

# Characteristics of optic neuropathy in Behçet disease

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## Abstract

### Objective

We present the clinical profile, features, and neuroimaging findings of 25 patients with Behçet disease (BD), and optic neuropathy (ON), which has been rarely reported in BD.

### Methods

Data from 5 university hospitals were retrospectively reviewed, and patients with BD and ON were evaluated. There were 2 groups: (1) those already diagnosed with BD when ON developed (BD→ON group) and (2) those diagnosed with BD during the evaluation of ON (ON→BD group).

### Results

There were 25 BD patients with ON (13 males). Among these, 13 had ON→BD, and 12 had BD→ON. Seventeen patients had unilateral ON, and 7 patients had recurrent ON. BD→ON patients were older. Disc edema was seen more in ON→BD than in BD→ON patients (10 vs 3). Fourteen patients also had uveitis, 7 with BD→ON and 7 with ON→BD. There was other neurologic involvement in 8 patients; in the BD→ON group, 4/4 had MS-like disease, in the ON→BD group, 3 had typical parenchymal BD, and 1 had MS-like disease. Twenty of 21 patients received immunosuppressive medications, corticosteroids, or both. Prognosis was favorable in most: vision improved in 20 patients, more often in those receiving combined therapies.

### Conclusion

BD may be diagnosed earlier if it is considered and investigated during the assessment of ON, particularly in high-risk regions. Prognosis of ON related to BD seems to be favorable. Immunosuppressants should be given along with corticosteroids. MS-like presentations should also be kept in mind in patients with BD and ON.

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## Glossary

DEU = Dokuz Eylül University; RMA = repeated-measures analysis of variance; LETM = longitudinally extending transverse myelitis; MOG = myelin oligodendrocyte glycoprotein; VA = visual acuity; RAPD = relative afferent pupillary defect; ON = optic neuropathy; BD = Behçet disease.

Behçet disease (BD) is an idiopathic autoinflammatory disease characterized by relapsing uveitis and genital and oral ulcers (aphthae).<sup>1,2</sup> BD can affect many other organ systems such as skin, musculoskeletal system, gastrointestinal system, cardiovascular system, and CNS.<sup>3</sup> Neurologic involvement in BD was first reported in 1944.<sup>4</sup> The CNS can be involved mainly in 2 ways: (1) as parenchymal involvement, usually presenting as a brainstem syndrome, and (2) as dural sinus thrombosis.<sup>2</sup> In the very first postmortem study of a patient with parenchymal BD, there was also round cell infiltration around the central retinal artery within the optic nerve.<sup>4</sup> Since then, optic neuropathy (ON) has rarely been reported in BD, probably as it was overshadowed by the more common ocular complication of BD, uveitis.<sup>5–9</sup>

ON in BD can occur alone, or with other CNS involvement, or secondarily due to other ocular involvement such as uveitis, papilledema from dural sinus thrombosis, obliterative retinal vasculopathy, or glaucoma secondary to uveitis.<sup>7,8</sup>

Here, we present a multicenter study of patients with BD who presented with ON or developed during the course of BD. Our goal was to define the timing of optic nerve involvement in BD, its association with other clinical features, and relevant laboratory and neuroimaging findings.

## Methods

### Standard protocol approvals, registrations, and patient consents

Data from neuro-ophthalmology clinics and uveitis clinics of 5 Turkish university hospitals were reviewed retrospectively for evidence of ON. The medical records of patients were first evaluated by a neurologist of each hospital and then reevaluated in joint sessions by 1 senior neurologist (G.A.) in one of the 5 hospitals, Dokuz Eylül University (DEU). The study was approved by the Ethical Committee of DEU Faculty of Medicine.

### Study participants

Patients were grouped into 2 according to the timing of the optic nerve involvement: (1) patients who had already been diagnosed as having BD when ON developed (BD→ON) and (2) patients who had ON before the diagnosis of BD was made or who were diagnosed as having BD during the etiologic evaluation of ON (ON→BD).

