

Case Report

Sebaceous Carcinoma of Eyelid: A Rare Cutaneous Malignancy with Brief Review of Literature

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ABSTRACT

Sebaceous gland carcinoma (SGC) in the eyelid is a malignant and relatively uncommon tumor accounting for less than 1% of all cutaneous malignancies. It is a slow growing tumor with a multifocal origin and predilection for upper eyelid. High recurrence rates and tendency for intra-epithelial spread, loco-regional and distant metastases makes it important to recognize SGC and consider it as a differential diagnosis in an eyelid lesion. We report a case of 66 year old female with SGC, where lesion recurred after excision. Early diagnosis of SGC, institution of aggressive primary treatment and follow up is of paramount importance to prevent its recurrence.

Key words: Carcinoma, epithelial membrane antigen, eyelid, sebaceous gland.

INTRODUCTION

Sebaceous glands are seen abundantly in the face and scalp region although found throughout all skin sites except for the palms and soles. Sebaceous gland proliferations include senile sebaceous gland hyperplasia, sebaceoma, sebaceous adenoma and sebaceous gland carcinoma. Sebaceous gland carcinoma (SGC) is a rare and aggressive cutaneous carcinoma accounting for less than 1% of all cutaneous malignancies. ⁽¹⁾ It occurs most frequently on the eyelids, where it comprises 4.7% of malignant epithelial tumors. ⁽²⁾

CASE REPORT

A 66 year old female presented with a history of swelling over the right upper eyelid since 1 year, insidious in onset and gradually progressive. There was no significant family history. The patient

underwent mass excision and lid reconstruction but it recurred after three months. O/E, a well defined, firm, nodular 8x3mm painless mass noted over the right upper eyelid. Rest of the ophthalmic examination was normal. There was no lymphadenopathy or organomegaly. An upper lid modified cutler beard surgery with a scleral graft was performed. Grossly, the external skin surface showed a polypoidal growth measuring 0.5x 0.3 cm. On cut section an ill defined proliferative growth with yellow and grey white areas noted measuring 1x 1cm (Fig 1). On microscopy a tumor composed of variably sized lobules with disorderly mixture of few basophilic sebaceous cells and mature tumor cells with distinct cell membrane, clear to vacuolated cytoplasm and pleomorphic vesicular nuclei with prominent nucleoli, luminal necrosis and 10 mitoses/10 HPF. No intra-epithelial

spread was noted (Fig 2, 3). Tumor cells were positive for fat stain and epithelial membrane antigen (EMA) (Fig 4, 5). A diagnosis of sebaceous carcinoma was given. The margins were free from tumor. Patient is been on regular follow up.



Fig 1- Gross- Ill defined proliferative growth

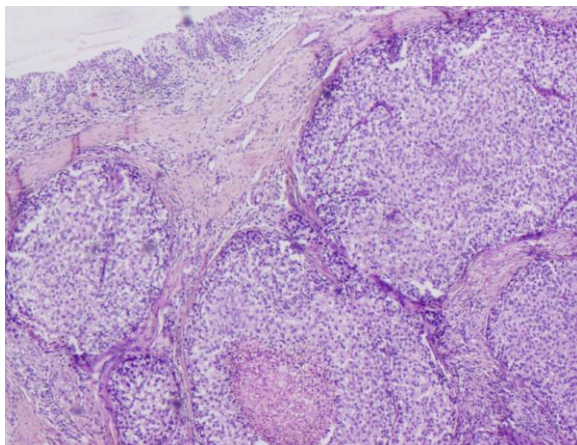


Fig 2- Variably sized lobules with disorderly mixture of few basophilic sebaceous cells and mature tumor cells, Luminal necrosis H& E X40

