Mucoepidermoid Carcinoma of the Lacrimal Gland: A Case Report

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Abstract

Background: Mucoepidermoid carcinoma (MEC), a tumour previously believed to occur exclusively in the major salivary glands, was atypically recognized in the lacrimal gland. Few ophthalmologists are familiar with this rare neoplasm. **Aim:** This case is being reported for its rarity and the association with human immunodeficiency virus (HIV). **Case Presentation:** High-grade MEC of the lacrimal gland of the right eye was diagnosed in an orbital exenteration specimen of a 60-year-old HIV-positive man. He presented to the eye unit of a mission hospital in Jos, Nigeria with significant visual impairment in the right eye and a rapidly growing painful superolateral orbital mass. The tumour invaded and completely obliterated the anterior chamber. The lens, ciliary body, and iris were displaced posteriorly but were free from tumour infiltration. The tumour was exenterated, and the patient was subsequently referred for adjuvant radiotherapy. No evidence of recurrence or metastasis has been noted 15 months post-surgery. **Conclusion:** Early and decisive surgical intervention in a case of high-grade MEC of the lacrimal gland is crucial in forestalling local invasion as well as distant metastasis. Because of the possibility of local recurrence, adjuvant radiotherapy is required and close follow-up of the patient is warranted.

Keywords: *High grade, HIV, lacrimal gland, mucoepidermoid carcinoma*

Abstrait

Base:

Mucoepidermoid carcinoma (MEC), une tumeur pensée auparavant ne survenir exclusivement que sur les principales glandes salivaires a été étrangement détecté dans la glande lacrimale. Peu d'Opthamologues sont habitués à ce néoplasme.

But:

Ce cas est rapporté pour sa rareté et son association avec le Virus de l'Immunodéficience Humaine (VIH).

Présentation de Cas:

MEC de grade élevé de la glande lacrimale de l'oeil droit a été diagnostiqué sur un specimen d'extenteration orbitale d'un vieil homme de 60 ans, séropositivif. Il présenta à la section des yeux d'un hôpital missionnaire à Jos, Nigeria, un sérieux problème de vision de l'oeil droit et une douleureuse masse orbitale superolatérale poussant très rapidement. La tumeur envahit complètement la membrane/chambre antérieure et l'oblitera. La lentille, le corps ciliaire et l'iris tous déplacés postérieurement étaient épargnés de l'infiltration de la tumeur. La tumeur était exenterisée et le malade recommendé pour une radiothérapie adjuvante. Aucune preuve de resurgence ou de metastasis n'a été enregistrée durant 15 mois d'après chirugie.

Conclusion:

Une intervention chirugicale décisive à temps dans un cas de grade élevé de MEC de la glande lacrimale est essentiel à empêcher l'invasion locale aussi bien que le metastasis éloigné. Vu la possibilité de résurgence locale, une radiothérapie adjuvante est nécessaire autant qu'un sérieux suivi du malade.

Mots-Clés: *Mucoepidermoid carcinoma VIH, glande lacrimale, grade élevé*

Introduction

Lacrimal gland lesions are relatively rare with majority of them being inflammatory.[1,2] Malignant lacrimal gland tumours are even rarer, accounting for approximately one-quarter of all lacrimal gland lesions. Mucoepidermoid carcinoma (MEC) of the lacrimal gland is a rare and aggressive tumour with only a few cases reported.[3-5] Histologically, MEC comprised varying proportions of epidermoid, mucous, intermediate, columnar, and clear cells in varying proportions and has been divided into low, intermediate, and high grade types based on cytological features.[6-8] Low-grade tumours are well-differentiated and

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are made up of over 50% of mucus-secreting elements and squamous epithelial cells.[3] High-grade MECs are aggressive and potentially lethal tumours, poorly differentiated and primarily made up of squamous epithelial and intermediate cells, containing less than 10% of mucus-secreting cells.[4] High grade tumours are characterized by predominant solid cellular proliferations of epidermoid and intermediate cells, with higher degrees of atypia, anaplasia, multiple mitoses, and necrosis when compared with the low and intermediate grades.[6-9] The histologic features of intermediate-grade tumours fall in between those of the low-grade and high-grade tumours.[4]