The diagnosis of BD was based on the criteria set by the International Study Group for Behçet's Disease.<sup>10</sup> Patients having recurrent oral aphthae at least 3 times in a year,

accompanied any 2 of the following: genital ulcerations; skin lesions such as erythema nodosum, folliculitis, and ulcerations; eye involvement such as anterior and posterior uveitis; and skin pathergy reaction, were diagnosed as having BD. The diagnosis of ON was more challenging in the patients who also had uveitis. Reduced visual acuity (VA) that could not be explained by the uveitis and significantly impaired color vision (tested using Ishihara color test plates) were the minimum requisites for the diagnosis of ON. Other probable manifestations were relative afferent pupillary defect (RAPD), prominent visual field defect, painful eye movements, optic disc swelling without intracranial hypertension, delay in visual evoked potentials, and optic nerve hyperintensity/enhancement on MRI. Patients with ON related to other causes or patients who had insufficient data recorded were excluded from the study.

Clinical records were scrutinized for the demographic characteristics, clinical findings, laboratory and neuroimaging features, and treatment regimens. Visual improvement was defined as  $\geq 2$  lines (score) of visual change in the logMAR scale. One-line improvement or no VA change in the logMAR scale was described as "no improvement." When visual improvement was recorded, the visual outcome was considered to be a favorable.

### Data analysis

In the descriptive analysis, data were presented as mean  $\pm$  SD if parametric distribution was provided; median (minimum–maximum) values were used for nonparametric distribution, and categorical data were defined by frequency and percentages. The *t* test, Mann-Whitney *U* test, Pearson  $\chi^2$  test, and Fisher exact test were applied to univariate comparison between BD- and ON-onset groups. The repeated-measures analysis of variance (RMA) test was used for repeated measurements and group effects (BD/ON). Statistical analyses were performed via the IBM SPSS 23.0 program. The statistical significance limit value was accepted as  $p < 0.05$ .

### Data availability

Detailed data about the patients are presented in table e-1 ([links.lww.com/NXI/A62](https://links.lww.com/NXI/A62)).

## Results

There were 13 male and 12 female patients aged 16–55 years (range  $35.6 \pm 11.1$  years), 13 with ON→BD (6 males and 7 females) and 12 (7 males and 5 females) with BD→ON. In the ON→BD group, the diagnosis of BD was made during the

etiologic evaluation of ON in 5 and later in 8. Figure shows the ophthalmologic details of a newly diagnosed BD patient with uveitis and ON.

