Posterior Uveitis and Cerebral Venous Sinus Thrombosis as the First Presentation of Behçet Syndrome: Case Report

Behçet Sendromunun İlk Başvuru Bulgusu Olarak Posterior Üveit ve Serebral Venöz Sinüs Trombozu Birlikteliği

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ABSTRACT The reported frequency of neurological involvement among Behçet syndrome patients is approximately 5%. In this paper, we reported posterior uveitis with cerebral venous sinus thrombosis as the first presentation of Behçet syndrome in a 19 year- old female patient. Our patient presented with a one-month history of headache, progressive loss of vision in the right eye. Her best corrected visual acuity was counting fingers at 1 meter in the right eye and 10/10 in the left eye. Funduscopy revealed vitreous cellular infiltrate and macular edema in the right eye in addition to papilledema (central macular thickness was 753 μm in the right eye and 168 μm in the left eye). Magnetic resonance imaging and magnetic resonance venography of the brain, were performed because of persistant headache, showed complete occlusion of left transvers and sigmoid sinus with thrombosis. A diagnosis of Behçet syndrome was made based on clinical criteria.

Key Words: Behcet syndrome; sinus thrombosis, intracranial

ÖZET Behçet sendromunda, nörolojik tutulumun sıklığı yaklaşık %5'dir. Biz bu çalışmamızda, Behçet sendromunun ilk başvuru bulgusu olarak posterior üveit ve serebral venöz sinüs trombozu birlikteliği gösteren, 19 yaşında kadın hastanın sunumunu amaçladık. Hasta kliniğimize bir aydır devam eden başağrısı ve sağ gözde ilerleyici görme kaybı şikayeti ile başvurdu. Düzeltilmiş görme keskinliği sağ gözde 1 mps, sol gözde 10/10 idi. Fundoskopik incelemede papilödeme ek olarak sağ gözde vitrede hücresel birikimler ve maküler ödem mevcuttu (santral maküler kalınlık; sağ gözde 753 μm, sol gözde 168 μm). İnatçı başağrısı nedeniyle çekilen beyin manyetik rezonans görüntüleme ve manyetik rezonans venografide, sol transvers ve sigmoid sinüsün, tromboz ile total tıkanıklığı saptandı. Hastaya klinik verilerine dayanarak Behçet sendromu tanısı kondu.

Anahtar Kelimeler: Behçet sendromu; sinüs trombozu, intrakraniyal

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Behçet syndrome (BS) is a systemic inflammatory vascular disease with several clinical manifestations and geographical differences in disease expression. Clinical features include oral and genital ulceration, skin lesions and the pathergy response, ocular involvement, arthritis, vasculitis, central nervous system (CNS) and gastrointestinal manifestations.[1,2]

The reported frequency of neurological involvement among BS patients ranges from 2,2 to 49.0%, but larger series have shown a rate of approximately 5%.[3-5] Neurological involvement is more common in men with a male to female ratio of up to 4:1.[3-5] Neurological involvement in BS may be subclassified into two major forms. One form is focal or multifocal central

nervous system parenchymal involvement, the other form is caused by cerebral venous sinus thrombosis (CVST). Papilledema and sixth nerve paresis are the most common signs reported and hemiparesis may develop in some.[4,6]

In this case report, we presented a 19-year-old female patient with posterior uveitis and CVST, as the first presentation of BS.

CASE REPORT

A 19-year-old female patient presented to the ophthalmology clinic with decreased vision in the right eye for 5 days and severe headache for 1 month. History of recurrent oral and genital ulcers was obtained.

At physical examination, oral or genital ulcer, skin lesions, neurological deficit were not present.

At ophthalmologic examination, her best corrected visual acuity (BCVA) was counting fingers at 1 meter in the right eye and 10/10 in the left eye. Funduscopy revealed vitreous cellular infiltrate and macular edema in the right eye in addition to papilledema (Figure 1,2). Fundus flouroscein angiography showed posterior vasculitis in the right eye. (Figure 3) At optical coherence tomography, central macular thickness (CMT) was 753 μm in the right eye (Figure 4) and 168 μm in the left eye. Posterior uveitis due to Behçet's disease was suspected. In addition, because the patient had a history of headache for a 1 month, neurology consultation was requested and magnetic resonance ımaging (MRI)- magnetic resonance angiography (MRA) of the head were applied.

