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Delayed-Onset White-Dot Syndrome in the Setting of Traumatic Choroidal Rupture

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Abstract

Purpose: To describe a patient with a traumatic choroidal rupture and a submacular hemorrhage, the course of which was complicated by delayed-onset posterior uveitis resembling a white-dot syndrome. **Methods:** A single case was evaluated. **Results:** A 34-year-old man presented after being struck in the left eye with a tennis ball. The visual acuity (VA) was 20/30 with otherwise normal ophthalmic vitals. An examination showed traumatic iritis and choroidal rupture with a submacular hemorrhage without subfoveal involvement. Despite treatment of anterior segment inflammation and a worsening hemorrhage with topical agents and intravitreal aflibercept, the VA decreased to 20/600. A repeat examination with optical coherence tomography showed new optic disc edema, placoid outer retinal lesions adjacent to the choroidal rupture, and corresponding ellipsoid zone atrophy. A broad workup was unremarkable, and the patient completed a long taper of high-dose oral prednisone without recurrence. **Conclusions:** Traumatic exposure of the immunologically privileged subretinal space to high-flow choroidal circulation likely triggered a pathway involving self-autoantigenicity and a uveitic response.

Keywords

choroidal rupture, submacular hemorrhage, posterior uveitis, white-dot syndrome(s)

Introduction

Choroidal rupture consists of a break in the choroid, retinal pigment epithelium (RPE), and Bruch membrane[1] that usually occurs in the setting of ocular trauma. Blunt or penetrating ocular trauma causes compression of the anterior-posterior globe with horizontal hyperextension, leading to breakage of the less elastic aforementioned structures at the site of impact, more commonly known as a contrecoup injury.[2,3] Complications include subretinal hemorrhaging, commotio retinae, and choroidal neovascularization (CNV).[4]

When the macula is involved, choroidal rupture may result in a submacular hemorrhage, inducing retinal damage via hemotoxic effects on photoreceptors, blockage of metabolic exchange between the RPE and photoreceptors, or avulsion of photoreceptors secondary to fibrin clot contraction.[5] Given the potential for vision loss, a submacular hemorrhage should be promptly managed; however, no optimal treatment has yet been established. Treatments for submacular hemorrhage include pneumatic displacement with or without tissue plasminogen activator (tPA) or antivascular endothelial growth factor (anti-VEGF) injections, surgical evacuation with or without tPA injection, macular translocation, RPE patching, or photodynamic therapy.[6] In contrast, choroidal rupture without foveal involvement is typically managed with observation, Amsler grid testing, and, in cases of CNV, anti-VEGF therapy.[4]

Although ocular trauma can result in immune-mediated disease, such as sympathetic ophthalmia[7] or autoimmune retinopathy,[8] choroidal rupture has rarely been reported as a cause of immune-mediated disease.[9] We describe a case of a 34-year-old man who developed multiple evanescent white-dot lesions approximately 2 weeks after a traumatic choroidal rupture and submacular hemorrhage.

Case Report

A 34-year-old man with no medical history presented to the retina clinic after a traumatic injury in which his left eye was struck by a tennis ball. He reported a gradually worsening crescent-shaped shadow in the left eye without photopsia. The initial examination showed an uncorrected visual acuity (VA) of 20/20

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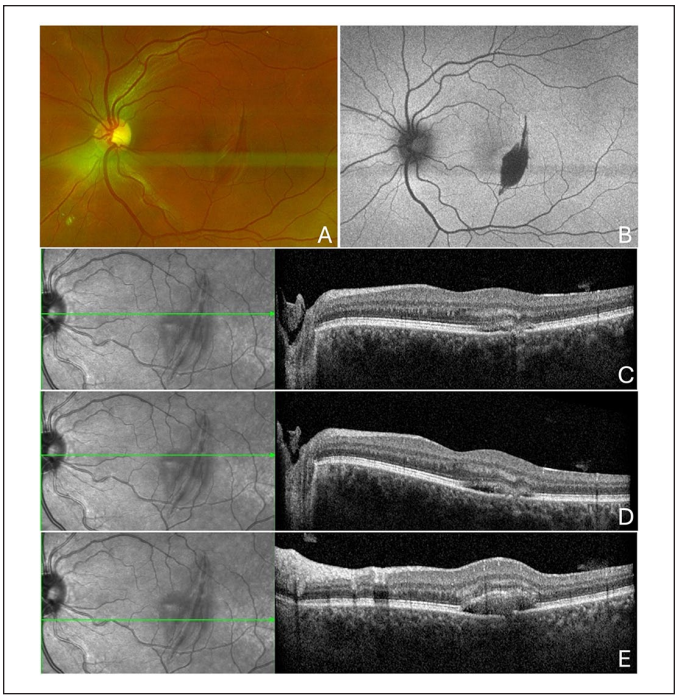

