roblastoma, the term ganglioneuroblastoma is appropriate. In the present case, clinical uncertainty regarding cellular composition coupled with progressive proptosis and its associated corneal complication prompted debulking of the lesion. The history of metastatic neuroblastoma and the presence of multiple ganglioneuromas lend credence to the theory purporting the presence of rests of metastatic neuroblastoma that subsequently undergo maturation. Ganglioneuromas, as fully differentiated neoplasms, do not have the capability to metastasize, so extensive surgical resections or chemotherapy is not normally necessary. Provided surgical sampling is sufficient to allow adequate histologic analysis and to assure no neuroblastic cellular elements are present. Excision may be considered when the pathologic diagnosis is uncertain or visual function is compromised by the neoplasm.

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Primary Epithelial-Myoepithelial Carcinoma of the Lacrimal Gland

Most lacrimal gland lesions are inflammatory or lymphoid neoplasms. Nonlymphoid neoplasms are less common, and most are primarily epithelial in origin. Among them, pleomorphic adenoma and adenoid cystic carcinoma are the most common benign and primary malignant tumors, respectively, accounting for 12% and 4% of all lacrimal gland lesions. Epithelial-myoepithelial carcinoma is an exceptional malignant epithelial tumor in view of its rarity and the relative lack of understanding of its clinical behavior. These rare tumors usually occur in the salivary gland, and, to our knowledge, only 2 cases in the lacrimal gland have been reported. One of these was a hybrid carcinoma and the other was an epithelial-myoepithelial carcinoma with pleomorphic adenoma background. We herein report a case of de novo epithelial-myoepithelial carcinoma of the lacrimal gland.

Report of a Case. An 80-year-old Chinese man had a painless, palpable subcutaneous mass in his left upper outer eyelid for 9 months. On examination, a contrast-enhancing mass of 1 cm in diameter was confirmed to arise from the left lacrimal gland, as demonstrated on the computed tomographic scan. The left eye was the patient's only functioning eye, with a visual acuity of 20/50; his right eye had been lost to trauma 15 years earlier. He was treated conservatively, as he refused any intervention for diagnosis. The mass gradually enlarged and displaced the left eye nasally and inferiorly. Adduction was reduced. There was also significant chemosis involving the upper bulbar conjunctiva. No lymph nodes could be palpated over the cervical and supraclavicular regions. Systemic reviews, including abdominal, respiratory, cardiovascular, and neurologic examinations, were unremarkable. The complete blood cell

Figure 1. Computed tomographic scan of the orbit showing a mildly enhanced lacrimal soft-tissue mass arising from the superolateral aspect of the left orbit.

Figure 2. Low-power photomicrograph showing a well-circumscribed encapsulated tumor (hematoxylin-eosin, original magnification ×2).
count, inflammatory markers (including erythrocyte sedimentation rate and C-reactive protein level), and biochemical profiles were all normal. A chest radiograph was clear, and results of an ultrasonographic examination of the abdomen were normal. Repeated computed tomographic scan (Figure 1) of the orbit 10 months after the first examination showed a mildly enhanced soft-tissue mass arising from the left lacrimal gland and pushing the globe posteriorly with bulking of the optic nerve. It measured $24 \times 15 \times 18$ mm. No intraocular invasion or bony erosion could be seen.

The patient eventually agreed to an excisional biopsy, and it was performed via a standard translid lateral orbitotomy. The tumor was a well-encapsulated mass and was removed en bloc uneventfully. There was no postoperative complication, and visual acuity returned to 20/50. Histopathological examination disclosed epithelial-myoeipithelial carcinoma of the lacrimal gland. Foci of capsular invasion and tumor nests at the surgical margin were present. Exenteration or external beam radiotherapy was offered to the patient but was declined, as this was his only seeing eye. He was kept under close observation. Two years after surgery, there was no evidence of local recurrence or metastasis.

Gross examination showed a firm, well-circumscribed mass measuring $2.3 \times 1.8 \times 1.8$ cm composed of homogeneously tan fleshy tissue without hemorrhage, necrosis, or calcification. Microscopic sections showed a fibrous encapsulated cellular tumor (Figure 2) with multiple foci of capsular invasion (Figure 3). The tumor was heterogeneous. In some areas, the tumor was in the form of glands that were lined by 2 cell types, the inner epithelial and the outer myoepithelial cells. Similar to a normal functional lacrimal acini unit, with an epithelial lining surrounded by myoepithelial cells, the immunohistochemical staining pattern of this tumor was reminiscent of such a pattern. The inner cuboidal cells did not show immunoreactivity for myoepithelial markers such as $S_{100}$ protein and smooth-muscle antigen (Figure 4 and Figure 5) but exhibited positivity with cytokera-

![Figure 3. Area of capsular invasion (hematoxylin-eosin, original magnification ×10).](image)

Figure 3. Area of capsular invasion (hematoxylin-eosin, original magnification ×10).

![Figure 4. Staining of inner and outer layers of cells for $S_{100}$ protein. The inner-layer cells are $S_{100}$ negative, whereas the outer-layer cells are $S_{100}$ positive (original magnification ×20).](image)

Figure 4. Staining of inner and outer layers of cells for $S_{100}$ protein. The inner-layer cells are $S_{100}$ negative, whereas the outer-layer cells are $S_{100}$ positive (original magnification ×20).

