

Blepharoptosis in Behçet's Disease

So-Min Hwang, On Lim,
Min-Kyu Hwang, Min-Wook Kim,
Jong-Seo Lee

Aesthetic, Plastic and Reconstructive
Surgery Center, Good Moonhwa Hospital,
Busan, Korea

No potential conflict of interest relevant to
this article was reported.

Behçet's disease is a systemic chronic disease that occurs in tissues such as eyes, joints, organs and nerves, and it has been noted that symptoms may be observed in a variety of tissues. In previous studies, reports of blepharoptosis observed in patients with Behçet's disease have been rare. We would like to report a case where a patient among those who visited our hospital with blepharoptosis had a history of Behçet's disease. This patient had been diagnosed with Behçet's disease, and complained of bilateral blepharoptosis even at the time of diagnosis. He complained of dysfunctions in vision and hearing, and upon eye examination, an eye movement disorder was found in his left eye. From the symptoms, neuro-Behçet's disease was diagnosed. The oculomotor and levator palpebrae superioris muscles are both controlled by cranial nerve III, which may suggest that Behçet's disease in this patient occurred in cranial nerve III. The patient received an oral steroid, and the symptoms have improved without surgery. Since we could identify the correlation between Behçet's disease and blepharoptosis, we considered that sharing this case and its outcome would be helpful for plastic surgeons who treat eyelids.

Keywords Behcet syndrome, Blepharoptosis

INTRODUCTION

Behçet's disease was named by Hulusi Behçet, a Turkish dermatologist, in 1937, and it is defined as a poorly understood multiple chronic inflammatory disorder [1,2]. Normally, Behçet's disease is accompanied by the 3 symptoms of oral ulcer, genital ulcer, and uveitis, and in addition, occasionally involves the central nerve system, articular tissues, and internal organs, resulting in the associated symptoms [3,4]. The most common diagnostic criteria of Behçet's disease are the Behçet's disease study group criteria (Table 1).

A patient among those who had visited our hospital with blepharoptosis as a major complaint had Behçet's disease, and subsequently, we investigated the correlation between Behçet's disease and blepharoptosis. In our literature survey, we found very few studies on

the correlation between the two diseases, and so we intended to report this case to demonstrate the relationship between them.

CASE REPORT

A 57-year-old male patient presented with blepharoptosis as a major complaint to our hospital in January 2015 (Fig. 1). The patient was diagnosed with Behçet's disease at a university hospital 5 years prior. At the time of diagnosis, he complained of oral aphthous ulcer and genital ulcer recurring every 2 to 3 months. The patient was also diagnosed with uveitis by ophthalmology, and those symptoms met the Behçet's disease study group criteria. After the diagnosis of Behçet's disease, the patient was given a steroid medication. However, he stopped regular medication voluntarily and was taking the medication only sporadically when he visited our hospital. He was diagnosed with bilateral blepharoptosis in the department of plastic surgery in our hospital, and levator muscle function was measured as 4 mm at the left and 5 mm at the right. Margin reflex distance-1 (MRD1) was 1 mm at the left and 2 mm at the right. During eye movement testing, reduced mobility of the left eye in the downgaze position was observed (Fig. 2). In addition, he complained of rapid deterioration of vision and hearing, which reportedly had begun at the time of the diagnosis of Behçet's disease.

Received: Apr 15, 2016 Revised: Aug 25, 2016 Accepted: Sep 19, 2016
Correspondence: So-Min Hwang Aesthetic, Plastic & Reconstructive
Surgery Center, Good Moonhwa Hospital, 119, Beomil-ro, Dong-gu, Busan
48735, Korea. E-mail: limon1980@naver.com

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Table 1. International Behçet disease study group criteria for the diagnosis of Behçet's disease

Must have had the following symptoms	
Recurrent oral ulceration	Minor aphthous, major aphthous, or herpetiform ulceration observed by physician or patient that recurred at least three times in one 12-month period
Plus two of the following	
Recurrent genital ulceration	Aphthous or scarring, observed by physician or patient
Eye lesions	Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination; or retinal vasculitis observed by ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis, papulopustular lesions; or acneiform nodules observed by physician in post-adolescent patients not on corticosteroids
Positive pathergy test	Read by physician at 24 to 48 hours



Fig. 1. Pictures of the patient with blepharoptosis taken 7 years ago and on the first visit to our hospital, respectively. The symptoms of blepharoptosis are observed to have occurred after the occurrence of Behçet disease.



Fig. 2. Picture of eye movement from the patient with blepharoptosis. The deterioration in eye movement is observed in the downgaze of his left eye.

We did not perform an operation immediately due to the history of irregular steroid administration, and decided to prescribe him a steroid, which is the standard treatment for Behçet's disease. He also refused surgery because of eye problems such as glaucoma and deterioration of vision. Subsequently, we were to make a decision on surgery after observing progress upon the regular administration of a steroid.

