

Globe-sparing Surgery and Adjuvant Radiotherapy for Lacrimal Gland Adenoid Cystic Carcinoma in a 37-Year-Old Filipino Female

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ABSTRACT

Objective: To report a case of lacrimal gland adenoid cystic carcinoma in a 37-year-old female managed with globe-sparing surgery and adjuvant radiotherapy.

Methods: This is a case report.

Case Presentation: This report details a 37-year-old Filipino female who presented with a 4-month history of right eye pain and non-axial proptosis. Computed tomography (CT) scan showed a right heterogenous extraconal mass with orbital roof remodeling. Lateral orbitotomy with excision biopsy was performed. Final histopathologic diagnosis of adenoid cystic carcinoma of the lacrimal gland was made based on the classic morphology and distinct arrangement of the neoplastic cells. Systemic surveillance revealed absence of metastases. Patient completed adjuvant radiotherapy four months post-operatively. Vision was preserved at 20/20 without signs of tumor recurrence five months post-operatively.

Conclusion: Adenoid cystic carcinoma is an aggressive lacrimal gland malignancy; hence, meticulous examination and high index of suspicion cannot be over-emphasized. Globe-sparing surgery with adjuvant radiotherapy could significantly preserve quality of life.

Keywords: Lacrimal Gland, Adenoid Cystic Carcinoma, Orbitotomy



INTRODUCTION

Lacrimal gland tumors have a yearly incidence of only one in one million, representing 10% of orbital space-occupying lesions.¹ Adenoid cystic carcinoma (ACC) is the most common lacrimal gland malignancy. Given its aggressive behavior, early management strategies traditionally relied on radical orbital exenteration to achieve local tumor control, prevent spread, and reduce risk for recurrence. To date, no treatment protocol has been established but current trends favor less mutilating globe-sparing surgery and adjuvant radiotherapy which exhibited favorable local control and long-term survival outcomes. Advances in multimodal globe-sparing treatment strategies have been described in recent literature; however, widespread adoption remains controversial due to limited data validating effectiveness. This report aims to present the clinical features and outcome of a 37-year-old female with lacrimal gland adenoid cystic carcinoma managed with excision biopsy via lateral orbitotomy and adjuvant radiotherapy to increase clinical awareness and aid in the diagnosis and management of this aggressive neoplasm.

CASE PRESENTATION

This is a case of a 37-year-old Filipino female with a 4-month history of right eye pain and non-axial proptosis. Facial asymmetry due to proptosis of the right eye prompted consult. Past medical, family, ocular, personal and social histories were non-contributory. Review of systems was unremarkable. On ophthalmologic examination, the uncorrected distance visual acuity for both eyes were 20/20. Pupils were 2 to 3 mm equally reactive to light. The right eye was displaced inferonasally (Figure 1A) with significant 3 mm proptosis on exophthalmometry, also evident on worm's view (Figure 1B). There was note of a firm, non-movable, non-tender, non-erythematous mass in the right superotemporal orbital rim without any discoloration, ulceration, and discharge, measuring approximately 2.00 cm x 2.00 cm in the widest diameters. The rest of the external eye and adnexal examinations were unremarkable. There was restriction in right upward gaze on motility testing. Patient denied pain and diplopia in all cardinal gazes. Slit-lamp examination, applanation tonometry, and

fundus examination were unremarkable. Plain orbital Computed Tomography (CT) scan (Figure 2) revealed a heterogenous extraconal lesion adjacent to the right lateral rectus and lacrimal gland measuring 2.00 cm x 1.50 cm x 2.40 cm with associated orbital roof remodeling, without involvement of the optic nerve.

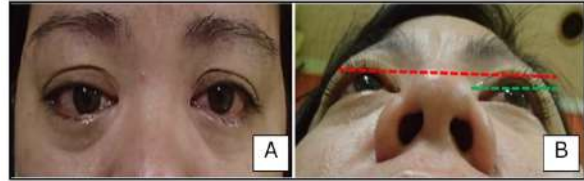


Figure 1. (A) The right globe displaced inferonasally. (B) Worm's view, showing proptosis of the right globe.

