



## Hemichorea: A Rare Manifestation of Acute Rheumatic Fever

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### ABSTRACT

*Acute Rheumatic Fever (Acute RF) is an immunological response to group A beta hemolytic streptococcal pharyngitis. There has been a decline in the incidence and severity of the disease in developed as well as developing countries. Sydenham's Chorea (SC) is one of the major clinical manifestation of Acute RF. The other major manifestations of Jones Criteria are carditis, migratory polyarthritits, subcutaneous nodules and erythema marginatum. We are reporting a rare case of a 9-year-old female child of Acute RF presenting as hemichorea.*

**Key Words:** *Sydenham's Chorea, Acute Rheumatic Fever, Hemichorea, Involuntary movements*

### INTRODUCTION

Sydenham's Chorea (SC) is reported in 1-8 percent of patients with Acute RF in Southeast Asia (worldwide incidence of 10-15 percent), and out of these patients only 15-20 percent have hemichorea.<sup>(1-5)</sup>

The disease was first named by Thomas Sydenham in 1686 as 'St. Vitus Dance' to differentiate it from dancing mania, a common

practice seen in the religious ceremonies in older days by those who danced to drive out prevalent epidemic illness.<sup>(6)</sup>

SC is commonly found in children between 5-15 years and is generally bilateral. It is twice as common in females as males.<sup>(2)</sup>

Chorea is a cardinal feature of Acute RF and is sufficient alone to make the diagnosis when other causes of chorea have been excluded. SC has

traditionally been considered to be a self-limiting benign disease, requiring no therapy. However, there are recent reports that a protracted course can lead to significant disability and/or social isolation.<sup>(12)</sup>

### CASE REPORT

A nine-year-old girl presented with two weeks history of acute onset, frequent involuntary, quasi-purposive, jerky movements of right upper and lower limbs. The movements worsened with anxiety and disappeared during sleep. It was associated with decreased scholastic performance, facial grimacing, difficulty in speech, deterioration of handwriting, walking and daily activities such as buttoning and feeding. She was also ridiculed in school for facial grimacing and abnormal movements by school teacher. The medical and family history was insignificant.

There was no history of fever, joint pain, exertional intolerance, chest pain, skin rash and subcutaneous nodules. Neurological examination showed hypotonia in right upper and lower limbs however power in all the muscles and deep tendon reflexes were normal. Signs of chorea such as

milkmaid sign (Figure 1), jack-in-the-box tongue (Figure 2) and pronator sign were present. Fundus examination of both eyes was normal and slit lamp examination did not show Kayser-Fleischer ring. Cardiovascular examination was within normal limit.

Laboratory tests showed normal ESR, ASO titers and C-reactive proteins. Chest radiograph, electrocardiography and 2D-echocardiography showed no evidence of carditis. Thyroid function test, anti-phospholipid antibodies, anti-nuclear antibodies, serum electrolytes, serum calcium, serum ceruloplasmin, urinary copper and magnetic resonance imaging of brain with angiography were unremarkable. The child was started on oral valproic acid (20mg/kg/day) and haloperidol (0.25mg/kg/day). She was also given one dose of intramuscular Benzathine penicillin (1.2 mega units) after test dose. She started responding within two weeks with complete resolution of chorea, normal daily activities and improvement in handwriting (Figure 3) in 4-6 weeks. Oral penicillin prophylaxis was initiated at the time of discharge.



Figure 1 – Milkmaid sign



Figure 2 - Jack-in-the-box tongue



Figure 3 – Showing improvement in Handwriting

## DISCUSSION

The onset of chorea in RF is usually 1-6 months after the episode of streptococcal pharyngitis explaining the lack of classical features of RF at the time of presentation<sup>(7,8)</sup>. SC is a neuro-psychiatric disorder consisting of choreiform movements, hypotonia, hyporeflexia and psychiatric symptoms like emotional lability and anxiety disorder<sup>(4,9)</sup>. It is proposed that when

genetically vulnerable individuals are attacked by group A beta hemolytic streptococcus, anti-streptococcal antibodies attack the basal ganglia causing inflammation and decreased blood flow<sup>(9)</sup>. When a child presents with chorea, preliminary evaluation for Acute RF should be focused on the cardinal features of modified Jones Criteria<sup>(6)</sup>. Hemichorea is involuntary, hypokinetic, unilateral disorder probably due to a decreased blood flow

to the contralateral basal ganglia<sup>(10,11)</sup>. It is a rare feature of Acute RF and may not strictly adhere to modified Jones Criteria. Our patient showed remarkable improvement in symptoms and signs post therapy.

Three special circumstances in which diagnosis of Acute RF can be made without strict adherence to Jones Criteria - <sup>(4,13)</sup>

1. Chorea – when other causes are ruled out.
2. Indolent carditis – insidious or late onset carditis.
3. Rheumatic Fever recurrence – in the presence of documented Rheumatic heart disease, even the presence of one criterion in the presence of supportive evidence of previous streptococcal infection suggests recurrence.

In (1) and (2) supportive evidence of previous streptococcal infection is not mandatory to diagnose Acute RF.

The other causes of chorea in children are Systemic lupus erythematosus, drug-induced (phenytoin, carbamazepine, lithium, L-dopa & phenothiazines), thyrotoxicosis, Wilson's disease and metabolic causes (hyponatremia, hypocalcemia)<sup>(8,14)</sup>.

In our knowledge, the only similar case reported by far was in Taipei in 2005. That patient had carditis, increased PR interval along with raised ASO titres and hemichorea.<sup>(15)</sup>

These patients of chorea are ridiculed in school and beaten at home (due to facial grimacing & clumsiness) with deterioration in handwriting.

Assessment of handwriting in chorea is an important tool to monitor patient's clinical status. Early diagnosis of the disease and timely

intervention helps the patient gain their lost confidence and self-esteem. To conclude, hemichorea as a sole feature of RF is rare but not uncommon especially in developing countries.

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