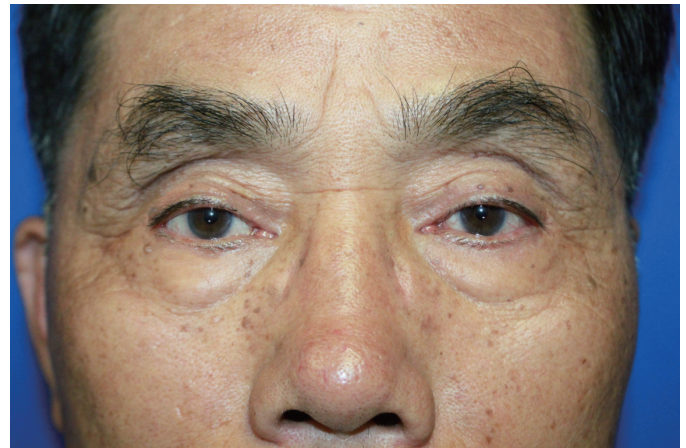


Fig. 3. Picture of the patient taken after an 8-month course of oral steroid treatment. Improvement in the symptoms of blepharoptosis is observed.



Fig. 4. Picture of the patient taken after 8-month course of oral steroid treatment. Improvement was observed in the downgaze of his left eye.

The patient visited the hospital again after 8 months, and improvement of the blepharoptosis could be observed (Fig. 3). In the le-

vator muscle function test, it was measured to be 7 mm at the left and 8 mm at the right, and MRD1 was 4 mm at both upper eyelids, which was a clear improvement in symptoms. The mobility of the left eye in the downgaze position was improved (Fig. 4).

DISCUSSION

Behçet's disease is a chronic systemic inflammatory disorder that involves the central nervous system, intestine system, and vascular system, leading to multiple clinical symptoms [1,4,5]. The most widely used diagnostic criteria are the Behçet's disease study group criteria (Table 1). Typical symptoms include oral ulcer, genital ulcer, and uveitis. Our patient had all of the aforementioned symptoms, which met the diagnostic criteria. Behçet's disease can be classified into vascular Behçet's disease, intestinal Behçet's disease, and neuro-Behçet's disease depending on the affected site, and among them, cases involving the nervous system are called neuro-Behçet's disease, which accounts for 5% of all cases of Behçet's disease [1,5-7].

Currently, neuro-Behçet's disease does not have widely used diagnostic criteria, and is normally diagnosed by medical imaging examinations and clinical symptoms [1]. Our patient complained of neurological symptoms such as vision loss, hearing loss, and deterioration in eye movement in addition to blepharoptosis, and was considered to have neuro-Behçet's disease as a subclass of Behçet's disease. The most common involved site of neuro-Behçet's disease is the brain stem; in addition, the hemisphere, meninges, and spinal cord may be involved [7]. Clinical symptoms vary according to each involved site, and among them, we noted a case where the brain stem was involved, in which case the symptoms appear in the organ where the cranial nerve branching off from the brain stem is innervated. The deterioration in eye movement is caused by the oculomotor muscle hypomotility that is controlled by the cranial nerve system (CNS) III. If neuro-Behçet's disease involves the CNS III, the levator muscle hypomotility of the upper eyelid may accompany this symptom, resulting in blepharoptosis. Our patient also showed deterioration in eye movement and blepharoptosis simultaneously, which could be considered a finding of involvement of the CNS III.

Treatment of Behçet's disease is aimed at alleviating the symptoms, reducing the infection, and controlling the immune system. Although the treatment differs slightly for each involved organ, corticosteroids and anti-tumor necrosis factor (anti-TNF) are commonly used. Our patient had been taking an oral steroid before visiting our hospital, although administration was not regular, as

he had stopped and then restarted the medication voluntarily. Our medical staff presented the accurate oral steroid dose to the patient, and he took 1 mg/kg/day of oral prednisone regularly for approximately 8 months. Five months later, when he visited the clinic again, a clear improvement of the blepharoptosis was observed, and thus it could be seen that the symptoms were improved without surgery.

Plastic surgeons who perform surgery on the eyelid due to blepharoptosis or other related diagnoses must carefully check the medical and medication histories of patients. For patients with Behçet's disease, the history of steroid medication absolutely must be checked before surgery, and it can be helpful for establishing a treatment direction to identify the extent of involvement of Behçet's disease by accurate eye movement and physical examinations. We focused on neuro-Behçet's disease among the subclasses of Behçet's disease, and could see that deterioration in eye movement and blepharoptosis could occur if the CNS is affected by neuro-Behçet's disease. Since no studies could be found that have provided an accurate description of the relationship between Behçet disease and blepharoptosis, it is suggested that more studies are needed in the future, for which this case can serve as a starting point.

PATIENT CONSENT

Patients provided written consent for the use of their images.

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