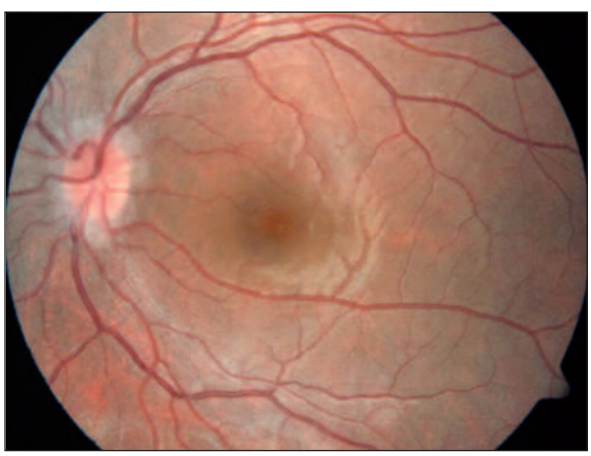

FIGURE 1: Fundus photograph of the left eye; At the first fundus examination, there is papilledema.
(See color figure at http://oftalmoloji.turkiyeklinikleri.com/)

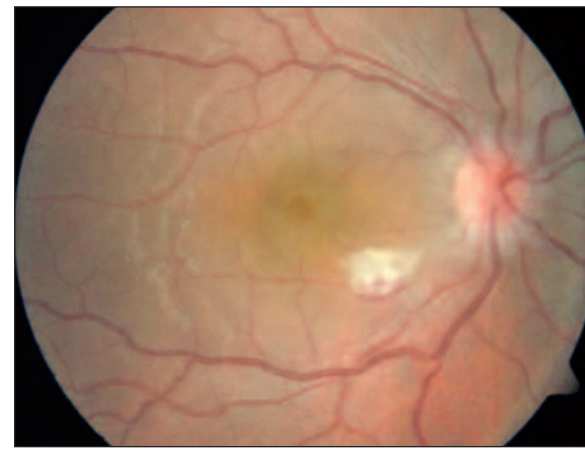

FIGURE 2: Fundus photograph of the right eye; At the first fundus examination, there is papilledema and exudative infiltrat at the inferonasal of the macula.
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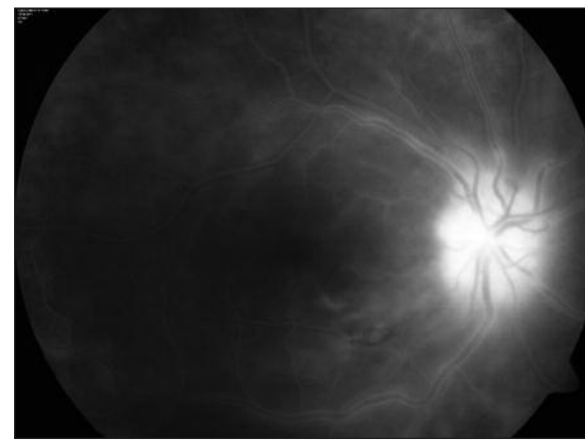

FIGURE 3: Fundus flourescein angiography of the right eye; there is posterir vasculitis.

MRI-MRA showed complete occlusion of left transvers and sigmoid sinus with thrombosis (Figure 5,6). Lumbar puncture opening pressure was 360 mm H20 with normal cells, glucose and protein. Prothrombin time (PT), partial thromboplastin time (PTT), fibrinogen, reptilase time, antithrombin III, protein C, protein S, homosistein were normal. Erythrocyte sedimentation rate (ESR) was 41 mm/h, CRP was 95,3 mg/L, platelets was 240x10^3/mL. It was thought that papilledema

caused by cerebral venous thrombosis, not posterior uveitis.