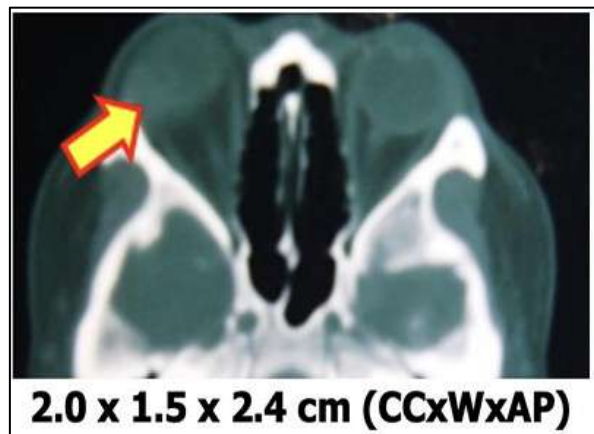


Figure 2. Plain Orbital CT-scan in axial view showing a heterogenous extraconal lesion adjacent to the right lateral rectus and lacrimal gland without involvement of the optic nerve, measuring 2.00 cm x 1.50 cm x 2.40 cm (yellow arrow)

The patient underwent excision biopsy of the tumor through lateral orbitotomy with beveled osteotomies under general anesthesia (Figure 3). A tan-red mass, measuring 5.50 cm x 1.20 cm x 1.00 cm along with 2 other specimens were obtained and sent for frozen section which revealed a malignant tumor, favoring ACC of the right lacrimal gland. The lack of true encapsulation and tumor configuration precluded en bloc excision. The lateral orbital bone was not repositioned to facilitate adjuvant radiotherapy. Final histopathologic diagnosis (Figure 4) of adenoid cystic carcinoma of the right lacrimal gland was made based on the classic arrangement of the neoplastic cells in tubular and cribriform patterns composed of nests with microcystic-like spaces filled with eosinophilic material. The neoplastic cells showed pleomorphism with coarse chromatin pattern and prominent

nuclei. There was no evidence of lympho-vascular invasion. Systemic surveillance utilizing cranial magnetic resonance imaging and chest CT scan, both with contrast, showed absence of regional and distant metastases. The American Joint Committee on Cancer (AJCC) staging for this case was T3aN0M0 based on the absence of bone or periosteal involvement in a tumor greater than 4mm in size (T3a), absence of regional metastasis (N0), and absence of distant metastasis (M0). As part of her comprehensive treatment plan, the patient received 70 Gy of adjuvant intensity-modulated radiotherapy in 35 fractions for enhanced local control which she completed four months post-operatively. Five months post-operatively, with good wound healing, uncorrected visual acuity of both eyes was 20/20 without any signs of tumor recurrence (Figure 5).

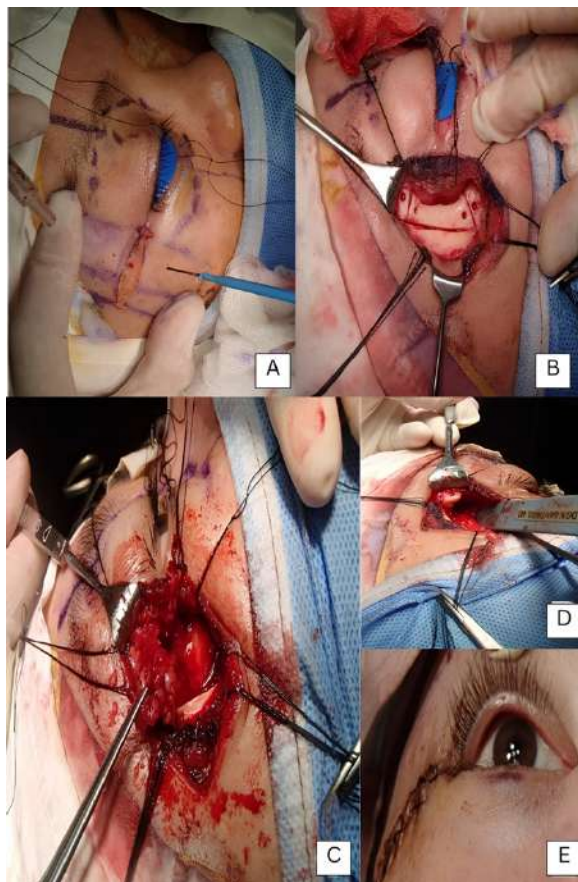


Figure 3. Surgical management. (A) Surgical markings placed as guide prior to surgery. (B) Beveled osteotomies created as described by Santiago et.al. in recent literature. (C) localization and excision of the lacrimal gland mass (D) Since frozen section revealed a malignancy, the lateral orbital bone was not repositioned in preparation for adjuvant radiotherapy. (E) Layer closure done. Right pupil that was reactive to light postoperatively is shown.

