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SHORT COMMUNICATION

Acute optic neuropathy in Behçet disease

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PURPOSE. *To report a patient with Behçet disease presenting with acute optic neuropathy.*
METHODS. *A 47-year-old man was admitted to the authors' clinic owing to sudden visual loss in the left eye. Ophthalmologic and systemic examinations were performed.*
RESULTS. *Visual acuity was 4/10 in the left eye. Fundus examination demonstrated swollen optic disc with blurred margins. There was relative afferent pupil defect in the same eye. Mega-dose corticosteroid treatment was started with the diagnosis of acute optic neuropathy. Ocular findings resolved within 2 weeks.*
CONCLUSIONS. *Behçet disease rarely presents with acute optic neuropathy. (Eur J Ophthalmol 2004; 14: 578-80)*

KEY WORDS. *Behçet disease, Neuro-ophthalmologic manifestation, Optic neuropathy*

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INTRODUCTION

Neurologic involvement in Behçet disease (BD) is well known, and has been classified by Pallis and Fudge (1) and Wadia and Williams (2) into three types: brainstem disturbance (cranial neuropathies, ocular motor dysfunction, nystagmus, gaze palsies), meningomyelitis, and a confusional syndrome (meningoencephalitis, dementia, parkinsonism). Involvement of the optic nerve is a rare neuro-ophthalmologic manifestation (3-6). We report a middle-aged man with BD presenting with acute optic neuropathy.

Case report

A 47-year-old man was admitted to Ankara University School of Medicine Department of Ophthalmology for sudden visual loss in the left eye. He had had chronic arthritis for the past 1.5 years. He had had relapsing oral ulcers for 10 years. Painful papulopustular lesions were present on his hands and legs. Visual acuity was 10/10 in the right eye and 4/10 in the left eye. Anterior segment structures were normal in both eyes, and there was no sign of previous uveitis. On Ishihara color vision testing, he missed all plates with the left eye. There was relative afferent pupillary defect. Fundus examination was normal in the right eye; the optic disc was swollen with blurred margins in the left eye (Fig. 1). Fluorescein angiography showed a hyperfluorescent optic disc in the left eye (Fig. 2). The visual field in the same eye also presented a paracentral scotoma (Fig. 3). Neurologic examination revealed no pathology. Magnetic resonance imaging of the brain demonstrated a few foci of previously incurred ischemic lesions in the left basal ganglia. Skin biopsy from the papulopustular eruptions on the hand showed leukocytoclastic angiitis (Fig. 4). HLA-class

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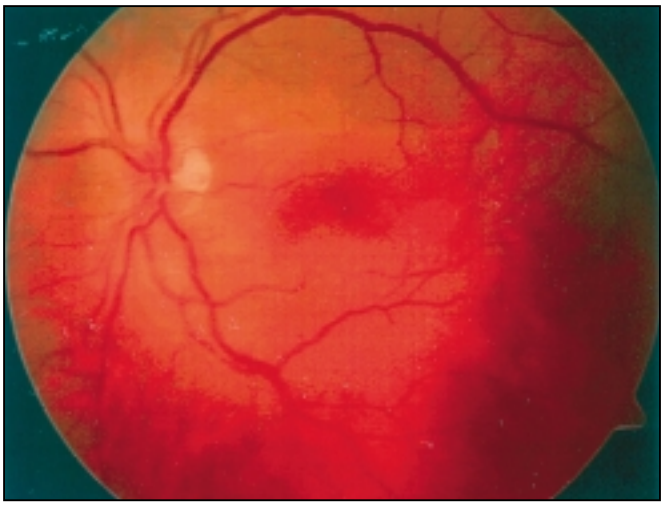

Fig. 1 - *Fundus examination of the left eye demonstrates swollen optic disc with blurred margins.*

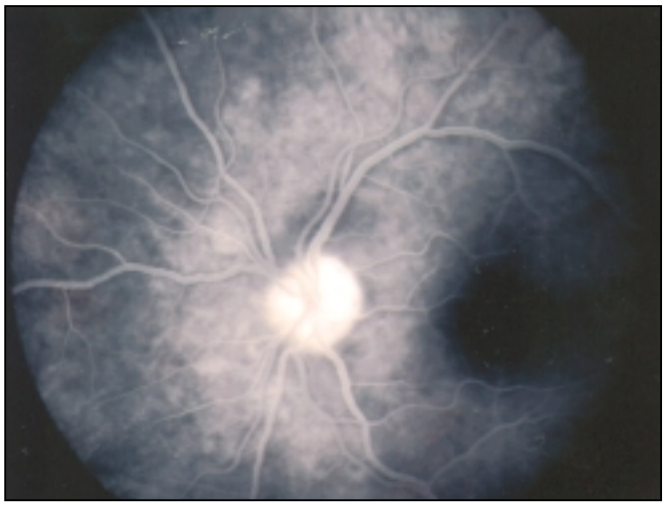

Fig. 2 - *Fluorescein angiography of the left eye showed a hyperfluorescent optic disc.*

