

Case Report

An Unusual Case of Sebaceous Gland Carcinoma of the Upper Eyelid – A Case Report

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ABSTRACT

Sebaceous gland carcinoma of the eyelid is an aggressive malignant tumor derived from adnexal epithelium of sebaceous glands. Sebaceous gland carcinoma took the second place of all malignant eyelid tumors and occurred in 19.1%. The upper lid involved by sebaceous gland carcinoma is 54.6% and has a predilection 3 times more than the lower lid. It presents as a poorly differentiated lesion^[1], which suggests a possibility of misdiagnosis because of its similarities to basal or squamous cell carcinoma. A 55-year-old male presented with a left upper eyelid swelling with an ulcerated wound over the lateral 2/3rd of the upper eyelid. A differential diagnosis of sebaceous gland carcinoma, squamous cell carcinoma and basal cell carcinoma of the eyelid was made. The X-ray orbit showed that there is a homogenous soft tissue density lesion along the lateral aspect of orbit on left side and a bony irregularity seen along the lateral margin of orbit on left side. B-scan showed mixed echogenic lesions in the subcutaneous plane involving the upper eyelid. A wide excision with lower lid switch and cheek rotation flap was performed. The diagnosis of sebaceous gland carcinoma was confirmed on histopathological findings. An early diagnosis and appropriate treatment may decrease the long-term morbidity. It may also extend the survival rates of such patients.

Key words: Sebaceous gland carcinoma, Pagetoid, Chalazion.

Section – Ophthalmology

INTRODUCTION

Sebaceous gland carcinoma is a very uncommon malignancy that is mostly found in the periorbital area, especially the eyelids. The tumor is also known as adenocarcinoma of sebaceous gland, meibomian gland carcinoma, or Zeis gland carcinoma. Sebaceous gland carcinoma (SGC) can be the third most common eyelid malignancy after squamous cell carcinoma and basal cell carcinoma (BCC) of the eyelid. SCG is a highly malignant eyelid tumor and an uncommon malignancy. They mostly

originate from the tarsal meibomian glands. They rarely arise from the gland of Zeis of the eyelashes, sebaceous gland of caruncle, eyebrows and periocular skin. It is a slow growing tumor with high recurrence rates. These periorbital primaries are locally highly aggressive tumors and can metastasize to the regional lymph nodes mostly the pre-auricular nodes and distant organs. Sebaceous gland carcinoma can be a challenging diagnosis for the clinicians as well as the pathologists. The correct diagnosis of Sebaceous Gland Carcinoma can be delayed because it masquerades as a variety of other ocular conditions. Malignant meibomian tumors can be composed of pavement cells arising from a duct, or present a basal-cell configuration. In this connexion Warvi and Gates pointed out that only a small area of a carcinoma may show a typically sebaceous structure and tumor may be mistaken for an ordinary squamous or basal-cell growth.^[2] It can have multifocal origin and pagetoid spread it has been given an unique place among the eyelid malignancies. SGC arises from sebaceous material secreting gland which are known to be occasionally multicentric. These eyelid tumors are reported to behave aggressively with the tendency to metastasize early with higher mortality rates especially if

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there is a delay in the correct diagnosis. In most of the cases the correct diagnosis of SGC of the eyelid is delayed because of its ability to masquerade as a variety of other ocular conditions such as a chalazion, chronic blepharoconjunctivitis, basal cell carcinoma or other eyelid tumors.