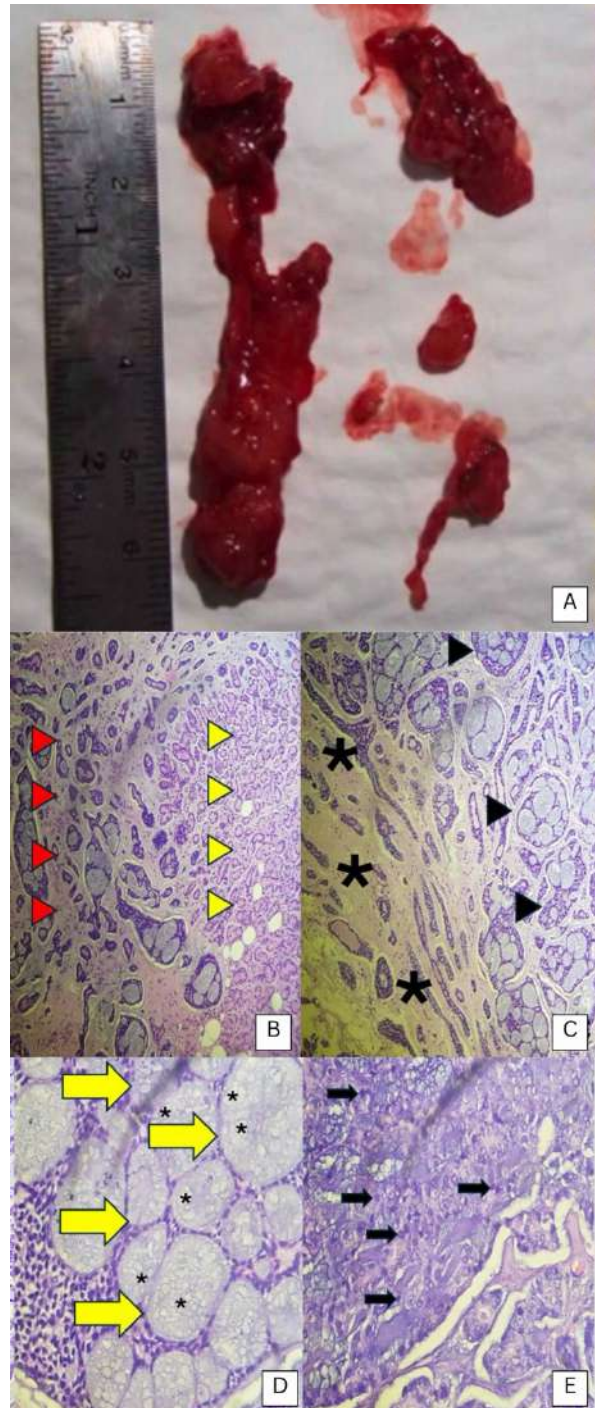


Figure 4. Histopathology. (A) Grossly, a tan-red, irregular, rubbery mass, with tan-red and tan-white cut surfaces, measuring 5.50cm x 1.20cm x 1.00cm. (B) Cut section showing normal (yellow arrow heads) and neoplastic cells (red arrow heads) (C) Microscopically, a neoplasm arranged in tubular (asterisks) and cribriform (black arrow heads) patterns was seen. (D) There are nests (yellow arrows) with microcystic like spaces (asterisks) filled with eosinophilic material. (E) The neoplastic cells showed pleomorphism with coarse chromatin pattern and prominent nuclei (black arrows).

