Bilateral Sequential Inflammatory Full Thickness Macular Holes in Birdshot Chorioretinopathy

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Introduction

Birdshot chorioretinopathy (BSCR) is a rare, insidious, chronic form of posterior uveitis characterized by progressive, bilateral hypopigmented choroidal lesions that can lead to vision loss and often require anti-inflammatory and immunosuppressive treatment[1]. A full-thickness macular hole (FTMH), typically caused by vitreomacular traction (VMT), is a break in the retina at the macula, leading to significant loss of central vision. We present the first case of bilateral FTMHs in a patient with BSCR.

Case

A 57-year-old woman presented in September 2020 with dry eyes and vision loss. Visual acuity (VA) was 6/7.5 OD and 6/6 OS. Examination revealed punctate epithelial erosions bilaterally, a mild cataract on the right, and mild vitritis bilaterally. Imaging showed choroidal granulomas, and she tested positive for HLA-A29, confirming BSCR. Initial optical coherence tomography (OCT) showed central foveal thicknesses of 223µm OD and 236µm OS, with minor epiretinal membranes (ERMs) and posterior vitreous detachments (PVDs) in both eyes (Fig. 1). Over several months, the patient experienced worsening bilateral photophobia, floaters, and nyctalopia. Mycophenolate mofetil improved her symptoms, and birdshot lesions remained stable, but it was later discontinued due to lymphopenia, with dexamethasone implants and adalimumab initiated in its place. By April 2023, left eye foveal thinning had progressed, and by July, vision had dropped to 6/12 OD and 6/24 OS. A 40µm FTMH was detected in the left eye, which expanded to approximately 550µm by November (Fig. 2), prompting a pars plana vitrectomy (PPV), internal limiting membrane (ILM) peeling, and immunosuppressants in December, improving VA to 6/30. She subsequently developed an approximately 810µm FTMH and significant cataract in the right eye, requiring surgery, which ultimately improved her VA to 6/38 in both eyes (6/24 best-corrected). Postoperative OCT showed closed FTMHs but residual foveal atrophy bilaterally and left eye cystoid macular oedema (CMO), which was treated with an Iluvien injection. VA improved to 6/18 OD and 6/24 OS the following month, with no active inflammation. Systemic anti-inflammatory therapy continues.

Imaging

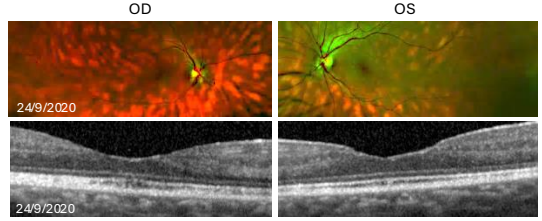

Figure 1: Initial widefield retinal imaging and OCT performed at the patient's presentation in 2020.

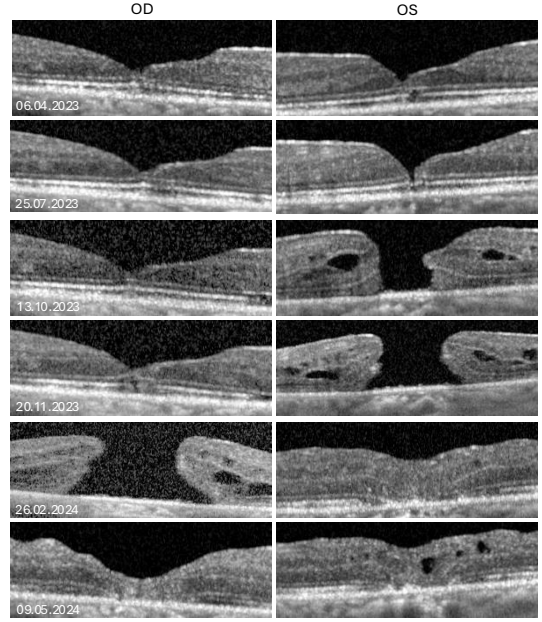

Figure 2: OCT scans showing progressive foveal atrophy over time, ultimately leading to FTMH formation in both eyes. Surgical intervention successfully closed both holes, with postoperative CMO in the left.

Discussion

The occurrence of macular holes in BSCR is rare. A longitudinal cohort study evaluating macular changes in patients with BSCR only identified one lamellar macular hole in 80 patients (160 eyes)[2]. To our knowledge however, there are no previously reported cases of FTMH associated with BSCR. The pathogenesis of primary (idiopathic) FTMHs involves anteroposterior and tangential traction at the vitreomacular interface, often due to anomalous PVD[3]. While most FTMHs are primary, some arise from postoperative CMO, trauma, or high myopia[4]. FTMH is a rare complication of posterior uveitis[4]. The pathogenesis of inflammatory macular holes is less understood and is thought to be multifactorial, involving inflammation-driven fragilization of retinal layers, chronic changes in the vitreous leading to PVD, and traction from inflammatory ERMs. Notably, OCT imaging in our patient showed full PVDs bilaterally in 2020, long before FTMH development, with no signs of opercula in the foveae. Minimal bilateral ERMs were unlikely to generate significant traction, and imaging ruled out inflammatory CMO as a cause. OCT images of the patient's macula showed no elements typically preceding macular hole formation, as well as a prior 'clean' detachment of the posterior vitreous from the retina. Macular atrophy is recognized in BSCR[5] and was observed in our patient. We propose that the FTMHs resulted from this progressive atrophy, leading to retinal fragility rather than conventional causes. Our patient experienced extensive BSCR, leading to years of posterior segment inflammation and retinal degeneration, as evidenced by generalized retinal atrophy and reduced central macular thickness. Morgan et al[6] described a thinned and atrophic macula with altered architecture as involutional macular thinning (IMT) and identified patients with IMT, in the absence of other FTMH risk factors, as being at significantly higher risk of developing idiopathic FTMHs. This IMT may parallel the inflammatory macular atrophy in our patient, predisposing her to FTMH formation. The authors proposed that the initial step in FTMH development may involve choroidal vascular changes causing ischemia in the fovea and retinal pigment epithelium, which can lead to macular thinning. In our patient's case, perhaps her extensive macular atrophy was due to choroidal ischemia caused by the chronic inflammation, however, we do not have angiographic evidence to corroborate this theory. FTMHs resulting from posterior uveitis can occasionally resolve spontaneously with medical treatment alone; however, we opted for surgical intervention after five months of unsuccessful non-surgical management of the first eye, as the FTMHs were large. The patient underwent PPV with ILM peeling and gas tamponade, successfully closing the left eye's FTMH and improving visual symptoms. A multicentre case series showed an 81% closure rate and a 75% improvement in visual acuity after similar surgeries for inflammatory FTMHs[7]. Unfortunately, the right eye subsequently developed a rapid enlargement of an FTMH. Given the patient's reduced vision in the first eye and the rapid progression of the second FTMH, we expedited surgical repair with a similar surgical technique to maximize visual and anatomical outcomes.

In conclusion, we present the first reported case of bilateral FTMHs in a patient with BSCR. We discuss the significance of inflammation-driven macular thinning in their pathogenesis, particularly in the absence of typical tractional VMT/ERM or inflammatory CMO, which are usually responsible for FTMH in posterior uveitis. We also demonstrate the successful surgical repair of the hole supplemented by periocular, topical and systemic immunosuppressive therapy.

References

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