Mucinous carcinoma of the eyelid: a case report

Adan AA, Jelle N

Department of Surgery, College of Health Sciences, University of Nairobi, P.O. Box 76239-00508, Nairobi, Kenya. Email: 1313dr@gmail.co

Corresponding author: Dr. Najma Jelle, P.O. Box 76239-00508, Nairobi, Kenya. Email: jnajma99@gmail.com

ABSTRACT

Primary cutaneous mucinous carcinoma is a rare malignant tumour which arises from the eccrine sweat glands most commonly in the head and neck region and more so in the eyelid. Ever since the first description by Lennox *et al.* in 1952, most of the information available on this rare malignant tumour with an incidence of 0.1 cases per a million individuals is from a few published case reports. Due to its slow growing nature, it is often misdiagnosed as other benign lesions of the eyelid. Diagnosis is made through histological analysis which reveals malignant epithelial cells surrounded by periodic acid Schiff positive mucin. Management is through excision of the tumour and due to its high recurrence rate, excision should be done with a wide margin of healthy tissue followed by reconstruction of the eyelid. We describe the case of a 75-year-old Kenyan Caucasian man who presented with a slow growing mass on the left lower eyelid for 10 years that was excised twice before and recurred. A diagnosis of mucinous carcinoma of the eyelid was made based on histology. The patient was managed successfully through excision of the mass with a 3mm margin of healthy tissue followed by reconstruction of the eyelid using a Mustarde flap and mucoperichondrial flap from the inner lining of the nasal septum to reconstruct the inner lining of the new eyelid.

Key words: Primary cutaneous mucinous carcinoma of the eyelid, Malignant tumour, Recurrence, Mustarde flap

INTRODUCTION

Primary Cutaneous Mucinous Carcinoma (PCMC) is a malignant and rare skin appendage tumour which arises from the sweat glands[1]. Whether this tumour has its origin from the eccrine sweat glands or apocrine glands has been a contested topic of discussion but it is mostly published as originating from the eccrine sweat glands[2]. Mucinous carcinomas of the skin mostly originate in the head and neck region with the eyelid as the most common site of occurrence[3]. An analysis of 289 PCMC cases of the head and neck region reported between the years 2004 and 2016 revealed that women are most commonly affected[3]. However, mucinous carcinoma specifically originating from the eyelid is more common in male patients with a mean age of occurrence at 61.3 years[4].

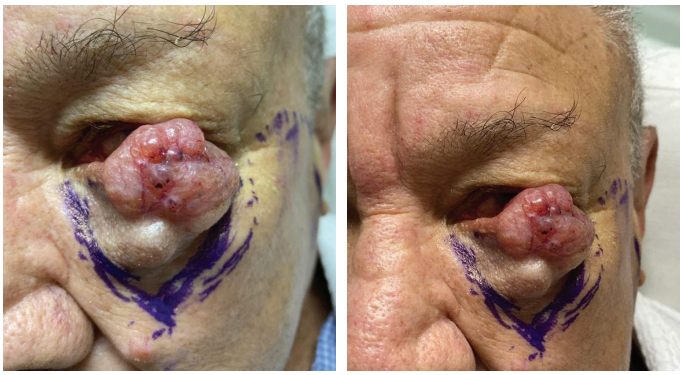
Mucinous carcinoma originating from the eyelid is slow growing and has an indolent course with a low metastatic potential, however, it is locally aggressive with a high recurrence rate[5]. Due to its slow growing nature, most cases of this malignant tumour are often misdiagnosed with variable initial diagnoses such as chalazion, myxoma, and epidermoid cyst prior to resection and histology[6]. Even though the tumour seems to have an excellent prognosis, it is important to have it as a differential diagnosis for eyelid lesions to ensure early diagnosis and management to increase the chance of optimal aesthetic outcome post excision and reconstruction of this sensitive region[3]. In this case report, we describe the case of a 75-year-old male Kenyan patient who presented with a 10-year history of a growth on the left lower eyelid which was excised two times before this current presentation. Histology revealed a diagnosis of mucinous carcinoma of the eyelid.

CASE REPORT

A 75 year old male patient presented to a private surgical facility specializing in plastic, aesthetic and reconstructive surgery with a history of a growth on the left lower eyelid which he first noticed 10 years ago. It began as an itch over the left lower eyelid which developed into a small mass gradually increasing in size. First excision of the mass was done 5 years prior but the mass recurred a year later. Second excision was done one year before this current presentation at the same center but the mass recurred a few months later, bigger in size according to the patient with associated ectropion and epiphora. The patient did not have a histological report of the mass from the previous excisions. The patient is a known hypertensive patient on antihypertensive medication and a history of gout on allopurinol. He used alcohol occasionally and stopped smoking 15 years before. Review of other systems was unremarkable.

On examination, there was a fungating mass measuring 2cm by 2cm and covering almost the entire length of the left lower eyelid but did not encroach into the conjunctiva (Figure 1). There was associated ectropion and redness of the left conjunctiva. The general as well as systemic examination findings were normal.

