

Isolated Hypoglossal Nerve Palsy in Behcet's Disease

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Abstract

Isolated hypoglossal nerve paresis as a manifestation of Behcet's disease has not been reported. We describe a 42-year-old man with isolated unilateral hypoglossal nerve paresis, who had had a five-year history of recurrent orogenital aphthae and relapsing arthritic manifestations. It is suggested that the nerve disorder represents a form of mononeuritis that may occur in Behcet's disease.

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Key words: Cranial nerve, Hemiglossal atrophy, Mononeuritis, Multisystem inflammatory disorder

Introduction

Behcet's disease is an idiopathic multisystem inflammatory disorder of young adults, most common in the Orient, Middle East and Mediterranean countries.¹ Involvement of the central nervous system occurs in 10% to 25% of cases,² usually following systemic manifestations by 1 to 10 years.³ The neurological manifestations may include aseptic meningitis, seizures, confusional state, bulbar or pseudobulbar palsy, pyramidal tract abnormality, cerebellar ataxia, ocular palsies, pseudotumour cerebri and cerebrovascular disease. However, isolated unilateral hypoglossal nerve dysfunction as a manifestation of Behcet's disease has not been reported. We describe a male patient with a five-year history of systemic manifestations of Behcet's disease, who was found to have isolated right hypoglossal nerve paresis, despite the treatment with systemic corticosteroid, azathioprine and cyclosporin A.

Case Report

A 42-year-old Chinese man had recurrent aphthous oral and pharyngeal wall ulcers, scrotal ulcers, arthralgia and joint swelling involving the knees for 5 years, despite treatment with oral prednisolone and azathioprine. He was also shown to have needle puncture pustules. In March 1996, cyclosporin A 100 mg daily was added to the treatment regime. In July 1996, he contracted a prolonged analgesic-refractory headache. He had no associated nausea or vomiting, nor any visual

symptoms. Furthermore, he complained that his tongue movement became clumsy. He was found to have right hypoglossal nerve paresis as evidenced by the presence of right hemiglossal atrophy, fasciculus and deviation of the tongue to the right on protrusion. The other cranial nerves were normal. Besides a large ulcer of the tongue and the scrotum, there were no additional abnormalities detected. Computerised tomographic (CT) scan and subsequent magnetic resonance imaging (MRI) of the head did not reveal any abnormalities or lesions compressing the hypoglossal nerve or lesions in the medulla such as a schwannoma or an aneurysm. An electromyographic (EMG) examination of the right genioglossus muscle showed presence of fibrillations and positive sharp waves, and motor units with a fast firing rate. Lumbar puncture opening pressure was 140 mm water. Cerebrospinal fluid (CSF) analysis revealed normal white cell count, protein 0.72 g/l, glucose 3.1 mmol/l (serum glucose 7.1 mmol/l), chloride 111 mmol/l, no oligoclonal bands, negative bacterial and fungal cultures. The sedimentation rate was 25 mm in 1 hour, while serum protein electrophoresis, immunoglobulins, C3 and C4 complements, pattern reversal visual evoked potential (VEP), brain-stem auditory evoked potential (BAEP) and medial and tibial somatosensory evoked potentials (SEPs) were normal. The other tests including CSF and serum fluorescent treponema antigen (FTA), rheumatoid factor (RF), antinuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA), and IgA capsular

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antigen of Epstein-Barr virus were negative.

He was treated with a non-steroidal anti-inflammatory drug (NSAID), and his headache gradually subsided. The hypoglossal nerve paresis persisted.

Discussion

Behcet's disease is an idiopathic inflammatory disorder, and thought to be due to an autoimmune vasculitis involving the small blood vessels.⁴ A recent study suggests that the Behcet's disease specific peptides stimulate the gamma delta T cell population.¹ The clinical course of Behcet's disease is very variable. Patients usually first notice mucocutaneous lesions, arthritic manifestations and ocular lesions, whereas neurologic involvement is usually delayed in onset if it occurs at all. Involvement of the central nervous system occurs in 10% to 25% of cases, on average from 1 to 10 years after onset of other manifestations,³ and is believed to be one of the major causes of death in Behcet's disease.⁵

Neurologic manifestations of Behcet's disease may include meningoencephalitis, seizures, bulbar or pseudobulbar palsy, pyramidal tract lesions, cerebellar ataxia, ocular palsies, strokes and pseudotumour cerebri. To our knowledge, isolated unilateral hypoglossal nerve paresis has not been reported. Herein we describe a male patient with Behcet's disease who developed the iso-

lated hypoglossal nerve paresis five years after the onset of orogenital aphthae and arthritic manifestations. A normal MRI, the presence of lower motor neuron signs and neurogenic changes in EMG support the hypoglossal nerve dysfunction as a form of mononeuritis. Other possible causes such as an aneurysm or a schwannoma have been excluded. In spite of the treatment of the patient with systemic corticosteroid, azathioprine and cyclosporin A, the development of the nerve disorder could not be prevented. The response of the disease to systemic steroids and immunosuppressive agents is unpredictable.

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