This case is being reported because of its rarity and lack of documented association with human immunodeficiency virus (HIV) in previous reports.[6]

Case Presentation

In April 2019, a 60-year-old man presented with a rapidly growing mass on the right eye which was associated with pain, tearing, redness, and loss of vision of 4 months duration. There were no other masses in any other parts of his body. He was diagnosed HIV-positive 2 years prior to presentation and had been regular on fumarate/lamivudine/efavirenz (TDF/3TC/EFV) fixed dose anti-retroviral combination therapy. The presenting visual acuity (VA) was perception of light (PL) in the right eye and counting fingers (CF) at a distance of 3 m in the left eye. VA in the left eye improved to 6/24 with refraction. He had a right-sided superotemporal orbital mass that extended inferolaterally. The right globe was inferonasally displaced and frozen in place [Figure 1A-C]. The cornea was hazy, the anterior chamber shallow, and the lens cataractous with no view of the fundus. Examination of other systems revealed no signs of metastasis.

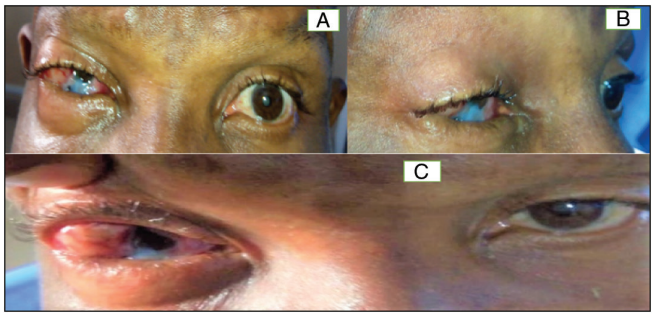

Figure 1: (A-C) A right superotemporal mass with globe infiltration and inferior-nasal displacement

Computer tomography (CT) scan of the brain could not be done due to financial limitation. A plain skull X-ray showed an ill-defined orbital mass in the right orbit with preservation of orbital lines [Figure 2]. Orbital ultrasound of the right globe revealed a mass on the superior and inferior lateral aspects of the globe with globe compression and invasion. At presentation, he had Centers for Disease Control (CDC) stage 2 HIV disease with a CD4 count of 442 cells/mm.[3] A pre-operative diagnosis of advanced conjunctival squamous cell carcinoma with ocular/orbital invasion was made.

A lid-sparing orbital exenteration was performed 3 weeks after presentation, which revealed a well-circumscribed mass firmly attached to a distorted globe [Figure 3]. Immediate post-operative course was uneventful and by 4 weeks thereafter he was referred for adjuvant radiotherapy. Subsequently, the patient was followed up every 2 months.

Histopathological examination of the specimen revealed a well-circumscribed tumour measuring 2.5 × 2 × 1.5 cm, greyish white, firm with tiny cystic spaces. It had a thin fibrous capsule overlying distinct nests of epidermoid, intermediate, clear, and mucinous cells. The malignant epithelial cells showed moderate-to-marked pleomorphism, prominent nucleoli, occasional mitosis, and increased nucleocytoplasmic ratio [Figures 4–6]. A cut section of the globe showed a distorted globe with a dumb-bell tumour extending from the conjunctiva into the globe. The tumour displaced the lens and ciliary body posteriorly but did not infiltrate these structures. The optic nerve, periorbital fat, and adjoining tissues were free of tumour. Lymphovascular invasion was not identified. A diagnosis of high-grade MEC (pT2NxMx), most likely of lacrimal gland origin, was made according to the 8th American Joint Committee on Cancer Staging System of salivary gland MEC.[10]

By the 7th month after surgery, the patient was yet to receive the recommended adjuvant therapy as he was unable to afford the cost of radiotherapy. Following further deterioration in vision of the left eye with maturation of the cataract, he had a manual small incision cataract extraction and a posterior chamber

