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Recurrence of Sydenham's Chorea

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Dear Editor,

Sydenham's chorea (SC) is an autoimmune movement disorder, which develops following group A beta-hemolytic streptococcal infections (GABHS) and is one of the major diagnostic criteria for acute rheumatic fever (ARF). Chorea is characterized by involuntary, short-term, erratic movements.

SC is the most frequent cause of acquired chorea in children and often affects children aged between 5-15 years. Usually, it is a benign, self-limiting disease, and spontaneous remissions are common. Children with persistent or recurrent chorea have high morbidity (1).

A 13-year-old boy was admitted with the complaints of getting fidgety, "wiggling" movements of both hands, dropping objects such as forks, spoons, and pencils from his hands, difficulty in writing, and stumbling feeling of right or left foot while walking, for 20 days. It was also noted that he had a feeling of restlessness and an urge to move at school, difficulty in writing his homework, and lack of communication with his friends during this period. It was also learnt that he had been diagnosed as ARF presenting with arthritis 2 years ago and 2 months following ARF, he presented with widespread involuntary movements predominantly affecting his left arm and leg, which were diagnosed as chorea and his symptoms improved with 0.8 mg/d haloperidol treatment for a couple of months. He also received penicillin G benzathine 1,200,000 IU injections for one year. When he was admitted to the hospital with arthralgia in his right knee 2 months ago, the pediatrician prescribed penicillin G benzathine 1,200,000 IU to be injected in every three weeks again. There was no movement disorder history reported in his relatives.

In neurological examination, erratic, rapid and involuntary choreiform movements in the distal parts of extremities, predominantly in upper extremities and especially on the left side, "milkmaid's grip" and mild hypotonia in both hands were observed. Laboratory tests including glucose, electrolytes, liver-kidney and thyroid function tests, lipid profile, sedimentation, C-reactive protein, creatine phosphokinase, hemogram, iron, total iron binding capacity, ferritin, copper and ceruloplasmin were all normal. Serological tests including rheumatoid factor, antinuclear antibody, and anti-double stranded DNA (anti-dsDNA) were negative, and the antistreptolysin-O (ASO) titer was 285 IU/mL (normal: 0-200 IU/mL). Electrocardiography, echocardiography, chest X-ray and cranial magnetic resonance imaging (MRI) were normal. He was diagnosed as having a recurrence of SC and haloperidol treatment was initiated. In the follow-up, haloperidol was discontinued due to improper use and unresponsiveness and valproic acid 500 mg/d was initiated. At the 2-month follow-up, clinical improvement was observed.

SC is one of the major diagnostic criteria of ARF and is sufficient for diagnosis alone. Primarily, it is seen in populations with untreated streptococcal infections. Cross-reaction between antibodies against streptococcal antigens produced following GABHS infections and basal ganglia antigens in children with genetic susceptibility is the main pathogenetic mechanism (2). In particular, antibodies against neurons in the caudate nucleus support this immune reactivity hypothesis. As a result,

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imbalance between dopaminergic and cholinergic systems causes involuntary movements (3). It was shown that the levels of gamma-aminobutyric acid and acetylcholine decrease and dopaminergic activity increases in the basal ganglia, which also may explain the mechanism for action of antiepileptic and neuroleptic drugs (4).

Chorea usually occurs several months after a GABHS infection and the ASO level is normal or slightly elevated in this period. The main characteristics of SC are chorea, hypotonia, dysarthria, and emotional lability. Difficulties in school life, irritability, and obsessive-compulsive behaviors can be seen. Chorea can be unilateral at first but becomes generalized eventually in most patients. Clinical features are the determining factors for the diagnosis of chorea (2). Systemic lupus erythematosus-associated chorea, drug-induced chorea, hyperthyroidism, Wilson's disease, antiphospholipid antibody syndrome (1,2) and pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) should be kept in mind for differential diagnosis (3). Hyperintensity in the putamen and globus pallidus in T2-weighted cranial MRI sections can be seen during the disease period and it disappears with improvement of the disease (2).

SC is mostly a benign disease with complete recovery (1,2). Treatment is necessary in patients who are affected moderately or severely (4). Antiepileptic drugs (valproic acid, carbamazepine), neuroleptic drugs (pimozide, haloperidol) and benzodiazepines are used in treatment (2,3). Immunotherapies including corticosteroids, intravenous immunoglobulins and plasmapheresis may be used in the treatment in selected patients (3). High-dose penicillin treatment for 10 days against active streptococcal infection and prophylactic penicillin treatment until the age of 21 are recommended (2).

Chorea recurrence is defined as re-emergence of chorea at least 2 months following the first episode and lasting more than 24 hours. Korn-Lubetzki et al. (5) reported the rate of chorea recurrence as 42%. Recurrence may be observed months and sometimes years after the first episode. Recurrence may be due to a basic abnormality that sensitizes the patients or a permanent subclinical damage to the basal ganglia following the first episode (5). Irregular antibiotic prophylaxis, inability to reach remission in 6 months, and persistence of symptoms longer than 1 year were reported as risk factors for chorea recurrence in a study (1).

In conclusion, ARF and SC are still frequently seen in our country. SC should be considered primarily in the differential diagnosis of school-age onset chorea.

Ethics

Informed Consent: Parent of involved to this study has given the informed consent.

Peer-review: Internally peer-reviewed.

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