

Managing Complications of Retinoschisis

Most patients with retinoschisis will remain asymptomatic without treatment. It is important to know when and how to intervene.

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Although degenerative retinoschisis is usually benign, vision-threatening complications can occur. It is important to appropriately manage these complications, which include:

- Posterior extension of the retinoschisis cavity.
- Outer wall breaks and “schisis detachment,”¹ which occurs when schisis fluid accumulates in the subretinal space.
- Progressive rhegmatogenous retinal detachment (RRD) associated with retinoschisis, in which breaks of both the inner and outer layers allow liquefied vitreous to gain access to the subretinal space.

POSTERIOR EXTENSION OF THE SCHISIS CAVITY

When a patient is found to have posterior retinoschisis (Figure 1), it is easy to get concerned because of the following logic:

- The retinoschisis was not there at birth;
- the retinoschisis started in the periphery;
- it is now near the arcades; and
- therefore, it will eventually wipe out the macula.

This logic has led to attempts to halt progression of retinoschisis using a wide array of techniques.¹ However, the temptation to lay down a strong chorioretinal scar with some “low risk” laser, for example, should be resisted. The natural history of retinoschisis suggests that it almost never progresses significantly more posterior from where it is first observed.¹ In fact, most cases of posterior retinoschisis will not progress beyond 3 disc diameters from the macula. Only a handful of cases of degenerative retinoschisis involving the macula have been reported.²⁻⁵

Additionally, no treatment, including laser, has been shown to halt the progression of retinoschisis. One possible explanation for this phenomenon is that retinoschisis, unlike retinal detachment, is multifocal in nature.⁶ Cases in which the disease appears to have



Figure 1. Superotemporal retinoschisis encroaching on the arcades.

progressed through laser scars may in reality be retinoschisis arising de novo in previously unaffected areas of the retina. Finally, although retinoschisis causes an absolute scotoma, patients are almost invariably asymptomatic, even with very posterior retinoschisis. Treatment for these patients should be observation.

Occasionally, a patient with retinoschisis will be referred for laser prophylaxis before cataract surgery. However, neither cataract surgery¹ nor posterior vitreous detachment (PVD)⁶ have any adverse effect on retinoschisis. This makes sense, as the pathology is within the retina, not at the vitreoretinal interface.

OUTER WALL BREAKS AND SCHISIS DETACHMENT

Outer wall breaks are usually large (greater than 3 disc diameters), round, and posterior, often with rolled edges. If they are chronic, a cuff of subretinal fluid may create a ring of pigmentation (Figures 2 and 3). This appearance is in contrast to inner wall breaks, which are typically small and round, reminiscent of atrophic holes. Outer wall

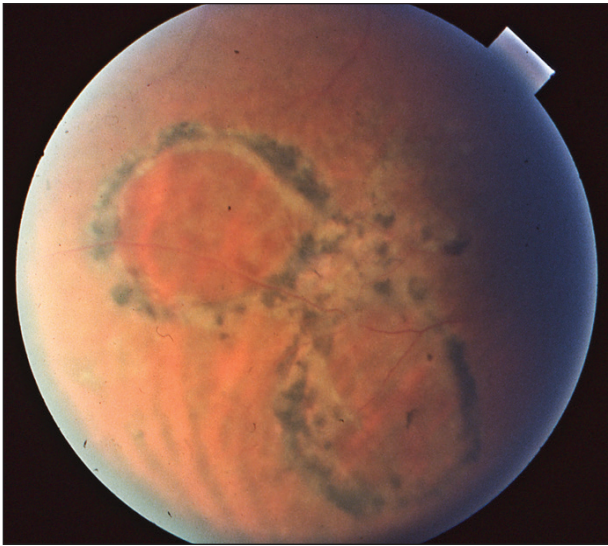


Figure 2. Two large, posterior, circular outer wall breaks. Pigmented cuffs denote chronicity.

breaks are common, found in 11% to 24% of retinoschisis cases.^{1,7,8} The large size and posterior location increase the perceived threat of outer wall breaks, and their amenability to laser prophylaxis increases the temptation to treat^{9,10}; however, studies suggest that outer wall breaks usually remain stable.¹