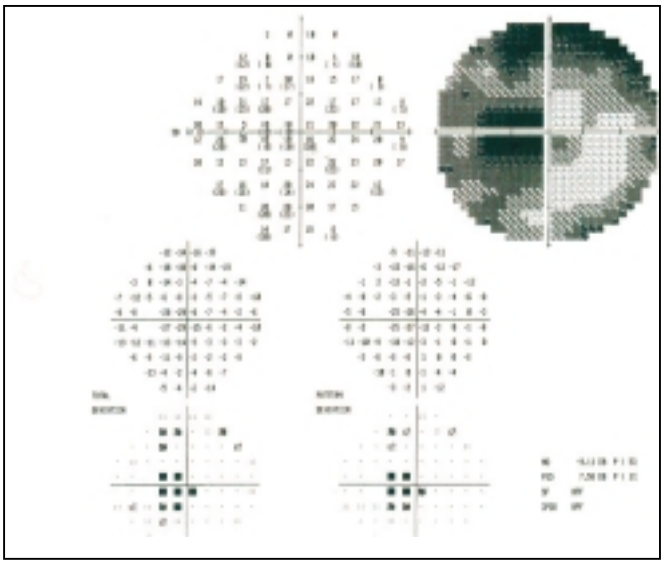

Fig. 3 - *Visual field of the left eye presented a paracentral scotoma.*

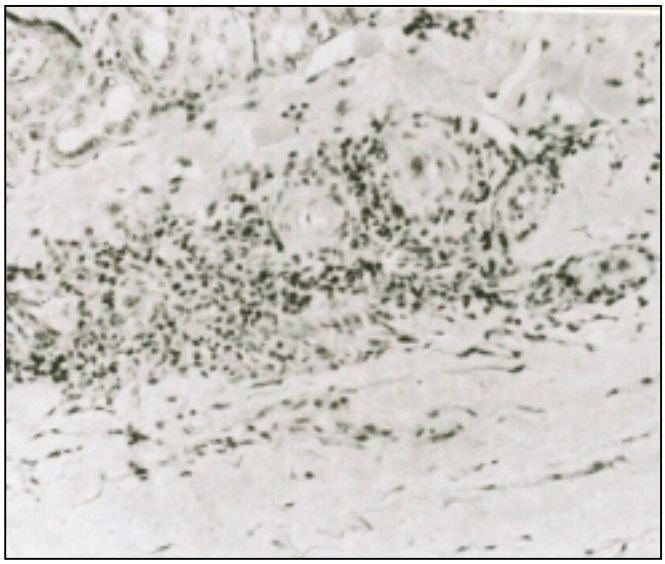

Fig. 4 - *Skin biopsy demonstrates inflammatory cellular infiltration, consisting mainly of leukocytes, around the small vessels (leukocytoclastic angiitis).*

l antigen testing revealed HLA-B51 positivity. Mega-dose corticosteroid treatment (1000 mg/day intravenous methylprednisolone for 3 days) was started with the diagnosis of acute optic neuropathy and BD. After mega-dose steroid treatment, oral corticosteroid therapy was initiated. Two weeks later, visual acuity was 10/10 in the left eye and the margins of the optic disc became clear.

DISCUSSION

Neuro-ophthalmologic signs in BD are known to be rare neurologic manifestations. The prevalence of neurologic involvement in Turkey has been reported as 5.3% by Serdaroğlu et al (7) and as 14% by Kansu et al (4). Scouras and Koutroumanos (3) reported two patients with ischemic optic neuropathy in BD and

Kansu et al presented three cases of optic neuropathy in BD (4).

Optic nerve damage in BD is believed to be caused by several mechanisms, such as the spreading of the inflammation from the uveal tract to the optic nerve (uveopapillitis), occlusion of small vessels of the optic nerve (vasculitis), or demyelinization induced by ischemia. The inflammation of the optic nerve may precede that of the uvea (3, 4). There was clinically no uveitis or neurologic symptoms in our patient but the magnetic resonance imaging of the brain demonstrated a few foci of previously incurred ischemic lesions. Thus, inflammatory involvement of the optic nerve without uveitis or acute inflammation of the nervous system is more probable. Colvard et al reported that inflammatory involvement of the optic nerve may be associated with generalized involvement of the central nervous system or may occur independently in the absence of neurologic signs (8).

To our knowledge, optic neuropathy cases were reported previously as either an early sign of BD or as a neuro-ophthalmologic sign during the disease course. Papillitis and retrobulbar neuritis were reported among these cases (3-6).

In conclusion, isolated optic neuropathy is a rare sign of Behçet disease in the absence of uveal inflammation and neurologic disorder.

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