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LETTER TO THE EDITOR

An Exceptional Case of Full-thickness Macular Hole Closure in a Patient with Behçet Disease

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ABSTRACT

Purpose: To present a case of a full-thickness macular hole closure in a patient with Behçet uveitis

Design: A 23-year-old-male patient with Behçet Disease had clinically inactive uveitis with topical steroids, oral azathiopurine and cyclosporine until he developed a retinal infiltrate in the left eye which evolved into a full-thickness macular hole during the follow-up.

Methods: Strict control of inflammation and subsequent vitrectomy was planned. Meanwhile another attack of panuveitis developed in the left eye and subcutaneous Interferon alfa-2b interferon treatment was initiated.

Results: After 2 months, the patient was clinically inactive with complete closure of the macular hole.

Conclusions: Strict control of inflammation may result in closure of the macular hole and avoid the need for vitrectomy.

Keywords: Behçet's disease, Behçet uveitis, macular hole, spontaneous closure

Behçet disease (BD) is a chronic inflammatory multi-system disorder of unknown cause. The disease is mainly characterized by ocular, oral, cutaneous, genital, vascular, and neurological manifestations.¹ Ocular pathologies, mostly bilateral panuveitis, are present in 60–80% of BD patients and are major causes of morbidity that may finally lead to blindness.² Among various associated ocular pathologies, development of a macular hole (MH) was reported in 2.6% and 3.4% of patients with BD in two large-scale studies.^{3,4} MH in BD may ultimately require vitreoretinal surgery, but a recently published study concluded that surgical intervention does not seem to have a potential beneficial effect on visual function.⁵ This case report presents closure of a full-thickness MH, with strict control of inflammation, in a patient with Behçet uveitis.

A 23-year-old male patient with a 2-week history of blurred vision in the right eye was referred to our clinic. His medical history revealed recurrent oral

aphthous ulcers for 1 year and a genital ulcer, which was present 2 months ago. On initial examination bilateral best-corrected visual acuity was 10/10. Slit-lamp biomicroscopic examination of the right eye revealed 2+ cells in the anterior chamber and 3+ in the vitreus. The left eye had trace amount of cells in the anterior chamber and vitreus. Fundus examination was unremarkable in the right eye, but small intraretinal hemorrhages with adjacent exudation in the nasal quadrant were observed in the left eye. Fundus fluorescein angiography showed diffuse dye leakage, signs of periphlebitis and cystoid macular edema in the left eye. Behçet disease was diagnosed based on the International Study Group for Behçet's Disease criteria.¹

Both eyes were prescribed hourly topical corticosteroids and cycloplegics TID in addition to oral azathiopurine (Imuran; Glaxo SmithKline, 2.5 mg/kg/day). The topical treatment was tapered gradually. During the follow-up, a retinal infiltrate developed

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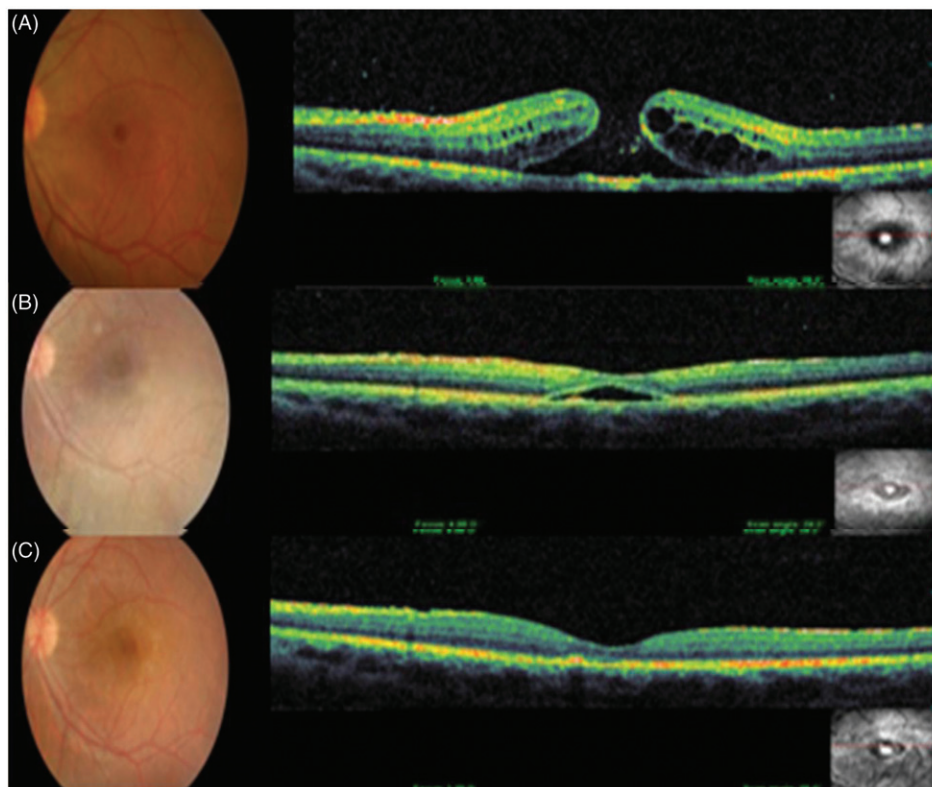