Pathergy response was resulted as negative and haplotype analyze of HLA-B5 was positive. In this patient, BS was diagnosed with the presence of the posterior uveitis and the history of oral, genital ulcers. Because of the CVSO, she was treated with pulse steroid for 7 days by neurology and her headache was interrupted with steroid therapy. Coumadin, azothioprine and colchicine were added to the treatment. At month 3 after treatment, her BCVA was 2/10 in the right eye, 10/10 in the left eye and there were neither papilledema nor macular edema, bilaterally (Figure 7,8).

Informed consent was obtained from the patient herself for publication of her findings as a case report.

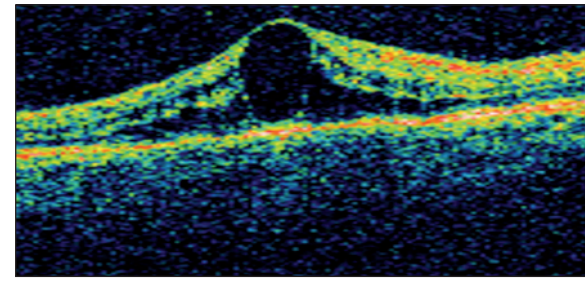

FIGURE 4: OCT photograph of the right eye; there is macular edema. OCT: optical coherence tomography.
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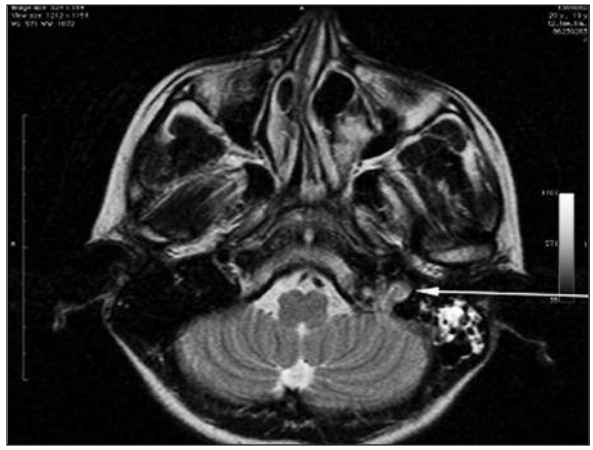


FIGURE5: Magnetic resonance ımaging of the head; T2 weighted image showing thrombosis in the left sagittal sinus.

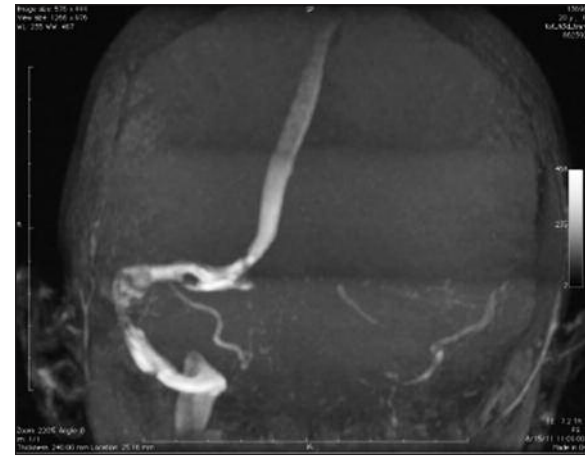
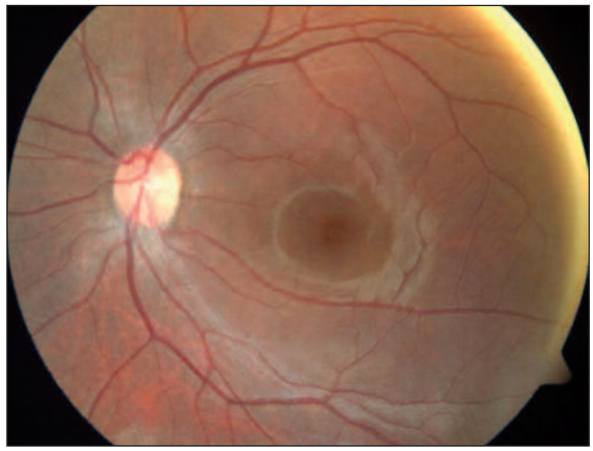


FIGURE 6: Cerebral magnetic resonance angiography; it confirms total occlusion of the left transvers and sigmoid sinus.



