Primary Ductal Adenocarcinoma of the Lacrimal Sac : A Case Report

Pawarit Wanichsetakul[1], Picha Thunpimon[1], Chatchai Thamwongskul[1]

[1]Department of Ophthalmology, Faculty of Medicine, Thammasat University, Thailand

Abstract

Primary lacrimal sac tumor is relatively rare and mostly malignant. Only limited cases have been reported. We herein presented a case of primary ductal adenocarcinoma of the lacrimal sac with androgen receptor expression. A 69-year-old Thai male presented with proptosis with a right medial canthal mass. Computed tomography demonstrated a heterogeneously enhancing mass at the right lacrimal sac region which involves the right upper nasolacrimal duct, right medial canthus, right lower and upper eyelid at the nasal side with skin involvement. The mass posteriorly involves right extraconal fat and insertion of the right medial rectus muscle with no bone remodeling or destruction. Biopsy from the right medial canthal mass was initially suggestive of oncocytic adenocarcinoma which can resemble primary ductal adenocarcinoma. There was no consensus on lacrimal sac tumor staging. After right eye exenteration was done, the final pathological findings and immunochemistry revealed a diagnosis of primary ductal adenocarcinoma of the lacrimal sac with androgen receptor expression. At first, the metastasis work-up was equivocal. Only a few nodules at the lung were observed which was suspected of lung metastasis. The tumor later showed a progression with multiple lymph nodes, lung and bone metastasis despite post-op radiation. This is the first case of primary ductal adenocarcinoma of the lacrimal sac with aggressive active disease at Thammasat University Hospital.

Keywords: Primary ductal adenocarcinoma, lacrimal sac, androgen receptor

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Introduction

Malignant epithelial lacrimal sac tumors are rare cancers with significant recurrence rates and are mostly primary in origin. There has been about 775 case reports from 1772-2017.[1] The most common presenting signs and symptoms were epiphora, recurrent dacryocystitis, and lacrimal sac mass.[1,2] Clinical presentation can resemble that of chronic dacryocysititis or dacryostenosis.[1,2] Benign and malignant lacrimal sac tumors can be epithelial or non-epithelial arising from mesenchymal components.[1,3] Among all the malignant epithelial lacrimal sac tumors, primary ductal adenocarcinoma of the lacrimal sac is found to be relatively rare. Only 28 adenocarcinoma cases have been reported according to the major review in 2020.[2,4] Herein, we present the first documented case of primary ductal adenocarcinoma of the lacrimal sac at Thammasat University Hospital.

Correspondence to:
Picha Thunpimon, *Department of Ophthalmology, Faculty of Medicine, Thammasat University, Thailand.*
E-mail: Pthunpimon@gmail.com
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Case History

A 69-year-old Thai male first came to the hospital with a chief complaint of right medial canthal palpable mass with no other signs of inflammation or infection.

(Figure 1A) He had a symptom of progressive painless blurred vision right eye which was thought to be due to globe displacement by the tumor. He had abnormal tearing and minimal irriation right eye. There was no pus discharge or bleeding from the tumor. He had no family history of orbital and its adnexal cancer. He had a history of cerebrovascular infraction about 7-8 years ago with no new events. He reported not taking any medication for almost 1 year. Upon examination, the tumor was fixed to the region, poorly defined border. Its size was about 1.5 cm in diameter. The tumor can be seen bulging from the lower fornix showing an erythematous appearance. Anterior segment and fundus examination were within normal range. Computed Tomography (CT) was done and demonstrated a heterogeneously enhancing mass at the right lacrimal sac region which involves the right upper nasolacrimal duct, right medial canthus, right lower and upper eyelid at the nasal side with skin involvement. The mass posteriorly involves right extraconal fat and insertion of the right medial rectus muscle with no bone remodeling or destruction. (Figure 1B-1C) Multiple lung nodules were also observed. Incisional biopsy of the mass was done for diagnosis. The pathological report was suggestive of oncocytic adenocarcinoma. Subsequently, orbital extended exenteration of his right eye with endoscopic right medial maxillectomy with right total ethmoidectomy was performed.

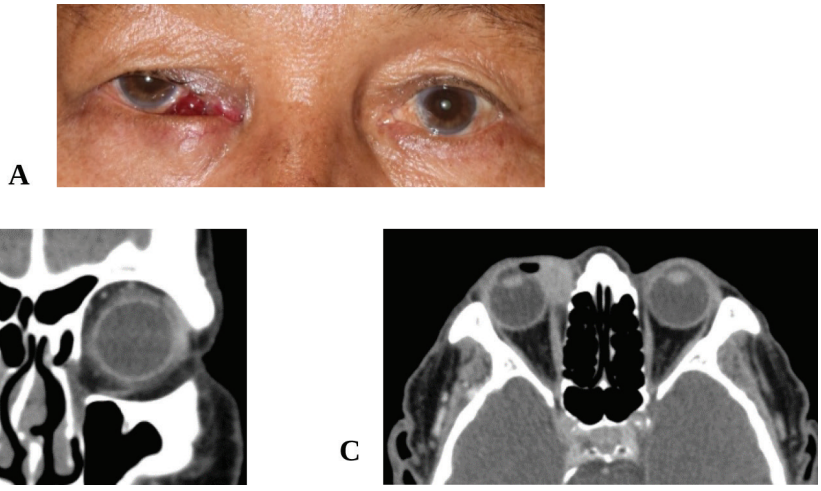

Figure 1: 69-year-old Thai male with right medial canthal mass (A). CT-scan of the orbits: coronal (B) and axial (C) views. A large mass presents in the area of the lacrimal sac extending to the nasolacrimal duct with superotemporal displacement of the right globe. (B) The mass involves the insertion of the right medial rectus muscle (C)

