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A Rare Case Report of Primary Cutaneous Mucinous Carcinoma of Eyelid in Seventy-Nine Years Old Male

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Abstract: *79 years old male presented to ophthalmology OPD for evaluation of a painless, progressive, superficial nodular lesion over his left upper eyelid for last 2 years. 5 years back he had similar lesion at same site which was excised after 2 years. The swelling recurred within 1 year and presented as painless, gradually progressive well-defined, irregularly marginated nodular lesion over the left upper eyelid. The overlying skin was freely mobile and there was no regional lymphadenopathy. Patient underwent excisional biopsy with 5 mm margin under local anaesthesia. Histopathology revealed a dermal tumor composed of pools of extracellular mucin and tumour cell islands having mildly pleomorphic nuclei floating into it consistent with the diagnosis of mucinous carcinoma. Primary mucinous carcinoma of the skin is a rare subtype of sweat gland tumour. Local recurrence occurs frequently following excision, but the rate of metastasis is low and most metastases are to regional lymph nodes. Treatment is wide local excision with at least 1 cm margins. This case brings forth a rare skin adnexal tumor involving the upper eyelid. Surgeons and ophthalmologists should be aware of this tumor in the periocular region and should consider these carcinomas in the differential diagnosis of cystic/solid eyelid lesions even though they appear benign on clinical course.*

Keywords: *Lymphadenopathy, Pleomorphic nuclei, Mucinous carcinoma, Local excision, Adnexal tumor.*

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I. Introduction

Primary mucinous carcinoma of the skin (MCS) is a rare subtype of sweat gland tumour. [1] While some debate exists as to the apocrine or eccrine origins of this tumour, most authors favor eccrine differentiation based on evidence obtained from immunohistochemical studies and electron microscopic ultra structural analysis. Most commonly it arises in the head or neck, with the eyelid being the most common site. [1] Men are more affected than women in a 2:1 ratio and it tends to occur in more elderly individuals (average age 62 years, range 34-84 years). [2]

Primary mucinous carcinoma of the skin typically has an indolent course. Local recurrence occurs frequently (29.4%) following excision, but the rate of metastasis is low (9.6%) and most metastases are to regional lymph nodes. [3] Primary MC has distinctive histochemical and ultrastructural features. The tumour is composed of small, irregular clusters of tumor cells around a ductal lumen in mucinous stroma. The tumor cells have a centrally placed, cuboidal nucleus and eosinophilic cytoplasm with little mitosis. [4] Mucin production is consistent with retained cellular function and an indication that the tumor is well-differentiated. Additionally, mucinous carcinomas are typically avascular, a factor that helps explain their low rate of metastasis. [5] The mucin is diastase-resistant, periodic acid Schiff-positive, hyaluronidase-resistant and alcian blue-positive (pH 2.5). [6] This histochemical profile is consistent with the presence of a non-sulfated mucoprotein, most likely sialomucin. Indeed, in the largest series on the subject described by Kazakov *et al.* [7] authors conclude that primary cutaneous MCs span a morphologic spectrum compatible with their mammary counterparts. It is recognized that distinguishing primary cutaneous adnexal neoplasms from metastatic carcinomas can be difficult and hence organ-specific immunostaining profiles [8] using multiple markers can be used with high sensitivity, specificity, and positive predictive value in detecting primary adenocarcinomas. Although multiple markers may help to differentiate primary MC from metastatic adenocarcinomas, histologic and immunohistochemical findings [9] of the two forms tend to overlap, and therefore, a careful workup to rule out metastatic tumors is necessary in all cases of primary cutaneous MC. To this end an extensive search for other possible primary site was done with relevant imaging procedures. [10]

Treatment for primary MC of the skin is wide local excision and since there is a significant risk of local recurrence, it has been recommended that the excision be done with at least 1 cm margins. Some authors have suggested Moh's micrographic technique as an alternative to wide local excision. [11] It is also recognized that

these tumors have a locally invasive natural history and there is a high risk of local recurrence despite Moh's surgery. [12] In our case as the upper eyelid was involved into entirety, such wide margins could not be possible. Nevertheless, the 5 mm margin we could achieve was sufficient to have clear lateral margins on histopathology. Opinion is divided over the use of adjuvant radiotherapy with some favoring it, while others not favoring it. We have referred this patient to oncologist for further management.

