Monitoring of non-progressive retinoschisis detachment with posterior outer leaf break

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DESCRIPTION

A 30-year-old emmetropic man presented with a 1-day history of left eye nasal shadow, without flashes, floaters or decreased vision. He had no relevant medical, ophthalmic or family history. Visual acuity was 6/6 bilaterally, with normal pupillary reactions and intraocular pressures. There was left inferotemporal retinoschisis (RS) detachment (RSD) extending to the inferior macular arcade with a posterior outer leaf break without posterior vitreous detachment (PVD) (figure 1). There was a small contralateral inferotemporal RS. Optical coherence tomography (OCT) of the RSD revealed a large outer leaf break with subretinal fluid (SRF) extension (figure 2A). Without treatment, RSD was non-progressive during the 6-month follow-up (figure 2B).

RS is a splitting of the neurosensory retina (NSR) into two layers with interface fluid. Juvenile RS is X-linked, affecting young males (incidence 1:15 000–30 000) due to mutated intercellular adhesion proteins (retinoschisin).^{1 2} Senile RS is degenerative, more common (3.9%–38%), and typically inferotemporal in older adults of either gender, though younger exceptions apply.^{3 4} Both forms may be bilateral and typically associated with hyperopia; however, senile RS can be seen with low myopia. Anatomically, RS has two subtypes: (1) flat RS (57%), being anterior to the equator without retinal breaks and (2) bullous-RS (43%), commonly posterior to the equator, associated with outer leaf breaks.⁵ OCT facilitates diagnosis, determines extent and demarcates inner leaf/outer leaf breaks in RS.⁴ RS rarely threatens vision, and must be

Figure 2 (A) Optical coherence tomography (OCT; Topcon Corporation, Tokyo, Japan) of the retinoschisis detachment area showing break in the outer leaf of retinoschisis resulting in some extension of fluid into the subretinal space resulting in retinoschisis detachment. (B) Follow-up OCT 6 months after presentation showing no posterior progression in the extent of retinoschisis detachment. The circular inset images show the plane of the OCT line scan. NB: Neither inner leaf breaks nor posterior vitreous detachment were seen clinically or on OCT.

distinguished from rhegmatogenous retinal detachment (RRD) due to breaks in both leaves of a RS.^{5–7} If fluid from an RS cavity enters the subretinal space via an outer leaf break, this is known as a RSD, which rarely extends beyond the RS margins due to the viscous nature of mucopolysaccharide-rich RS cavity fluid.^{4 8 9} RSD is easily overlooked; however, lack of uniformity and apparent heterogeneity of retinal density/transparency due to compartmentalised SRF may aid detection.⁶

Though subjective visual field may help differentiate RS from RRD, OCT is an objective tool for accurate assessment of NSR position relative to the retinal pigment epithelium.⁴ RS is not typically associated with flashes and floaters, which may be due to unrelated PVD with coincidental diagnosis of a pre-existing RS,⁶ though subsequent PVD may destabilise RS/RSD to RRD. Outer leaf breaks (24% of RS) are more important than inner leaf breaks as they may lead to RSD (56%–60% of RS) or RRD (0.05% of RS).^{6 8} RSD rarely extends into the macula, at which stage, surgical intervention is warranted.^{7 8} A long-term study of untreated asymptomatic RSD eyes (6.3-year follow-up, n=13) showed stability in 85% with regression in the remainder, suggesting observation as safe first-line management.^{6 8} Treatment options include cryotherapy, photocoagulation, scleral buckling±SRF drainage, and vitrectomy±gas/oil tamponade±schisis deroofing or retinotomy; however, these interventions may be complicated by RRD, maculopathy, vitreous haemorrhage, new retinal breaks and proliferative vitreoretinopathy.^{6 10} Considering the typically non-progressive behaviour of RSD and the visually significant risks

Figure 1 Colour fundus photograph (Optos, Scotland) of the left eye showing inferotemporal retinoschisis detachment. Of note is the dome-shaped contour with clear demarcation at the posterior extent.

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of treatment, observation is first-line management with intervention reserved for cases in which RSD progression endangers the macula.

Learning points

- Acquired retinoschisis (RS) prevalence is 3.9%–38% in adults, typically in hyperopes, and may be complicated by localised RS detachment (RSD) if outer leaf breaks are present (56%–60%) or progressive rhegmatogenous retinal detachment (RRD) (0.05%) if inner leaf and outer leaf breaks are present.
- RS is translucent, inflexible (ie, does not flatten with scleral depression), smooth, well-demarcated and causes an absolute scotoma while RRD is irregular, opaque, flattens with scleral depression (with the exception of bullous RRD)^{4 6} and gives rise to a relative scotoma.
- RSD is a rare complication of RS and is very slowly progressive or non-progressive due to viscous schisis fluid modified by the presence or absence of posterior vitreous detachment and both inner and outer leaf breaks. First-line management is close observation with invasive treatment (eg, vitrectomy, scleral buckle, laser or cryotherapy) reserved for progressive cases, which endanger the macula.

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