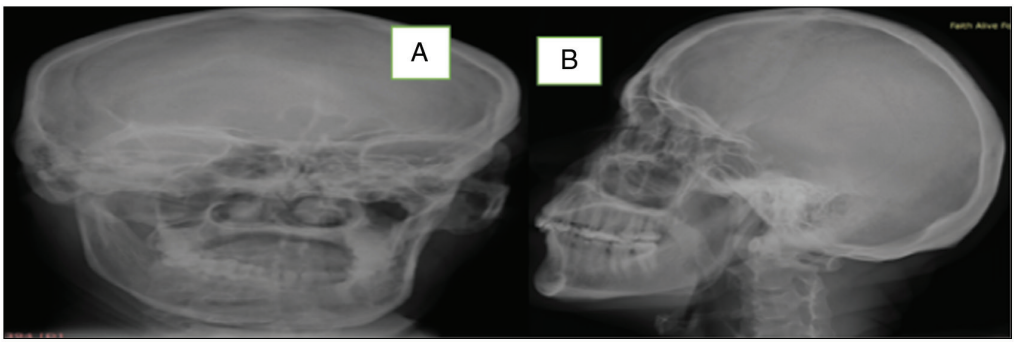

Figure 2: Plain skull X-ray; AP (A) and lateral (B) showing an ill-defined right orbital mass with distinct orbital lines

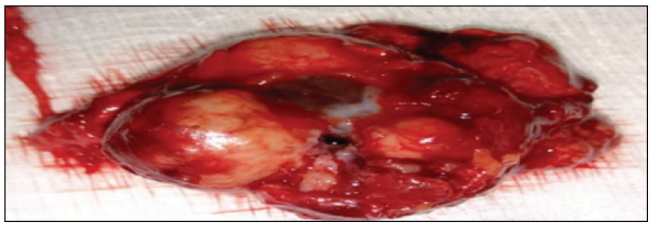

Figure 3: Right exenteration specimen

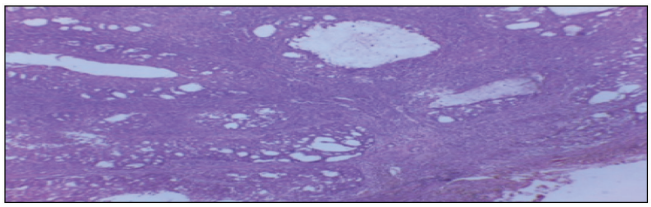

Figure 4: Hematoxylin and eosin staining showing tumour mass with ductal spaces (×4)

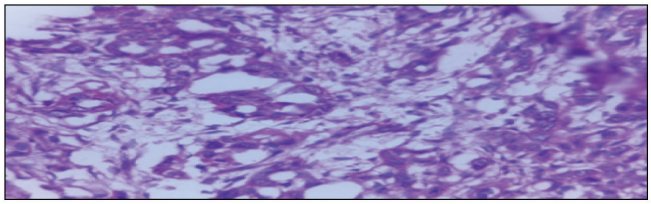

Figure 5: Hematoxylin and eosin staining showing tumour mass with extravasated mucin and duct formation lined by pleomorphic mucous cells (×40)

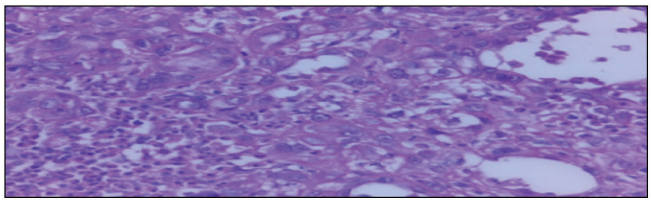

Figure 6: Hematoxylin and eosin staining showing pleomorphic epidermoid cells, intermediate cells, and clear cells (×40)

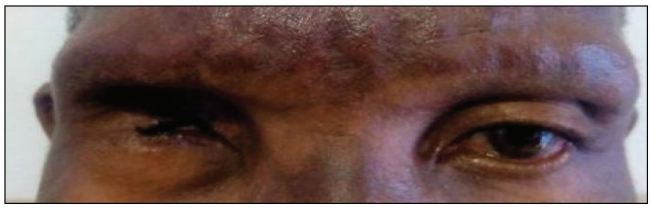

Figure 7: Healthy right orbit 15 months post-primary surgery

intraocular lens implantation with an early post-operative VA of 6/12 and 6/6 at 6 weeks and had been on 3-monthly review. At 15 months following the primary surgery, there was no clinical evidence of local recurrence [Figure 7] or distant metastasis and the patient is still yet to receive adjuvant radiotherapy.

