

Case Report

Bilateral optic neuritis in a patient with Behçet's disease who respond to therapeutic plasma exchange



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Abstract

Bilateral optic neuritis has been reported very rarely as a manifestation of neuro-Behçet's disease. We present a 50 year old woman who had 20-year history of Behçet's disease presented with acutely blurred vision associated with orbital pain in both eyes. Visual acuity was 0.4 in the right eye and light perception in the left eye; afferent pupillary defect was detected in the left eye. Bilateral swelling of the optic disk was found. The cerebrospinal fluid sample tests were within normal limits. Brain magnetic resonance imaging, magnetic resonance venography and fundus fluorescein angiography were normal. She was diagnosed with bilateral optic neuritis and treated with intravenous methyl prednisolone for 10 days. As there was no response to the treatment, therapeutic plasma exchange was started and the patient's visual acuities improved moderately. We suggest that when high dose steroid is failed to treat ON in BD, treatment with TPE may be considered.

Keywords: Bilateral optic neuritis, Behçet's disease, Therapeutic plasma exchange (TPE)

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Introduction

Behçet's disease (BD) is a multisystem disease of unknown cause in which an inflammatory perivasculitis can arise in almost any tissue. It is manifested by a triad of relapsing uveitis, aphthous stomatitis and genital ulcers.¹ Central nervous system (CNS) involvement is first described in 1941.² Optic neuritis (ON) has been reported rarely and bilateral simultaneous ON has been reported very rarely as a manifestation of neuro-Behçet's disease (NBD).^{3–10} Here we present a case who had bilateral synchronous optic neuritis due to BD who responds to therapeutic plasma exchange (TPE).

Case report

Fifty-year old woman who had 20-year history of BD presented with acutely blurred vision associated with orbital pain. She said that her vision became worse over two weeks. Visual acuity was 0.4 in the right eye and light perception in the left eye; afferent pupillary defect was detected in the left eye. There was no finding of uveitis in ophthalmologic examination. Bilateral swelling of the optic disk and peripapillary splinter hemorrhages in both eyes was found in her examination. The other neurological examination findings were all normal.

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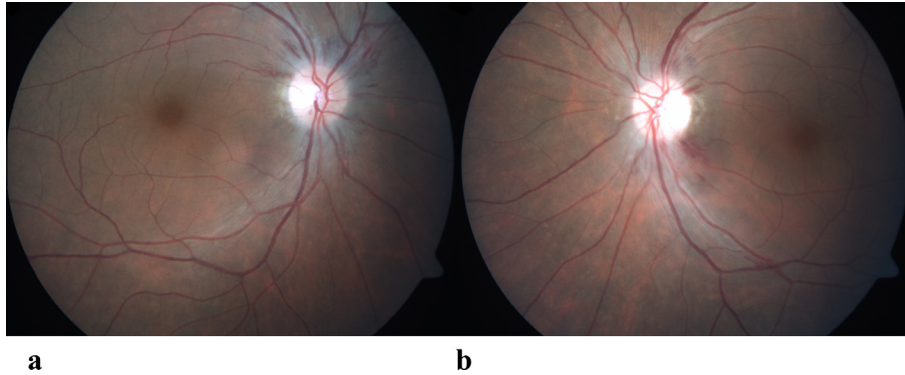


Fig. 1. Resolution of optic disk swelling in both eyes after TPE [(a): right, (b): left].

In laboratory tests, blood count, renal and liver functions tests, erythrocyte sedimentation rate, serum levels of C-reactive protein and angiotensin converting enzyme were all within normal limits. Serum neuromyelitis optica antibody was negative. Magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) of the brain were inconclusive. Lumbar puncture was performed and the opening pressure was 19 cm H₂O. The cerebrospinal fluid sample (CSF) was acellular, the protein level was 25 mg/dl and the glucose level was 66 mg/dl. Fundus fluorescein angiography was normal. The patient was diagnosed with bilateral ON secondary to BD. She was treated with intravenous methyl prednisolone for 10 days at a dose of 1 gram daily. However, the visual acuities became even worse during the treatment. Just after steroid treatment TPE was started, totally 5 sessions of TPE were carried out in every other days, and one plasma volume was exchanged for each session. Following TPE treatment bilateral optic disk swelling was improved on fundus examination (Fig. 1). Three months after TPE the visual acuity was 0.7 in the right eye and 0.1 in the left eye.

