Temporary resolution of foveal schisis following vitrectomy with silicon oil tamponade in X-linked retinoschisis with retinal detachment

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X-linked retinoschisis (XLR) is an uncommon bilateral vitreoretinal dystrophy characterized by typical foveoschisis in all patients that may be associated with peripheral retinoschisis. A young male with XLR with retinal detachment in his right eye underwent 23 gauge pars plana vitrectomy with silicone oil tamponade. Postoperatively, best-corrected visual acuity (BCVA) improved to 20/120 with an attached retina. Spectral-domain optical coherence tomography showed macular thinning with the collapse of the schitic cavities with silicone oil in situ. Following silicone oil removal at 6 months follow-up, the retina remained attached with a BCVA of 20/80 however the foveal schitic cavities reappeared. This unusual course has not been described previously.

Key words: Foveoschisis, retinoschisis, silicone oil, vitreectomy

X-linked retinoschisis (XLR) is a rare, bilateral, hereditary disorder affecting males caused by mutation of the RS1 gene on chromosome Xp22. It is characterized by bilateral foveal schisis that may be associated with peripheral retinoschisis in about half of the patients. Roughly, 10–20% patients may progress to retinal detachment (RD).

Case Report

A 22-year-old male presented with decreased vision in his right eye since 1 month. He reported subnormal vision bilaterally since childhood. Family history for similar complaints was negative. Best-corrected visual acuity (BCVA) was hand motions close to face with accurate projection of rays in his right eye, and 20/60 in his left eye. Both anterior segments were unremarkable. On fundus examination, he was found to have XLR with typical foveal schisis bilaterally [Figs. 1a, b and 2a]. The right eye had subtotal RD with inferotemporal retinoschisis and retinal breaks and grade C-2 posterior proliferative vitreoretinopathy [Fig. 1a]. The left eye demonstrated peripheral schitic cavities located temporally and inferotemporally [Fig. 1b]. Spectral domain optical coherence tomography (SD-OCT) confirmed the presence of foveal schisis with a split in the outer plexiform layer in both eyes [Fig. 1c and d]. The right eye showed a large intraretinal cyst with a detached retina [Figs. 1c and 2b].

The patient underwent 23 gauge pars plana vitrectomy with induction of posterior vitreous detachment, subretinal fluid drainage, endolaser, and silicone oil tamponade in the right eye. Postoperatively, BCVA improved to 20/120 with an attached retina [Fig. 2c]. SD-OCT showed macular thinning with the collapse of the schitic cavities in the presence of silicone oil [Fig. 2d]. At 6 months follow-up, silicone oil was removed. While the retina remained attached postoperatively with a BCVA of 20/80, the foveal schitic cavities reappeared with a split in the outer plexiform layer at 3 weeks following silicone oil removal [Fig. 2e and f].

Discussion

In XLR, accumulation of defective retinoschisin protein within and around Muller cells results in the formation of cystic spaces in multiple retinal layers, which coalesce to form larger schisis cavities. SD-OCT has demonstrated that the foveal schisis cavities are commonly located at the inner nuclear layer, outer plexiform layer or the outer nuclear layer. This is contrary to previous histopathologic reports that showed the involvement

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of the superficial retina (retinal nerve fiber layer and ganglion cell layer). Over time, with the structural loss of Muller cells, defective retinoschisin might no longer be released but would likely be degraded and as a consequence cystic spaces would no longer be sustained. It has been observed that in adulthood, some patients may have a collapse of the retinoschisis and development of macular atrophy.

Surgery is indicated for XLR complicated by RD or vitreous hemorrhage. The ideal treatment for schisis involving or threatening the macula remains unclear. The removal of vitreous and the internal limiting membrane (ILM) could be helpful for resolution of the foveal schisis and restoration of the retinal structure. It has been shown that following the complete removal of the vitreous cortex, retinoschisis resolves, suggesting that vitreous traction may play a role in the development of the foveal schisis.

The RS1 gene encodes a secretory protein, retinoschisin that is secreted from the bipolar cells and rod and cone photoreceptors and functions as a cellular adhesion protein. The absence of retinoschisin as in XLR may cause intraretinal cell-to-cell dehiscence. Thus, elimination of vitreous traction and gas tamponade may promote structural restoration of the retina. Temporary tamponade with silicone oil has also been shown to stabilize the retinoschisis, persisting even after oil removal. Our case had recurrent foveal schisis following removal of silicone oil. This has not been reported previously. It has been noted that ILM peeling may be a risk factor for a macular hole or retinal breaks in XLR due to the weakened retinal structure in the schitic area. Hence, we did not perform ILM peeling in our case.

This case adds to the limited literature elaborating on the course and outcomes of foveoschisis in XLR following vitrectomy. The unusual sequence of events that occurred in the above case have not been described earlier. This may further shed light on the pathophysiology of this rare inherited disorder.