This case brings forth a rare skin adnexal tumor involving the upper eyelid. Surgeons and ophthalmologists should be aware of this tumor in the periocular region and should consider these carcinomas in the differential diagnosis of cystic/solid eyelid lesions even though they appear benign on clinical course.

II. CASE REPORT

A 79 years old male presented to Ophthalmology OPD of Agartala government medical College for evaluation of a painless, superficial nodular lesion over his left upper eyelid that had slowly grown over the course of approximately 2 years, to measure 4.3 x2.5 x 1.3 cm. Patient gave history of a similar swelling at the same site which had appeared in 2012 and after a similar slow, painless progressive increase in size, it had been excised in 2014. The histopathology report from the previous surgery was not available for review. The swelling recurred within 1 year and displayed as painless, gradual, progressive increase in size till he presented at our centre. On examination, he had a well-defined, irregularly marginated nodular lesion over the left upper eyelid [Fig. 1].

Fig 1: *Well-defined, irregularly marginated nodular lesion over the left upper eyelid*

The overlying skin was normal in appearance and freely mobile over the underlying nodular lesion. There was no regional lymphadenopathy. The lesion appeared free from the underlying orbital ridge. The lesion was excised with 5 mm margins under local anaesthesia. The resultant defect involved the entire upper eyelid. 2 weeks postoperatively, there was marginal ectropion and lateral tissue sag [Fig. 2].

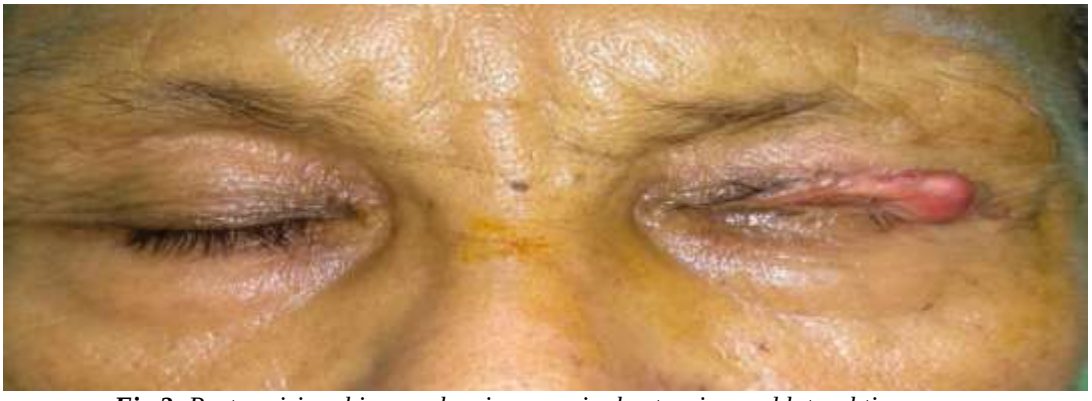

Fig 2: *Post excisional image showing marginal ectropion and lateral tissue sag.*

Gross pathology revealed a soft glistening mucoid tumour measuring 4.3 × 2.5 × 1.3 cm. Microscopic examination revealed a dermal tumor composed of pools of extracellular mucin and tumour cell islands having mildly pleomorphic nuclei floating into it consistent with the diagnosis of mucinous carcinoma [Fig. 3]. The tumour reaches up to inked base. Closest skin cutting and epidermis was free of tumour.

