

Isolated lesion of the cervical spine in a case of neuro-Behçet

Murat Terzi, MD, Hande Türker, MD, Nilgün Cengiz, MD, Musa K. Onar, MD.

ABSTRACT

Behçet's disease is characterized by small vessel vasculitis and involvement of multiple systems. In neuro-Behçet cases, spinal cord involvement is approximately 10-18%. Spinal cord lesions are often associated with cerebral or brainstem lesions, and isolated spinal cord involvement in Behçet's disease is very rare. Here, we report a case with an isolated lesion of the cervical spine.

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From the Department of Neurology, Ondokuz Mayıs University School of Medicine, Samsun, Turkey.

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Address correspondence and reprint request to: Dr. Murat Terzi, Department of Neurology, Ondokuz Mayıs University School of Medicine, 55139 Kurupelit-Samsun, Turkey. Tel. +90 (532) 3156884. Fax. +90 (362) 4576029. E-mail: mterzi@omu.edu.tr

Behçet's disease is characterized by small vessel vasculitis and involvement of multiple systems.¹ It was first described by the Turkish dermatologist Hulusi Behçet with a triad of oral aphthous ulcerations, genital ulcerations, and recurring uveitis attacks.² The central nervous system (CNS) involvement includes chronic meningoencephalitis, intracranial hypertension, cerebral hemisphere, brainstem, and spinal cord lesions. Neuro-Behçet's disease is seen rarely, and spinal cord involvement may appear even more rarely.³⁻⁶ Here, we report a case of an isolated lesion of the cervical spine, and hope this case may be an example for decision making in the differential diagnosis of such cases.

Case Report. A 24-year-old male patient was admitted to our clinic with complaints of urinary incontinence and difficulty in walking. He had had recurring oral aphthous ulcers for approximately 10

years, and genital ulcerations for approximately 6 months. He also had urinary incontinence for the last 6 months with constipation. He complained of blurring of vision occasionally in both eyes lasting for 15-20 days for the last 2 years. A week before his admittance to our clinic, he felt numbness and weakness in his feet and had difficulty in walking. He said he could not walk without support in the last 3 days before his admittance. The patient had a habit of smoking (a packet of cigarettes/day for 8 years), but he did not use any medications apart from colchicine, which was initiated 2 months ago with a diagnosis of Behçet's disease. On systemic examination, he had oral aphthous ulcers and scars of genital ulcerations. His ophthalmological examination revealed signs of an old uveitis. His pathergy test was negative. He did not have any skin lesions. In the lower extremities, the proximal muscle groups were 4/5 and the ankle dorsi and plantar flexors were 3/5 in muscle strength (according to the Medical Research Council). Upper extremities were normal in strength. There was hypoesthesia on the right side of the body, excluding the face. He had pyramidal ataxia and used a walker. There was mild spasticity in the lower extremities. Tendon reflexes were normal in the upper extremities, but increased in the lower. Plantar reflexes were flexor bilaterally, and there was also bilateral ankle clonus. In the laboratory investigations, complete blood count, renal and liver function tests, electrolytes, serum creatine kinase, creatine kinase-myocardial bound, thyroid hormones, vitamin B12, folic acid, serum immunoglobulin, and complement levels were all normal. Erythrocyte sedimentation rate was 16 mm/h. Rheumatoid factor, lupus cell and serologic studies for HIV, hepatitis B and C, venereal disease research laboratory, *Treponema pallidum* hemagglutination, toxoplasmosis, other [congenital syphilis, and viruses], rubella, cytomegalovirus, and herpes simplex virus, and brucella were all negative. The protein and glucose levels of the CSF were normal. The oligoclonal band in the CSF was negative. The cranial MRI was normal. In the cerebral single photon emission computed tomography (SPECT), the whole left parietal lobe was found to be hypoperfused. The left occipital and temporal lobes contained large hypoperfused areas. In the cervical MRI, there was a lesion of 2 cm diameter that was hyperintense on T2 weighted images at the level of the fourth cervical vertebra (Figure 1). The patient fulfilled the criteria of the International Study Group for Behçet's disease, and the clinical signs of cervical myelopathy were consistent with the cervical MRI lesion so



Figure 1 - An MRI showing a 2 cm diameter lesion, hyperintense on T2 weighted images of the 4th cervical vertebra.

the patient was diagnosed as neuro-Behçet. A course of steroid therapy, first intravenous then oral with a dose of 64 mg/day was initiated. Later, the steroid was tapered off by 8 mg/week and the therapy was stopped by the end of the second month. Meanwhile, a rehabilitation program was initiated. The patient was also continued on tablet colchicine 1.5 mg/day. The cervical MRI was normal at the end of the second month's follow-up. The patient walked without support, but there was a mild distal weakness of the lower extremities.

Discussion. According to the criteria of the International study group, diagnosis of Behçet's disease's can be made by the positivity of 2 of the 3 criteria, which are skin lesions, ophthalmic involvement, and positive pathergy test.² Our case fulfilled these diagnostic criteria by recurring oral and genital ulcerations and ophthalmic involvement. The rate of neurological involvement in Behçet's disease is 10-49%.⁵ The rate of onset of the disease by neurological symptoms is only 5-15%. The most frequent neurological symptoms are headache, meningoencephalitis, meningomyelitis, brain stem involvement, and organic confusional syndrome.⁷

In neuro-Behçet cases, the spinal cord involvement is supposedly 10-18%, whereas in autopsy cases this rate increases up to 28%.⁵ Spinal cord lesions are often associated with cerebral or brainstem lesions and isolated spinal cord involvement in Behçet's disease is very rare. The MRI characteristics of spinal cord lesions in Behçet's disease are similar to those of cerebral lesions. They are iso-hypointense in T₁w images and hyperintense in T₂w images.^{6,8} Gadolinium enhancement shows the dysfunction of the blood-brain barrier. Regression of this

enhancement is correlated with the repair of the barrier. The MRI findings reported up to now are non-specific. Other diseases, for example, some tumors, myelopathy due to multiple sclerosis, radiation myelopathy, spinal vascular diseases, and myelitis due to infections may have similar MRI findings.⁹ Although non-specific, MRI findings are significantly sensitive in the diagnosis of neuro-Behçet's disease.

The most frequently affected site is accepted to be the mesodiencephalic junction; the pontobulbar region comes second.⁹ Hypothalamus, basal ganglions, cerebral hemispheres, cerebellum, and spinal cord are rarely affected.^{6,9} It is not infrequent to see SPECT pathologies in neuro-Behçet's disease, although cranial MRI is normal. Our case is likewise. Multiple hypoperfusion sites in SPECT studies of neuro-Behçet's disease show a decrease of metabolic activity, but such findings are non-specific for neuro-Behçet's disease. Hypoperfusion in SPECT contributes to the understanding that there is early functional dysfunction in the CNS in Behçet's disease.

An MRI is used not only in the diagnosis, but also as a guide in the follow up of progression and the response to therapy. In cases with reversible lesions of the spinal cord, it is shown that the improvement observed in imaging studies, may not always correlate with the clinical improvement. The MRI lesions of the cervical spinal cord in our case also vanished after therapy at the end of the second month, but the clinical improvement was not complete. Isolated spinal involvement may be seen rarely in neuro-Behçet's syndrome and becomes a challenge in differential diagnosis. The case presented here may be an example for decisions of both diagnosis and clinical follow-up of such cases.

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