Eyelid adenoid cystic carcinoma: An analysis of one case with clinicopathological features mimicking sebaceous gland carcinoma

Bharadia Aadil Mohamed[1], Nyenze Emmanuel Muindi[1]
[1]Department of Ophthalmology, Faculty of Health Sciences, University of Nairobi, Kenya

Corresponding author:
Dr. Aadil Mohamed Bharadia,
Department of Ophthalmology, Faculty of Health Sciences, University of Nairobi,
Email: baradiaaadil@gmail.com.

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Abstract

Background: Adenoid cystic carcinoma (ACC) is a rare malignant epithelial tumor representing 1–2% of all head and neck tumors and 10–15% of malignant salivary gland tumors. In the eyelid, it can arise from the glands of Moll, the palpebral lobe of the lacrimal gland, the accessory lacrimal glands in the conjunctiva, or from ectopic lacrimal gland tissue. It is the most common primary malignant epithelial tumor of the lacrimal glands and accounts for approximately 1.6% of all orbital tumors. In this case, we see adenoid cystic carcinoma arising from the skin of the eyelid with features simulating the more commonly seen sebaceous gland carcinoma.

Case: A 74-year-old male patient presented with a two-year history of swelling in the left upper lid. The lesion was excised without being sent for histology. Subsequently, there was a recurrence of the swelling, and the patient was referred to Kenyatta National Hospital. Upon admission, the working diagnosis was left upper lid sebaceous gland carcinoma, and the patient underwent a wide full-thickness excisional biopsy of the left eyelid. The histological diagnosis revealed adenoid cystic carcinoma of the eyelid. The patient then underwent lid reconstruction surgery.

Conclusion: Malignant eyelid lesions may masquerade as several different clinically benign conditions and all excised eyelid lesions should be submitted for histopathologic confirmation.

Key words: Eyelid tumor, adenoid cystic carcinoma, sebaceous gland carcinoma.

Introduction

Adenoid cystic carcinoma (ACC) is a rare malignant epithelial tumor representing 1–2% of all head and neck tumors and 10–15% of malignant salivary gland tumors(1). In the eyelid, it can arise from the glands of Moll, the palpebral lobe of the lacrimal gland, the accessory lacrimal glands in conjunctiva, or from ectopic lacrimal gland tissue but very rare to arise from the eyelid skin(2). It is the most common primary malignant epithelial tumor of the lacrimal glands and accounts for approximately 1.6% of all orbital tumors(3,4). The differential diagnosis clinically chalazion and sebaceous gland carcinoma, and histologically basal cell carcinoma(5).

The preferred treatment for this condition is surgical excision with clear margins, followed by long-term monitoring. In cases where complete excision is unattainable, chemotherapy and radiation therapy are advised(6).

We present a rare case of adenoid cystic carcinoma arising from the skin of the left upper eyelid with features simulating the more commonly seen sebaceous gland carcinoma.

Case report

A 74-year-old male patient presented with a 2-year history of left upper eyelid swelling. The swelling was not associated with pain, redness, discharge, or tearing.
He went to a local health facility where he received unspecified eye drops but no improvement was noted. The swelling gradually increased in size and the patient complained of functional discomfort during palpebral opening and closing that motivated his re-consultation.

Excision of the lesion was carried out but was not taken for histology.

A few weeks later he noted a recurrence of the swelling which was now associated with pain and redness but no discharge. He was then referred to our department at Kenyatta National Hospital.

He is a known asthmatic patient on Ventolin inhaler. He also has a reducible inguinoscrotal hernia. No known food or drug allergies.

The clinical examination at consultation showed that he was in a fair general condition with a reducible inguinoscrotal hernia and a few broken teeth with dental carries. The ophthalmological examination of visual acuity, pupils, motility, and intraocular pressures were normal. His left upper lid showed a mass on the upper medial third of the eyelid measuring 2cm by 1.5cm with firm consistency, nodular surface, free from overlying skin, not mobile and inhibited the eversion of the upper eyelid. There was loss of lashes in the medial third of the upper lid with some telangiectasis but no ulceration of the skin overlying the lesion.