FIGURE 1. Color fundus photograph and spectral domain-optical coherence tomography (OPKO/OTI Spectral OCT/SLO, OPKO Health, Inc. Miami, Florida) images of the left eye show (A) a full-thickness macular hole before, (B) macular hole closure with subretinal fluid accumulation 1 month after, and (C) complete macular hole closure 4 months after the initiation of interferon alfa-2a therapy.

in the right eye, which prompted the addition of oral cyclosporine A (Sandimmun Neoral; Novartis Pharma, 5 mg/kg/day) and oral corticosteroids (Deltracortril; Pfizer, 1 mg/kg/day, tapered gradually) to the ongoing treatment. Three months after the first ocular manifestation, a panuveitis attack was observed in the left eye. The dosage of oral corticosteroids was increased to 1.5 mg/kg/day and the inflammation was brought under control. In the follow-up, visual acuity in the left eye decreased to 2/10 due to the development of a full-thickness macular hole (Figure 1A). Strict control of inflammation and subsequent vitrectomy were advised by the vitreoretinal surgeon. While the patient was waiting for vitrectomy, another panuveitis attack developed in the left eye. Due to the inadequacy of the ongoing treatment, subcutaneous interferon alfa-2b treatment at a daily dosage of 5×10^6 units (Intron A; Schering Plough) was initiated and tapered accordingly (5×10^6 2×/week after 1 month).⁶ The patient was clinically inactive with complete closure of the macular hole after 2 months. Best-corrected visual acuity increased to 7/10 in the left eye (Figure 1B, C).

Antero-posterior traction, tangential traction, and degeneration of the inner retinal layers have been suggested as possible mechanisms of idiopathic

macular holes.⁷ The same principles can be applied for macular holes observed as a complication of posterior pole involvement in uveitis disease. Recurrent inflammation associated with edema and cystoid degeneration of the macula is suggested to contribute to macular hole development by causing disruption of retinal layers. Changes on the vitreoretinal interface, abnormal vitreoretinal tractions, and development of an epiretinal membrane secondary to inflammation are also thought to contribute to macular hole development by causing antero-posterior and tangential traction.⁸

Several hypothetical mechanisms have been proposed as explanations to spontaneous closure of a macular hole. These include spontaneous release of vitreous traction, shrinkage of a developed epiretinal membrane, glial cell or RPE cell proliferation, and bridging of retinal tissue over the macular hole.⁹ Although inconclusive, the following mechanisms can be adapted to this case as possible explanations to the macular hole closure: induction of glial or RPE cell proliferation at the base of the hole secondary to the severe inflammation in the last attack might have caused an approximation of hole borders; control of inflammation might have resulted in the resolution of cystoid macular edema and release of vitreous traction with subsequent

flattening of the macula. Overall, strict control of inflammation in Behçet uveitis patients might promote the closure of the hole and avoid the need for vitrectomy.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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