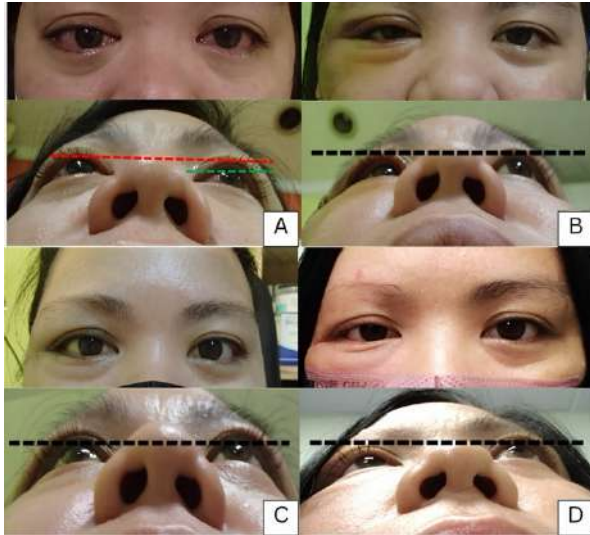


Figure 5. The patient is shown (A) preoperatively, (B) 1 week, (C) 2 weeks, and (D) 5 months postoperatively. There is marked resolution of proptosis as seen on worm's view without signs of infection and recurrence five months postoperatively.

DISCUSSION

A 10-year retrospective review at the Philippine Eye Research Institute conducted by Domingo et.al. revealed that among the 1,551 histologically confirmed ocular and adnexal tumors that were included, only 23 (1.48%) were lacrimal gland ACC.² It is the most frequently encountered malignant epithelial tumor of the lacrimal gland but accounts for less than 2% of tumors in the head and neck area. Maintaining a high index of suspicion cannot be overemphasized given its relative rarity.

ACC affects younger individuals in the 4th decade of life with male preponderance.³ It is an aggressive malignancy that typically presents with pain due to perineural invasion and bone destruction, which help differentiate it from pleomorphic adenoma. Penetration and extension into the posterior orbit are not uncommon due to lack of true encapsulation. Due to the gland's anatomic location, ocular motility restriction and non-axial globe displacement may be observed which vary depending on tumor size and depth of extension.

Imaging is vital in initial evaluation and surgical planning for lacrimal gland lesions. Epithelial neoplasms tend to appear as isolated globular masses that displace and indent the globe. For this

case, plain orbital CT scan demonstrated a heterogenous extraconal lesion located in the right superotemporal orbit, displacing the globe but did not appear to be indenting it. Orbital roof remodeling was observed but does not seem to coincide with the clinical picture. These, coupled with the ophthalmologic exam findings, guided pre-operative diagnosis which leaned toward a lacrimal gland neoplasm, with benign and malignant causes equally plausible at that point.

Although most lacrimal gland masses represent inflammatory conditions, malignancy remains a possibility. An American Society of Ophthalmic Plastic and Reconstructive Surgery Society (ASOPRS) Foundation lecture noted that lesions involving the lacrimal gland have lower threshold for biopsy to exclude a malignancy.⁴ Hence, en bloc excision biopsy through a lateral orbitotomy with beveled osteotomies was offered to the patient and subsequently performed after thorough discussion.

To date, there is neither consensus nor an established treatment protocol for lacrimal gland ACC due to its rarity. Traditionally, radical orbital exenteration with adjuvant radiotherapy was the standard but current trends favor globe-sparing surgery with adjuvant radiotherapy. In a retrospective review by Han et.al. Ten patients with orbit-confined ACC who had globe-sparing surgery with adjuvant radiotherapy exhibited favorable local control and long-term survival outcomes. Eight were alive until the last follow up (Follow up: median = 89.5 months, range = 37 to 217 months) and only one had recurrence at 53 months post-operatively. All ten did not have systemic metastasis. Similarly, in a case report by Lewis et.al., a monocularly-functioning patient with lacrimal gland ACC adjacent to the only seeing eye demonstrated favorable local control without evidence of recurrence four years post-operatively after local tumor excision and adjuvant radiotherapy. Adjunctive intra-arterial cytoreductive chemotherapy stands out as a pivotal advancement in the management of adenoid cystic carcinoma.⁷ It demonstrated potential in reducing recurrence rates and improving long-term survival and may serve as an effective component of multimodal globe-sparing therapeutic strategies.⁸ However, owing to disease rarity and the lack of robust randomized controlled trial data, substantial evidence remains

inadequate and its adoption continues to be controversial.