Discussion

MEC is the most common primary epithelial tumour of the salivary gland in both children and adults.[6,7] However, it is a rare finding in the ocular region, where it can arise in the conjunctiva, lacrimal gland, or lacrimal sac.[11,12] The lacrimal gland is considered to be a minor salivary gland that shares histologic features with the major salivary glands.[13] MEC of the lacrimal gland has been reported to account for between 3.6% and 4% of all malignant epithelial lacrimal gland tumours.[3,9,11,14] This report is peculiar in that the presentation is in an HIV patient who is also on regular antiretroviral therapy.

Some literatures report that the peak age for MEC is the third and fourth decades of life.[6,11] Although our patient does not fall within this peak age for MEC, he is within the 10–73 years age range for lacrimal gland tumours as noted by Andreoli *et al*.[14]

Chemical carcinogens and oncogenic viruses have not been linked with MEC as is the case with some other malignancies, but prior exposure to ionizing radiation has been implicated as a risk factor.[6] Although the history of previous exposure to ionizing radiation could not be established in our patient, the possibility that HIV played a role in the aetiology and/or the course of MEC in this case cannot be disregarded. Data from a review of carcinomas arising from the head and neck region suggest that there is a rise in the rate of these tumours among HIV-positive patients in whom they present with more aggressive disease and worse prognosis when compared with those who are HIV-negative.[15] Immunosuppression and oncogenic viruses have been linked to the rising cases of malignancies among HIV patients, despite the advent of highly active retroviral therapy.[15,16] An increase in the rate of salivary gland tumours has been well established in patients with HIV.[15] Although the relationship between CD4 count and ocular surface squamous neoplasms (OSSNs) in HIV patients is yet to be clearly understood, Rathi *et al*.[16] noted that majority of HIV-positive patients with OSSN were significantly immunosuppressed at the time of tumour detection with a CD4 count <200 cells/mm^3. Furthermore, it has been observed that majority of the head and neck tumours in HIV-positive patients are AIDS defining.[15] In keeping with this finding, the diagnosis of MEC in our patient was AIDS defining, moving the patient from symptomatic HIV stage (B2) to AIDS stage (C2), despite the fairly good CD4 count at presentation. We did not come across a reported case of MEC of the lacrimal gland in HIV patients in our literature search. Hence, the implication of HIV in the aetiology, stage, grade, and prognosis of MEC in our patient remains unclear.

The presentation of our patient with a unilateral tumour in the superotemporal orbit associated with pain, inferior media dystopia, and extra-ocular motility defect is in keeping with previous reports.[3,12] Some patients have also reported the presence of proptosis and diplopia.[3]

The diagnosis of MEC can only be confirmed histologically, but supportive investigations such as orbital ultrasound scan, CT scan, and magnetic resonance imaging are necessary

for determining the extent of intraocular/orbital invasion, detection of bony involvement and metastasis, and for planning the extent of surgical intervention.[4,5,12] We had to depend exclusively on findings of orbital ultrasound scan and plain X-rays of the skull, with their diagnostic limitations, due to financial constraints. These challenges highlight some of the everyday difficulties we face in the management of ocular malignancies in our practice.

High-grade MEC of the lacrimal gland can metastasize to the lung, brain, parotid gland, and mediastinum.[4] Recurrence is also a common feature of high-grade MEC, reported to be as high as 70–100% within the first 6 months of follow-up.[11] For this reason, high-grade MEC needs careful exenteration, resection of the involved orbital bone, and radiation.[4] Chemotherapy becomes necessary only when metastasis has been established.[3] Patients are more likely to experience a recurrence if the margins of resection are positive, regardless of the tumour grade. It is expected that despite our patient's tumour being high grade, the fact that there was no histological evidence of posterior segment, neural, or vascular infiltration makes the chance of recurrence low to negligible.

Limitation

Immunohistochemistry which would have been useful for further characterization of the tumour could not be done due to financial constraints.

Conclusion

Early and decisive surgical intervention in a case of high-grade MEC of the lacrimal gland may be crucial in forestalling local invasion as well as distant metastasis. Because of the possibility of local recurrence, adjuvant radiotherapy is required and close follow-up of patients is warranted.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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