

Primary mucinous adenocarcinoma of the eyelid- a rare tumor**Sivagurunathan Premala-Devi^{1,2}, Raja-Omar Raja-Norliza¹, Jalaluddin Juliana¹, Embong Zunaina^{2,3}**¹Department of Ophthalmology, Hospital Besar Melaka, Melaka, Malaysia.²Department of Ophthalmology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia.³Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia.***Corresponding author**

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Abstract: Primary mucinous adenocarcinoma is a rare, mucin-secreting adnexal neoplasm. We report two cases of successfully treated primary mucinous adenocarcinoma. The first patient is a 66-year-old gentleman presented with painless, left upper eyelid mass for 6 months duration. Histopathological report of the excisional biopsy revealed mucinous adenocarcinoma with margins close to tumour (< 1 mm). Patient was subsequently referred for radiotherapy but he declined. Till date there is no local recurrence. The second patient is a 55-year-old Malay gentleman presented with painless, left lower eyelid mass for 3 years duration. An excisional biopsy was performed and the histopathological report showed mucinous adenocarcinoma with involvement of surgical margins. Subsequently he underwent a repeat excisional biopsy. Full oncological screen excluded presence of primary mucinous carcinoma elsewhere and of any metastatic spread in the two patients.**Keywords:** primary tumour, mucinous adenocarcinoma, eyelid mass.

INTRODUCTION

Primary mucinous adenocarcinoma is a mucin secreting adnexal neoplasm of rare pathologic entity. This tumour was first described in 1952 by Lennox [1]. Since then, there has been 25 reports describing 55 patients with primary mucinous adenocarcinoma of the eyelid in the medical literature and mostly as case reports [2]. Of all these patients, only 2 patients were of Asian origin (5.7%) [2]. Though this tumour has low metastatic potential, it does have a significant recurrence rate. We present the occurrence, clinical, histological features, and management of this tumour in 2 Malay patients.

CASE REPORT**Case 1**

A 66-year-old Malay gentleman with underlying diabetes and hypertension presented with left upper eyelid mass for 6 months duration. The mass was painless, and progressively increasing in size. This was the first presentation. He had no similar mass elsewhere. He had no other systemic manifestation and no family history of malignancies.

On examination, there was a 2 cm x 3 cm, non tender, nodular, mobile mass palpable on the left upper eyelid. The mass resembled a benign granuloma. On eye examination, visual acuity was 6/9 in both eyes. Anterior and posterior segment examination was normal in both eyes and did not show any other mass. On

systemic examination, there was no lymphadenopathy or hepatosplenomegaly. The provisional diagnosis at this point of time was benign cystic mass or epidermoid cyst. Patient subsequently underwent excisional biopsy. Histopathological report revealed cohesive clusters and islands of malignant epithelial cells floating in the lakes of mucin which corresponded to the diagnosis of mucinous adenocarcinoma with margins close to the tumour (< 1 mm)(Figure 1).

Due to the close surgical margins, patient was referred for radiotherapy but he declined. Full oncological screen which include tumour markers and computerized tomography (CT) scan excluded presence of primary mucinous carcinoma elsewhere and of any metastatic spread in this patient. Till date, there is no local recurrence.

Case 2

A 55-year-old Malay male with underlying diabetes presented with a mass at the lateral part of the left lower eyelid for 3 years duration. The mass was painless and was gradually increasing in size. He had no similar mass elsewhere in his body and this was the first presentation. Patient had no other ocular or systemic manifestation and no family history of any malignancies.

On examination, there was a mass located at the lateral part of the left lower eyelid measuring 2 cm x

2 cm (Figure 2A). It was firm, non tender with irregular surface. There were no underlying skin changes. Transillumination test was positive. On eye examination, visual acuity was 6/9 in both eyes. Anterior and posterior segment examination was normal in both eyes and did not show any other mass. On systemic examination, there was no lymphadenopathy or hepato-splenomegaly. He was diagnosed as having epidermoid cyst. An excisional biopsy was performed and showed a well-encapsulated mass (Figure 2B).

Histopathological report showed malignant epithelial cells arranged in cords and islands surrounded by abundant extracellular mucin which corresponded to

the diagnosis of mucinous carcinoma (Figure 3). Immunohistochemical stains for neoplastic cells were positive for cytokeratin (CK) 7 but negative for CK20, prostate-specific antigen (PSA), CDX2 and carcinoembryonic antigen (CEA). However, there was involvement of the surgical margins.

Subsequently he underwent a repeat excisional biopsy which yield similar findings and free of surgical margins. Full oncological screen excluded presence of primary mucinous carcinoma elsewhere. Review at one year post excision showed no recurrence of the tumour at excision site (Figure 4) and there was no metastatic spread.

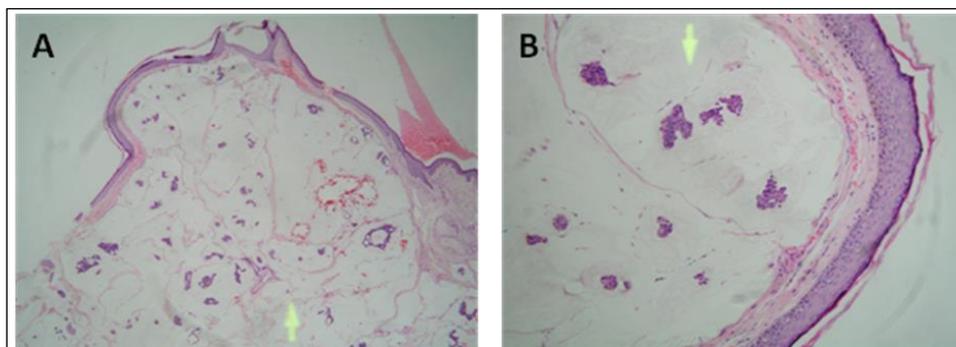


Fig-1: Histopathology specimen of left upper eyelid mass in case 1 showed floating clumps of malignant epithelial cells in the abundant mucin lakes (arrow) (A: lower magnification, B: higher magnification).

