CASE REPORT

Atypical Cystic Carcinoma: A Rare Malignancy of The Lower Eyelid

Janah R[a], Boesoirie SF[b]

[a]Department of Anatomic Pathology, Cicendo Eye Hospital, Bandung, Indonesia
[b]Department of Ophthalmology, Cicendo Eye Hospital, Bandung, Indonesia

Keywords
Adenoid cystic carcinoma, Lower Eyelids, Rare Case

Corresponding Author
Dr Raudatul Janah
Department of Anatomic Pathology, Cicendo Eye Hospital, Bandung, Indonesia.
E-mail: raudatul.janah1@gmail.com,

Received: 24th June 2024; Accepted: 22th April 2025

Doi: https://doi.org10.31436/imjm.v24i03/2654

ABSTRACT

Adenoid cystic carcinoma (ACC) is a rare malignancy of the secretory glands, most commonly originating in the lacrimal gland but rarely involving the eyelids. ACC is known for its aggressive behaviour, high recurrence rate, and potential for metastasis, leading to a poor prognosis. Histopathological examination is essential for the diagnosis, and early detection with complete excision offers the best chance for a favourable outcome. This case highlights the importance of comprehensive management to reduce the recurrence risk and improving patient survival. The authors present a 56-year-old man with ACC of the left lower eyelid, who underwent surgical excision followed by reconstruction, ensuring tumour-free margins.

INTRODUCTION

A rare type of eyelid malignancy that occurs includes sebaceous cell carcinoma, and malignant melanoma, including adenoid cystic carcinoma (ACC).[1,3] ACC is a subtype of cancer of the secretory glands. ACC of the eye generally manifests in the lacrimal gland, but it can also develop from Moll glands, sweat glands on the eyelids, or accessory lacrimal glands covering of the conjunctiva. ACC is rare in its prevalence but has local and regional recurrence rates and high levels accompanied by metastases. Appropriate, fast management and enforcement of the diagnosis through histopathological examination is an important stage in determining the therapy, and prognosis, preventing recurrence and metastasis so that patients get better results.[2,4,5] ACC involving the lower eyelids is a rare presentation, as most cases originate in the lacrimal gland. The importance of early diagnosis and complete surgical excision of the mass after histopathological confirmation is to improve prognosis. Reconstructive surgery plays a role in restoring function after tumour excision. Whilst the need for long-term follow-up is to monitor for recurrence or metastasis. This case report aims to explain the diagnosis and management of cases of ACC on the inferior eyelid with the involvement of the punctum and canaliculi.

CASE REPORT

A 56-year-old man with the main complaint of a lump on the lower eyelid of his left eye which he felt has become enlarged for the last 3 years. The lump was initially the size of a green bean and grew bigger until it was the size of a marble. The lump was reddish in color, sometimes accompanied by itching and lumpiness. The lump never bled or festered. No lymph node enlargement was observed in the head, neck, or axillary regions. The patient had no history of malignancy, chronic diseases such as diabetes or hypertension, immunosuppression, or prior radiation exposure. There were no known exposure to carcinogens, chemicals, or UV radiation. Additionally, no systemic symptoms, including weight loss, night sweats, or fatigue, were reported. Examination of the left eye (Figure 1) showed that in the lower eyelid there was a mass measuring 2 x 1.5cm which affected the inferior punctum and covered the medial canthus area. The mass had an uneven surface, was soft in consistency, had firm boundaries with a fixed base. It also had a partly red and yellowish central mass. The mass did not bleed, and no pus or ulcers were present, and there was also madarosis. A thorax x-ray examination performed before surgery showed that there were no visible active pulmonary TB

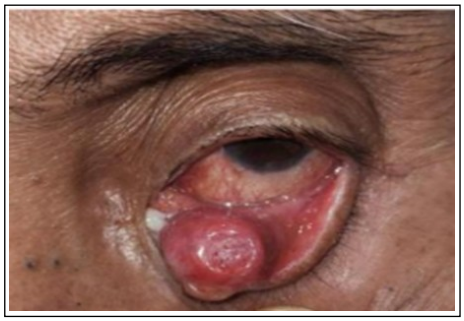

Figure 1: Mass on the lower eyelid

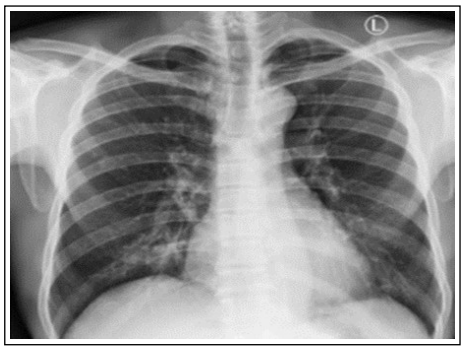

Figure 2: No mass in the pulmonary

The patient was planned for a wide excision of the inferior eyelid mass with reconstruction of the eyelid defect using the Tenzel semicircular flap technique to preserve function and cosmesis and placement of a silicone tube to maintain the patency of the remaining inferior canaliculi. The results of histopathological examination (Figures 3 and 4) showed adenoid cystic carcinoma in the lower palpebra of the left eye. The base of the incision and the 4-1 thread border are not yet free of malignant tumor cells. Incision margins 1-2, 2-3 and 3-4 were free of malignant tumor cells. Since one surgical margin was positive for tumour cells, re-excision was planned. Regular clinical assessments every 3-6 months for the first 2 years was planned, following that, annual follow-up.