![Figure 5. Staining of inner and outer layers of cells for smooth-muscle antigen. The inner-layer cells are negative for smooth-muscle antigen, whereas the outer-layer cells are smooth-muscle antigen positive (original magnification ×20).](image)

Figure 5. Staining of inner and outer layers of cells for smooth-muscle antigen. The inner-layer cells are negative for smooth-muscle antigen, whereas the outer-layer cells are smooth-muscle antigen positive (original magnification ×20).
tin (clone AE1/AE3) and epithelial membrane antigen. In contrast, the outer layer of cuboidal or flattened cells was positive for S100 protein and SMA immunostain (Figures 4 and 5), confirming that they were myoepithelial. In other areas, the tumor was in the form of oval and spindled cells forming nests and sheets. We noted moderate cellular pleomorphism and 5 mitoses in 40 high-power fields (Figure 6). The nuclear pleomorphism, frequent mitoses, and capsular invasion were features of a malignant tumor. Extracellular globules of hyaline material were focally present. Focal clear cell change was seen. These oval and spindled cells were also positive for the myoepithelial immunomarkers, indicating that they were derived from overgrowth of the myoepithelial component (Figure 7 and Figure 8). Above all, these immunohistopathological features were diagnostic of epithelial-myoepithelial carcinoma. No perineural or perivascular invasion could be seen. There was no other preexisting abnormality associated with this tumor, suggesting it was a de novo, isolated tumor. The characteristic biphasic cell arrangement and immunostaining features helped to distinguish this tumor from common differential diagnoses such as adenoid cystic carcinoma with an infiltrative cribriform growth pattern and pleomorphic adenoma with melting of the epithelial cells in myxoid or chondroid stroma.

Comment. Epithelial-myoepithelial carcinoma is a rare malignant tumor that is more commonly encountered in salivary glands and comprises 1% of all salivary gland tumors.8-10 Its occurrence in the orbit is extremely rare.2 Myoepithelioma can be subclassified histologically into different morphologic types such as chondromyxoid, spindle, hyaline, and epithelial, including the clear-cell variants.7 The epithelial type is believed to behave in a more malignant fashion than the former 2 groups. In the English literature, we found only 1 case of hybrid carcinoma occurring as a mixed tumor, and 1 associated with a preexisting pleomorphic adenoma in a 63-year-old man with an 8-year history of painless proptosis.2 To our knowl-
epithelial lesion is known to be indistinguishable from a benign one. In cases of malignant from a benign epithelial tumor, it is important to consider the possibility of malignancy, especially in cases of rapidly growing masses.

However, the reliability of radiologic features alone may not be sufficient to differentiate between benign and malignant conditions, as some benign tumors may mimic malignancy, and vice versa. In such cases, a biopsy may be necessary.2,11

In our case, sudden rapid growth of a painless and slowly growing mass suggested a malignant lesion. The patient had undergone a limited procedure of tumor excision only. During 24 months of follow-up, there was no recurrence or metastasis, and useful vision was maintained.

Recurrence and metastasis rates of epithelial-myoepithelial carcinoma from salivary gland have been reported to be from 35% to 50% and from 8.1% to 25%, respectively.3 In this case, the presence of tumor at the surgical margin suggests that radiotherapy or orbital exenteration may be necessary. However, the ophthalmic complications or morbidity of the treatment should be discussed with the patient, especially in considering the patient’s expectations, age, and overall health.

The patient was treated with large spot–size laser ablation by indirect ophthalmoscopy to the anterior iris plane. Despite this treatment, the patient showed symmetric anterior ischemic retinopathy and secondary ridge neovascularization consistent with FEVR. Focal traction in this region was noted. The patient did not develop progression of familial exudative vitreoretinopathy in the opposite eye. Four months later, there was minimal progression of traction in the left eye with a localized area of extramacular schisis formation. The right eye showed progressive tractional alterations through the macula with development of a falciform retinal fold.

**Progression of Familial Exudative Vitreoretinopathy After Laser Treatment**

Familial exudative vitreoretinopathy (FEVR) is an inherited vitreo-retinal dystrophy with a variable clinical course. Early disease with no retinal detachment has been shown to respond well to primary laser treatment.2 The following case manifested symmetrical but responded asymmetrically to appropriate and aggressive laser treatment.

**Report of a Case.** A 17-month-old girl was referred to the Vitreoretinal Surgical Service of the Bascom Palmer Eye Institute, Miami, Fla, for a strong family history of FEVR. The patient had diabetes at delivery, her medical history was unremarkable, and her developmental milestones were intact. She had no previous ocular history, but her mother noticed that the patient was bringing objects close to her face, squinting, and bumping into things. External examination revealed primary alternating esotropia. Dilated retinal examination of both eyes showed symmetric anterior ischemic retinopathy and secondary ridge neovascularization consistent with FEVR. Focal traction in this region was noted. The patient was treated with large spot–size laser ablation by indirect ophthalmoscopy to the anterior iris plane. Despite this treatment, the patient showed minimal progression of traction in the left eye with a localized area of extramacular schisis formation. The right eye showed progressive tractional alterations through the macula with development of a focal retinal fold.

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