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Case Report

Pathology Section

Sebaceous Carcinoma of the Eyelid- A Rare Aggressive Malignant Neoplasm Worth Early Recognition

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ABSTRACT

Sebaceous carcinoma of the eyelid is an uncommon malignant adnexal neoplasm accounting for 1 - 5.5% of eyelid malignancies. Diagnosis and therapy of their aggressive malignancy often get delayed since it is often misdiagnosed both clinically and histopathologically because of its varied clinical appearance and deceptive histopathological features.

In this case report, we discuss one such rare occurrence of sebaceous carcinoma in the lower eyelid of an elderly lady aged 70 years along with the key diagnostic features and differential diagnosis

Keywords: Lsebaceous carcinoma, Sebaceous hyperplasia, Sebaceous cell adenoma

CASE SUMMARY

A 70 year old female patient presented to the Ophthalmic department with a gradually progressive, painless swelling in the right lower eyelid present since 3 years. On examination, the swelling was hard, non- tender and with a smooth surface. Skin over the swelling was pinchable. General examination did not reveal any significant findings. A clinical diagnosis of carcinoma of the lower eyelid was arrived at and a wide triangular excision was done.

HISTOPATHOLOGICAL FINDINGS

The specimen was a skin- covered grey- white nodule which measured 1 x 0.7 x 0.5 cms. Cut- section showed thinned out overlying skin and well- circumscribed, grey- white, lobulated mass in the subcutis [Table/Fig-1].

Microscopy- Showed an asymmetrical, partly circumscribed lobulated tumor. Tumor lobules were separated by thin septae [Table/Fig-2]. Majority of the tumor cells showed clear cytoplasm, large round to polygonal, vesicular nuclei and prominent nucleoli [Table/Fig-3] Occasional large undifferentiated tumor cells with large and hyperchromatic nuclei were seen [Table/Fig-4] Occasional atypical mitoses were seen

Histopathological diagnosis- Sebaceous carcinoma of eyelid

Histochemistry- Showed positivity for Oil Red O in the cytoplasm of tumor cells.

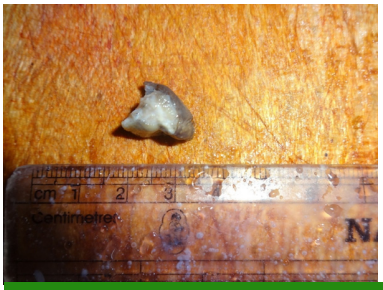
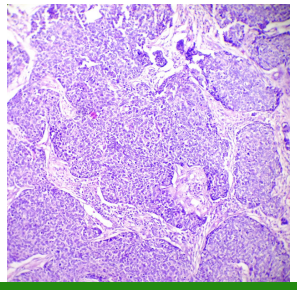
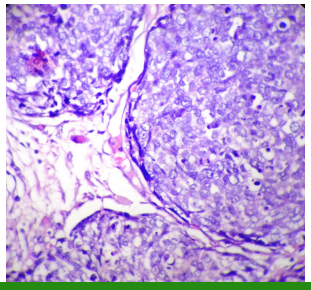
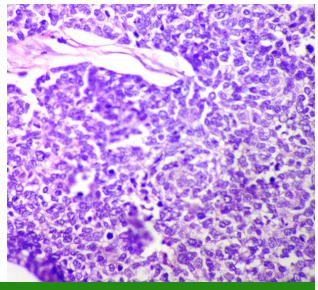
A thorough evaluation was undertaken and regional and systemic metastases were ruled out.

DISCUSSION

Sebaceous carcinomas of the eyelid are uncommon malignant adnexal tumors. They account for 1- 5.5% of malignant eyelid neoplasms. They are the third most common eyelid malignancies with basal cell carcinoma and squamous cell carcinoma occupying the first and the second places respectively. Sebaceous carcinoma of the eyelid usually occurs in elderly women, as in the present case. It is commoner in the upper eyelid [1]. In contrast, the tumor in the present case was seen in the lower eyelid.