Discussion

Optic neuritis has been reported very rarely as a manifestation of NBD. In 1989, Kansu et al. reported three cases of ON in BD.³ One of these cases was presented with bilateral synchronous optic neuritis. Gallinaro et al. reported an acute presentation with unilateral neuropapillitis in a 34-year-old woman with BD.⁴ Tarzi et al. reported a 32-year-old man with a 4 year history of BD who had visual disturbance developed over a period of hours with bilateral papillitis.⁵ Cranial MRI, MRV and also the analysis of CSF were noted to be normal. A well response to the treatment with intravenous methylprednisolone at a dose of 1 g/daily was reported. Yalcindag et al. reported a 47-year-old man of BD who had acute unilateral ON.⁶ The authors described that the patient showed improvement in two weeks after the treatment with intravenous prednisolone at a dose of 1 gram daily. Two cases with the recurrent ON were reported by Voros et al.⁷ The times of ON attacks were noted similar with systemic relapses and one attack was presented as bilateral ON. The authors prescribed oral prednisolone for each attack and only one attack did not respond to the treatment and permanent visual loss occurred. Nanke et al. reported a 27-year-old man of BD who had recurrent unilateral ON and responded to oral prednisolone treatment.⁸ Cetin et al. reported unilateral optic

neuropathy in a 57-year-old man of BD.⁹ He was treated with systemic steroid but there was no response to the treatment. A 12-year-old boy with acute episcleritis and papillitis as presentations of pediatric BD was reported by Parentin et al.¹⁰ After treatment with oral prednisone 1 mg/kg, visual acuity and fundus appearance returned to normal within one week. The authors reported that in the following six months the disease relapsed four times in both eyes and oral cyclosporine was given. Under oral cyclosporine treatment another two episodes occurred so, intravenous infliximab treatment was started. A well response was noted to the treatment. In a review published in 2013, 20 cases of BD with primary inflammatory optic neuropathy were reported. Four of these cases were bilateral and synchronous.¹¹ Recently, Shokoohi and Yakhooobi reported a 28-year-old Iranian man of Behçet's Disease presenting with bilateral papillitis who he showed improvement with steroid treatment.¹²

In summary bilateral synchronous optic neuropathy has been rarely reported in the literature.^{10,12} To our knowledge, this is the sixth case with bilateral synchronous optic neuropathy due to NBD. It may present solitary without parenchymal NBD, such as our patient.⁵⁻¹² The severity of visual loss and the degree of response to the treatments can vary but patients usually benefit from the steroid treatment.

It has been suggested that TPE is an effective treatment in neurologic diseases that autoimmunity plays an important role in the pathogenesis.¹³ TPE can be used in several neurological diseases such as acute disseminated encephalomyelitis, Guillain-Barré Syndrome, Myasthenia Gravis and neuromyelitis optica according to the Guidelines on the Use of Therapeutic Apheresis in Clinical Practice-Evidence-Based Approach from the Writing Committee of the American Society for Apheresis.¹⁴ NBD is not in any of its recommendations. However, there have been some reports suggesting TPE is effective when used in a group of autoimmune diseases including Behçet's Disease.¹⁵ Since the visions of the patient decreased during high dose steroid treatment, the patient was put on TPE just after the treatment with steroid. After the beginning of TPE, initially visual acuity improved slightly, and then improvement continued, finally the visual acuity improved moderately. The improvement may be secondary to TPE or to the treatment with intravenous methyl prednisolone, or it may be spontaneous improvement over time. However, it may be helpful to keep in mind that when high dose steroid fails to treat ON in BD, TPE may be tried as a second line treatment.

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