Sydenham Chorea with a Rare Presentation

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Rheumatic fever is delayed sequel to upper respiratory tract infection with group A Beta hemolytic streptococci rheumatogenic strains; the gram positive organism having M protein in their outer protein layer which determines virulence of organism\(^1\) and stimulates formation of opsonizing and precipitating antibodies which not only impedes phagocytosis but also result in diffuse auto-immune inflammatory disease of connective tissue that mainly involves heart, blood vessels, joints, CNS and sub-cutaneous tissues\(^1\). Symptoms usually begin 5-30 days after streptococcal sore throat and then one or more major system is involved mostly joints and heart.

This disease has been on declining trend in western countries and in America but this is not true for developing countries where although burden of disease might have reduced than seen in earlier 20\(^{th}\) century but it still is a major concern for health professionals throughout the third world countries like Pakistan\(^3\). Not only incidence of chorea is highly variable amongst different studies and populations but clinical features of chorea also show considerable variation. Some studies state that its incidence is decreasing while others indicate that prevalence of disease is way more than what is actually being reported although this case report is being written because of its unique presentation.

**Case Report**

A Four year old boy presented with complaints of frequent falls and abnormal movements for last 10 days. He had experienced falls while playing ‘walking’ during routine daily activities and even on standing still although he had not suffered from any fractures. Abnormal movements involving his whole body were irregular, rapid and semi-purposeful which continued during rest or periods of inactivity but settled during sleep. These complaints were associated with emotional lability explained by uncontrolled and un-provoked episodes of crying and laughing. This whole symptom complex was preceded by low grade fever. There was no history of sore throat, joint pains, rash, fits, headache, vomiting, chest pain, palpitations, cyanosis, edema, jaundice, abdominal distension, and fluctuating consciousness. His past medical and drug history was unremarkable. He was born at 7\(^{th}\) months of gestation by spontaneous vaginal delivery; no complications in ante-natal, natal and post-natal period. Feeding, vaccination and developmental history was un-remarkable. His anthropometric measures were between 25\(^{th}\) to 50\(^{th}\) percentiles and were plotted on standard growth chart according to age and gender. Patient was 12\(^{th}\) among sib ship of 12; belonged to low socioeconomic class and lives in an over-crowded place. On examination, patient was conscious and alert with rapid involuntary semi-purposeful jerky movements of whole body, hemodynamic ally stable and afebrile with unremarkable cardiovascular, respiratory and gastrointestinal examination. His higher mental functions and cranial nerves were intact with no signs of meningeal irritation. Patient had explosive bursts of dys-arhythmic speech; generalized hypotonia and hyporeflexia with decreased muscle power (4/5). Milk-maid sign, pronator sign, spooning, wormian darting tongue movements, clumsy gait; all these signs were positive leading us to our differentials of Rheumatic chorea, Wilson disease, Systemic lupus erythematos, Huntington disease. Investigations ordered included complete blood count, chest X-ray, liver function tests (LFTs), echocardiography, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), antibodies to streptolysin-O titer (ASOT), throat swab, anti-nuclear antibodies (ANA), anti-Double stranded Antibodies, serum ceruloplasmin and urinary copper estimation. All the ordered test reports came out to be normal except raised ESR, CRP, ASOT; providing us with our final diagnosis of Rheumatic chorea also known as Sydenham chorea or St. Vitus dance made on basis of his chorea, raised ESR, history of fever and ASOT levels above 2000( normal<300). He was started on benzathine penicillin injection I.2 million unit's i/m and haloperidol 0.02 mg/kg/day BD and was advised benzathine penicillin injection after every 3 weeks for secondary prophylaxis. On his follow-up after 3 weeks, choreiform movements had improved with marked improvement in his gait and speech but cardiac examination on follow-up showed tachycardia with visible pulsations in apical area, muffled first heart sound; pan-systolic murmur grade IV at apex radiating to axilla. Echocardiography showed mitral regurgitation and pan-carditis. Final diagnosis of Acute Rheumatic Fever with Chorea and Carditis was...
established and was managed by anti-inflammatory drug aspirin; initially 70mg/kg/day 6 hourly for 5 days then 50mg/kg/day 6 hourly for 3 weeks and then 25mg/kg/day 6 hourly for 3 weeks along with secondary prophylaxis by penicillin.

Discussion
Sydenham chorea is the neurological manifestation of rheumatic fever. Cross-reactive anti basal-ganglia antibodies are formed in predisposed individuals leading to neurological insult. Most of literature concerning Sydenham chorea is very old whereas disease itself has modernized, has become less common, less severe with less frequent relapses. Coming onto comparing features of our case with other reported cases, Sydenham's chorea mainly affects female patients, the female predilection of chorea is an almost constant finding but in our case, patient was a boy. Second most important unique feature is the age; this disease is mostly seen in those aged 5-15 years but age of our patient was less than 5. Third unique feature is the pattern of disease; younger children less than 5 years old are more likely to have carditis and arthritis and less likely to have chorea but chorea was the prime presenting complain in our case. Sydenham chorea is more prevalent in low socio-economic conditions as in our case. 2% of children may become bed-ridden called as chorea paralytic but prognosis in our case was very encouraging. There was no motor impairment, tics or disability. Psychological symptoms included only emotional labiality but no obsessive compulsive disorder, cognitive defects, personality changes and that too improved with treatment. The youngest child with Sydenham's chorea who was aged three and half years; case report of India published in 2009 is comparable to our case report. Usually it is a self-limiting disease with remission in a week or two but symptoms may wax and wane for up to 10 years.

References