Electron microscopy of the anterior and posterior capsule of the lens showed aberrant epithelial cells with dilated endoplasmic reticulum, formation of collagenous fibrous tissue, and dystrophic calcification. These findings are reminiscent of the changes seen in cartilage. The histopathologic changes in the lens indicate that it is involved in Kniest dysplasia either primarily or secondarily. Formation of abnormal subcapsular collagen plaques beneath the capsule together with dystrophic calcification may be responsible for the abnormally hard cataracts. The cataract could also develop as a posterior capsular opacity and progress to a total cataract. Another possible mechanism for cataract development could be abnormal lens fiber formation due to the metaplastic lens epithelial cells. Dilated endoplasmic reticulum in cartilage chondrocytes and lens epithelial cells may be indicative of synthesis of abnormal protein and extracellular matrix material. Although changes in Kniest dysplasia appear to be developmental, unilateral cases of cataract and retinal detachment have been reported.

Our observation supports the theory that the underlying defect causing cataracts is primarily a collagen synthesis defect. Production of abnormal glycosaminoglycans probably has a minimal role in the pathogenesis of the ocular defects. Our findings bear great similarity to the histopathologic features described in Lowe syndrome. The abnormalities noted in Kniest dysplasia could be developmental, and a periodic comprehensive ophthalmic evaluation of the patient is warranted.

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**Polymorphous Low-Grade Adenocarcinoma of the Lacrimal Gland**

Malignancies constitute half of all epithelial tumors of the lacrimal gland, with the majority being adenoid cystic carcinoma. Adenocarcinomas are much less common and generally demonstrate a poor prognosis. However, the recent application of histological subtyping to this group is leading to a better characterization of what appears to be a heterogeneous collection. We present a rare case of polymorphous low-grade adenocarcinoma (PLA) of the lacrimal gland and discuss the clinical manifestation and prognosis in the context of similar tumors arising within the salivary glands.

**Report of a Case.** A 67-year-old man was initially seen with painless right upper eyelid swelling and conjunctival hyperemia across a 2-day period. The patient had been previously asymptomatic, and old photographs were not available for review. Examination revealed him to be afebrile with a visual acuity of 6/12 OD and 6/9 OS, 6 mm of proptosis, 3-mm inferior globe displacement, temporal conjunctival hyperemia, and a tender mass in the superotemporal orbit. Extraocular movements were limited in lateral and up gazes. The erythrocyte sedimentation rate was 61 mm/h and white blood cell count, 13 200/µL (13.2 × 10^9/L). Computed tomography findings showed an ill-defined mass in the lacrimal gland with a central radiolucency, rim enhancement, and no bony changes (Figure 1).

The mass was surgically explored via a skin-crease incision and appeared to consist of an abscess containing yellow-green viscous material. A culture yielded no growth, but biopsy findings of the cavity wall revealed adenocarcinoma thought to represent a primary lacrimal neoplasm rather than metastasis because the tumor expressed both cytokeratins (CAM 5.2) and S100 protein immunoreactivity. This immunoprofile, while not diagnostic, is typical of salivary gland tumors including PLA.

The patient underwent thorough systemic evaluation, including computed tomography of the chest, abdomen, and head. All investigation results were normal, and a lid-sparing exenteration, including the biopsy track, was performed.

**Results.** Histopathologic results revealed PLA of the lacrimal gland (Figure 2). The tumor cells were cytologically bland, showing mild nuclear pleomorphism, and no mitoses were identified. However, there was extensive infiltration of the adjacent tissue, and the tumor displayed a variety of architectural patterns, including cribriform, tubular, and fascicular areas along with solid cell nests. Some necrosis was seen centrally, and infiltration of the extraocular muscle was present. How-

**Figure 1.** Axial computed tomographic scan shows a mass in the right lacrimal gland with a central radiolucent area.
ever, there was no perineural or vascular invasion or evidence of encapsulation, and the residual gland was atrophic with evidence of chronic inflammation and fibrosis. The patient made a good recovery and at last follow-up, 6 years after initial examination, there was no recurrence.

Comment. Adenocarcinomas constitute 5% to 7% of epithelial tumors arising in the lacrimal gland. Until recently, these neoplasms were considered a single entity, classified under the rubric of “adenocarcinoma” or “adenocarcinoma not otherwise specified.” The advent of the World Health Organization Classification of Salivary Gland Adenocarcinomas proffered a framework for the categorization of these uncommon lacrimal tumors. This classification system is of significance because certain subtypes are associated with differing biologic behaviors and outcomes. Three of these subtypes, salivary duct carcinoma, epithelial-myoepithelial carcinoma, and PLA, have been reported within the lacrimal gland.

The histological subtype of PLA seen in our case occurs almost exclusively in the minor salivary glands and is distinguished by cytologically bland, uniform cells with few or no mitoses. These cells assume a variety of growth patterns including cribriform areas, solid nests, tubular regions, clusters, and “Indian file” columns. The cribriform areas are distinguished from adenoid cystic carcinoma by the relative lack of atypia of the component cells. In contrast to pleomorphic adenomas, PLAs show infiltration into adjacent tissues as well as a propensity for perineural invasion.

Within the lacrimal gland, PLA has only been reported on one previous occasion. Ni et al mentioned 3 cases in their series of 272 epithelial lacrimal tumors but did not document clinical manifestation, management, or outcome. The clinical manifestation of our patient was distinctly unusual for an epithelial malignancy of the lacrimal gland. While inflammatory manifestations have been described in a few benign mixed tumors, usually on the basis of mucinous cyst rupture, such a manifestation for malignant epithelial tumors is distinctly rare. Mucoepidermoid carcinomas in the lacrimal gland rarely cause inflammation, and with regard to adenocarcinomas, an acute inflammatory process has not previously been described. Heaps et al, in a series of 13 lacrimal adenocarcinomas, found a palpable mass, proptosis, and pain to be the most common manifestation.

In our case, the constellation of acute inflammation in conjunction with the central low-density area on computed tomographic scan was suggestive of a lacrimal abscess. However, the absence of growth in the necrotic cavity contents mitigated against an infective etiology. In addition, the central area was neither mucinous nor hemorrhagic, leading us to postulate a mechanism of central necrosis secondary to infarction to account for both the inflammatory response and the radiological appearance.

While microscopic focal necrosis has been described in PLA, the occurrence of significant central cavitation is unusual behavior in a low-grade tumor. In fact, within the orbit, central necrosis is generally a feature of aggressive, rapidly growing malignancies such as sarcomas or metastases. In comparison, PLA arising in the salivary glands has an indolent growth pattern and a relatively low incidence (6.5%) of metastasis to the regional lymph nodes. Local recurrence occurs in
22% but can usually be managed by further resection. Distant metastasis or death due to tumor is extremely rare, and as a consequence, PLA has a much better prognosis than other salivary adenocarcinomas.10

The extrapolation of the course of salivary gland PLA to those originating in the lacrimal gland was one of the factors considered in treating our patient. Although exenteration followed by radiotherapy has been advocated for lacrimal adenocarcinomas as a group,9 we elected to treat with exenteration alone on the basis of the known behavior of this tumor. This was in concordance with the management of salivary PLA, which carries an excellent long-term prognosis. This was in concordance with the known behavior of this tumor.

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