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.

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. 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.

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

Correspondence: Dr Mitry, Princess Alexandra Eye Pavilion, Ophthalmology, Chalmers Street, Edinburgh EH3 9HA, Scotland (mitryd@gmail.com).

Financial Disclosure: None reported.


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

Of all intrinsic lacrimal gland masses, 28% are epithelial neoplasms. Epithelial-myoelepithelial carcinoma (EMC) and myoepithelial carcinoma (MC) are uncommon epithelial malignancies of the salivary gland that have been rarely reported in the lacrimal gland. 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 hand motions OS. A full-diameter corneal ulcer with impending perforation was present in the left eye, which was protopathic 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. Histopathological examination revealed MC arising in a pleomorphic adenoma (Figure 2B, D, and F), with a predominant epi-

©2009 American Medical Association. All rights reserved.
thelioid pattern (Figure 3C) and focal clear cell areas (Figure 3D). There was a moderate degree of cytologic pleomorphism and atypia with vesicular nuclei, prominent nucleoli, and necrosis in a comedolike pattern (Figure 2F). The mitotic rate was 3 per 10 high-power fields. The bone was invaded by carcinoma, leading to a classification of T4bN0M0. The patient refused follow-up and further treatment.

Comment. Epithelial-myoepithelial carcinoma is a rare neoplasm accounting for approximately 1% of all salivary gland neoplasms, with most cases arising in the parotid gland. Only 2 cases of primary EMC of the lacrimal gland have been reported. Classically, this neoplasm shows a biphasic pattern of a central inner layer of cuboidal ductal epithelial cells surrounded by a peripheral outer layer of myoepithelial cells, often with clear cytoplasm. In the largest series, reported by Seethala et al, there was a biphasic pattern in some areas in all tumors. The mean epithelial to myoepithelial ratio was 0.56 but ranged from 0.05 to 2.33. Epithelial-myoepithelial carcinoma of the salivary gland is considered to be a low-grade malignancy, with a recurrence rate of 36.3% and a survival rate of 93.5% and 81.8% at 5 and 10 years in the series of Seethala et al. Factors significantly affecting disease-free survival were positive margins, lymphovascular invasion, necrosis, and myoepithelial anaplasia. Three of

Figure 1. Epithelial-myoepithelial carcinoma with myoepithelial anaplasia. A, Coronal computed tomographic scan showing a hyperattenuating mass in the region of the left lacrimal gland. B, Area of epithelial-myoepithelial carcinoma showing classic bilayered pattern of inner cuboidal ductal cells and outer clear myoepithelial cells (hematoxylin-eosin, original magnification ×400). C, Juxtaposition of predominant pattern of inner clear cells with outer cuboidal cells (left) next to an area of solid myoepithelial overgrowth (right) (hematoxylin-eosin, original magnification ×100). D, Area of myoepithelial anaplasia with numerous plasmacytoid cells and necrosis (top left) (hematoxylin-eosin, original magnification ×400).

Figure 2. Immunohistochemical staining patterns: epithelial-myoepithelial carcinoma (A, C, and E) and myoepithelial carcinoma (B, D, and F). (All photomicrographs are avidin-biotin immunoperoxidase with hematoxylin counterstain, original magnification ×100.) A, Pankeratin staining showing strong staining of the classic pattern of epithelial-myoepithelial carcinoma (EMC) (lower right) and lesser staining of an area of solid myoepithelial overgrowth (upper left). B, More diffuse and moderate staining of myoepithelial carcinoma (MC) for low-molecular-weight keratin with Cam 5.2. C, Outer myoepithelial cells in EMC are stained by p63. D, Variable staining of MC for S100. E, Staining for Ki-67 in EMC showing a low proliferative rate in the classic areas and a higher rate in the solid areas. F, Staining for Ki-67 in MC showing a moderate proliferative rate that was uniform throughout the tumor. A focus of necrosis is present centrally.
45 patients died of disease, 2 with local recurrences and 1 of distant metastasis. Our patient had 3 of 4 poor prognostic factors, including positive margins, necrosis, and myoepithelial anaplasia.

Myoepithelial carcinoma has also been reported rarely in the lacrimal gland in large series, but not illustrated. It also occurs most commonly in the parotid gland and is defined as a malignant neoplasm with histologic evidence exclusively of myoepithelial differentiation. It does not show tubule formation or the biphasic pattern characteristic of EMC. The largest series of 25 patients with salivary gland MC, reported by Savera et al, had 10 high-grade and 15 low-grade lesions. Fifteen tumors arose in the background of a preexistent benign lesion, 2 benign myoepitheliomas and the remainder pleomorphic adenomas. This neoplasm is characterized by all of the different histologic patterns that myoepithelial cells assume: epithelioid, clear, hyaline (plasmacytoid), spindle, and mixed, with most neoplasms having 2 or more patterns. Ten of 17 had recurrences and 8 had metastases. Five patients (29%) died of disease after a mean of 32 months, 2 were alive with metastases, and 10 were alive or died without disease after a mean of 42 months. No histologic factors correlated with outcome statistically. Thus, the prognosis for MC is generally worse than that for EMC.

Most rare epithelial neoplasms of the salivary glands have now been reported in the lacrimal gland. Consequently, it is important for the ophthalmic pathologist to be cognizant of this and familiar with the most recent World Health Organization classification of salivary gland neoplasms when diagnosing an unusual lacrimal tumor. It is important to study the entire tumor pathologically. Further investigation will be required to determine if recognition of the same entities is as important for prognostication in the lacrimal gland as in the salivary gland. It is also imperative that any bone removed be examined pathologically for proper staging, as demonstrated by our 2 cases.

Correspondence: Dr White, Department of Pathology and Laboratory Medicine, Vancouver General Hospital, 910 W 10th Ave, Vancouver, BC V5Z 4E3, Canada (val.white@vch.ca).

Financial Disclosure: None reported.