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Case Report

Sydenham's Chorea

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Abstract
Sydenham's chorea is a delayed complication of group Aß haemolytic streptococcal infections and forms one of the major criteria of acute rheumatic fever. It is characterised by chorea, muscular weakness, and a number of neuropsychiatric symptoms. It is considered to be an autoantibody mediated disorder with the evidence suggesting that patients with Sydenham's chorea produce antibodies that cross react with streptococcal, caudate, and subthalamic nuclei. However, documented evidence of previous streptococcal infection is found in only 20%–30% of cases. It has a good prognosis for full recovery so treatment is not warranted in most cases. Following case is a high school student presented with progressive changes in his handwriting during two months before. Chief complaint of this 15-year-old boy was difficulty in writing at classroom. His physics notebook has been shown as interesting figures.
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Keywords ● Sydenham's chorea ● acute rheumatic fever ● handwriting

Introduction

Sydenham's chorea is a neurological movement disorder characterized by irregular, abrupt, relatively rapid involuntary movements of muscles of the face, neck, trunk, and limbs.[1] It most frequently occurs in children or adolescents between the ages of 5 to 15 with female preponderance following acute rheumatic fever (ARF). Choreic movements usually begin gradually, progressively worsening over a few weeks to a month. Associated findings may be extremely variable, ranging from relatively mild incoordination to severe disruption in conducting voluntary movements of multiple muscle groups, potentially affecting speech, arm movements, walking, and the ability to perform certain activities of daily living.[1-3]

Case Report

A 15-year-old boy presented with difficulty in writing and some purposeless involuntary movements in his face and hands. He was visited in rheumatology clinic on November 28th 2005. He was a high school student and complained of progressive changes in his handwriting during two months before (figures1-4). Last February, he had a bad throat and lumps in the neck that was treated with diagnosis of streptococcal pharyngitis. Then after about two weeks he involved migratory polyarthritis in his large joints that was kept him in bed for six days. His temperature was 38.4°C ,ESR was 70mm/h, ASO was 950 U, CRP was positive and there was leukocytosis with left shift. On that time according to Jones criterion he was

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Figures 1-4: The physics notebook of presented case: show progressive and interesting changes in his handwriting.

treated with diagnosis of acute rheumatic fever. His family history for rheumatic fever was negative.

Now he has some involuntary movements in hands; his face also similarly affected. He has been restless at night but when asleep there is no twitching. The legs are to a less extent affected. The right side of the body seems most affected. All other musculoskeletal and neurological examinations including cerebella, sensory system and pyramidal tract were normal. Sphincters act properly either bladder or anal. Intelligent, emotional, memory was good. Joints not swollen and in heart auscultation grade II systolic murmur was heard in left sternal border. Mild mitral regurgitation in echocardiography was detected. Erythrocyte sedimentation rate was 11mm/hour, ASO titer was 230^{U} and CRP was negative. This young boy treated with diazepam as Chorea minor and benzathine was utilized as a prophylaxis for relapses of rheumatic fever. His short term follow up after six weeks showed diminished chorea movement and he obtained full recovery after 4 months.

Discussion

Sydenham's chorea was described in the medical literature in 1686 by Thomas Sydenham.[1] The disorder has also been referred to as St. Vitus' dance, Acute chorea, chorea minor, Rheumatic chorea. It is considered a neurological complication following infection with group A beta-hemolytic streptococci.[1-3]

In about 20% of cases, Sydenham's chorea occurs as the only manifestation of acute rheumatic fever, however, in others, it develops as a late feature of ARF following other characteristic manifestations, such as fever and migratory polyarthritis.[4] Choreic movements usually developing by 1 to 6 months (or more) following pharyngitis.[1] In most children, irregular, involuntary, jerky movements may initially appear as increasing awkwardness or clumsiness, such as difficulty writing as described in our case. More specifically, the choreic movements consist of relatively fast, irregular, uncontrollable, jerky motions that disappear with sleep and may increase with stress, fatigue, excitement, or other factors. The neuromuscular abnormalities associated with Sydenham's chorea may lead to facial grimacing, significant deterioration in handwriting (in school-aged children), slight or significant difficulties dressing, feeding, and walking, slurred, and slowed speech.[1-4] Associated symptoms may tend to begin relatively subtly, progressively worsen over a few weeks to months (usually over 2 to 4 weeks), and gradually spontaneously resolve within approximately 3 to 6 months. In spite of the more prevalence of Sydenham's chorea is in females particularly in the years around puberty we report it in a young boy. Sydenham's chorea appears to result from an autoimmune or antibody-mediated inflammatory response involving certain regions of the basal ganglia. Antibodies against group A beta-hemolytic streptococcus cross-react with basal ganglia.[5-7] Positron emission tomography (PET) scanning has shown increased glucose metabolism within major substructures of the basal ganglia, a finding that was reversed with clinical improvement.[8,9] In addition, magnetic resonance imaging (MRI) has shown abnormally increased size of the caudate nuclei, the globus pallidus, and the putamen.[10] A diagnosis of Sydenham's chorea is primarily based upon a thorough clinical evaluation, detection of characteristic symptoms and a careful patient history. Once a diagnosis of Sydenham's chorea is considered, a thorough cardiac evaluation should also be conducted to rule out or confirm possible cardiac involvement. Many individuals with Sydenham's chorea may have negative results of CRP, ESR and have normal ASO. Patients with chorea are exception to the Jones criteria.[1,3] As Sydenham's chorea may spontaneously resolve or not cause significant functional impairment, treatment with certain medications, such as dopamine antagonists, neuroleptics, tetrabenazine or valproic acid should be avoided unless associated chorea is functionally disabling and severe as in generalized form. Patients who have Sydenham's chorea should receive ongoing therapy to help prevent recurrences of rheumatic fever.[2-4]

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