The worst typical outcome of outer wall breaks is the development of an asymptomatic schisis detachment. Byer found that schisis detachment occurred in

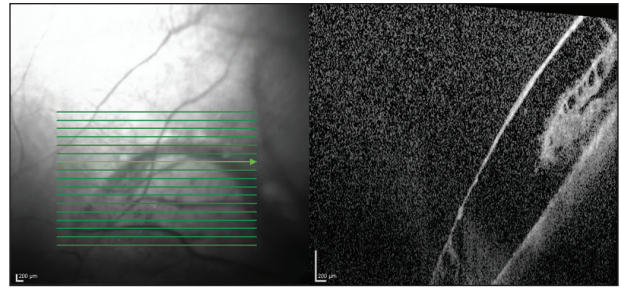


Figure 3. Optical coherence tomography shows the scrolled edge of an outer wall break.

58% (14 of 24) of patients with outer wall breaks during long-term follow up.¹ Fortunately, the subretinal fluid does not typically extend beyond the borders of the schisis cavity itself, likely due to the highly viscous nature of the intraschisis fluid and its high mucopolysaccharide content.⁶ None of the patients in Byer's natural history study received treatment or developed symptoms due to retinoschisis complications over an average of 9 years of follow-up.¹ However, until more comprehensive data is available, surgeons' judgments and patients' wishes should guide discussion over whether to barricade outer wall breaks and localized schisis detachments.

Rarely, the subretinal fluid of a schisis detachment will leak into the macula and cause symptomatic vision loss (Figure 4).^{9,11,12} However, even very posterior subretinal fluid may remain stable for years.¹³ For example, despite the juxtafoveal fluid in Figure 4, the vision remained



Figure 4. Posterior extension of subretinal fluid in a case of schisis detachment. Note the 2 large superotemporal outer wall breaks. The posterior extent of the schisis cavity is seen at the temporal macula (arrowhead). The posterior extent of the subretinal fluid (arrowhead) involves the fovea.

