Photoreceptor Outer Segment Glaucoma in Rhegmatogenous Retinal Detachment

Schwartz-Matsuo syndrome describes a combination of rhegmatogenous retinal detachment (RRD) with oral dialyses or tears of the non-pigmented 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 and uneventful intracapsular cataract extraction without an intraocular lens implant 3 years before her arrival at our clinic. Anterior chamber activity persisted, and preoperative IOP remained elevated at 36 mm Hg despite treatment with topical steroids and standard oral and topical IOP-reducing agents. She had undergone a vitrectomy procedure with 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...

Figure 1. Relatively intact lamellar outer segments in case 2 (original magnification ×15,000). Bar indicates 1 µm.

Figure 2. Membrane-bound aggregate (A) and partially destroyed outer segments (original magnification ×15,000) in case 1. Bar indicates 1 µm.
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. \(^3,4\) 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 reduction 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.

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Unusual Carcinomas of the Lacrimal Gland: Epithelial-Myoepithelial Carcinoma and Myoepithelial Carcinoma

Of all intrinsic lacrimal gland masses, 28% are epithelial neoplasms. \(^1\) Epithelial, myoepithelial carcinoma (EMC) and myoepithelial carcinoma (MC) are uncommon epithelial malignancies of the salivary gland that have been rarely reported in the lacrimal gland. \(^2\) Herein, we report 2 patients with each of these tumors in the lacrimal gland and compare and contrast these unusual neoplasms.

Report of Cases. Case 1. An 86-year-old man presented with painless double vision for 6 months. His visual acuity was 20/30 OD and 20/50 OS. He had a palpable mass under the left superolateral orbital rim and bilateral symmetrically decreased tear function. His palpebral fissure was 9 mm OD and margin reflex distance, 5 mm OD, compared with 6 mm and 3 mm, respectively, OS. Exophthalmometry was 13 mm OD and 16 mm OS. He had limited elevation, abduction, and adduction of the left eye with an 8-diopter left hypotropia. Computed tomographic scan of the orbits showed a 2.5 × 1.8-cm, moderately homogenous, hyperattenuating structure in the region of the left lacrimal gland with an area of scalloping in the lacrimal fossa (Figure 1A). The appearance was suspicious for transformation of a pleomorphic adenoma to malignancy. The patient underwent a lateral orbitotomy for resection of the lacrimal mass that was found to be adherent to the periorbital fat, with irregularity of the adjacent bone. Because of this, the underlying bone of the lacrimal fossa was sent for histopathologic examination. The gland revealed a biphasic tumor containing the typical bilayered pattern of EMC (Figure 1B and Figure 2A, C, and E), with areas of myoepithelial anaplasia (Figure 1C). The latter occupied approximately half of the tumor volume and was composed of a solid component of cells with myoepithelial features and prominent atypia, including nuclear pleomorphism, enlarged nuclei, mitotic figures, and focal necrosis (Figure 1D). Extension to the anterior, superior, and inferior resection margins was present. The bone specimen was negative for invasion; therefore, the tumor was classified as T2N0M0. Surgical treatment was followed by adjuvant radiotherapy of 6000 cGy because of positive margins. Repeat computed tomographic scan at 8 months postoperatively showed no evidence of recurrence. The patient died 15 months after excision, with no clinical evidence of recurrent disease.

Case 2. An 84-year-old man presented with severe left ocular pain and visual loss for 2 weeks. His visual acuity was 20/30 OD and hand motions OS. A full-diameter corneal ulcer with impending perforation was present in the left eye, which was proptotic by 10 mm and displaced inferonasally. Computed tomographic scan demonstrated a 3.2 × 2.6 × 2.2-cm, well-circumscribed, calcified lacrimal gland mass extending to the apex, displacing the globe inferiorly and medially with irregularity in the adjacent bony orbital wall (Figure 3A). Incisional biopsy of the lacrimal gland revealed MC. Metastatic workup findings were negative so left eyelid-sparing exenteration was performed along with excision of the adjacent bone. Histopathologic examination revealed MC arising in a pleomorphic adenoma (Figure 2B, D, and F and Figure 3B), with a predominant epi-