Currently, safe access to the orbital space necessitates removal of the lateral orbital wall. Hence, the innovative beveled osteotomy approach for lateral orbitotomy technique described by Santiago et.al. was performed (Figure 3B). The beveled osteotomies were created by positioning a customized 21 mm rotating circular bone saw at an oblique angle relative to the bone surface. The angled cuts enabled the separated bone segments to be extended while preserving greater contact between the bony surfaces. As a result, there was a broader posterior surgical opening, optimizing the surface area for secure repositioning of the bone flap following tumor excision. Additionally, the beveled configuration enhanced the inherent stability of the osteotomies, eliminating the need for fixation devices since the precise beveled cuts minimized bone loss. The beveled cuts would have allowed the separated bone segments to be repositioned with an increased surface area for contact facilitating better stability and healing.⁹ However, for facilitation and better penetration during radiotherapy, the lateral orbital bone was not repositioned.

The final histopathologic report revealed a neoplasm arranged in tubular and cribriform patterns composed of nests with microcystic like spaces filled with eosinophilic material which is consistent with non-basaloid type ACC (Figure 4). Gamel and Font emphasized the need to differentiate between basaloid and non-basaloid types for prognostication. The patient had the non-basaloid type of tumor with a 5-year survival rate of up to 71%.¹⁰ In addition, a retrospective review by Ahmad et. al. revealed that AJCC classification of more than T3 disease at initial diagnosis had worse outcomes.¹¹ Timely surgical planning allowed prompt diagnosis for our patient whose tumor classification fell under T3a (Figure 6). The short post-operative surveillance period is an explicit limitation of this report as it precludes thorough assessment of long-term disease control, survival outcome, and delayed complications.

CONCLUSION

Although ACC is the most encountered malignancy of the lacrimal gland, it remains an infrequent pathology both globally and locally;

hence, meticulous examination coupled with advances in orbital imaging to arrive at an accurate diagnosis cannot be overemphasized. The successful removal of the neoplasm through globe-sparing surgery combined with adjuvant radiotherapy, in contrast to a disfiguring exenteration, enabled the patient to maintain a positive outlook despite the diagnosis of malignancy. It offered a sense of hope and control, preserving her quality of life.

Primary tumor	T0	No evidence of primary tumor
	T1	Tumors with a size <2 cm with or without invasion of the soft orbital tissue moreover
	T1a	Absence of periosteal/bone involvement
	T1b	Presence of periosteal involvement
	T1c	Both periosteal and bone involvement
	T2	Tumors with a size between 2 cm and 4 cm
	T2a	Absence of periosteal/bone involvement
	T2b	Presence of periosteal involvement
	T2c	Both periosteal and bone involvement
	T3	Tumors greater in size than 4 cm
	T3a	Absence of periosteal/bone involvement
	T3b	Presence of periosteal involvement
	T3c	Both periosteal and bone involvement
	T4	Tumor extension into adjacent structures
Regional lymph nodes	Nx	No assessment of regional lymph nodes
	N0	No involvement of regional lymph nodes
	N1	Presence of regional lymph nodes metastasis
Metastasis	M0	Absence of distant metastasis
	M1	Presence of distant metastasis

Adapted from AJCC Cancer Staging Manual, 8th ed. Springer International Publishing; 2017: 1032.

Figure 6. American Joint Committee on Cancer Staging for Lacrimal gland ACC. The patient is classified under T3aN0M0 based on the following: absence of bone or periosteal involvement in a tumor greater than 4mm in size (T3a), absence of regional metastasis (N0), and absence of distant metastasis (M0).

ETHICS COMPLIANCE STATEMENT

The authors affirm that this case report was prepared in accordance with the ethical standards and publication policies of the Philippine Journal of Ophthalmology, as well as applicable institutional and international ethical guidelines governing the reporting of clinical cases.

Written informed consent for the publication of the patient's clinical information and accompanying images was obtained prior to manuscript submission. All reasonable measures were undertaken to safeguard patient privacy, and all identifying information has been omitted or anonymized to ensure confidentiality.

As this manuscript describes a single clinical case managed according to standard clinical practice and does not involve experimental interventions or research procedures, formal institutional review board approval was not required under the policies of the authors' institution.

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