Figure 1. (A) Fundus imaging at presentation shows a choroidal rupture with a subretinal hemorrhage. (B) Fundus autofluorescence shows linear hypoautofluorescence consistent with choroidal damage and subretinal blood. (C–E) Optical coherence tomography of adjacent regions of rupture shows subretinal fluid and subretinal hyperreflective material suggestive of hemorrhage adjacent to a break in Bruch membrane and an irregular choroidal contour with otherwise intact adjacent inner and outer retinal laminations.

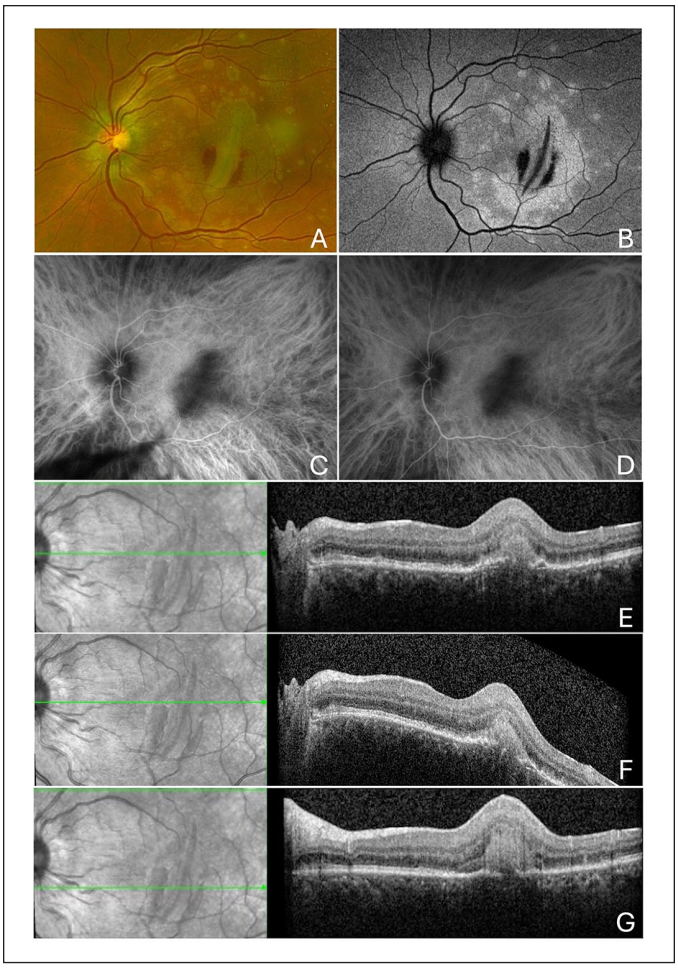

Figure 2. Imaging approximately 2 weeks after initial presentation. (A) Fundus imaging shows a choroidal rupture and submacular hemorrhage with an annular distribution of the surrounding placoid lesions. (B) Fundus autofluorescence shows linear hypoautofluorescence consistent with choroidal damage. (C and D) Indocyanine green angiography shows a large central mixed filling and blocking defect. (E–G) Optical coherence tomography of adjacent regions of rupture shows persistent subretinal hyperreflective material suggestive of hemorrhage, a new expanded attenuation of the ellipsoid zone, and outer retinal hyperreflective deposits and subfoveal hyperreflectivity.

OD and 20/30 OS with an intraocular pressure (IOP) of 16 mm Hg and 13 mm Hg, respectively. Extraocular movements and visual fields were full bilaterally, and an external examination was unremarkable. A slitlamp examination of the left eye was significant for conjunctival injection, 2+ anterior chamber cells, angle recession, and vitreous syneresis. Optical coherence tomography (OCT) and a fundus examination of the left eye showed a choroidal rupture with a subretinal hemorrhage without subfoveal involvement. Fundus autofluorescence showed corresponding linear hypoautofluorescence consistent with the presence of subretinal blood and a choroidal rupture (Figure 1).