CASE REPORT

A 55-year-old male presented to us with a swelling of the left upper eye lid for the past five years. There was a 5-year history of a small swelling on the left upper eyelid which gradually increased in the past 5 months to present size. It was associated with drooping of the left upper eyelid since 1½ years. There was heaviness of the left upper eyelid and diminution of left side vision due to drooping of left upper eyelid. It was associated with distortion of left upper eyelid skin and discharge since 4 months. The discharge was watery, non-purulent and non-foul smelling. There was bleeding from the lower part of the swelling. He had 2 episodes of bleeding from the swelling in last 3 months. History of matting of the eyelashes in the morning due to over-night collection of discharge. On ocular examination temporal field of vision was hampered. A left upper eye lid swelling of 2.5 cm × 2 cm with an ulcerated wound on the lateral aspect of lid margin was seen (Fig 1a). On general examination, no lymphadenopathy or organomegaly was detected. There was a history of significant weight loss. A differential diagnosis of sebaceous gland carcinoma, squamous cell carcinoma, or basal cell carcinoma was made. X-ray orbit showed that there is a homogenous soft tissue density lesion along the lateral aspect of orbit on left side and there is a bony irregularity seen along the lateral margin of orbit on left side (Fig 1b). B-scan showed mixed echogenic lesions in the subcutaneous plane involving the upper eyelid.

A Fine Needle Aspiration Cytology (FNAC) confirmed the diagnosis of Meibomian gland carcinoma. A wide excision with lower lid switch and cheek rotation flap was carried out. Aspiration performed on left upper eyelid tumour revealed highly cellular smears. The smear contains sheets of cells as well as the neoplastic cells arranged in a glandular pattern. The neoplastic cells are pleomorphic contain abundant vacuolated cytoplasm with markedly pleomorphic nuclei. The background shows mixed inflammatory cells and red blood cells. Based on history, clinical and histopathological findings a diagnosis of Meibomian gland carcinoma of upper lid was made.

DISCUSSION

Meibomian gland carcinoma was first described more than a century ago by Fuchs. Sebaceous Gland Carcinoma has to be given special attention because of its masquerading tendency and also because of its much higher prevalence in the Indian subcontinent in contrast to the Western world.^[3]



Figure 1(a) Left upper eyelid swelling with an ulcerated wound on its lateral aspect.



Fig 1b: X-ray orbit shows there is a homogenous soft tissue density lesion along the lateral aspect of orbit on left side. There is a bony irregularity seen along the lateral margin of orbit on left side.

Sebaceous gland carcinoma of the eyelid is a rare but a potentially fatal neoplasm. It represents approximately 5 percent of malignant eyelid tumors. Its incidence varies depending on demographics, ranging from 0.5 per million in Caucasians aged 20 and older.^[4] It has higher rates in Asian populations. In India there was a report that 28 percent of 85 malignant eyelid tumors were diagnosed as sebaceous gland carcinoma.^[5] The incidences of SGC among all eyelid tumors are 0.2% to 0.7%, and among eyelid malignancies is 1% to 5.5%.^[6]



Fig 1c: On B-scan There is evidence of mixed echogenic lesions approximately measuring 3.0×2.3×1.7 cm in the subcutaneous plane involving the upper eyelid. No obvious necrotic areas/calcifications. On color Doppler lesion shows increased vascularity.

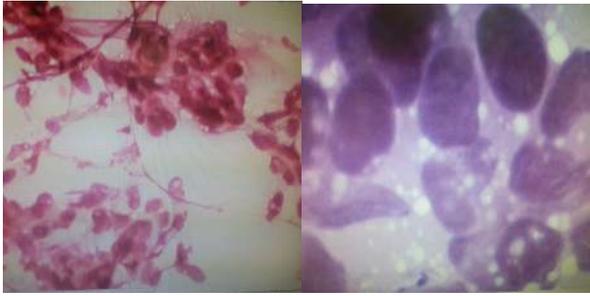


Fig 2(a)

Fig 2(b)

Histopathological slide showing (a) The smear contains sheets of cells as well as the neoplastic cells arranged in a glandular pattern. (b) The neoplastic cells are pleomorphic contain abundant vacuolated cytoplasm with markedly pleomorphic nuclei (haematoxylin and eosin, ×200)



Fig 3(a)

Fig 3(b)



Fig 3(c)

Fig 3 (a) Post-Operative photo showing lower lid switch with cheek rotation flap.