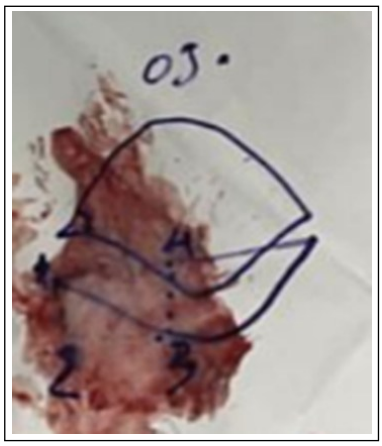

Figure 3: Macroscopic mass

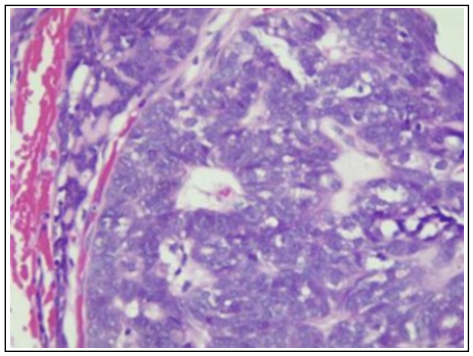

Figure 4: Microscopic cribriform pattern seen in ACC. Basaloid cells with hyperchromatic nuclei, small gland-like spaces filled with eosinophilic material. No perineural invasion is a hallmark of aggressive behaviour. (400x H&E)

DISCUSSION

Adenoid cystic carcinoma is a rare secretory gland cancer with an incidence of 4-5 cases per 100,000 individuals. Epidemiological data shows that ACC is only 1% of all cancers and are located in the head and neck area. ACC is a type of malignancy with relatively slow clinical progression; however, it has high local and regional recurrence rates. ACC cases often metastasize including distant metastases such as lung metastases or liver. ACC that occurs in the eye area generally affects the lacrimal gland including the lacrimal system which has a histological structure resembling the saliva gland. ACC can also appear in other glands such as sweat glands, glands of Moll and other accessory glands scattered in the eyelids and conjunctiva. Manifestations of ACC on the inferior eyelid with involvement of the punctum and canaliculi are not common. The mass that arises can be a primary tumour or secondary tumours due to metastases from other organs.[1,6,7] In our patient the mass presented on the inferior eyelid and involved the punctum and inferior canaliculi. It has progressively grown in size for the last 3 years. The lump which sometimes felt itchy has an irregular surface that indicates one of the clinical signs of malignancy. The patient is likely a case of primary ACC because no other local or regional mass other than the inferior eyelid was found.

Signs and symptoms that can be found in cases of ACC resemble malignancy of the outer eyelid. The clinical manifestation of ACC may present as a firm, nodular lesion, which is typically painless, with possible madarosis. The mass located in the lacrimal system can cause epiphora. Other accompanying symptoms depend on the

location and size of the mass. Manifestations in the lacrimal glands can cause ptosis.[2,8] Our patient's tumour mass was located in the inferior palpebra accompanied by inferior punctum distension. The mass measured 2x1.5cm, had an uneven surface, was soft in consistency, had firm boundaries with a fixed base. It also had a partly red and yellowish central mass. The mass did not bleed, and no pus or ulcers were present, and there was also madarosis. Making a definite diagnosis in ACC cases is through examination of histopathology of the tumour. The results of histopathological examination on ACC of our patient showed epithelial cells basaloid with pleomorphic nuclei (Figure 4). Histologically there are three types of ACC, namely tubular, solid, and cribriform with the most common type being cribriform.[9] In the patient the results of histopathological examination showed group tumour masses that were hyperplastic. Tumour cells with round oval shapes, pleomorphic nuclei, hyperchromatic, coarse chromatin, abnormal mitoses were found to form cribriform and cystic structures and solidify diffusely. This histopathological description aligns with the characteristic features of ACC.

