

A novel mechanism of macular holes in patients with Behçet's uveitis

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Abstract:

To describe two cases of Behçet's retinitis lesions in the macula causing sloughing of retinal tissue forming a full-thickness macular hole. This was a case series study. Case 1 was a 26-year-old presented, known case of Behçet's disease, presented with a large area of retinitis involving the center of the macula with overlying vitritis. One day after the initiation of treatment, vitritis improved, but the retinitis patch sloughed and created a full-thickness macular hole. Case 2 was a 31-year-old male, known case of Behçet's uveitis, who presented vitritis and multiple retinitis patches involving the macula of the left eye. Two weeks after infliximab infusion, the inflammation resolved with medical management, however, the retinitis patch was complicated by a full-thickness macular hole with an inferior rhegmatogenous retinal detachment. Retinitis at the macula can be complicated by a full-thickness macular hole.

Keywords:

Behçet's uveitis, macular hole, posterior uveitis, retinitis, uveitis

INTRODUCTION

Macular hole is reported in 3.4% of patients with Behçet's uveitis.^[1] The mechanism of macular hole was thought to be related to posterior vitreous detachment and macular edema.^[2,3] In this report, we described two unique cases of a Behçet's retinitis causing sloughing of retinal tissue forming a full-thickness macular hole.

CASE REPORTS

Case 1

A 26-year-old male patient presented to the ophthalmology emergency room with gradual decreased vision in the left eye for 1 month. The patient reported a history of recurrent painful oral and genital ulcers and deep venous thrombosis in both legs. On examination, visual acuity was 20/20 in the right eye and counting fingers near the face in the left eye. The intraocular pressure was 12 mmHg in both eyes. Examination of the right eye was

unremarkable. Anterior segment examination of the left eye showed 1 + cells in the anterior chamber. Fundus examination of the left eye revealed the presence of vitritis along with a large area of retinitis involving the central macula with small patches of retinitis in the parafoveal area [Figure 1a]. Fundus fluorescein angiography (FFA) of the left eye confirmed the presence of diffuse capillary leakage with disc leakage [Figure 1b]. Optical coherence tomography examination revealed the presence of attachment between the vitreous and the central macula at the site of the retinitis inducing avulsion of the retina [Figure 1c].

Systemic workup including purified protein derivative test, venereal disease research laboratory test, rapid plasma reagin, and computerized tomography of the chest was unremarkable. The diagnosis of Behçet's uveitis was made, and medical management with infliximab infusion (5 mg/kg) and mycophenolate mofetil (1 g twice a day) was started. One day after the initiation of treatment, vitritis improved, but the retinitis patch sloughed and created a full-thickness macular hole [Figure 2]. Eventually, the inflammation was controlled

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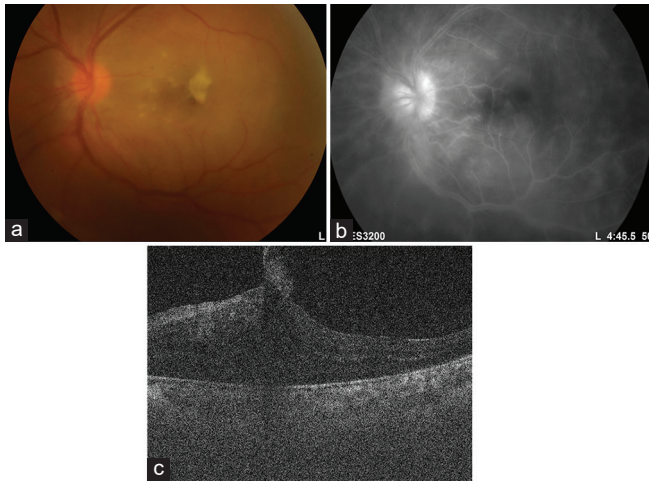


Figure 1: (a) Fundus photography of the left eye demonstrating the presence of vitritis along with a large area of retinitis involving the center of the macula and small patches of retinitis in the parafoveal area. (b) Fundus fluorescein angiography showing diffuse capillary leakage and disc leakage. (c) Optical coherence tomography illustrated the attachment of the vitreous to the retina at the site of the retinitis creating vitreomacular traction

with infliximab (5 mg/kg) infusion every 8 weeks and mycophenolate mofetil 500 mg twice per day, and visual acuity was stable at 20/200.

