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Case Report

Amyotrophic Lateral Sclerosis in a Patient with Behçet's Disease

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Abstract
Behçet's disease is a multisystem vasculitis. Its neurological involvement mostly includes parenchymal and non-parenchymal central nervous system manifestations. Peripheral nervous system presentations are rare. A 32-yr-old male patient who fulfilled the international study group criteria for Behçet's disease, referred to our center with walking difficulty and repeated falling downs. Neurological examination revealed weakness, wasting and fasciculation of distal muscles associated with corticospinal signs. Neuroimaging data were insignificant but electrodiagnostic studies were in favor of motor neuron disease. To best of our knowledge, this is the first report of association of amyotrophic lateral sclerosis (ALS) and Behçet's disease.
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Keywords ● Behçet's syndrome ● amyotrophic lateral sclerosis ● Iran

Introduction

Neurological manifestations of Behçet's disease mainly include parenchymal and non-parenchymal involvement of central nervous system. Peripheral nervous system presentations are relatively rare. Herein, we report a case of Behçet's disease with amyotorphic lateral sclerosis (ALS).

Case presentation

A 32-yr-old Iranian man presented with complaint of weakness of hands and legs, walking problems and repeated fallings. He was admitted in neurology ward of the Nemazee Hospital of Shiraz University of Medical Sciences, in Shiraz, south of Iran. His history went back to two years before admission which he developed decreased vision of left eye and uveitis was detected by ophthalmologist. With obtaining the history of recurrent oral and genital ulcers, acne-like cutaneous lesions, polyarthralgia; positive pathergy reaction and consistent biopsy of tongue ulcers diagnosis of Behçet's disease had been considered for him. He was on oral cyclosporine 200mg three times a day, azathioprine 100mg/day and prednisolone 10mg/day at the time of admission. He developed weakness and wasting of muscles of distal extremities since nine months prior to admission, which become associated with walking difficulties and falling downs.

He had no significant personal, occupational and family history. In general examination he had only decreased visual acuity of left eye. In neurological examination, he had fasciculation of muscles of the tongue, shoulder arms and decreased strength

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of muscles of proximal and specially distal extremities, profuse atrophy of muscles of hand and foot (specially in first dorsal interossei), generalized hyperreflexia and bilateral Hoffman's and Babinski's signs and clonus of ankle, preserved abdominal reflex and normal sensory examinations.

Routine biochemistries, urine analysis, creatine kinase level, liver and thyroid function tests, all were normal. Hematologic studies only showed slight leukocytosis but normal sedimentation rate and C-reactive protein. Rheumatoid factor, LE cell, anticardiolipin antibodies, anti neutrophil cytoplasmic antibodies and syphilitic serology test were negative.

Anti GM1 antibody and hexosaminidase level cannot be measured in our center. In HLA typing, he was positive for HLA-B51 but negative for HLA-B27 and -B8. Brain, cervical and thoracic MRI showed no abnormality and lumbosacral MRI revealed only mild degenerative changes of intervertebral disks. Nerve conduction studies and late responses showed no abnormality but electromyography revealed increased insertional activity, fibrillations, positive sharp waves and fasciculations and high amplitude polyphasic motor unit potentials in proximal and distal appendicular and paraspinal muscles.

The patient was discharged with the augmented dosage of the above drugs and colchicine one mg daily. Then he discontinued his medication and became completely bedridden. After one year he expired due to respiratory infection but his family disagreed with autopsy. The written consent was taken from his family for this report.

Discussion

Our case fulfilled both El Scorial criteria for ALS,[1] and international study group criteria for Behçet's disease.[2] Corticospinal signs are of the most common presentations of neuro-Behçet's disease.[3] There is also at least one report of neurogenic muscular atrophy associated with disease that was confirmed by electrophysiologic, histologic and histochemical studies.[4] Furthermore, association of ALS and other autoimmune diseases like systemic lupus erythematosus,[5] scleroderma,[6] and Wegener's granulomatosis,[7] were previously reported. But as far as we know, this is the first report of association of Behçet's disease and ALS.

One can encounter similarities in pathogenesis of ALS and neuro-Behçet's disease. In immunologic studies CSF IL-6 may be elevated in both ALS and neuro-Behçet's disease.[8,9] Increased serum TNF-α is also reported in both ALS and Behçet's disease.[10,11] Superoxides which are the probable cause of cell damage in ALS,[12] were also reported to contributing in pathogenesis of Behçet's disease.[13] All of the above evidences made us to think that association of ALS and Behçet's disease, seen in this case, was not fortuitous and a causal relationship may be present.

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