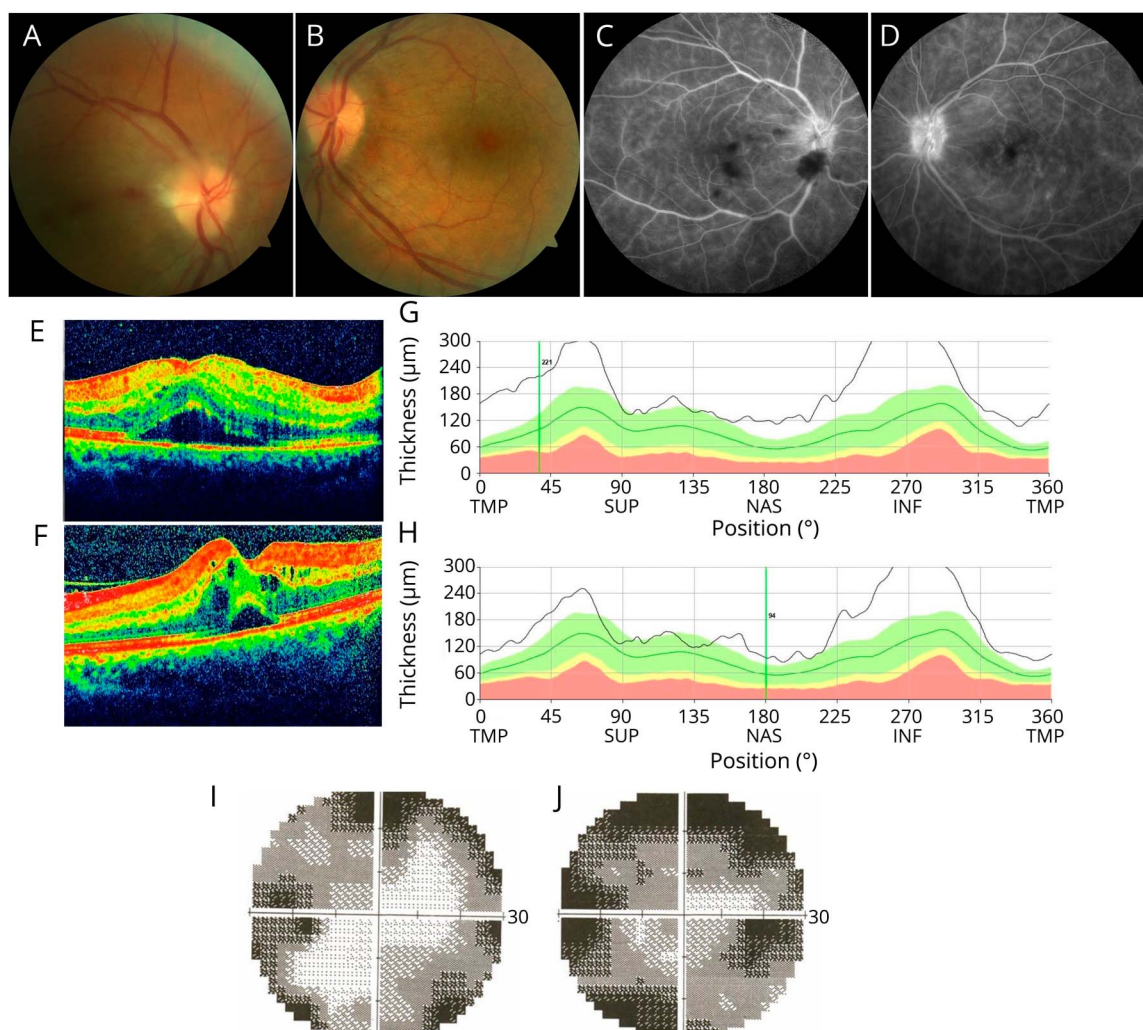
In the BD→ON group, ON was diagnosed  $6.6 \pm 5.3$  years (range 1–14 years) after the diagnosis of BD, whereas the diagnosis of BD was made  $1.8 \pm 0.5$  years (range 1–2 years) after the diagnosis of ON in the ON→BD group. The follow-up period ranged between 1 and 36 years ( $11.7 \pm 9.5$ ).

Of the 25 patients, 17 patients had unilateral ON, 8 patients, 3 with BD→ON and 5 with ON→BD, had bilateral ON. Eighteen patients experienced only 1 attack of ON, and the other 7 had 2 or more attacks. Six patients had orbital pain during the attack. Thirteen patients had disc edema, 10 with ON→BD and 3 with BD→ON. Uveitis was observed in 7/12 BD→ON and

7/13 ON→BD patients. Uveitis was unilateral in 6 of these 14 patients, and the ON occurred in the eyes that already had uveitis or then developed uveitis in 4. Vision improved in 9/12 BD→ON patients and in 11/13 ON→BD patients.

Eight of the 25 patients had other types of neurologic involvement: 5 had an MS-like presentation, 4 of whom were in the BD→ON group; 3 had typical BD parenchymal brainstem involvement, 1 with an additional longitudinally extending transverse myelitis (LETM), all 3 in the ON→BD group. The remaining 17/25 patients had no other neurologic involvement; their brain MRIs were normal (1 BD→ON patient had asymptomatic periventricular lesions). None of the patients had dural sinus thrombosis. None of the patients had aquaporin or myelin oligodendrocyte glycoprotein (MOG) antibody testing.

**Figure** Color fundus photographs



Color fundus photographs of a 35-year-old woman with newly diagnosed Behçet disease showing slight haze in vitreous, prominent disc edema, cotton wool spots, and some retinal hemorrhages in the right (A) and left (B) eyes. Venous phase of the fluorescein angiogram depicting marked disc leakage, scattered retinal capillary leakage, and hypofluorescent areas corresponding to the cotton wool spots and retinal hemorrhages in the right (C) and left (D) eyes. Optical coherence tomography of the macula exhibiting subfoveal serous neurosensory retinal detachment and thickened retina in the right (E) and left (F) eyes. The retinal nerve fiber layer was thickened in the right (G) and left (H) eyes. Humphrey 30.2 visual field test showing mainly peripheral visual field defects with visual field constriction in the right (I) and left (J) eyes.