The patient was prescribed topical prednisolone and cyclopentolate for traumatic iritis. Surgical evaluation with or without tPA was discussed. Given the presence of a fovea-sparing hemorrhage, the retina and glaucoma services recommended close observation with serial examinations.

One week later, the patient returned reporting worsening vision and dizziness. The VA was 20/25 OS in the setting of a worsening submacular hemorrhage with juxtafoveal involvement, for which he received intravitreal aflibercept without complications.

Six days after the injection, the patient again returned reporting an enlarging central scotoma and new central and peripheral photopsias. At the time, the VA was counting fingers (20/600 pinhole), and formal visual field testing was completed. The IOP was stable, and the slitlamp examination was significant for improving anterior inflammation, with few pigmented cells seen in the anterior vitreous. A dilated funduscopy examination showed new Frisen grade 1 left optic disc edema, papillitis, and innumerable outer retinal white-dot lesions in an annular distribution surrounding the choroidal rupture with placoid consolidation around the rupture site. The submacular hemorrhage was smaller and remained fovea-sparing. The new lesions were found to be hyperautofluorescent with corresponding areas of ellipsoid zone (EZ) attenuation and subretinal hyperreflective deposits on OCT, which also showed rare vitreous cells. Indocyanine green angiography showed central mixed blocking and filling defects in the left eye (Figure 2).