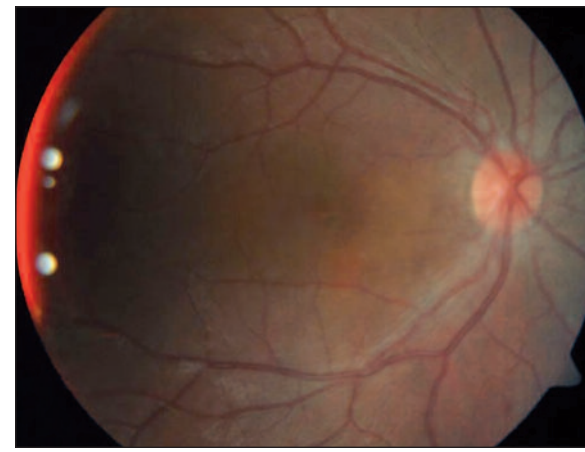


FIGURE 7,8: Fundus photograph; At month 3 after treatment, disc with clear boundaries, no papilledema.
(See color figure at http://oftalmoloji.turkiyeklinikleri.com/)

DISCUSSION

The key clinical manifestations of BS are recurrent aphthous ulcers, recurrent genital ulcers, recurrent uveitis and skin disorders. But the disease may also affect the CNS, the major vessels, gastrointestinal system and the joints.[6]

The reported frequency of oral ulcers, eye disease and neurological involvement among BS patients is approximately 97-100%, 50% and 5%, respectively.[7] In a recent study, the frequency of CNS involvement was 13,0% among men and 5,6% among women after two decades of follow-up.[8]

CVST is seen in 10-20% of BS patients in whom neurological involvement occurs.[7] The superior sagittal sinus is the most commonly thrombosed.[9,10] CVST can cause severe headache, mental changes and motor ocular palsies because of the increased intracranial pressure, but headache is the most common neurological symptom seen in BS.[5,7] Papilledema and sixth nevre paresis are the most common signs reported and hemiparesis may develop in some.[4,6,10]

Our patient presented with persistant headache and visual loss in the right eye. At ophthalmological examination, there was posterior uveitis in the right eye and papilledema. The further evaluation was applied because of the persistant headache and CVST was diagnosed.

Some cases presented with CVST, as the first presentation of BS, were reported in the literature. Swerdlow et al reported a 35-year-old woman with a recent diagnosis of pseudotumor cerebri who presented with headache, emesis, and blurring of vision. An MRI supported the presence of sagittal sinus thrombosis, a finding which was confirmed by MRA. Further workup for an underlying cause of sinus thrombosis disclosed symptoms and signs fulfilling the diagnostic criteria for BS.[11] Ascaso et al described a 30-year-old Algerian man with BS who developed, as first symptom of BS, a typical intracranial hypertension picture with headaches, bilateral papilledema and raised cerebrospinal fluid pressure. MRA revealed a CVST.[12] Chaloupka et al. reported a 13- year- old boy with recurrent venous thrombosis of the dural sinus. BS was diagnosed based on the medical history (recurrent oral ulcers, pseudo-folliculitis) and clinical examination (pathergy test was positive). As a result, CVST was accepted as the first manifestation of the BS.[13] Rahil et al reported CVST as the initial presentation of BS in a 40 year old man. The patient presented with a six-week history of headache, progressive loss of vision in the right eye and recurrent oral ulcers. MRI and MRA of the brain showed superior sagittal, left transverse, and left sigmoid sinus venous thrombosis. A diagnosis of BS was made based on clinical criteria.[14] Lizarazo-Barrera et al reported a 29-year-old female admitted for acute headache and vomiting. MRI showed a large thrombosis of sagital and transverse sinuses. She developed oral and genital ulcers a week later. Ophthalmologic examination revealed left anterior uveitis and ipsilateral papilledema. BS was diagnosed.[15] While there was no uveitis at presentation in cases of Swerdlow, Ascaso and Chaloupka, panuveitis and anterior uveitis were present in Rahil case and Lizarazo-Barrera case, retrospectively, in addition to CVST.