Clinical findings, duration of BD and ON, follow-up period, interval between BD and ON, and visual acuities at presentation and after treatment were compared (table 1). Patients with BD→ON were significantly older than patients with ON→BD ( $p = 0.004$ ). The duration of BD in the BD→ON group was significantly longer than that in the ON→BD group ( $p = 0.005$ ). The follow-up period was significantly longer in the BD→ON group than in the ON→BD group ( $p = 0.014$ ). The interval between BD and ON was significantly longer for patients with BD→ON than for patients with ON→BD ( $p = 0.001$ ).

Six BD→ON patients were under colchicine treatment when ON developed, 4 of them also received oral corticosteroids. One patient was not on any BD treatment when ON developed. After ON developed, 18 patients received combined therapies with corticosteroids and azathioprine and other immunosuppressive drugs. Azathioprine was used in patients with bilateral ON, other

neurologic involvement, or with relapses. One patient was treated with corticosteroids alone. Five patients were treated with only azathioprine; 1 did not receive any treatment, and in 4 patients, medical records were not informative.

Although this is a retrospective analysis, which was not designed to evaluate the effects of therapies, significantly better visual improvement was found with combined therapies than with monotherapies ( $p = 0.012$ ). No significant difference between the therapeutic approach was noted with regard to sex, sequence of BD and ON, uveitis, disc edema, and pain ( $p > 0.05$ ). Twenty patients showed visual improvement and 5 patients showed minimal or no change; therefore, visual prognosis was considered to be good in most BD-ON patients.

RMA analysis (for 2\*2 design) revealed that there was significant difference between the first and last scores in 2 models

**Table 1** Comparison of clinical findings, duration of BD and ON, follow-up period, interval between BD and ON, and visual acuities at presentation and after treatment between 2 groups according to the sequence of BD and ON

	<b>BD→ON</b> n = 12	<b>ON→BD</b> n = 13	<b>p</b>
<b>Sex, n (%)</b>			0.54 <sup>a</sup>
<b>Male</b>	7 (58.3)	6 (46.2)	
<b>Female</b>	5 (41.7)	7 (53.8)	
<b>Mean age ± SD, y</b>	<b>42 ± 11</b>	<b>30 ± 7</b>	<b>0.004<sup>b</sup></b>
<b>ON attacks ≥2, n (%)</b>	3 (25.0)	4 (30.8)	0.54 <sup>c</sup>
<b>Bilateral involvement, n (%)</b>	3 (25.0)	5 (38.5)	0.39 <sup>a</sup>
<b>Pain, n (%)</b>	2 (15.0)	4 (30.8)	0.36 <sup>a</sup>
<b>Disc edema, n (%)</b>	3 (25.0)	10 (76.9)	<b>0.009<sup>a</sup></b>
<b>Uveitis, n (%)</b>	7 (58.3)	7 (53.8)	0.82 <sup>a</sup>
<b>Recovery, n (%)</b>	9 (75.0)	11 (84.6)	0.46 <sup>c</sup>
<b>Duration of BD median (minimum–maximum), y</b>	15.0 (3.0–36.0)	3.0 (0.0–16.0)	<b>0.005<sup>d</sup></b>
<b>Duration of ON median (minimum–maximum), y</b>	10.0 (0.0–23.0)	5.0 (0.0–16.0)	0.28 <sup>d</sup>
<b>Follow-up period median (minimum–maximum), y</b>	15.0 (3.0–36.0)	5.0 (0.0–16.0)	<b>0.014<sup>d</sup></b>
<b>Interval (BD-ON) median (minimum–maximum), y</b>	7.0 (1.0–14.0)	0.0 (0.0–2.0)	<b>0.001<sup>d</sup></b>
<b>Number of ON attacks median (minimum–maximum), y</b>	1.0 (1.0–2.0)	1.0 (1.0–3.0)	0.68 <sup>d</sup>
<b>R-Onset VA(logMAR) median (minimum–maximum), y</b>	0.5 (0.0–1.0)	0.4 (0.0–1.0)	0.95 <sup>d</sup>
<b>L-Onset VA(logMAR) median (minimum–maximum), y</b>	0.4 (0.0–1.0)	0.5 (0.0–1.0)	0.93 <sup>d</sup>
<b>R-Last VA(logMAR) median (minimum–maximum), y</b>	0.2 (0.0–1.0)	0.0 (0.0–1.0)	0.45 <sup>d</sup>
<b>L-Last VA(logMAR) median (minimum–maximum), y</b>	0.2 (0.0–1.0)	0.0 (0.0–1.0)	0.21 <sup>d</sup>

