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Case report

Visual recovery after surgical repair of chronic macular detachment associated with peripheral retinoschisis

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ABSTRACT

Purpose: To report 2 cases of chronic macular detachment associated with peripheral retinoschisis in which surgical repair resulted in significant visual recovery.

Observations: A 44-year-old man and 60-year-old woman were evaluated for chronic macular detachment, with a duration of 5 years and 6 months, respectively. In each case, optical coherence tomography was used to establish a diagnosis of full-thickness macular detachment resulting from peripheral retinoschisis and to confirm or identify the pathogenic outer layer breaks. After surgical repair with pars plana vitrectomy, endolaser photocoagulation of outer layer breaks, and gas tamponade, both patients had significant improvement in vision. The best-corrected visual acuity improved from 20/50- to 20/20 at one year post-operatively in the first patient and from 20/1250 to 20/200 at 8 months post-operatively in the second.

Conclusion and importance: In cases of chronic schisis-detachment involving the macula, surgical intervention can sometimes result in unexpected levels of visual recovery.

1. Introduction

Peripheral degenerative retinoschisis is the end result of cystoid peripheral degeneration of the retina and involves progressive retinal splitting, typically at the level of the outer plexiform layer.[1] Patients with this form of retinoschisis often remain stable and asymptomatic, although some may develop an associated retinal detachment. Byer et al.[2] reported that the incidence of progressive symptomatic schisis-detachment is quite rare, with a ratio of asymptomatic schisis-detachment to progressive symptomatic schisis-detachment of 178:1. Progressive symptomatic schisis-detachment is one of the few indications for surgical repair. On the other hand, degenerative retinoschisis without breaks, with asymptomatic outer layer breaks, or with localized schisis-detachment rarely requires treatment.[2] A long duration of macular detachment is generally expected to cause permanent photoreceptor injury and loss, regardless of the etiology of the detachment. We report two cases of chronic macular detachment associated with peripheral retinoschisis in which surgical repair with pars plana vitrectomy led to substantial improvement in visual function.

2. Findings

2.1. Case 1

A 44-year-old man reported poor central vision in the right for the previous 5 years. He had been diagnosed at the age of 39 with peripheral retinoschisis in the right eye and macular elevation presumed to represent extension of retinoschisis into the macula. No treatment had been recommended. Our evaluation revealed visual acuity of 20/50-2 in the right eye and 20/20 in the left eye. There was moderate myopia (approximately −6.00 D) in both eyes. Fundus examination of the right eye showed peripheral retinoschisis inferotemporally with an outer layer break temporal to the macula causing full-thickness macular detachment. The detached neurosensory retina in the macular area appeared thin and transparent. Fundus examination of the left eye showed shallow peripheral retinoschisis inferotemporally with no macular involvement. Optical coherence tomography (OCT) of the right eye confirmed the peripheral retinoschisis, outer layer break, and full-thickness macular detachment with outer retinal atrophy (Fig. 1).

After discussion of the uncertain visual prognosis, the patient underwent right pars plana vitrectomy with induction of posterior vitreous detachment, drainage retinotomy over the outer retinal break, endolaser photocoagulation around the outer layer break, and gas tamponade. One year post-operatively, the visual acuity had improved

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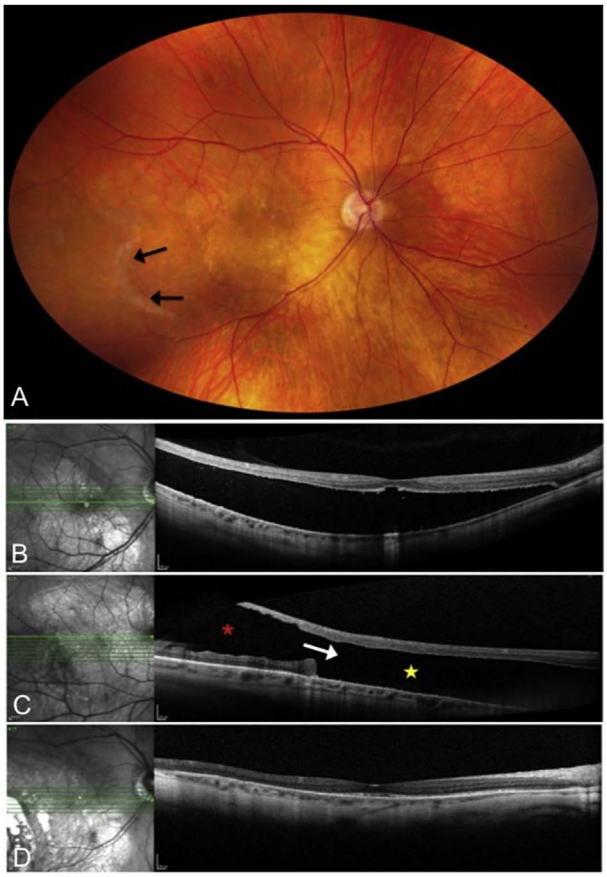