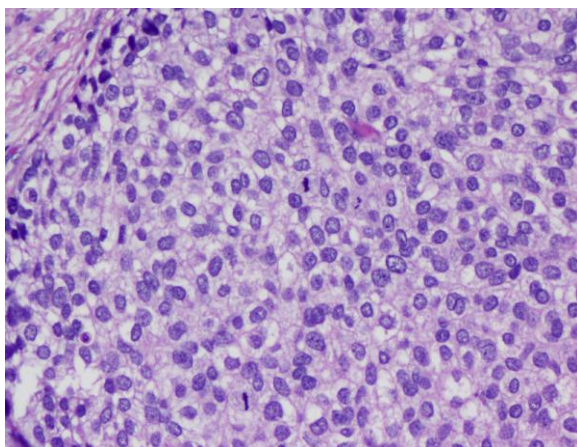


Fig 3- Cells with distinct cell membrane, clear to vacuolated cytoplasm, pleomorphic vesicular nuclei, prominent nucleoli and mitosis. H& E X200

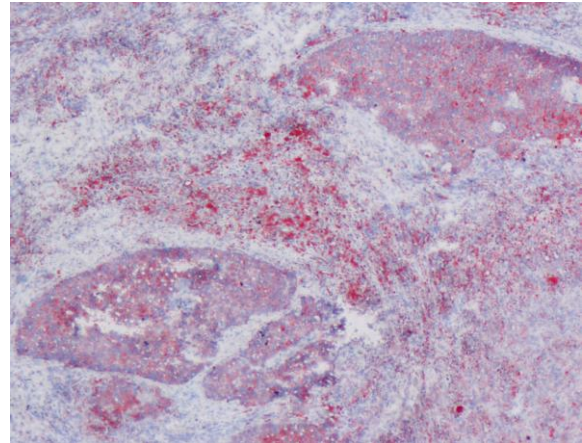


Fig 4- Tumor cells positive for fat stain Sudan IV X40

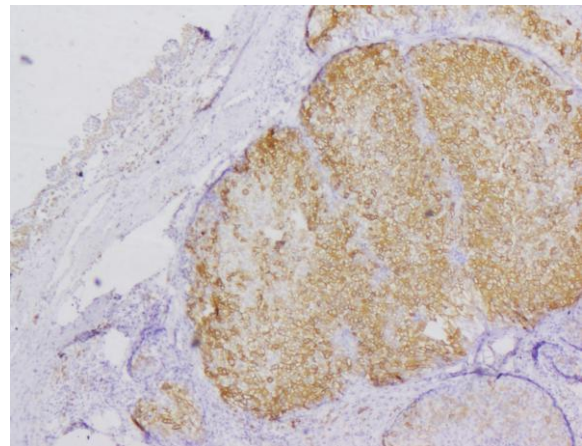


Fig 5- Tumor cells positive for EMA X40

DISCUSSION

Periocular SGC accounts for 75% of all SGC. It usually arises most commonly from the meibomian glands, then from the glands of Zeis and the sebaceous glands of the eyelid skin. SGC arises more frequently in the upper eyelid due to the presence of more meibomian glands in the upper eyelid compared to the lower eyelid. The ratio varies from 1.3 to 3.0 in the literature. Other periocular sites include the eye-brow, caruncle, lacrimal gland and conjunctiva. ⁽¹⁻³⁾ In 1956, Straatsma ⁽⁴⁾ published the first extensive series describing the natural history and prognosis of SGC in the eyelid.

The incidence of SGC of the eyelid has considerable geographical and ethnic variation. Among whites it is rare representing 0.2%-1.2% of all lid lesions and 1.13%-3.2% of all lid malignancies. ⁽²⁾ In China, Japan and other Asian countries,

the incidence appears to be much higher, probably due to genetic or racial reasons or alternatively, the rates of SGC could be higher due to the lower incidence of other eyelid malignancies. ⁽¹⁾