Abbreviations: BD = Behçet disease; ON = optic neuropathy; VA = visual acuity.

<sup>a</sup> Pearson  $\chi^2$ .

<sup>b</sup> *t* test.

<sup>c</sup> Fisher exact test.

<sup>d</sup> Mann-Whitney *U* test.

Bold values are statistically significant.

(RMA-VA-logMAR right and RMA-VA-logMAR left) ( $p = 0.004$  and  $p < 0.001$ , respectively). There was no significant difference between the 2 groups (BD→ON and ON→BD groups) in terms of these parameters.

## Discussion

In the present article, we evaluated 25 BD patients presenting with ON in a multicenter Turkish cohort. To draw attention to the timing of the ON attack, we divided our group into 2: patients with known BD who develop ON during the course of BD (BD→ON group) and patients who present with ON and get the diagnosis of BD during etiologic workup (ON→BD group). The 2 groups were distributed evenly, and sex distribution was also similar, with the M:F ratio nearly 1. Eight patients had bilateral ON, and 7 patients had recurrent ON attacks. In the end, 20 of our 25 patients had recovery. Because BD is fraught with various types of ocular inflammation, it is important to differentiate ON from other ocular inflammation patterns in patients with BD. As hot disc appearance (staining of the optic disc in late phases of fluorescein angiogram) is a common fluorescein angiographic finding in Behçet eyes, the clinical signs indicating the optic nerve involvement such as the presence of RAPD, acquired color vision defect, impaired visual field test, etc. should be looked meticulously in every eye to reach a correct diagnosis of ON. For instance, afferent pupillary defect cannot be evaluated properly because of already present extensive posterior synechiae. High level of clinical suspicion is a must to detect optic nerve disturbance.

Other than case reports, to our knowledge, there are 2 relatively large studies evaluating ON in BD. Kidd<sup>8</sup> reported 20 BD-ON cases; 4 of his own, and 16 from the literature. Eleven cases were unilateral, 4 bilateral simultaneous, and the rest bilateral sequential. There were no ocular abnormalities in 9, vitreous cells in 2, retinal vascular sheathing in 2, optic disc swelling in 9, anterior uveitis in only 1. Patients improved spontaneously or with corticosteroids, and overall, the visual outcome was good; 67% of the cases recovered normal vision. A more recent article from Tunisia described 10 cases with inflammatory ON among a single-center series of 440 patients with BD.<sup>9</sup> In that series, 50% of the cases had also various forms of other ocular involvement (retinal vasculitis in 1 and anterior/posterior/pan uveitis in 4). In the end, only 20% had vision loss, 40% had improvement, and 40% was stable. In our study, 14 patients had uveitis; 6 patients had ON concurrently with uveitis, 4 in the same eye. There was no statistically significant relationship between the presence of uveitis and recovery from ON. Also, there was no significant relationship between the number of ON attacks and visual improvement. Of the 5 patients who did not have visual improvement, only 1 patient had more than 1 ON attack.