There has been no consistent primary abnormality of the coagulation, anticoagulation or fibrinolytic systems identified in BS patients.[16] HLA-B5 have been found to be a risk for thrombophlebitis.[17]

In the treatment, while some authors use a combination of anticoagulation with steroids, others use corticosteroids alone.[9,10] We used a combination of coumadin and corticosteroid in our patient for CVST and azothioprine and colchicine for BS.

As a result, CVST can be seen in the first presentation of BS. So,for a patient presenting with persistant headache and papilledema in addition to the posterior uveitis, CVST should be thought in the differential diagnosis. Persistant headache is an important sign that leads us to the diagnosis of CVST.

REFERENCES

1. Kontogiannis V, Powell RJ. Behçet's disease. Postgrad Med J 2000;76(900):629-37.
2. Goker B, Goker H. Current therapy for Behçet's disease. Am J Ther 2002;9(5):465-70.
3. Akman-Demir G, Serdaroglu P, Tasçi B. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. The Neuro-Behçet Study Group. Brain 1999;122 (Pt 11):2171-82.
4. Siva A, Kantarci OH, Saip S, Altintas A, Hamuryudan V, Islak C, et al. Behçet's disease: diagnostic and prognostic aspects of neurological involvement. J Neurol 2001; 248(2):95-103.
5. Saip S, Siva A. [Neuro-Behçet's syndrome]. Turkiye Klinikleri J Int Med Sci 2005;1(25):32-41.
6. Okada AA. Behçet's disease: general concepts and recent advances. Curr Opin Ophthalmol 2006;17(6):551-6.
7. Siva A, Altintas A, Saip S. Behçet's syndrome and the nervous system. Curr Opin Neurol 2004;17(3):347-57.
8. Kural-Seyahi E, Fresko I, Seyahi N, Ozyazgan Y, Mat C, Hamuryudan V, et al. The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. Medicine (Baltimore) 2003;82(1):60-76.
9. Akman-Demir G, Bahar S, Baykan-Kurt B, Gürvit IH, Serdaroğlu P. Intracranial hypertension in Behçet's disease. Eur J Neurol 1996;3(1):66-70.
10. Wechsler B, Vidailhet M, Piette JC, Bousser MG, Dell Isola B, Blétry O, et al. Cerebral venous thrombosis in Behçet's disease: clinical study and long-term follow-up of 25 cases. Neurology 1992;42(3 Pt 1):614-8.
11. Swerdlow RH, Hanna GR. Behcet's disease: presentation with sagittal sinus thrombosis diagnosed noninvasively. Headache 1996; 36(2):115-8.
12. Ascaso FJ, Rodriguez A, Cristóbal JA. Cranial hypertension as first manifestation of Behçet's disease: a case report. Doc Ophthalmol 2002;105(3):291-9.
13. Chaloupka K, Baglivo E, Hofer M, Chizzolini C, Delavelle J, Rossillion B, et al. [Cerebral sinus thrombosis in Behçet disease: case report and review of the literature]. Klin Monbl Augenheilkd 2003;220(3):186-8.
14. Rahil AI, Errayes M, Salem KM. Cerebral venous thrombosis as the initial presentation of Behcet's disease. Chang Gung Med J 2009; 32(2):220-3.
15. Lizarazo-Barrera JC, Jacobelli S, Mellado P, González S, Massardo L. [Extensive cerebral vein thrombosis as first manifestation of Behçet's disease. Report of one case]. Rev Med Chil 2010;138(6):746-51.
16. Leiba M, Seligsohn U, Sidi Y, Harats D, Sela BA, Griffin JH, et al. Thrombophilic factors are not the leading cause of thrombosis in Behçet's disease. Ann Rheum Dis 2004;63(11): 1445-9.
17. Calamia KT, Schirmer M, Melikoglu M. Major vessel involvement in Behçet's disease: an update. Curr Opin Rheumatol 2011;23(1):24-31.