Behcet’s disease is an inflammatory disorder of unknown cause, characterized by recurrent oral aphthous ulcers, genital ulcers, uveitis, and skin lesions [1,2]. Diagnosis is based on the presence of a constellation of clinical findings [3]. Behcet’s disease can affect both the anterior and posterior segments of the globe. Posterior segment manifestations include retinal vasculitis involving both arteries and veins leading to arterial and venous occlusions, retinal hemorrhage, retinal infiltrates, edema, and an inflammatory response in the vitreous [4,5]. Posterior segment involvement leads to irreversible alterations and serious vision loss. Macular hole, though rarely reported, is a major cause of irreversible vision loss [5–8]. In this report, we present two cases of macular hole in the worse eye of bilateral cases of Behcet’s disease.

**Case Presentation**

**Case 1**
A 37-year-old male was referred to us for blurred vision in the left eye for 9 months; it was diagnosed as optic neuritis and he received steroid treatment for 3 months. Intermit-
tent redness was noted in both eyes in the 4 months prior
to referral. His history revealed recurrent oral ulcers and
genital ulcers for more than 1 year. On initial examination,
best-corrected visual acuity was 20/25 in the right eye and
counting fingers at 5 cm in the left eye. Slit-lamp biomicros-
copy revealed trace cells in both anterior chambers. Indirect
ophthalmoscopy revealed 2+ vitreous haziness in both
eyes, a slightly hyperemic disc in the right eye, retinal
hemorrhage in both eyes, and a full-thickness macular hole
in the left eye. Cystoid change was noted in the surrounding
retina. Fundus fluorescein angiogram (FAG) showed dye
leakage from the optic disc and capillaries in both eyes,
cystoid macular edema in the right eye, and a central win-
dow defect in the macular center of the left eye (Figure 1).
An erythematous nodule was noted after needling in the
forearm. Behcet’s disease was diagnosed based on the
International Study Group for Behcet’s Disease (ISGBD)
criteria [3]. Other laboratory data were negative, including
routine blood, HIV, Venereal Disease Research Laboratory
(VDRL), antinuclear antibody (ANA), and immunoglobu-
lin tests.

Topical and systemic corticosteroid and oral cyclosporin
A (200 mg/day) treatment was initiated. The inflamma-
tion was controlled with resolution of vitreous inflammation
and retinal infiltrates. Visual acuity had improved to 20/20
in the right eye and 20/800 in the left eye 2 months later. Un-
fortunately, secondary glaucoma and complicated cata-
A 32-year-old male came with the chief complaint of blurred vision and redness in his right eye for 1 week. In the past 3 years, he had suffered from intermittent blurred vision and pain in both eyes. He had a history of recurrent episodes of genital ulcers, oral ulcers, and acne-like skin lesions. On the initial examination, best-corrected visual acuity was 20/200 in the right eye and 20/300 in the left eye. Slit-lamp biomicroscopy revealed 2+ cells in the right anterior chamber and trace cells in the left eye. Indirect ophthalmoscopy revealed 4+ vitreous haziness in the right eye and 2+ vitreous haziness, retinal hemorrhage, and sheathing on the distal segment of the temporo-lower branch of the retinal arteriole, as well as a full-thickness macular hole with an edematous edge, in the left eye. FAG showed leakage from the optic disc and capillaries in both eyes (especially the right eye), cystoid macular edema in the right eye, and a central window defect in the macular center of the left eye (Figure 2). Physical examination revealed erythema nodosum at the wrist and oral ulcer. Behcet’s disease was diagnosed based on the ISGBD criteria [3]. Other laboratory data were negative, including routine blood and urine, HIV, VDRL, ANA, rheumatoid factor, and immunoglobulin tests.

Topical and systemic corticosteroid as well as oral cyclosporin A (200 mg/day) treatment was initiated. The inflammation was controlled with resolution of vitreous inflammation and retinal infiltrates. Visual acuity improved to 20/20 in the right eye 1 month later. The vision did not improve in the left eye due to the macular hole. No macular hole had developed in the right eye by the fifth year of follow-up.

**DISCUSSION**

Ocular involvement occurs in 60–80% of patients with Behcet’s disease and presents as panuveitis in most cases. Although the hallmark of intraocular inflammatory disease is the presence of cells and flare, it is often the sequel of inflammation that appears to be sight threatening. Vision loss
in Behcet’s disease usually results from long-standing inflammation and consequent sequelae in the posterior segment, macular lesions, retinal and optic nerve atrophy, vitreous hemorrhage, and neovascular glaucoma [4,5,7,8]. Although most macular aspects of visual morbidity have been reported, macular hole is rarely discussed.

The development of a partial- or full-thickness macular hole in uveitis is relatively uncommon. Careful three-mirror examination of putative holes very often reveals a thinly covered cystoid lesion rather than a hole. Macular hole is thought to be related to posterior vitreous detachment (PVD) [9]. A previous study reported a high prevalence of PVD in Behcet’s disease and other uveitis cases [10]. While the exact mechanisms are not clear, an increased number of inflammatory cells in the vitreous is thought to be the primary reason for vitreous gel shrinkage in uveitis, and this shrinkage induces PVD with minimal vitreous liquefaction. In these cases, vitreous opacity was noted in the active stage. In our cases, macular hole was noted in the eye with more condensed vitreous opacity on the first visit. The surrounding retina appeared to have cystoid changes, instead of the cuff of subretinal fluid seen in idiopathic macular hole. According to the history, the eye with the macular hole was the one involved first. The vision of the eye with the macular hole did not improve after control of inflammation by intensive medical treatment, although no macular hole had developed in the fellow eye at the time of the last follow-up. We speculate that aggressive treatment decreased the intensity of vitreous inflammation and macular edema, preventing the development of a macular hole in the fellow eye.

Macular hole is a rare complication of panuveitis in Behcet’s disease and can ultimately lead to severe loss of vision. From our cases, macular hole might be prevented by early diagnosis and aggressive treatment.

REFERENCES


黃斑裂孔在貝西氏症患者之表現

許淑娟 楊正安
高雄榮民總醫院眼科

貝西氏症為一原因不明之發炎性疾病，以反覆性口腔潰瘍、生殖器潰瘍、葡萄膜炎及皮膚病變為特徵。百分之六十至八十的患者會有眼表現，其中以雙眼全葡萄膜炎主。眼後葉發炎常造成嚴重視力損傷，黃斑部裂孔雖不常見，卻是可能導致嚴重視力喪失的原因之一。作者提出兩位雙眼全葡萄膜炎併單眼黃斑裂孔之病例，探討其臨床表現、可能之成因及治療方式。

關鍵詞：貝西氏症，黃斑部裂孔
（高雄醫誌 2004;20:558—62）