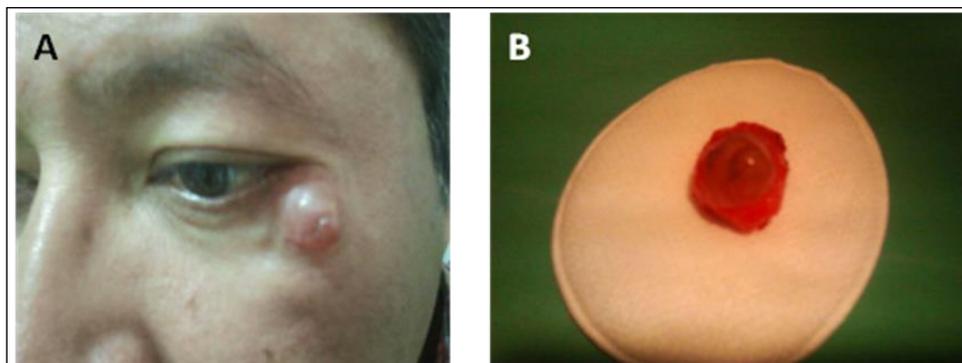


Fig-2: Mass located at the lateral part of the lower eyelid at presentation (A). Gross pathological appearance showed a well-encapsulated mass after excision (B).

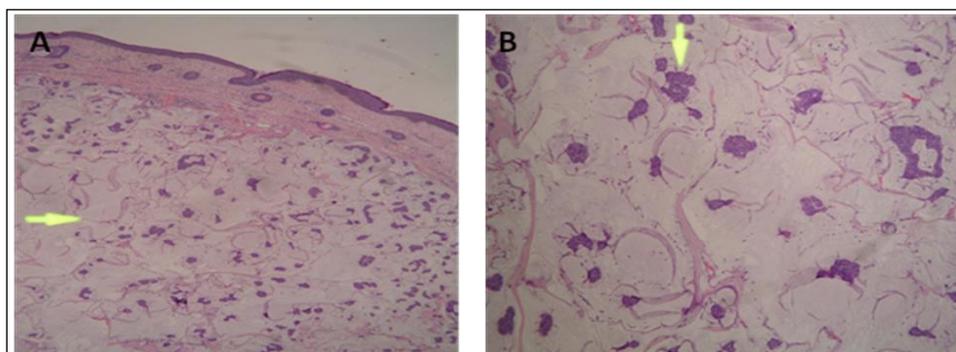


Fig-3: Histopathological specimen of left lower eyelid mass in case 2 showed nests of malignant epithelial cells within mucin matrix (arrow) (A: low magnification, B: high magnification).



Fig-4: Left lower eyelid showed no recurrence of the tumour at one year post excision.

DISCUSSION

Primary mucinous adenocarcinoma of the eyelid is a rare clinical entity. It's a rare subtype of a sweat gland tumour first described by Lenox et al in a paper published in 1952 [1]. Mucinous carcinoma most commonly arises in the head and neck and the eyelid is regarded as being the most common site [3]. It can arise from eccrine or apocrine glands. Studies showed that this tumour is more prevalent in men than women with a ratio of 2:1 and tends to occur in elderly individuals with the mean age of 62 years [2]. This eyelid tumour is usually unilateral although bilateral cases have been reported [4]. Mucinous carcinoma usually has no distinctive features and usually appears as solitary, slow growing mass with either smooth, irregular or crusted surface. The lesion can range from 3 mm to 20 mm in size [4]. In view of the variable clinical appearance, the diagnosis can include benign lesions such as chalazion, epidermoid cyst, lipoma, papilloma to malignant conditions such as sebaceous carcinoma, basal cell carcinoma, or adenoid cystic carcinoma [5].

Diagnosis of mucinous adenocarcinoma is usually made and confirmed with histopathological report as these tumours have distinctive histochemical and ultrastructural features. The tumor is composed of small, irregular clusters of tumour cells around ductal lumen in mucinous stroma. Distinguishing primary neoplasm from metastatic carcinomas can be difficult. Therefore organ specific immunostaining profiles with multiple markers can be done to exclude metastatic tumours which commonly arise from gastrointestinal tract, lung, prostate, and thyroid. Although these markers may be helpful in differentiating primary from metastatic adenocarcinoma, a full workup to rule out metastatic tumours is necessary in all cases of primary mucinous adenocarcinoma.

The treatment of choice for primary mucinous adenocarcinoma is wide local excision. The excision

should be done with at least 1 cm free of surgical margins in view of significant risk of local recurrences [1,6]. Mohs micrographic technique is the other alternative. These tumours have locally invasive natural history and there is high risk of local recurrence despite excision (29.4%) but rate of metastasis is low (9.6%) [3]. Following excision, continued follow up is required to monitor for recurrence.

CONCLUSION

Early recognition and early treatment of mucinous adenocarcinoma of the eyelid provides good outcome for patients. However, since this tumour has high risk of local recurrence, therefore close monitoring and follow up is vital.

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