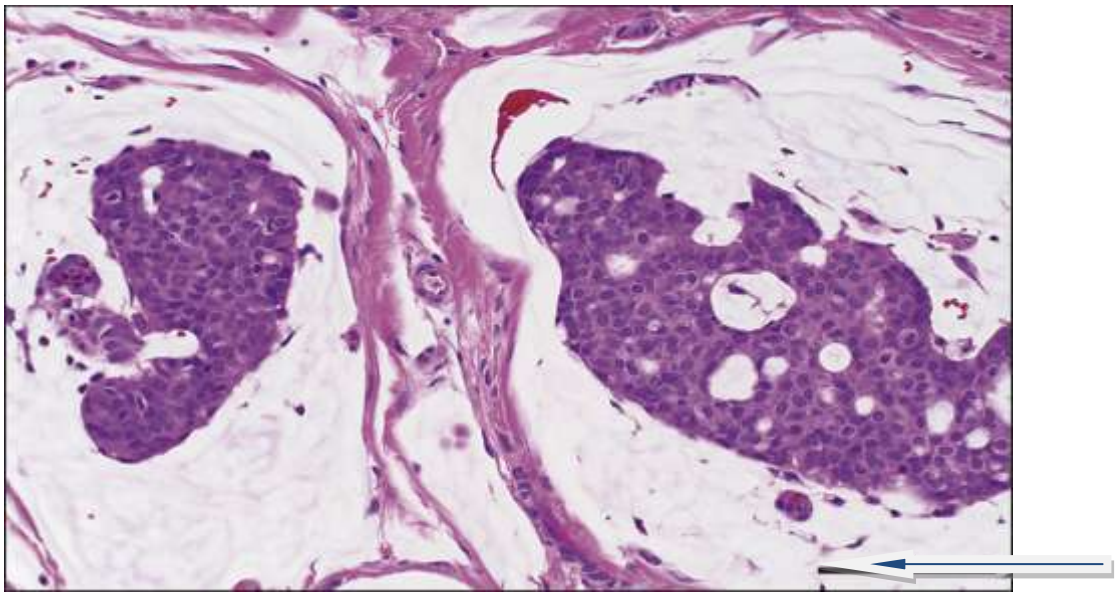

Fig 3: *Histopathology showing dermal tumor composed of pools of extracellular mucin and tumour cell islands having mildly pleomorphic nuclei (Arrow).*

III. Discussion And Conclusion

79 years old male presented to ophthalmology OPD for evaluation of a painless, progressive, superficial nodular lesion over his left upper eyelid for last 2 years. 5 years back he had similar lesion at same site which was excised on the same year. The swelling recurred within 1 year and presented as painless, gradually progressive well-defined, irregularly marginated nodular lesion over the left upper eyelid. The overlying skin was freely mobile and there was no regional lymphadenopathy. Patient underwent excisional biopsy with 2 mm margin under local anaesthesia. Histopathology revealed a dermal tumor composed of pools of extracellular mucin and tumour cell islands having mildly pleomorphic nuclei floating into it consistent with the diagnosis of mucinous carcinoma. Primary mucinous carcinoma of the skin is a rare subtype of sweat gland tumour. These are low-grade tumors. Local recurrence occurs frequently following excision, but the rate of metastasis is low and most metastases are to regional lymph nodes. Treatment is wide local excision with at least 1 cm margins. Morbidity in these tumors is associated with incompletely resected lesions, and tumors located at the inner canthus may be especially difficult to excise completely. Rarely, primary cutaneous mucinous carcinoma of the eyelid will metastasize to preauricular or submandibular nodes. Paramount in the clinical evaluation of patients with cutaneous mucinous carcinoma are a detailed history, physical examination, and appropriate radiographic studies to eliminate the possibility of a visceral mucinous tumor (mainly from the breast and intestinal tract) that may have manifested with cutaneous metastasis. This case brings forth a rare skin adnexal tumor involving the upper eyelid. Surgeons and ophthalmologists should be aware of this tumor in the periocular region and should consider these carcinomas in the differential diagnosis of cystic/solid eyelid lesions even though they appear benign on clinical course. Due to the difficult clinical diagnosis of this often benign appearing lesion, it is imperative that physicians send all specimens for histopathological and immunohistochemical correlation.

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