Why we thought it was a sebaceous gland carcinoma is because there was:

- Slow growing tumor on the upper eyelid
- Painless firm mass
- Loss of cilia
- Recurrence
- Nodular surface
- Telangiectasis

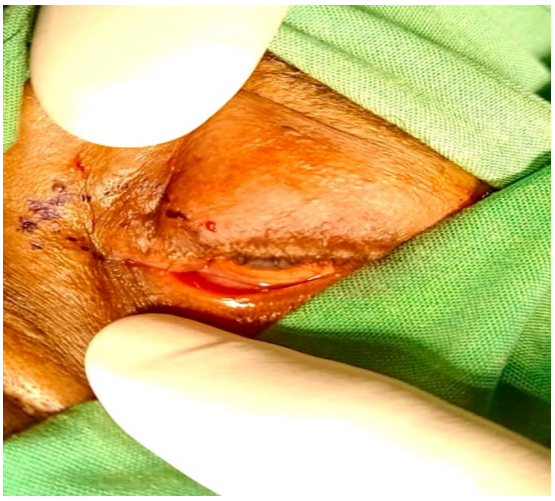

Figure1: Left eye upper lid adenoid cystic carcinoma (© Aadil Bharadia)

A head and orbit CT scan was performed, which revealed a fairly well-defined enhancing mass in the medial aspect of the upper lid measuring 0.75cm by 0.99cm with no bone erosion or lysis. There was no intraorbital extension. The images could not be retrived from the patient.

The patient underwent a wide full-thickness excision biopsy with a 4mm margin from the mass and skin margins apposed to the medial canthal region awaiting biopsy results for lid reconstruction.

Histology results from the patients excision biopsy

Gross – The specimen consists of a skin ellipse (eyelid) measuring 20x10x10mm with a suture attached to the medial margin. Cut surfaces show a white tumor 5 mm across extending into skeletal muscles. Radial margins are 3-5mm from the nearest margins.

The sections show skin-lined tissues from the eyelid showing nests and lobules of a malignant epithelial tumor as shown in *Figure 2*. The tumor had 2 patterns; solid and cystic which was hypercellular as seen in *Figure 3* and *Figure 4*.

The malignant cells are of small round cell type with a cribriform pattern bearing hyperchromatic nuclei as seen in *Figure 5* and *Figure 6*. *Figure 7* displays nodules and sheets of a malignant epithelial tumor infiltrating skeletal muscles. There is invasion of perineural tissues. The surgical margins are tumor-free.

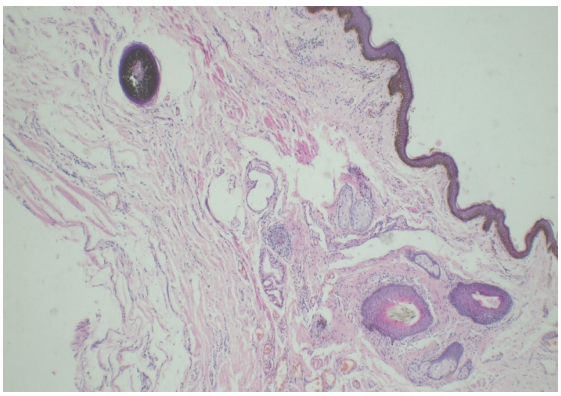

Figure 2: Eyelid skin with appendages (© Aadil Bharadia)