Fig. 1. Fundus photograph and optical coherence tomography (OCT) of Case 1. (A) Fundus photograph of the right eye shows an outer retinal layer break (black arrows). The chronic shallow macular detachment and peripheral retinoschisis are not visible due to transparency of the retina. (B) OCT image shows a full-thickness retinal detachment involving the macula. There is atrophy of the outer retinal layers due to chronicity of the detachment. (C) OCT image through the outer layer break in the temporal macula demonstrates a communication (white arrow) between the peripheral retinoschisis cavity (red asterisk) and macular detachment (yellow star). (D) OCT image 5 years post-operatively shows the macula to be attached with only mild attenuation of the ellipsoid zone in the foveal region and absence temporally. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

to 20/20 in the right eye and the macula was flat. OCT imaging 5 years postoperatively showed the outer retinal layers to be largely intact in the central macula, but atrophic temporally (Fig. 1). The shallow peripheral retinoschisis in the left eye had not progressed.

2.2. Case 2

A 60-year-old woman with a history of gyrate atrophy, myopia and type 2 diabetes mellitus with mild non-proliferative retinopathy was referred for evaluation of macular retinal detachment and vitritis in the left eye. The macular detachment had been present for at least 5 months and thorough evaluations at other institutions had failed to elucidate its cause. The visual acuity measured 20/40 in the right eye and 20/1250 in the left eye. Slit lamp biomicropscopy was notable for 1+ vitreous cell in the right eye and 2+ vitreous cell in the left eye, without posterior vitreous detachment in either eye. Fundus examination showed extensive peripheral chorioretinal atrophy and pigment derangement (Fig. 2) with localized chorioretinal atrophy in the temporal macula in both eyes. Additionally, there was cystoid macular edema and retinal detachment involving the entire macular area in the left eye. Meticulous contact lens biomicroscopy of the left eye revealed no retinal breaks, but suggested the presence of shallow retinoschisis extending over the chorioretinal atrophy in the inferonasal periphery of the left

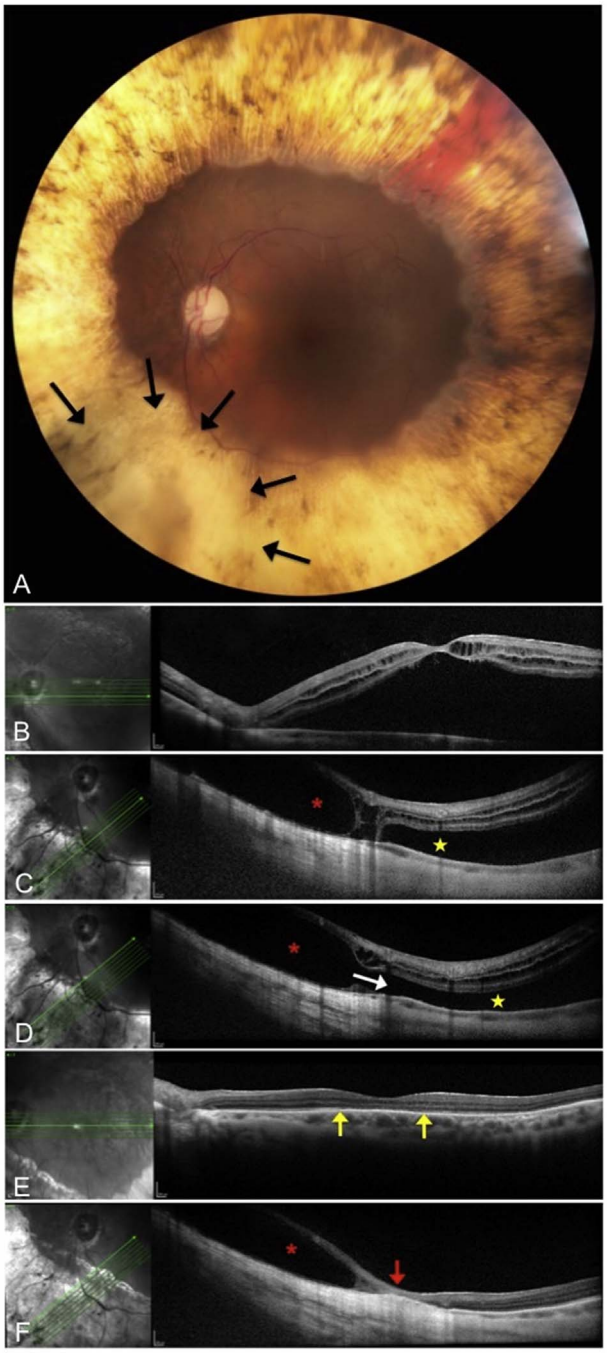

Fig. 2. Fundus photograph and optical coherence tomography (OCT) of Case 2. (A) Fundus photograph of the left eye shows extensive chorioretinal atrophy associated with gyrate atrophy. The posterior retina was detached out to the edge of the atrophy and the peripheral retina overlying the atrophy was attached. Although not visible in the photograph, biomicrosopy suggested the presence of shallow retinoschisis overlying chorioretinal atrophy in the inferonasal quadrant (black arrows). (B) Preoperative OCT image confirms retinal detachment involving the macula with associated retinal edema. (C) OCT image at the edge of the chorioretinal atrophy confirms the presence of peripheral retinoschisis (red asterisk) and retinal detachment (yellow star) involving the macula. (D) OCT image shows a single tiny communication (white arrow) between the peripheral retinoschisis cavity (red asterisk) and macular detachment (yellow star). (E) OCT image 3 months post-operatively demonstrates that the macula is attached with resolution of macular edema. Outer retinal signals are preserved except in the foveal area (between yellow arrows) and temporally. (F) OCT image through previous communicating channel shows persistence of the retinoschisis cavity (asterisk) inferonasally and sealing of the communication by laser scarring (red arrow) with complete attachment of the macula. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