The final pathological study was suggestive of ductal adenocarcinoma with perineural and lymphovascular invasion. (Figure 2A-2C) Additional immuno-histochemistry is as follows; Androgen receptor (positive), GCDFP-15 (positive), HER-2 (membranous positive), ER (negative), PR (negative), CK7 (positive), CK20 (negative), TTF-1 (negative), Ki67 index = 40% (Figure 2D-2E). Based on histomorphology, immunostaining results and clinical correlation, the final diagnosis was made as "Primary ductal adenocarcinoma of the lacrimal sac" with suspected lung metastasis. The patient received a postoperative radiotherapy treatment, a total of 66Gy/33Fx. He experienced weight loss and refractory epistaxis during the course. He was scheduled to follow up every one to two weeks with the internal medicine department due to the radiotherapy side effects. Later, he was found to have

multiple sites metastases including lung and bone. His clinical worsened and was not fit for another course of chemotherapy. With a progressive clinical course, he was finally taken care of by the palliative team. He died one year after the diagnosis.

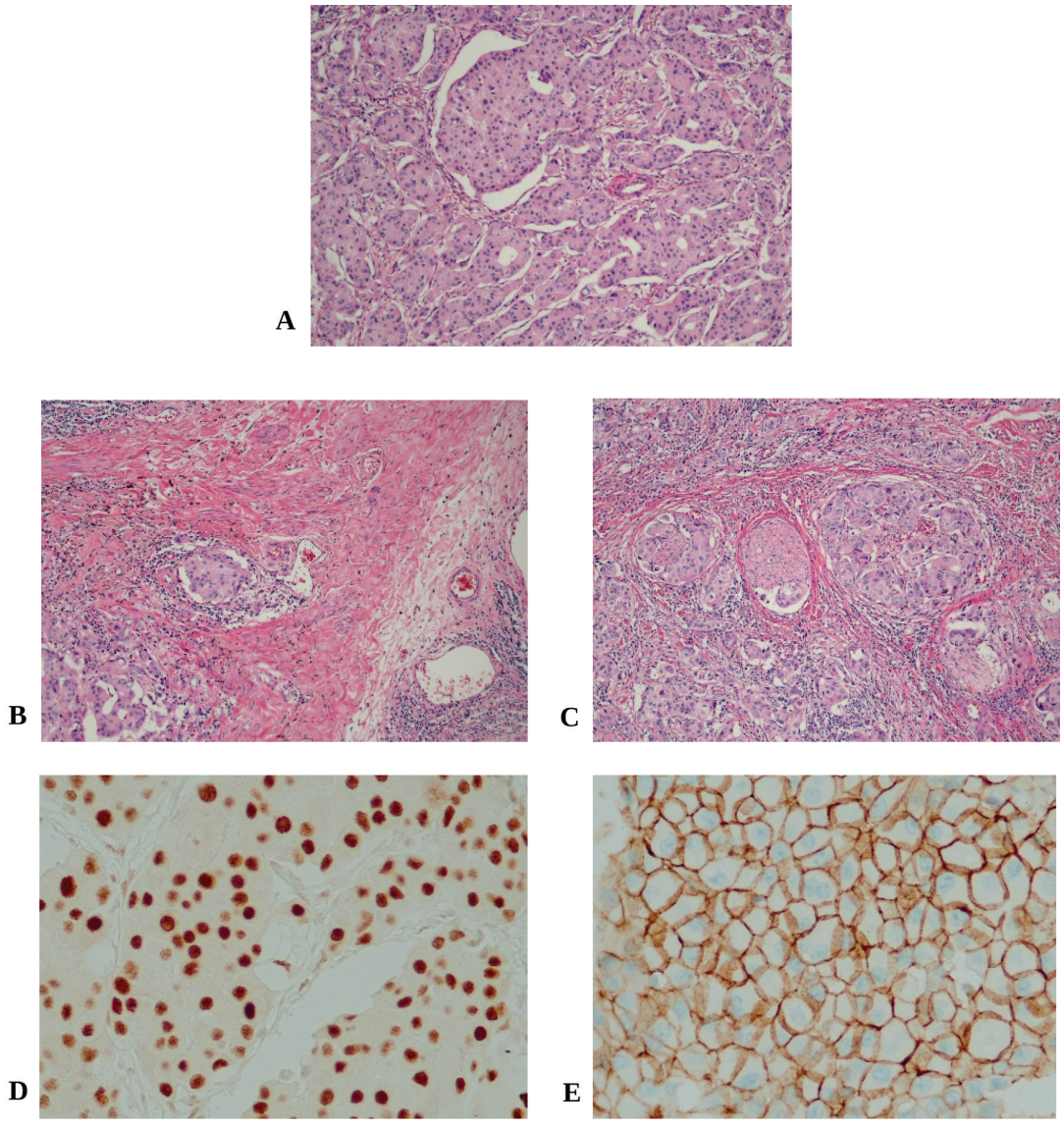

Figure 2: Histopathologic evaluation of the exenteration specimen. (A) Low-power view of the tumor arising from the lacrimal sac showing malignancy feature (stain, hematoxylin-eosin). (B) Lymphovascular invasion (stain, hematoxylin-eosin). (C) Perineural invasion (stain, hematoxylin-eosin). (D) Immunohistochemistry positive for androgen receptor. (E) Immunohistochemistry positive for HER-2.

Discussion

In this report, we described a case of primary ductal adenocarcinoma of the lacrimal sac presenting with medial canthal mass. The differential diagnosis of masses in the medial canthus is crucial for proper management. This includes dacryocystitis, nasolacrimal duct blockage, and less commonly lacrimal sac and duct neoplasms.[1,2,5]

Orbital imaging is mandatory when neoplasm is suspected and useful for disease staging and preoperative evaluation.[1,2] Both CT and MRI can be used. However, MRI is more accurate in soft-tissue evaluation. In this case, only orbital CT was done and adequate for pre-operative evaluation. For a definitive diagnosis, a tissue biopsy must be obtained. The biopsy of the medial canthal mass was done suggesting oncocytic adenocarcinoma possibly due to the small amount of poorly stained specimen. The exenteration was done due to the tumor extension to the fornix and orbit. After the exenteration, the tumor histopathology showed granular differentiation with malignant features. The tumor has positive immunoreactivity for Androgen receptor, GCDFP-15, CK-7 and HER-2 and negative immunoreactivity to ER, PR, CK20, TTF-1 which corresponds to ductal adenocarcinoma. Therefore, the final diagnosis of primary ductal adenocarcinoma of the lacrimal sac was made in correlation with clinical presentation and pathological findings.