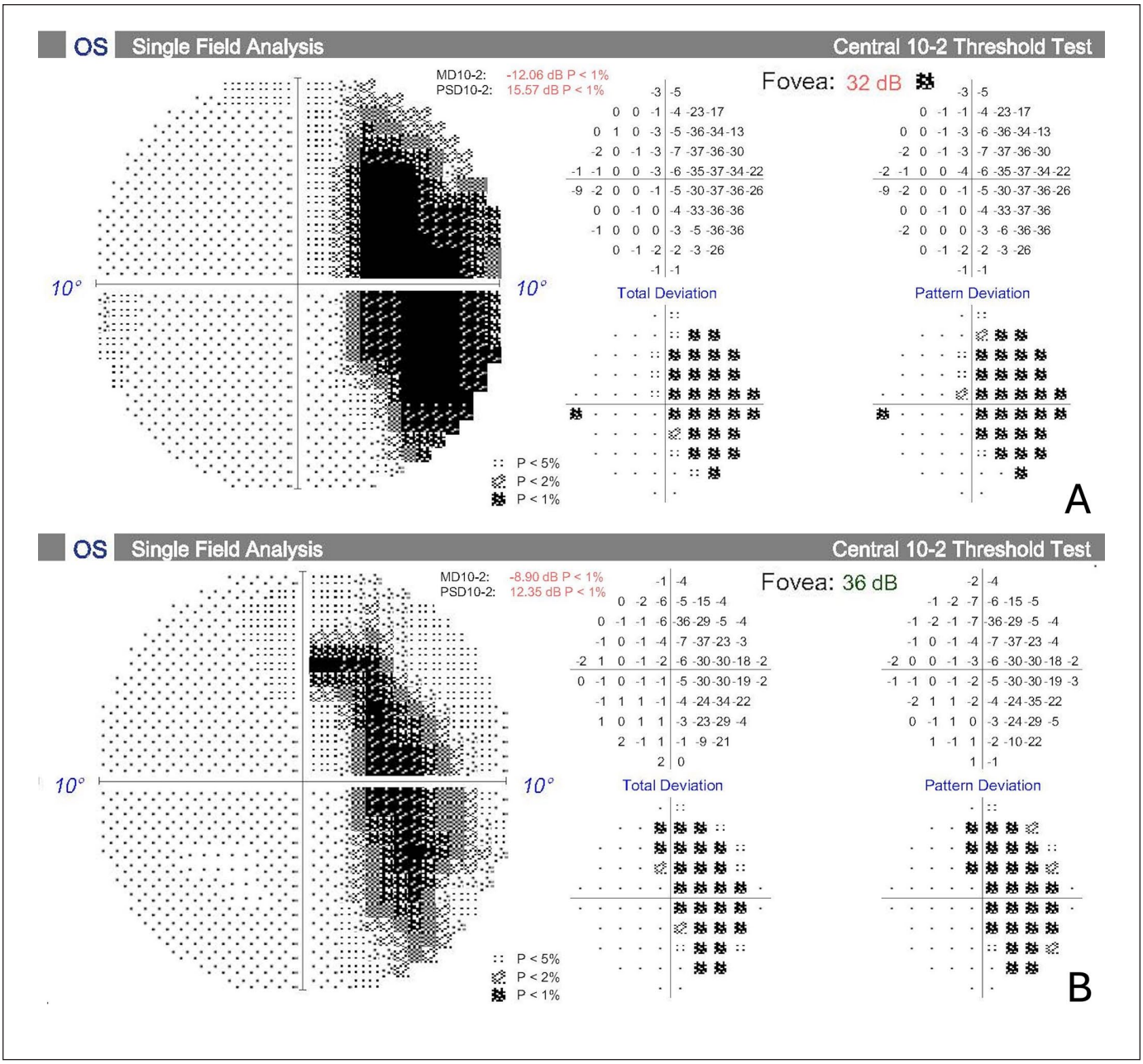

Figure 3. 10-2 Humphrey visual field tests of the left eye. (A) The eye has a dense nasal paracentral scotoma approximately 3 months after initial presentation. (B) An improvement in visual field loss is seen approximately 9 months after initial presentation.

Given the patient's improving subretinal hemorrhage, his worsening VA and visual field deficits (Figure 3) were attributed to the new inflammatory outer retinal lesions, which involved the fovea and peripapillary region. Before systemic steroid therapy was initiated, a broad infectious and inflammatory workup was performed to rule out other causes of chorioretinitis. Testing, including the erythrocyte sedimentation rate, C-reactive protein, rapid plasma reagin, QuantiFERON Gold, double-stranded DNA, antineutrophil antibody, human leukocyte antigen-B27, and *Bartonella* screening, was negative or within normal limits. The patient was prescribed prednisone 60 mg daily and gastrointestinal prophylaxis with continuation of topical prednisolone and cyclopentolate drops.

The patient's following course was notable for traumatic mydriasis and multifactorial ocular hypertension in the setting of angle recession and a presumed steroid response, which was managed with topical drops. In the setting of new photopsias, a small anterior retinal tear was identified 1 month after the initial trauma and was treated with laser barricade the same day. Gradual recovery of the EZ was noted on serial OCT follow-up, as was recovery of the patient's nasal paracentral scotoma (Figure 3). After completion of a 5-month steroid taper, the

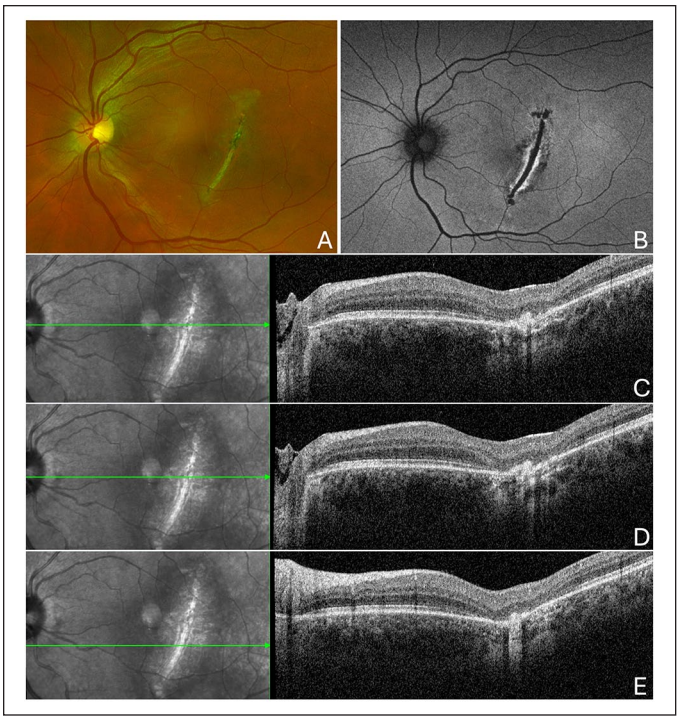

Figure 4. (A) Fundus imaging approximately 11 months after initial presentation. (B) Fundus autofluorescence shows hypoautofluorescence and hyperautofluorescence at the previously ruptured region. (C–E) Optical coherence tomography of adjacent regions of rupture shows an interval of improvement of subretinal hyperreflective material and ellipsoid zone recovery adjacent to the site of a break in Bruch membrane.

best-corrected VA improved to 20/60 OS. The outer retinal inflammatory lesions resolved with further EZ recovery and resolution of ocular hypertension (Figure 4).