Fig 3(b) Release of flap 3 weeks post-operatively.

Fig 3 (c) Post-operative photo after 6 weeks.

The prognosis of SGC has been reported as poor compared with most other malignant eye lid tumors with a mortality rate secondary to malignant melanomas.^[4] Tumors which are in excess of 10 mm in size are associated with a poor outcome as well as survival rate.^[4] Tumors of the upper eye lid are associated with an adverse outcome than to those affecting the lower eyelid. There are a various other clinicopathologic features that indicate a worse prognosis in such tumors. These features include vascular, lymphatic and orbital invasion, multicentric origin, poor differentiation, duration of symptoms more than 6 months, a highly infiltrative pattern and pagetoid invasion of the overlying epithelium of the eyelids.^[4]

The two most common pathologic misdiagnoses made are basal cell and squamous cell carcinomas. But, the correct diagnosis can be made when lipid material is recognized within the cytoplasm of these cells.^[6] If the clinician suspects the possibility of sebaceous gland carcinomas, a frozen section of formalin-fixed tissue stained with oil red-0 will confirm the diagnosis. Dehydration with alcohol should not be used because extraction of lipid material will

occur. Tumor cells originating from the normal sebaceous gland are also a valuable indicator of sebaceous gland carcinoma.^[6]

It is believed that the intraepithelial component originates with the invasive tumor and spreads in a centripetal fashion from the underlying carcinoma.^[7] This hypothesis is supported by the observation that the number of intraepithelial pagetoid cells tends to gradually decrease farther from the nidus of invasive tumor.^[7]

The most acceptable management of periorbital sebaceous carcinoma is complete surgical removal. If there is a localized eyelid lesion that is well circumscribed, removal by eyelid resection along with frozen-section control of surgical margins can be done. In the same surgical procedure, small map biopsies of the palpebral and bulbar conjunctiva should be performed, even if no distinct conjunctival involvement is seen clinically.^[8]

If the circumscribed lesion is larger and more extensive eyelid repair is anticipated, then excision of the main lesion is attempted, small map biopsies of the palpebral and bulbar conjunctiva should be done, and definitive surgery should be planned, based on the results of those biopsies.^[8]

If the lesion is ill defined and diffuse, without a nodular component, then eyelid and conjunctival map biopsies should be done and submitted for permanent histopathology slides.

Complete surgical excision should be planned, depending on the results of those biopsies. Supplemental cryotherapy to the bulbar conjunctiva should then be applied in the same operation, even though histopathologic results are not yet available. If there is a question of residual involvement of the bulbar conjunctival by the neoplasm, topical chemotherapy using Mitomycin C may be a useful supplemental treatment.^[8]

Some authors advocate the use of Moh's micrographic surgery in patients with SC, this technique is questioned due to chances of misinterpretation of subtle intraepithelial pagetoid spread with frozen section analysis. To preserve the function of the eyelid and ease of reconstruction, it is important to try and preserve as much healthy tissue as possible during a successful excision. Excision and delayed reconstruction offer excellent option for the management of this rare and highly malignant tumor.^[9] The majority of recurrences in SGC appear within the first 4 years of treatment. In our case, distant metastasis was not seen. Surgical treatment may range from a local excision of the tumor to orbital exenteration.^[5] A wide excision with lower lid switch and cheek rotation flap was performed in our patient. Patients with SGC must be followed up at short intervals post-operatively because the tumor has a very fast growth potential and high recurrence rate. Adequate follow-up includes meticulous inspection of the local site of involvement of the eyelid. It is mandatory to palpate the pre-auricular, submandibular and other neck lymph node chains. The guidelines for follow up are 3 monthly interval during the first year, 6 monthly during the second year and then on a yearly basis for life.^[4]

CONCLUSION

Early diagnosis and treatment of these patients may decrease the long-term morbidity and extend the survival rate.

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