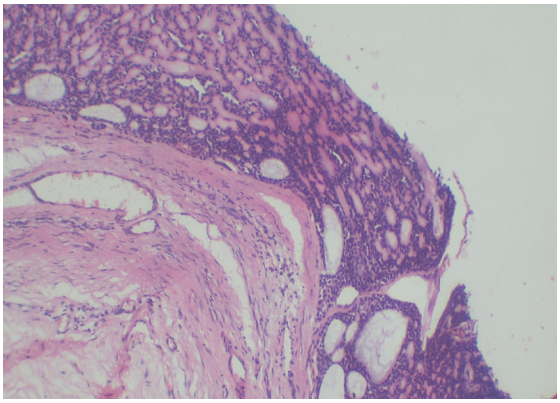

Figure 3: Hypercellular tumor noted (© Aadil Bharadia)

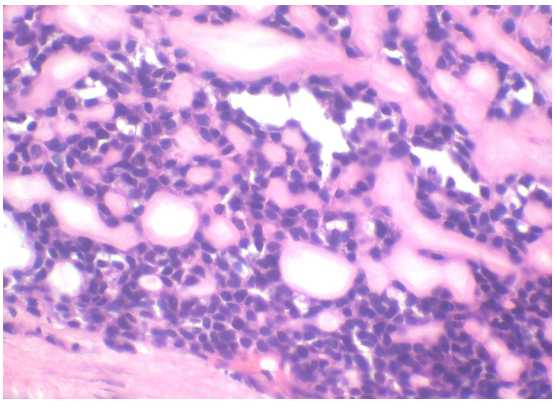

Figure 4: The tumor had 2 patterns; solid and cystic (© Aadil Bharadia)

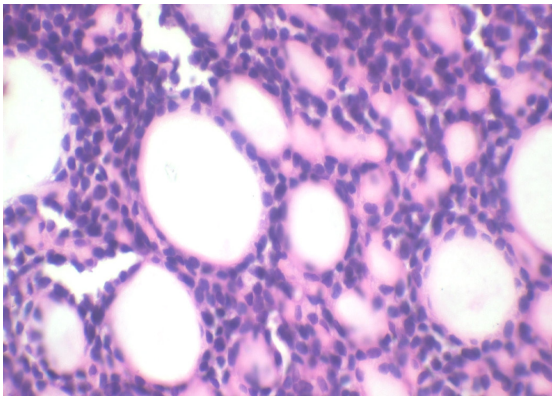

Figure 5: Cribriform pattern; cysts close together were noted (© Aadil Bharadia)

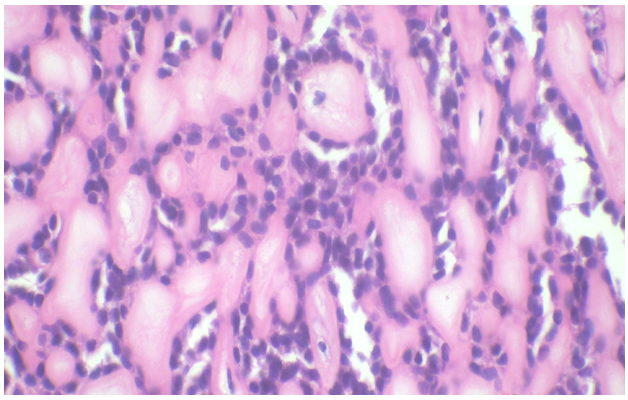

Figure 6: Histology demonstrated small round epithelial cells (© Aadil Bharadia)

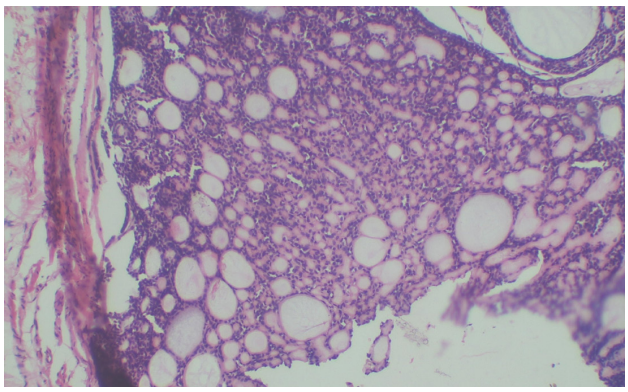

Figure 7: Muscle invasion by the tumour (© Aadil Bharadia)

A diagnosis of a completely excised adenoid cystic carcinoma of the left upper eyelid was made.