Conclusions

Although a rare process,[9] the exposure of the immunologically privileged subretinal space to high-flow choroidal circulation led to the development of posterior uveitis resembling a white-dot syndrome with features of multiple evanescent white-dot syndrome (MEWDS) and acute posterior multifocal placoid pigment epitheliopathy. A traumatic choroidal rupture disrupts the tight junctions between RPE cells that form the outer blood–retina barrier[10] by physically blocking entry of bloodborne toxins into the subretinal space and inhibiting effector T cells to promote systemic tolerance. RPE cells express few or no major histocompatibility complex class II molecules for initiation of immune responses, and they also secrete anti-inflammatory cytokines such as interleukin-10 to induce immunosuppression and express proteins, such as Fas ligand, to prevent immune cell activation.[11] Under the right conditions, RPE damage and subsequent exposure of the subretinal space can promote self-autoantigenicity and inflammation, as in our patient's case.

The inflammatory chorioretinopathy seen in our patient shared features with several white-dot syndromes, specifically MEWDS and acute posterior multifocal placoid pigment epitheliopathy. MEWDS and acute posterior multifocal placoid pigment epitheliopathy are both classically associated with viral illnesses; however, MEWDS typically occurs unilaterally in myopic women between 20 years and 50 years of age, while acute posterior multifocal placoid pigment epitheliopathy occurs bilaterally in men and women between 20 years and 30 years of age. Both syndromes present with lesions at the level of the outer retina and RPE of the posterior pole. Disc edema and lesion consolidation, as observed in our patient, is also seen in MEWDS.[12]

Choroidal rupture has been rarely reported as a cause of immune-mediated disease. Fung et al[9] described a similar atypical white-dot syndrome reminiscent of MEWDS and multifocal choroiditis and panuveitis that occurred 10 weeks after a traumatic choroidal rupture and subretinal hemorrhage in a 24-year-old woman. Gray–white dots at the level of the outer retina and RPE, a subretinal inflammatory mass adjacent to a resolving subretinal hemorrhage, vitritis, papillitis, vasculitis, foveal granularity, and blind-spot enlargement improved with systemic prednisone. The authors suspected an inflammatory chorioretinopathy incited by choroidal antigens or dehemoglobinized blood. In addition, although there are reports of MEWDS arising at the site of atrophic scars,[13,14] to our knowledge no report has described MEWDS arising secondary to the use of anti-VEGF therapy. Therefore, we attribute the patient's inflammatory response to his choroidal rupture rather than his aflibercept injection.

Although in general MEWDS and acute posterior multifocal placoid pigment epitheliopathy are self-resolving (albeit with the latter having a worse prognosis), foveal involvement may warrant treatment with systemic steroids.[12] Given the foveal involvement of the inflammatory lesions, our patient was treated with a prolonged high-dose steroid taper, and there was nearly complete resolution of lesions 5 months later.

In summary, we present a case of a white-dot syndrome that occurred after a traumatic choroidal rupture with a submacular hemorrhage with features similar to MEWDS and acute posterior multifocal placoid pigment epitheliopathy. We posit that these findings were precipitated by the exposure of the immunologically privileged subretinal space to antigens in the high-flow choroidal circulation and resulted in the development of self-autoantigenicity. Our findings provide further insight into the function of the outer retinal barrier and the mechanism behind other inflammatory chorioretinopathies.

Ethical Approval

This case report was conducted in accordance with the Declaration of Helsinki. The collection and evaluation of all protected patient health information were performed in a US Health Insurance Portability and Accountability Act–compliant manner. A single case report does not constitute human subjects research per the Weill Cornell Medicine Institutional Review Board; therefore, ethical approval was not obtained.

Statement of Informed Consent

Consent was obtained from the patient for clinical care. Given that the present report does not contain features, images, or clinical information that could result in patient identification, informed consent for the report was not obtained.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of the article.

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