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A Disease to Remember: Sydenham's Chorea

Hatırlanması Gereken Bir Hastalık: Sydenham's Koresi

Fizik Tedavi ve Rehabilitasyon

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Özet

Sydenham'ın koresi, A grubu β-hemolitik streptokok enfeksiyonlarından sonra gelişen bir otoimmün hareket bozukluğu olup, çocuklukta edinilmiş korenin en yaygın nedenidir. Bu raporda, sık düşme atakları, sağ alt ve üst ekstremitelerde anormal hareket paternleri, konuşma ve yazmada bozulma, düşük akademik performans ve duygusal değişiklikler öyküsü ile fizik tedavi polikliniğine başvuran bir hastayı sunuyoruz.

Anahtar kelimeler: *Sydenham'ın koresi*, *A grubu β-hemolitik streptokok*, *Nörolojik hastalık*

Abstract

Sydenham's chorea is the most common cause of acquired chorea in childhood and is an autoimmune movement disorder developing after group A β-hemolytic streptococcus infections. In this report, we present an patient who was admitted with a history of frequent falling attacks, abnormal movement patterns in the right lower and upper extremities, impaired speech and writing, decreased academic performance, and emotional changes.

Keywords: *Sydenham's chorea*, *group A β-hemolytic streptococcus*, *neurological disease*

Introduction

Sydenham's chorea (SC) is the most common cause of acquired chorea in childhood [1,2] and is an autoimmune movement disorder developing after group A β-hemolytic streptococcus (GABHS) infections [3]. This clinical condition is among major diagnostic criteria of acute rheumatic fever (ARF) and is characterized by choreiform movements, hypotonia, and emotional changes. The diagnosis is often made clinically and there is still no confirmation test used in clinical practice [4]. Initial manifestations of CS include attention deficit, increased physical activity, dropping objects frequently, decreased academic performance, impaired writing, clumsiness, poor relationships with peers, impaired speech, and frequent falling attacks [5].

In this report, we present an SC patient who was admitted with a history of frequent falling attacks, abnormal movement patterns in the right lower and upper extremities, impaired speech and writing, decreased academic performance, and emotional changes.

Case Report

A 13-year-old girl presented to the physical therapy outpatient clinic with a one-month history of involuntary movements in the right lower and upper extremities, falling objects frequently, slurred and impaired speech, poor relationships with peers, and frequent falling attacks. The patient had been diagnosed with tonsillopharyngitis two or three months earlier. Physical examination showed abnormal movement patterns in the right lower and upper extremities and ataxic gait. Cerebellar tests were positive. Moreover, the patient had a slurred and impaired speech.

Laboratory parameters were as follows: white cell count (WBC) 8.5 /mm^3, hemoglobin 12.62 gr/dL, platelet

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308,000/mm^3, erythrocyte sedimentation rate (ESR) 9 mm/h, and C-reactive protein (CRP) 0.134 mg/dl. Liver and kidney functions tests, creatinine kinase, electrolytes, and urinalysis were normal. The anti-streptolysin O (ASO) level was 531 IU/ml. Antinuclear antibody, anti-dsDNA, anticardiolipin, antiphospholipid, p-ANCA, c-ANCA, Borrelia burgdorferi, herpes simplex virus, and hepatitis were negative. Ceruloplasmin, ferritin, plasma levels of lead and copper, thyroid function tests, urinary and plasma amino acid analyses, and spinocerebellar and spinal muscular atrophy gene analyses were normal. Transthoracic echocardiography findings for carditis or silent endocarditis were normal. In addition, no abnormality was detected in cranial magnetic resonance imaging (MRI) and electroencephalography.

Based on clinical findings, the patient was diagnosed as having SC and was initiated amoxicillin+calavulanic acid 400/57 mg suspension for 10 days; 5 drops of haloperidol therapy and coordination and balance exercises. After this therapy, the patient didn't improvement. Then, 300 mg/twice of day valproic acid therapy was initiated. At 3-month follow-up, the complaints regressed.

Case Discussion

Sydenham's chorea was first described by Thomas Sydenham in 1686 and the relationship between SC and streptococci was documented in later years [6] . SC is typically characterized by motor and mental symptoms including choreiform movements in the extremities, hypotonia, hastiness, attention deficit, and emotional, character, and behavioral changes. In 1992, SC was accepted as a major diagnostic criterion of ARF and was described in the Revised Jones Criteria [7,8]. Although its exact etiology remains unclear, SC is considered to result from secondary immune reactions against the central nervous system (CNS) including the basal ganglia [9].

SC is more common in girls aged 5-15 years. Due to the long latency period between the inciting streptococcal pharyngitis and the onset of clinical signs of chorea, SC may not be accompanied by the other findings of ARF [10,11]. Our patient had been diagnosed with tonsillopharyngitis two or three months prior to admission and her complaints had emerged one month earlier.

Chorea refers to rapid, irregular, purposeless, and involuntary movements that begin in the fingers and hands and spread to the arms, legs, and trunk and also affect the face and tongue. It usually involves one side of the body and then spreads to the other side. Cocontraction of the agonist-antagonist muscles and the abrupt, purposeless, involuntary jerking movements in the extremities lead to dropping objects frequently, limping, and frequent falling attacks. In addition, daily life activities such as buttoning, dressing, undressing, and writing are impaired due to the deterioration of fine motor skills [4,5] . A chorea can be accompanied by neuropsychiatric disorders, which may lead to deterioration in the child's relationship with family members and peers. In severe cases, chorea can be accompanied by psychosis, attention deficit, hyperactivity disorder, obsessive-compulsive disorder, or severe tic symptoms [2,12] . Moreover, it worsens with emotional changes such as stress and disappears in sleep [13-15] .

Patients with SC should receive high-dose penicillin for 10 days and should continue preventive prophylaxis until the age of 21 against active streptococcal infections[16] . Since it is a self-limiting disease, most of the mild cases heal without treatment [17]. Treatment is generally preferred in moderate-to-severe cases. Choreiform movements can be treated with valproic acid, carbamazepine, or haloperidol therapy [18]. Treatment options for severe cases include corticosteroids, intravenous immunoglobulins, and plasmapheresis[19-21] . In our patient, the administration of haloperidol therapy provided beneficial outcomes.

Differential diagnosis of SC includes various clinical conditions such as systemic lupus erythematosus, Wilson's disease, hemochromatosis, drug reactions, cerebrospinal ataxias, metabolic diseases, and porphyria. Additionally, increased ASO values and increased metabolic activity in basal ganglia detected by MRI or positron emission

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tomography during the active phase of the disease support the diagnosis of SC [11,22] . In our patient, ceruloplasmin, ferritin, spinocerebellar ataxia gene mutation analysis, and serology tests and the MRI findings were normal, which could be associated with the fact that the patient was not in the active phase of the disease.

In conclusion, GABHS infections are highly common in our country. SC should be considered in the differential diagnosis of these infections, particularly in school-age patients presenting with a history of impaired communication and motor skills.

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