Case 2

A 31-year-old male, known case of Behçet's uveitis, presented with sudden decreased vision in the left eye. On physical examination, visual acuity was 20/20 in the right eye and counting finger at the near face in the left eye. Anterior segment examination of the left eye showed the presence of 3 + white cells in the anterior chamber. On fundus examination of the left eye, there was vitritis with multiple patches of retinitis [Figure 3a]. Examination of the fellow eye was unremarkable. FFA confirmed the presence of diffuse capillary leakage with an inflamed optic disc. The patient received immediately infliximab (5 mg/kg/day) infusion. Two weeks later, the patient presented with decreased vision in the same eye. On fundus examination, there was a full-thickness macular hole with an inferior rhegmatogenous retinal detachment [Figure 3b and c]. Subsequently, he underwent pars plana vitrectomy with encircling band and silicone oil tamponade.

DISCUSSION

Macular involvement, which was reported in 24% of the eyes with Behçet's uveitis, includes retinitis, macular ischemia, macular atrophy, vascular occlusion, macular edema, and rarely macular hole.^[4] Vitreoretinal tractions, epiretinal membrane, and macular edema all contribute to the formation of macular holes in Behçet's uveitis.^[4,5] Vitreous cells cause vitreous shrinkage and the induction of posterior vitreous detachment.^[5] In the two presented cases, the retinitis resulted in sloughing of retinal tissue and the formation of

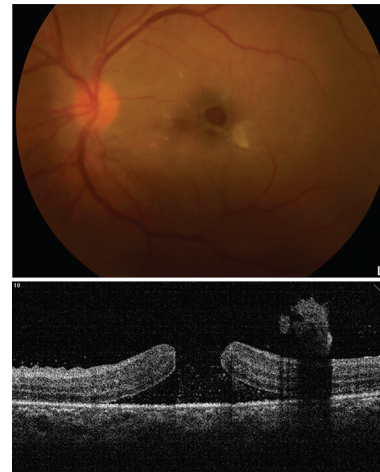


Figure 2: One day after treatment, the retinitis patch sloughed and created a full-thickness macular hole as shown clinically and illustrated with optical coherence tomography

a full-thickness macular hole. Up to our knowledge, this unpredictable mechanism has never been described in the literature.

Surgical intervention did not have any significant improvement in Behçet's macular holes, most likely due to macular damage secondary to recurrent inflammation and macular ischemia.^[2] Nevertheless, a case was described in the literature where a macular hole closed spontaneously with adequate medical management for Behçet's uveitis.^[3] This stresses on the importance of early diagnosis and initiation of treatment. Although immediate treatment was started in the first case, the retinitis focus became necrotic and sloughed within 24 h of treatment administration. This may be explained by the fact that the patient had presented late, and the retinitis patch already became necrotic and the macular hole was inevitable even with appropriate medical management. However, in the second case, despite that the patient presented 1 day after the onset of his symptoms, the retinitis patch eventually formed a full-thickness macular hole. Hence, retinitis patches at the macula carry the risk of forming full-thickness macular holes regardless of the onset of presentation.

In conclusion, the development of full-thickness macular holes in patients with Behçet's uveitis is multifactorial and cannot be attributed to macular edema and vitreoretinal tractions alone.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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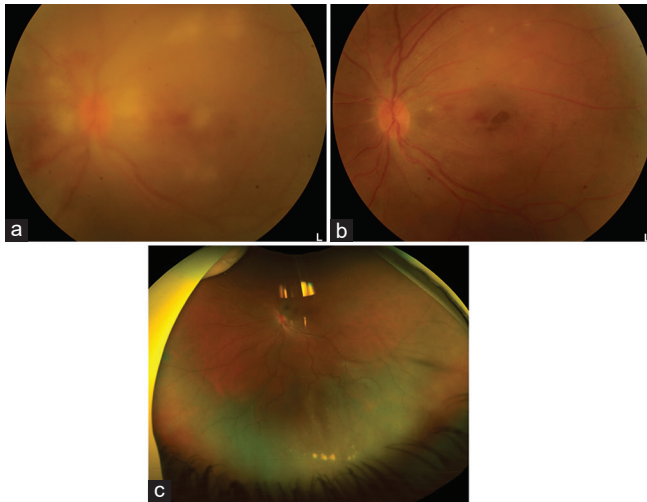


Figure 3: (a) Fundus photography of the left eye of case 2 showing vitritis with multiple patches of retinitis. (b) Fundus photography of the left eye of case 2 showing the presence of full-thickness macular hole 2 weeks later. (c) Ultra-widefield fundus photo showing the presence of shallow inferior rhegmatogenous retinal detachment

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Conflicts of interest

There are no conflicts of interest.

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