We grouped our patients into 2 groups according to the timing of ON and BD because we have noted that not very few people who presented with ON were eventually found to have BD. Almost half of our patients had ON attack before the diagnosis of BD. Although the numbers are relatively small,

BD should be kept in mind in Mediterranean, Middle Eastern, and Far Eastern regions where it is rather prevalent. However, we did not find any major demographical differences between the 2 groups. Patients with BD→ON were significantly older, and the duration of BD was significantly longer, as might be expected. Moreover, the follow-up period was significantly longer, and the interval between BD and ON was significantly longer for patients with BD→ON. Patients with ON→BD might have milder mucocutaneous BD, so they might not have sought medical attention before the onset of ON. In that group, 7 patients were female, whereas in the BD→ON group, only 5 were female, who more commonly have milder mucocutaneous disease.<sup>11</sup> However, parenchymal brain involvement is also seen in the ON→BD group and that is contradictory to this assumption.

In large neuro-BD series, 10%–15% present with an MS-like clinical picture<sup>2,6</sup> with clinical features in between parenchymal neuro-BD and MS.<sup>12</sup> Most of our patients with an MS-like presentation were in the BD→ON group. One of our ON→BD patients had LETM besides typical parenchymal involvement. BD spinal cord involvement of BD may present as LETM,<sup>13</sup> and up to now aquaporin or MOG antibodies could not be demonstrated in any case (Akman-Demir, unpublished data). However, unfortunately, in this series, our patients were not tested for aquaporin or anti-MOG antibodies; it would be worthwhile to check these patients for aquaporin or MOG antibodies. Theoretically, though, it is possible to have overlapping presentations of NMO in patients with BD because both disorders are relatively common in similar geographical areas. This needs to be clarified in further studies.

As 20 of our 25 patients experienced a visual recovery from ON, the prognosis of BD associated ON might not be as poor as the prognosis of BD uveitis. Moreover, our results indicate that BD associated with ON responds to the combined therapies with corticosteroids and other immunosuppressive agents. Combined therapies with different immunosuppressive agents and corticosteroids might prevent or reduce visual impairment. Although ON is rare in BD, it could present as a solitary finding without parenchymal CNS and should be considered as part of the neuro-BD disease spectrum. New visual symptoms and signs in BD, which cannot be explained by ocular involvement, should increase the possibility of ON.

The diagnosis of ON should lead the clinician to avoid cyclosporine, sometimes used to treat BD uveitis since cyclosporine can promote the development of neurologic involvement in BD patients.<sup>14</sup> ON in BD should be considered in the spectrum of neuro-BD and treated accordingly. Potential limitations of our study are (1) its retrospective nature and (2) the diversity of data collection in different university hospitals. Therefore, future prospective multicenter studies are needed.

In a patient presenting with ON, the clinician should consider BD in the differential diagnosis, especially if uveitis is present, and ask about recurrent mucocutaneous ulceration. When

a patient with established BD loses vision, the clinician should consider ON and uveitis as the possible cause. Color vision testing, careful observation of the optic discs and nerve fiber layer, visual field evaluation, RAPD testing, visual evoked potentials, and orbital MRI would all help to make the diagnosis of BD-ON.

### Author contributions

G. Akdal: data acquisition, drafting/revising the manuscript, study concept or design, analysis or interpretation of data, contribution of vital reagents/tools/patients, statistical analysis, and study supervision. H. Ertaşoğlu Toydemir: data acquisition, drafting/revising the manuscript, study concept or design, analysis or interpretation of data, and statistical analysis. O. Saatci: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, and acquisition of data. U. Uygunoğlu: drafting/revising the manuscript, analysis or interpretation of data, and acquisition of data. B. Altunrende: data acquisition, drafting/revising the manuscript, study supervision, and principal investigator/guarantor: no. S. Saip: data acquisition. A. Yaman: data acquisition and principal investigator/guarantor: yes. P. Keskinoğlu: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, statistical analysis, and study supervision. S. Güven Yılmaz: data acquisition, principal investigator/guarantor: yes, and study sponsor role—acquisition of data. N. Çelebisoy: data acquisition, principal investigator/guarantor: yes, and corresponding author, study group—responsible for determining whether members qualify as authors: yes. M. Söylev Bajin: data acquisition. A. Siva: drafting/revising the manuscript, study concept or design, and contribution of vital reagents/tools/patients. G. Akman-Demir: data acquisition; drafting/revising the manuscript, study concept or design, analysis or interpretation of data, study supervision, and principal investigator/guarantor: yes.

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