's chorea is a rare presentation of acute rheumatic fever. Early diagnosis and management of Sydenham's chorea and Rheumatic heart disease are very crucial. Long term follow-up and antibiotic prophylaxis are required to prevent its recurrence. Although rare in developed countries, Sydenham's chorea is still present in developing countries and should not be neglected.

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