SGC is generally considered to be a tumor of the older age group, mostly occurs in sixth to eighth decades of life. There has been no consensus established yet if any gender predilection exists. ⁽⁴⁾ The aetiology of SGC is predominantly unknown. Reported risk factors include advanced age, Asian race, women, previous irradiation to the head and neck region and a genetic predisposition. ⁽⁵⁾ Muir-Torre syndrome, a rare genodermatologic disorder which is characterized by autosomal dominant non-polyposis colorectal carcinoma, sebaceous gland tumours and visceral malignancies is a risk factor for SGC. Other reported associations include Rb and p53 mutations, HIV and HPV. ⁽¹⁾

SGC presents most commonly as a slow growing, discrete, firm and immobile yellow nodule, which can mimic several ophthalmologic or dermatologic conditions. ⁽⁵⁾ The pagetoid spread of SGC occurs with intraepithelial infiltration of the lid margin or conjunctiva causing diffuse thickening and loss of eyelashes. ⁽¹⁾ The tumor may have a papillomatous appearance or that of a diffuse plaque like thickening of the tarsus causing lid eversion. ⁽⁶⁾ Rarely the presenting feature may be enlarged cervical or preauricular lymph nodes. ⁽²⁾ Clinically, SGC is characterized by the “masquerade syndrome”. It masquerades not only inflammatory conditions like blepharoconjunctivitis or chalazion but also benign, premalignant and malignant tumors. ⁽⁶⁾ The average time between the presentation and diagnosis may extend from 1 to 3 years. ⁽⁴⁾

Hence it's important to consider SGC as a differential diagnosis in eyelid lesions, particularly in Asian population. Definitive diagnosis is only by

histopathology. On microscopy, the tumor cells are arranged in the form of sheets or lobules, sometimes with central comedo necrosis. Individual tumour cells have distinct cell membranes, clear to vacuolated cytoplasm and vesicular nuclei with prominent nucleoli, numerous mitoses and apoptotic cells. Intra-epithelial spread is a characteristic feature of SGC seen in up to 39% patients. Intra-epithelial spread can have a Bowenoid, pagetoid or mixed pattern. ⁽¹⁾ The demonstration of intracytoplasmic lipid vacuoles by Oil red O or Sudan IV stains can be performed on frozen sections. On light microscopy diagnosis is missed in 23–77% of cases especially the poorly differentiated SGC. ^(1,7) On immunohistochemistry (IHC), SGC are immunoreactive for epithelial membrane antigen (EMA), BRST-1 and Cam 5.2. Also immunostaining for androgen receptor (AR) suggests AR to be a reliable marker of sebaceous differentiation. ⁽¹⁾

Poor prognostic factors include involvement of both upper and lower eyelids concomitantly, upper eyelid location of the tumor, size of 10 mm or more, a duration of >5 months, lymphovascular and orbital invasion, multicentric disease, poorly differentiated tumors, pagetoid spread and an infiltrative growth pattern. ^(1,5,8)

Standard surgical resection, radiotherapy and chemotherapy are used to manage SGC based on tumor stage at presentation. Wide excision with at least 4 mm margin and radical neck dissection in cases with loco-regional metastases is required. However, 32% cases recur even with a margin clearance of 5-6 mm. recently potential role of hormonal therapy in patients with SGC is suggested. ^(1,5)

SGC is reported to recur in 6% to 29% of cases. Distant metastasis affects 14-25% of cases and may involve lymph node or liver, lungs, brain and bones through hematogenous spread. Majorly all recurrences appear within the first 4 years

after treatment. ⁽²⁾ According to literature, mortality rates in primary SGC vary from 9 to 40%, and the 5-year mortality in cases with metastatic disease is been estimated to be 50 - 67%. ⁽⁷⁾

Post operatively patients must be followed up at short intervals as the tumor has a fast growth potential. Adequate follow up must include examination of the local site and the regional lymph nodes.

CONCLUSION

SGC is a rare cutaneous tumor predominantly periocular in location and affecting upper eyelid. Its diagnosis is challenging for both clinicians and pathologists. SGC has high recurrence rate and metastasis potential, hence high index of suspicion, accurate histopathological diagnosis, early treatment and adequate follow-up are of extreme importance to decrease the long term morbidity and prolong the survival of the patients.

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