ACC is a malignant tumour with a generally poor long-term prognosis. Although ACC is a slow-growing epithelial malignancy, it has a high tendency for recurrence and metastasis. The estimated survival rates for ACC patients are 68% at 5 years, 52% at 10 years, and 28% at 20 years. Early detection, accurate diagnosis and appropriate therapy is necessary for the management of patients with ACC to ensure a better prognosis. Definitive therapy for ACC is excision of the mass until the excision margins are free of tumour cells. Excision of the mass with a frozen section is recommended to ensure the excision area is completely tumour free. Other more aggressive measures such as exenteration can be considered depending on the size and distribution of the mass. Approach to the management with globe-sparing interventions can be carried out and should be accompanied by adjuvant radiotherapy.[4,9-10] Our patient underwent a wide excision of the mass until the margins were tumour-free. The tumour mass in the patient appeared to be localized with clear boundaries so the intervention chosen is wide excision which maintains the integrity of the eyeball. Frozen section is performed intraoperatively to ensure that the excision margin is free from tumour mass.

ACC commonly originates in the lacrimal gland, but our case involves the lower eyelid, which is an extremely rare site for this malignancy. Most ACC cases of the eyelid affect the upper eyelid or tarsal region, making this presentation unique. Eyelid involvement poses challenges in achieving clear surgical margins while maintaining function and cosmesis. This case demonstrates that precise surgical excision with reconstruction can achieve both oncologic control and good cosmetic outcomes. Many ACC cases present late with perineural invasion and distant metastases (e.g., lungs, bones), but this patient had no lymph node involvement or distant spread, making early intervention highly effective.

Since ACC of the lower eyelid is rare, this case alerts clinicians to consider ACC in differential diagnoses of persistent, firm eyelid masses, ensuring timely biopsy and histopathological confirmation. Early detection and wide excision with tumour-free margins remain the gold standard for improving survival. This case highlights surgical precision in balancing oncologic safety and functional/cosmetic reconstruction. The need for collaboration among ophthalmologists, pathologists, oncologists, and reconstructive surgeons is reinforced to achieve optimal patient outcomes. Given the rarity of eyelid ACC, this case adds valuable clinical data to enhance future guidelines and treatment protocols. It highlights the need for long-term follow-up and consideration of adjuvant therapy in selected cases.

CONCLUSION

ACC is managed by complete tumour removal. A multidisciplinary approach is needed to manage patients with ACC to achieve a better prognosis for functional and aesthetic outcomes.

REFERENCES

1. Korn BS, Burkat CN, Carter KD, et al. Orbital neoplasm and malformations, reconstructive eyelid surgery. Dalam: Basic and clinical science course: oculofacial plastic and orbital surgery. San Fransisco:

American Academy of Ophthalmology 2023; 213–26. (2-1)
2. Syed NA, Berry JL, Heegaard S, et al. Ophthalmic pathology and intraocular tumors. Dalam: Basic and clinical science course. San Fransisco: American Academy of Ophthalmology 2022; 237–83. (3-5) (3-2)
3. Baş Z, Sharpe J, Yaghy A, et al . Prevalence of and associated factors for eyelid cancer in the American academy of Ophthalmology intelligent research in sight registry. Ophthalmol Sci 2022; 3(1): 100227. (5-2) (5-3)
4. Lorini L, Ardighieri L, Bozzola A, et al. Prognosis and management of recurrent and/or metastatic head and neck adenoid cystic carcinoma. Oral Oncology. 2021; 115: 105213 (6-7) (6-4)
5. Saleh E, Ukwas A. Adenoid cystic carcinoma of salivary glands: a ten-year review and an assessment of the current management, surgery, radiotherapy, and chemotherapy. Int J Otolaryngol 2023; 29: 7401458 (7-3) (7-5)
6. Iro S, Raiteb M, Maadane A, Elmrini S, Slimani F. Cutaneous adenoid cystic carcinoma of lower eyelid: case report of a rare malignant entity. Annals of Medicine and Surgery 2021; 67: 102467 (8-8) (8-6)
7. Ishida E, Ogawa T, Rokugo M, et al. Management of adenoid cystic carcinoma of the head and neck: A singleinstitute study with over 25-year follow-up. Head Face Med 2020;16 (1): 14. (9-10) (9-7)
8. Liang M, Yu Z, Wang F. A case report: An unusual presentation of adenoid cystic carcinoma of the lacrimal gland. Medicine 2023;102(13): e33446. (12-4) (12-8)
9. Lee RH, Wai KC, Chan JW, Ha PK, Kang H. Approaches to the management of metastatic adenoid cystic carcinoma. Cancers 2022; 14(22): 5698. (14-6) (14-9)
10. Nightingale J, Lum B, Ladwa R, Simpson F, Panizza B. Adenoid cystic carcinoma: a review of clinical features, treatment targets and advances in improving the immune response to monoclonal antibody therapy. Biochimica et Biophysica Acta - Reviews on Cancer 2021; 1875(2):188523. (15-