The patient was taken for lid reconstruction 7 days later and left eye canthotomy plus cantholyis plus glabella flap was done successfully.

The patient was reviewed and the wound was clean with no ectropion, the patient was satisfied and comfortable. He was discharged for follow-up in the oculoplastics clinic.

Discussion

Adenoid cystic carcinoma (ACC) is a rare type of cancer that originates from the secretory glands, often affecting the salivary glands. It makes up about 1% of all head and neck malignancies. However, it is the most common tumor of the minor salivary glands and the second most common tumor of the major salivary glands(7). In the orbit lacrimal gland tumors comprise of about 10% to 15% of space occupying tumors, adenoid cystic carcinoma has been reported to be the most common form of malignant epithelial lacrimal gland tumor, comprising approximately 66% of malignant lesions(8,9). In the eyelid, it can arise from the glands of Moll, the palpebral lobe of the lacrimal gland, the accessory lacrimal glands in conjunctiva, or from ectopic lacrimal gland tissue but very rare to arise from the eyelid skin(2).

The tumor is typically slow-growing compared to other carcinomas and has a tendency for peri-neural spread in these tumors, this is the most common mode of metastasis and also increases the rate of local recurrence(10). Due to its rarity, there is limited data on risk factors and management of advanced disease. Fewer than 50 cases of adenoid cystic carcinoma primarily of the eyelid skin have been reported(11).

Adenoid cystic carcinoma presents as a painless, firm nodular mass and is associated with loss of cilia in elderly patients(12). It can resemble basal cell carcinoma, sebaceous gland carcinoma, or a chalazion(2).

Complete resection of the tumor via surgery, if possible, is the mainstay for treatment. Adjuvant radiotherapy reduces the risk of local recurrence, especially with perineural invasion, while chemotherapy is an option for metastatic cases(6). Despite good surgical techniques the 5 to 10 year recurrance rate is about 75%(13). Features associated with poor prognosis are positive surgical margins, perineural invasion and positive lymph nodes(14). More than 50% of primary adenoid cystic carcinomas have perineural invasion, which is the most common form of dissemination and raises the risk of local recurrance following excision(15). The similarity with sebaceous gland carcinoma in this case was that it was a slow-growing tumor on the skin of

the upper eyelid which was a painless firm mass on the lid margin associated with loss of cilia which was initially thought to be a chalazion(16). Basal cell carcinoma frequently arises from the lower eyelid. It is also a slow growing, lacally invasive but non-metastasizing. It can present as ulceration and bleeding and usually redish in colour(17). Seemingly benign lid lesions which demonstrate unusual characteristic on examination, investigation, at surgery or recurrence should be biopsied.

Conclusion

Cutaneous adenoid cystic carcinoma of the eyelids remains a very rare entity and requires long-term surveillance.

Malignant eyelid lesions may masquerade as several different clinically benign conditions and all excised eyelid lesions should be submitted for histopathologic confirmation.

Ethical considerations

Patient particulars and facial appearance were not disclosed. The patient gave consent for the photographs to be taken and for publication of this case report.

