Schwartz-Matsuo syndrome describes a combination of rhegmatogenous retinal detachment (RRD) with oral dialyses or tears of the nonpigmented ciliary epithelium, the presence of photoreceptor outer segments in the aqueous humor, and raised intraocular pressure (IOP), which normalizes after surgery.1 We report 2 cases of longstanding RRD with midperipheral retinal tears and degenerating aggregates of photoreceptor outer segments in the aqueous humor.

Report of Cases. Case 1. A 36-year-old white woman with no significant medical history had a 3-week history of a painful left eye and visual acuity reduced to the ability to view only basic hand movements. The eye was aphakic with an IOP of 64 mm Hg with 3 or more cells in the anterior chamber without other signs of ocular inflammation. Fundal examination revealed a temporal horseshoe tear in the midperiphery with the presence of a longstanding total RRD with moderate proliferative vitreoretinopathy. Significant ocular history included a penetrating corneal injury 30 years previously for which she underwent a primary repair. She subsequently developed a mature cataract and had undergone a planned paracentesis through the peripheral cornea before the procedure. The obtained aqueous sample was processed for transmission electron microscopy according to a standard technique.2 On her latest visit 6 months postoperatively, no anterior chamber activity was found, and the IOP was 16 mm Hg without treatment.

Case 2. A white, 51-year-old man with pseudophakia but otherwise no significant medical history had a 5-week history of floaters in the left eye. When he arrived at our clinic, his visual acuity was 6/60 in the affected eye with 2 or more cells in the anterior chamber and an IOP of 40 mm Hg. Fundal examination revealed a superior bullous RRD with macular involvement, a subretinal demarcation line, and 2 superotemporal horseshoe tears. As in case 1, the IOP and anterior chamber activity were refractory to treatment, and the patient underwent a non-drainage scleral buckle procedure. An anterior chamber paracentesis was performed for aqueous humor analysis. Nine months after surgery, no anterior chamber activity was detected, and the IOP was normal at 14 mm Hg.

Results of Electron Microscopy. In both cases, examination of the aqueous humor revealed photoreceptor outer segments. In case 1 the segments were relatively intact, whereas in case 2, degenerating photoreceptor segments predominated. No inflammatory cells were found. The intact segments consisted of 2 membranes that comprised regularly arranged lamellae of approximately 20-nm thickness, which were not continuous with the plasma membrane. This finding suggests that these segments might be from rod photoreceptors (Figure 1). Aggregation of degenerating photoreceptor outer segments was also evident in the anterior chamber (Figure 2).

Comment. Schwartz-Matsuo syndrome was described by Matsuo1 as a clinical entity that consists of 3 major signs: RRD with tears around the ora serrata, secondary occurrence of aqueous cells, and elevated IOP. In the review by Matsuo, the author...
states that tears posterior to the ora serrata rarely cause the syndrome.

Our atypical patients had posterior horseshoe retinal tears, exhibited cells in the anterior chamber without other signs of ocular inflammation, and had elevated IOP, which normalized after retinal detachment repair. Although no macrophages were identified, the cells observed in the anterior chamber of the affected eyes may represent an anterior migration of photoreceptors and other cell types. The pathway into the anterior chamber has been well described previously. Numerous mechanisms have been proposed for the elevated IOP in this syndrome, but mechanical obstruction owing to photoreceptor outer segment deposition in the trabecular meshwork remains the most widely accepted mechanism, although it is unproven by histopathologic studies.3,4

Electron microscopic analysis of the aqueous humor in our cases revealed photoreceptor outer segments at varying stages of degeneration and sizable lipid-containing aggregates (Figure 2). Although the origin of these structures is uncertain, photoreceptor segment membranes contain a high content of cholesterol and lipid.3 The presence of these membrane-bound structures may represent an aggregation of photoreceptor outer segments, which in their bulky nature contribute to a mechanical obstruction in aqueous outflow through the trabecular meshwork.

These cases comprise an atypical form of Schwartz-Matsuo syndrome with posterior retinal tears and electron microscopic evidence of degenerating photoreceptor outer segments, which appear to aggregate in the anterior chamber of affected eyes. Our findings add to the case documentation of Schwartz-Matsuo syndrome and provide more insight into the origin and mechanism of glaucoma associated with RD.

Danny Mitry, MD
Ian Constable, FRANZCO
Jaswinder Singh, FRCSEd

Correspondence: Dr Mitry, Princess Alexandra Eye Pavilion, Ophthalmology, Chalmers Street, Edin-

burgh EH3 9HA, Scotland (mitryd@gmail.com).

Financial Disclosure: None reported.