Sebaceous carcinoma may deceptively masquerade as a benign condition both on clinical and histopathological examination which often leads to inappropriate management [2]. Since it is characterized by a high rate of local recurrence with regional and distant metastases, it qualifies as an exclusive entity worthy of early recognition in order to facilitate appropriate management and to avoid undue morbidity and mortality.

The clinical presentation may be variable mimicking a chalazion and blepharoconjunctivitis. The patients may receive multiple course of incision and drainage for recurrent chalazions before a biopsy is performed thus causing delay in the diagnosis and initiation of proper treatment [2]. So, the clinician should consider the possibility of sebaceous carcinoma in any patient with conjunctivitis, chronic or recurrent chalazion or persistent blepharoconjunctivitis. Any of these conditions that are not improving after three months of observation have to be invariably scheduled for a biopsy [3]. Once diagnosed, an intensive search for metastases is warranted since the tumor is known to spread regionally into the lacrimal glands and excretory systems, to regional lymph nodes and may rarely

[Table/Fig-1]: Cut- section showed thinned out overlying skin and well- circumscribed, grey- white, lobulated mass in the subcutis
[Table/Fig-2]: Tumor lobules separated by thin septae, (H & E, ×100)
[Table/Fig-3]: Tumor cells showed clear cytoplasm, large round to polygonal, vesicular nuclei and prominent nucleoli, (H & E, ×400)
[Table/Fig-4]: Large undifferentiated tumor cells with large and hyperchromatic nuclei, (H & E, ×400)

even metastasize hematogenously to the lungs, liver and bones [3]. Though histopathology is the gold standard for the diagnosis of sebaceous carcinoma, it may occasionally be mistaken for sebaceous hyperplasia or sebaceous gland adenoma.

Sebaceous carcinoma may arise from the Meibomian glands, glands of Zeis or glands in association with caruncle [4]. They are grouped under epidermal appendageal tumors. They deserve a unique place and attention amongst eyelid tumors owing to their multifocal origin and the pagetoid spread that they are known to possess [1]. Periorbital primary sebaceous carcinomas are known to behave aggressively with a tendency for early metastasis and significant mortality [5].

In contrast to squamous cell carcinoma or basal cell carcinoma, sebaceous carcinoma of the eyelid is three times more common in the upper eyelid than in the lower eyelid since a higher number of Meibomian glands are situated there [6,7]. The lower eyelid and the caruncle are the other common sites involved [8]. The tumor in the present case involved the lower eyelid.

Though the clinical appearance may not be characteristic, pagetoid infiltration of the conjunctival epithelium or the epidermis of the skin are the histological hallmarks of the tumor unlike the squamous cell carcinoma or basal cell carcinoma where the spread is radial [9,10]. The pagetoid spread may be mistaken for carcinoma in situ.

Sebaceous carcinoma is considered as one of the most dangerous eyelid tumors due to many reasons including-

a) Its capability to mimic inflammatory conditions like blepharoconjunctivitis, chalazion and superior limbic keratoconjunctivitis [11-13] or other eyelid cutaneous malignancies like squamous cell carcinoma with hydropic change or basal cell carcinoma with sebaceous differentiation because of which the diagnosis may be delayed until metastasis has set in.

b) High incidence of metastasis (41%) 6,9

c) Difficulty in delineating tumor margins either due to multifocality or pagetoid spread [7,10,14-16].

d) Other benign and malignant histopathological mimics.

Keeping these facts in view, it has been recommended that, any chalazion of unusual consistency or its recurrence more than three times should be subjected to full thickness resection and histopathological examination [1]. Any unilateral blepharoconjunctivitis with loss of eyelashes, thickening of the lid margin and any suspicious area in the conjunctiva or limbus has to be biopsied [1].

The prognosis is still regarded as poor as compared to the other malignant eyelid tumors with a mortality rate second only to malignant melanoma [10].

CONCLUSION

Sebaceous carcinoma can be a notorious diagnostic pitfall for the clinicians and pathologists alike. Timely and accurate diagnosis and treatment is essential since this neoplasm is the most aggressive of all the epithelial tumors of the eyelid.

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