eye. The inner layer appeared extremely thin, but no inner or outer retinal breaks could be visualized on examination. Extensive OCT imaging revealed a single tiny communication between the inferonasal retinoschisis cavity and the subretinal fluid in the macula (Fig. 2).

Laboratory evaluation of the vitritis was unrevealing and the macular detachment persisted several weeks after an empirical intravitreal triamcinolone injection. After discussion of the uncertain visual prognosis, pars plana vitrectomy with induction of posterior vitreous detachment, endodrainage, fluid-gas exchange, and endolaser photocoagulation of the communication between the retinoschisis cavity and subretinal fluid was performed. Eight months postoperatively, visual acuity in the left eye had improved to 20/200 and the macula was completely attached, with attenuation of outer retinal signals in the central macula (Fig. 2).

3. Discussion

Each of our patients was referred for an unusual macular elevation and found to have a macular detachment resulting from a communication between a peripheral retinoschisis cavity and the subretinal space. In one patient, the communication could only be identified by meticulous OCT imaging of the border between the schisis cavity and the full thickness macular detachment. Because schisis-detachments commonly develop in eyes with peripheral retinoschisis complicated by outer layer breaks,[2] patients diagnosed with retinoschisis should be educated in the signs and symptoms of retinal detachment and monitored regularly to prevent progression of detachment into the macula.

The management decision-making in our patients was complicated by the chronicity of the detachments—5 years in one patient and approximately 6 months in the other. Each patient elected to proceed with surgical repair despite the likelihood of permanent retinal damage and the uncertain prognosis for significant visual recovery. Despite the long chronicity of macular detachment, surgery was effective in reattaching the retina and inducing substantial visual recovery, with one patient achieving a best-corrected visual acuity of 20/20 by one year postoperatively.

The unexpected degree of visual recovery in these cases prompts speculation about why these eyes seemed to fare better than eyes with chronic macula-involving rhegmatogenous retinal detachment. One possible explanation is that shallow retinal detachments, as in these cases of chronic schisis-detachment, generally have a better prognosis than bullous detachments.[3] It is plausible that the closer proximity of the retina to the retinal pigment epithelium/choriocapillaris in an eye with a shallow retinal detachment[3] results in less outer retinal ischemia and subsequent degeneration. Another possible explanation involves the difference in the composition of the submacular fluid in schisis-detachment (schisis fluid) versus rhegmatogenous retinal detachment (liquid vitreous). It is reasonable to speculate that schisis fluid may not be as harmful to photoreceptors as liquid vitreous, resulting in better preservation of photoreceptors in chronic schisis-detachment compared with rhegmatogenous detachment of similar duration.

Joshi and co-workers[4] and Drenser and colleagues[4] analyzed schisis fluid in patients with X-linked retinoschisis and found 2 unique proteins, tenascin-C and cystatin C. Neither of these proteins was found in the vitreous of the same patients nor in normal control vitreous.[4,5] Tenascin-C is an extracellular matrix glycoprotein that modulates cell adhesion to fibronectin in the extracellular matrix.[6] It is upregulated by factors involved in inflammation resolution, suggesting that it may have a role in controlling inflammation.[7] It has been found to loosen extracellular matrix adhesions in cells exposed to tractional force. Such changes in cell adhesion may protect against retinal stretch trauma.

Cystatin C is a protease inhibitor that likely protects tissues during pathologic conditions.[8] It is thought to modulate photoreceptor outer segment degradation by inhibiting C1 cysteine proteases. Such proteases cause tissue destruction during inflammation and infection.[8] Cystatin C in submacular retinoschisis fluid may potentially play a role in protecting the detached macula from tissue degradation.

4. Conclusion

In summary, both of our cases of chronic schisis-detachment were successfully treated with vitrectomy surgery and had substantial visual improvement despite long-duration macular detachment. The precise explanation for the favorable visual outcomes is unknown, but may involve differences in the composition of retinoschisis fluid compared with liquid vitreous that favor photoreceptor preservation. We believe these results should encourage surgical repair of similar cases in the future and promote additional studies of the composition of retinoschisis fluid.

Patient consent

As no identifying information is disclosed, patient consent was not obtained.

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Conflict of interest

The following authors have no financial disclosures: VSD, DHG. Dr. Johnson serves on Data and Safety Monitoring Boards for Tyrogenex and Ohr Pharmaceuticals and is site principal investigator for a clinical trial sponsored by Hoffman La-Roche.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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