References

1. Dillon PM, Chakraborty S, Moskaluk CA, Joshi PJ, Thomas CY. Adenoid cystic carcinoma: A review of recent advances, molecular targets, and clinical trials. Head Neck. 2016 Apr;38(4):620–7.
2. Marback EF, Costa AL, Nossa LMB, Marback RL, Rao NA. Eyelid skin adenoid cystic carcinoma: a clinicopathological study of one case simulating sebaceous gland carcinoma. Br J Ophthalmol. 2003 Jan;87(1):118–9.
3. Ellington CL, Goodman M, Kono SA, Grist W, Wadsworth T, Chen AY, et al. Adenoid cystic carcinoma of the head and neck. Cancer. 2012;118(18):4444–51.
4. von Holstein SL, Coupland SE, Briscoe D, Le Tourneau C, Heegaard S. Epithelial tumours of the lacrimal gland: a clinical, histopathological, surgical and oncological survey. Acta Ophthalmol (Copenh). 2013;91(3):195–206.
5. Ronquillo CC, Vegunta S, Ellis N, Mamalis N, Patel BCK. Adenoid Cystic Carcinoma of the Eyelid Can Locally Recur and Mimic Basal Cell Carcinoma: Case Report and Review of the Literature. Ophthal Plast Reconstr Surg. 2020;36(5):e112–6.
6. Am F, Tj B, Ma B, Ja T, Br N. Perineural invasion of cutaneous malignancies. Dermatol Surg Off Publ Am Soc Dermatol Surg Al [Internet]. 2001 Jun [cited 2024 Oct 14];27(6). Available from: https://pubmed.ncbi.nlm.nih.gov/11442588/
7. Din MAU, Shaikh H. Adenoid Cystic Cancer. In: StatPearls [Internet] [Internet]. StatPearls Publishing; 2023 [cited 2024 Oct 14]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK557855/
8. Chen TY, Keeney MG, Chintakuntlawar AV, Knutson DL, Kloft-Nelson S, Greipp PT, et al. Adenoid cystic carcinoma of the lacrimal gland is frequently characterized by MYB rearrangement. Eye. 2017 May;31(5):720–5.
9. Adenoid Cystic Carcinoma of the Lacrimal Gland - EyeWiki [Internet]. [cited 2024 Oct 29]. Available from: https://eyewiki.org/Adenoid_Cystic_Carcinoma_of_the_Lacrimal_Gland
10. Chummun S, McLean NR, Kelly CG, Dawes PJ, Meikle D, Fellows S, et al. Adenoid cystic carcinoma of the head and neck. Br J Plast Surg. 2001 Sep;54(6):476–80.
11. Mencía-Gutiérrez E, Gutiérrez-Díaz E, Ricoy JR, Madero-García S. Primary cutaneous adenoid cystic carcinoma of the eyelid. Am J Ophthalmol. 2001 Feb 1;131(2):281–3.
12. Cavazza S, Laffi GL, Lodi L, Collina G. Primary cutaneous adenoid cystic carcinoma of the upper lid: a case report and literature review. Int Ophthalmol. 2012 Feb;32(1):31–5.
13. Chen AM, Bucci MK, Weinberg V, Garcia J, Quivey JM, Schechter NR, et al. Adenoid cystic carcinoma of the head and neck treated by surgery with or without postoperative radiation therapy: prognostic features of recurrence. Int J Radiat Oncol Biol Phys. 2006 Sep 1;66(1):152–9.
14. van Weert S, Bloemena E, van der Waal I, de Bree R, Rietveld DHF, Kuik JD, et al. Adenoid cystic carcinoma of the head and neck: a single-center analysis of 105 consecutive cases over a 30-year period. Oral Oncol. 2013 Aug;49(8):824–9.
15. Sammour R, Lafaille P, Joncas V, Krasny M, Wong J, Allaire G, et al. Adenoid Cystic Carcinoma of the Eyelid: A Rare Cutaneous Tumor Treated with Mohs Micrographic Surgery. Dermatol Surg. 2009 Jun;35(6):997.
16. Sebaceous Carcinoma - EyeWiki [Internet]. [cited 2024 Oct 14]. Available from: https://eyewiki.org/Sebaceous_Carcinoma
17. Shi Y, Jia R, Fan X. Ocular basal cell carcinoma: a brief literature review of clinical diagnosis and treatment. OncoTargets Ther. 2017 May 8;10:2483.

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