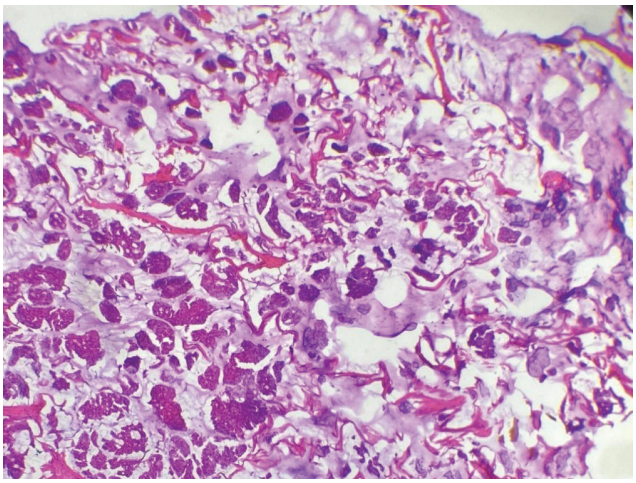
Figure 1: Fungating mass on the left lower eyelid diagnosed as mucinous carcinoma of the eyelid after histology



Investigation and diagnosis

Investigations done included full haemogram, uric acid levels, liver function tests and renal function tests which showed no abnormalities. Echocardiography was done that concluded aortic valve sclerosis with no gradient or regurgitation. Electrocardiography done revealed incomplete right bundle branch block but an otherwise normal ECG. A section of the mass was excised and histology done which showed skin bearing mucinous carcinoma characterized by malignant epithelial cells bathed in lakes of mucin with margins positive for tumour (Figure 2). A diagnosis of malignant skin appendage tumour consistent with mucinous carcinoma of the eyelid was thus made. Based on the histology findings and diagnosis, a decision to excise the mass and reconstruct the lower eyelid was made.

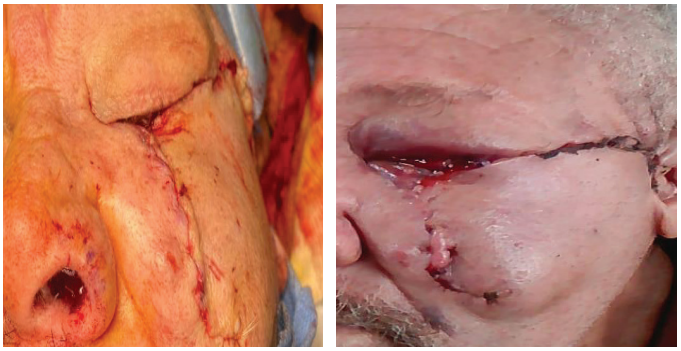
Figure 2: Magnification ×10, histology of the resected tumour showing pools of extracellular mucin and nests of malignant cells. The arrow is pointed at a lake of extracellular mucin



Management

Excision was done with a 3mm margin of healthy tissue. Mustarde flap was used in the reconstruction of the lower eyelid and a mucoperichondrial flap harvested from the inner lining of the nasal septum and used as the inner lining of the new eyelid. During the post operative review before discharge, the patient was doing well with no ectropion and was able to close the eyelid well.

Figure 3: Immediate post-operative image after excision of the tumour with 3mm margin of healthy tissue and reconstruction of the eyelid using a Mustarde flap



DISCUSSION

Primary Cutaneous Mucinous Carcinoma (PCMC) was first described by Lennox *et al.* back in 1952[7]. Since the first description in literature, most of the knowledge available on PCMC is found from case reports, series and literature reviews. PCMC is a rare malignant tumour which originates from the eccrine sweat glands. It occurs mostly in the head and neck region with the eyelid as the most commonly affected site[3]. PCMC of the eyelid is most often missed and misdiagnosed as other benign tumours due to its slow growing nature. These tumours have a low metastatic potential but unpredictable locally infiltrative growth and a high rate of recurrence[8,9]. They most often present as painless slow growing masses that sometimes may ulcerate and recur especially if wide excision with margin control was not done[10]. As seen in this case, the mass had recurred twice in our patient after excision.

Most of the patients with mucinous carcinoma present between the ages 50-70 years with eyelid carcinomas having a slightly higher male predominance[11,12]. The prevalence is higher in Caucasian patients as compared to Asians and African populations. It is important to be aware of mucinous carcinoma of the eyelid and have it as a differential diagnosis for benign appearing tumours of the eyelid. Diagnosis is made through excisional biopsy and histological analysis of the mass characterized by malignant epithelial cells surrounded by periodic acid-Schiff positive mucin[13]. Further investigation with immunohistochemical staining for CK7 and CK20 can be done with most tumours staining positive for CK7 and negative for CK20[6]. While metastasis is rare, it is possible and the most common site of metastasis is to the lymph node, so it is also important to do a proper and thorough physical examination of the patient. However, in most cases further investigations such as CT scan to rule out metastasis is not needed.

After diagnosis and when making a decision on the choice of management, consideration should be given to preventing recurrence as well as optimizing cosmetic outcomes for the patient. Management options include wide local excision of the tumour followed by meticulous reconstruction or Mohs angiographic surgery with frozen section control to minimize the amount of healthy tissue excised[14]. In our case, a decision was made to excise the mass together with a 3mm margin of healthy tissue to prevent recurrence. Reconstruction was done with a Mustarde flap to reconstruct the eyelid and a mucoperichondrial flap from the nasal cavity to reconstruct the inner lining of the eyelid.

CONCLUSION

Mucinous carcinoma of the eyelid is slow growing which makes it often misdiagnosed as other benign lesions of the eyelid. It is important for physicians to have mucinous carcinoma as a differential diagnosis for eyelid tumours as the tumour is locally infiltrative and has a high rate of recurrence. Management poses a challenge as to finding the balance between wide excision of the tumor while also preventing adverse cosmetic outcomes for the patient.

Declaration

Ethical consideration: Consent to publish these findings and use the photos was obtained. However, no personal identifiers have been used in the report and the images have been anonymized.

Conflict of interest statement by authors: None.

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