The treatment for lacrimal sac tumors consists of surgery, radiotherapy, and infrequently, chemotherapy.[1,2,6-8] Our patient had gone through the extended exenteration of the right orbit and post-operative radiotherapy. In the literature, 47 percent of patients underwent surgery and radiotherapy treatment, another 53 percent underwent surgical treatment alone. Only 33 percent of patients underwent exenteration.[1] Despite the provided treatment, he was later found to have multiple site metastases and was not tolerable to the chemotherapy. As described in most studies, the malignant lacrimal sac tumor has a high rate of morbidity and mortality rate. The prognosis in this patient is poor due to multiple distant metastases. He had only 1-year survival after the diagnosis. According to the literature, the overall survival rate in non-adenocarcinoma was only 26 percent.[1] The best palliative care was accepted by the patient and his family members before he passed.

The limitation of this study is that there is no consensus on staging or treatment protocol. The treatment was based on the expert's opinion and previously reported data. The technique done for the biopsy was also missing.

Conclusion

Primary ductal adenocarcinoma of the lacrimal sac is relatively rare but fatal. Our patient only had 1 year survival after the diagnosis. The differential diagnosis of medial canthal mass must be carefully made. Imaging and biopsy must be done for all the cases suspected for neoplasms so that the early definitive diagnosis can be made for a proper treatment.

References

1. Singh S, Ali MJ. Primary Malignant Epithelial Tumors of the Lacrimal Drainage System: A Major Review. Orbit. 2020;40(3):179-92.
2. Krishna Y, Coupland SE. Lacrimal Sac Tumors-A Review. Asia Pac J Ophthalmol (Phila). 2017;6(2):173-8.
3. Heindl LM, Junemann AGM, Kruse FE, Holbach LM. Tumors of the Lacrimal Drainage System. Orbit. 2010;29(5):298-306.
4. Bi YW, Chen RJ, Li XP. Clinical and pathological analysis of primary lacrimal sac tumors. Zhonghua Yan Ke Za Zhi. 2007;43(6):499-504.
5. El-Sawy T, Frank SJ, Hanna E, Sniegowski M, Lai SY, Nasser QJ, Myers J, Esmaeli B. Multidisciplinary management of lacrimal sac/nasolacrimal duct carcinomas. Ophthalmic Plast Reconstr Surg. 2013;29(6):454-7.
6. Pe'er J. Lacrimal Sac Tumors. Clinical Ophthalmic Oncology. Springer; 2014.
7. Parmar DN, Rose GE. Management of lacrimal sac tumours. Eye (Lond). 2003;17:599-606.
8. Montalban A, Lietin B, Louvrier C, Russier M, Kemeny JL, Mom T, et al. Malignant lacrimal sac tumors. European annals of otorhinolaryngology, head and neck diseases. 2010;127:165-72.