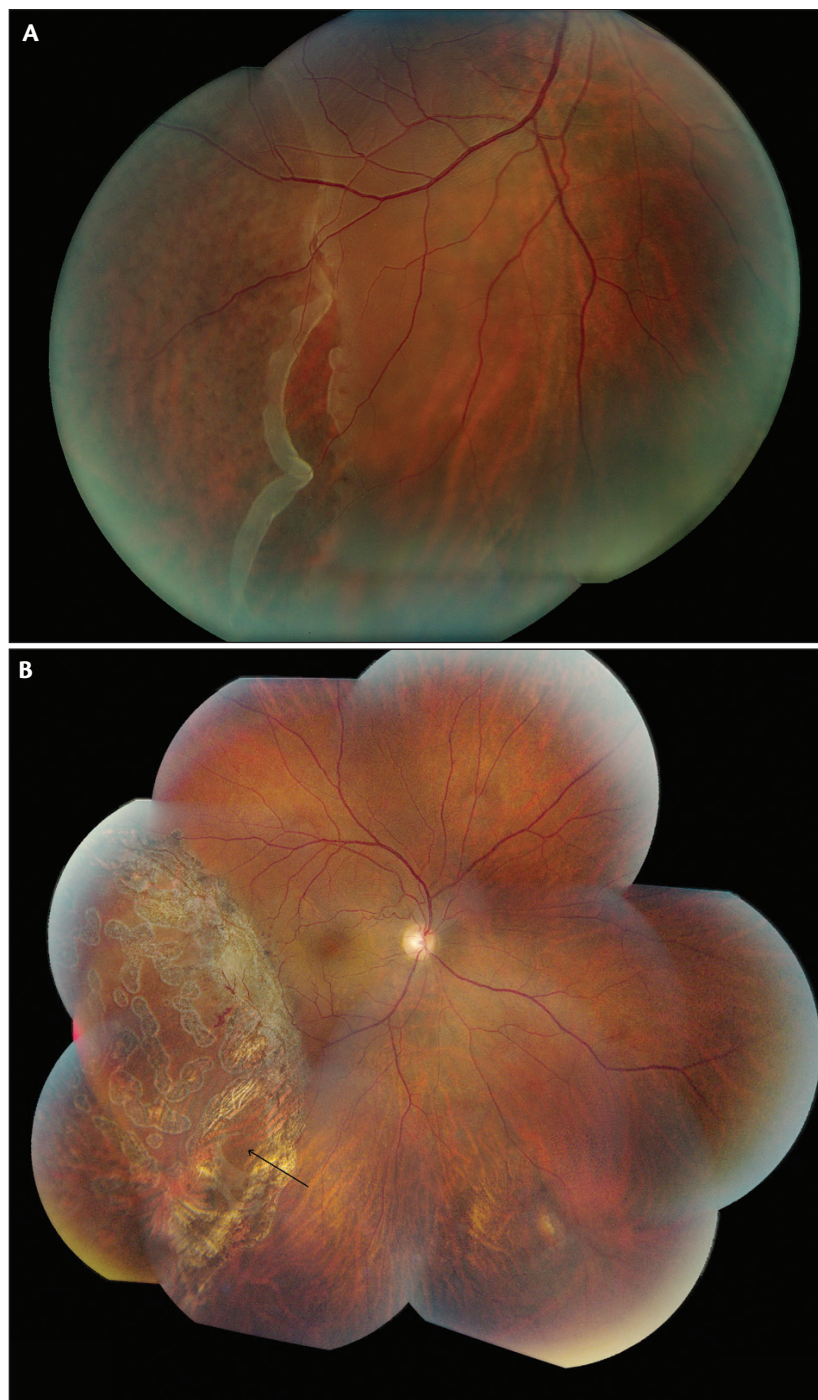


Figure 5. Progressive RRD associated with retinoschisis. A large posterior outer wall break appears to be nearly linear in this oblique view. The schisis cavity overlying subretinal fluid is seen temporal to the outer wall break. Fluid beneath full-thickness retina is seen nasal and posterior to the outer wall break (A). Postoperatively, in the same eye, the inner wall of the schisis cavity has been resected, demonstrated by the lack of retinal vessels in this area (B). The entire outer wall of the (former) schisis cavity has been treated with laser retinopexy. The inferotemporal outer wall break has been barricaded (arrow).

20/50 over 4 years of follow-up without treatment. Because cases of posterior extension of schisis detachment are rare, surgeons must use their best judgment on a case-by-case basis. Laser barricade can be attempted in asymptomatic patients, but vitreoretinal surgery may be required. The principles of vitreoretinal surgery in the setting of retinoschisis are discussed below.

PROGRESSIVE RRD

Progressive RRD associated with retinoschisis occurs when breaks in both the inner and outer layers permit vitreous fluid into the subretinal space (Figure 5A). Outer wall breaks will be evident, although inner wall breaks may be difficult to find.¹⁴ In contrast to the smooth, immobile surface of retinoschisis, progressive RRD associated with retinoschisis will feature opacified and often corrugated retina. Also, the patient will be symptomatic due to the progressive scotoma. Fortunately, this complication is rare, occurring in only 0.05% of patients with retinoschisis.¹

Treatment of progressive RRD associated with retinoschisis follows the same principles as treatment of any RRD, but with a few special considerations. First, the primary surgical goal is to close the outer wall breaks; treating inner wall breaks and collapsing the schisis cavity is optional.¹⁴ If the schisis cavity is collapsed intraoperatively, it will likely recur postoperatively. Even if the schisis cavity remains collapsed, the scotoma from this area is not reversed.^{15,16}

Pars plana vitrectomy and/or scleral buckle may be used to fix the RRD. Factors favoring pars plana vitrectomy include posterior location of outer wall breaks and presence of PVD. Anterior location of outer wall breaks and absence of PVD favor scleral buckle.¹⁷ When performing pars plana vitrectomy, a drainage retinotomy in the inner wall overlying the outer wall

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break may be created to drain sub-retinal fluid (Video).¹⁴ Alternatively, the inner wall of the schisis cavity may be resected entirely (Figure 5B).



CONCLUSIONS

Apart from posterior extension of schisis detachment and progressive RRD, in the majority of cases the complications of degenerative retinoschisis require observation only. Surgical treatment of posterior schisis detachment and progressive RRD requires closing the outer wall breaks using vitreoretinal surgical techniques. ■

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