Correspondence

A Case of Lacrimal Gland Myoepithelial Carcinoma Managed with Surgical Excision and Radiation

Hoang Nhat Mai Nguyen, MD[1], Hee Sung Kim, MD, PhD[2], Jeong Kyu Lee, MD, PhD[1,3]

[1]Department of Ophthalmology, College of Medicine, Chung-Ang University, Seoul, Korea

[2]Department of Pathology, College of Medicine, Chung-Ang University Hospital, Seoul, Korea

[3]Department of Ophthalmology, College of Medicine, Chung-Ang University Hospital, Seoul, Korea

Corresponding author Jeong Kyu Lee, MD., PhD.

Department of Ophthalmology, College of Medicine, Chung-Ang University, 102 Heukseok-ro, Dongjak-gu, Seoul, 06974, Republic of Korea.

Tel: 82-2-6299-1688; Fax: 82-2-825-1666, E-mail: lk1246@gmail.com

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Dear Editor,

Myoepithelial carcinoma is characterized by the presence of neoplastic myoepithelial cells that exhibit infiltrative growth [1]. This malignancy predominantly originates in the salivary glands and is relatively uncommon in the lacrimal gland. Typically, it remains asymptomatic until it exerts a mass effect. However, some individuals may experience significant proptosis, diplopia, and displacement of the eyeball. This report presents a case of myoepithelial carcinoma diagnosed through a comprehensive approach that included ophthalmic examination, radiological imaging, histopathological evaluation, and immunohistochemical analysis.

A 58-year-old male patient presented with edema of the right upper eyelid, which had developed over the preceding two to three weeks (Fig. 1A). The patient reported no significant prior ophthalmological history. Upon ophthalmic examination, visual acuity was assessed at 20/20 in both eyes. The examination revealed bulging of the right upper eyelid, accompanied by 2 mm of exophthalmos. Extraocular movement was found to be restricted in both upward and lateral gaze of the right eye. Computed tomography imaging identified a lobulated mass measuring 3 x 2 cm with calcification in the right lacrimal gland (Fig. 1B). T2-weighted magnetic resonance imaging demonstrated a well-circumscribed mass in the right orbit, which was exerting pressure on the eyeball (Fig. 1C).

The entire mass was excised with the capsule intact (Fig. 1D). Histopathological examination identified round to polygonal epithelioid cells exhibiting nuclear pleomorphism and atypical mitotic figures (Fig. 1E). The tumor demonstrated focal necrosis and perineural invasion; however, lymphovascular invasion was not detected. Immunohistochemical analysis indicated positive expression of p63, calponin, smooth muscle actin (SMA), and S-100 protein, which are indicative of myoepithelial differentiation (Fig. 1F). Additionally, CK8/18 staining highlighted the ductal component of the tumor, while Ki-67, a proliferation marker, was significantly elevated at 30%,

suggesting a high proliferative index and invasive characteristics of the tumor (Fig. 1G). The patient was diagnosed with myoepithelial carcinoma, classified as T2N0M0 per the TNM staging system. Adjuvant radiotherapy was administered, delivering a total dose of 6000 cGy over 30 fractions. Despite months of managing dry eye and keratoconjunctivitis post-radiotherapy, a two-year follow-up revealed no recurrence (Fig. 1H).

Lacrimal gland tumors are epithelial in approximately 50% of cases, with benign mixed tumors comprising half. Unlike the indolent and asymptomatic nature of benign mixed tumors, malignant tumors tend to progress rapidly and often cause pain. In this case, the mass demonstrated rapid growth over 2-3 weeks. Imaging showed no bone destruction, but calcification and the considerable size of the mass led to the decision to proceed with surgery. Since malignancy could not be definitively established, the entire mass was excised with the capsule preserved rather than performing an en bloc resection with margin control.

The diagnosis of myoepithelial carcinoma is confirmed through histopathological and immunohistochemical analyses. Previous studies have suggested that myoepithelial tumors exhibiting significant hypercellularity or pleomorphism, as well as perineural invasion, should be classified as neoplasms with malignant potential [2, 3]. Furthermore, myoepithelial tumors with a Ki-67 labeling index exceeding 10% are indicative of malignancy. The histopathological findings in this patient demonstrated several parameters associated with malignant potential, including markedly elevated Ki-67 levels.

Myoepithelial carcinoma may arise either spontaneously or as a result of the progression from a pleomorphic adenoma. Previous study has indicated that myoepithelial carcinoma originating from a pleomorphic adenoma tends to exhibit low-grade malignancy, whereas de novo cases may exhibit a more aggressive growth pattern [4].

Malignant neoplasms of the lacrimal gland are primarily managed through comprehensive

surgical resection. In certain circumstances, orbital exenteration may be warranted. Notably, there exists a singular documented case of myoepithelial carcinoma of the lacrimal gland in Korea, which was managed via orbital exenteration [5]. However, in cases where the tumor is locally confined, as observed in this instance, it may be possible to avoid exenteration by utilizing radiation therapy as an adjunctive treatment modality. This case is particularly significant as it represents the first documented instance in Korea of myoepithelial carcinoma of the lacrimal gland being treated with a combination of surgical resection and radiation therapy.

In conclusion, lacrimal myoepithelial carcinoma should be considered in the differential diagnosis of lacrimal gland neoplasms. The correlations among histopathological and immunohistochemical findings, along with clinical presentations and radiological findings, may aid in establishing a definitive diagnosis.

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Figure 1. (A) A preoperative image of a patient exhibiting swelling of the right upper eyelid. (B) Computed tomography (CT) imaging identified a lobulated mass measuring 3 x 2 cm with calcification (arrow) in the right lacrimal gland. (C) T2-weighted magnetic resonance imaging illustrated a well-circumscribed mass in the right orbit, exerting pressure on the eyeball. (D) A tumor that had been entirely resected was observed. (E) The histopathologic examination showed round to polygonal epithelioid cells exhibiting nuclear pleomorphism and atypical mitotic figures (H&E, x40). (F) Immunohistochemical analysis revealed a positive expression for calponin, which serves as a marker for myoepithelial differentiation. (G) The proliferation marker Ki-67exhibited a notable increase, reaching 30%, indicating a high proliferative index and invasive characteristics of the tumor. (H) Postoperative CT scan confirmed complete tumor excision with no evidence of recurrence.