

Sebaceous Carcinoma of Eyelid: A Report of Two Cases with Review of Literature

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Abstract

Background: Sebaceous carcinoma of the eyelid is rare. The diagnosis might be difficult because of its ability to masquerade as other periocular lesions. Prognosis is still regarded as being poor compared with most other malignant eyelid tumors with a mortality second only to malignant melanoma. The present reported cases analyses clinical and histopathological findings. *Case Reports:* Two case reports were presented here. One presented as primary complaint of progressive painless mass in left lower eyelid in a 75 year female and another case in young 30 year old with history of recurrence, operated previously for the same complaint with unavailable previous report. Both cases on histopathology reported as sebaceous gland carcinoma. *Conclusion:* Early diagnosis and consequent surgical therapy of sebaceous carcinoma of the eyelid leads to a better outcome and higher survival rates than generally assumed. Even local recurrences can be treated successfully. However, sebaceous carcinoma remains a threatening disease, which leads to death in 9% and to mutilating exenteration in 23% published in an English literature.

Keywords: Sebaceous carcinoma; Meibomian gland; Eyelid.

Introduction

The sebaceous carcinoma is a very rare malignant tumor primarily found in the area of the eyelid. Most of these carcinomas originate in the tarsal meibomian glands although they may in extremely rare cases originate in the glands of Zeis of the eyelashes or the sebaceous glands of the caruncle [1]. Sebaceous carcinoma primarily periorbital and comprise the fourth most common neoplasms of the eyelid. It is believed that metastases spread typically to regional lymph nodes. Reported risk factors for sebaceous carcinoma include advanced age, Asian or South Asian race, women, previous irradiation to the head and neck, and a genetic predisposition for Muir-Torre syndrome or possibly familial retinoblastoma [1-4].

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Case Reports

Case 1: A 75 year old female patient presented with painless progressive pinkish swelling in the left lower eyelid for four months. On clinical examination revealed exophytic, firm mass measuring 3.2 x 2.2 cm, with regular shiny surface in the left lower eyelid involving both palpebral and bulbar conjunctiva reaching upto the cornea (Figure 1). Rest of her medical examination was within normal limit. CECT orbit showed 3.6 X 3.5 cm well defined heterogeneously enhancing soft tissue mass seen in inferolateral extra-corneal compartment of left eye growing exophytically. She underwent excision of the lower eyelid mass with eyelid reconstruction. Histopathological examination revealed tumor cells were arranged in sheets and lobules (Inset of Figure 2). The tumor cells had moderately pleomorphic vesicular nuclei, having prominent nucleoli, moderate to abundant amount of eosinophilic to vacuolated cytoplasm with increased vascularization (Figure 2). Histopathological features were of sebaceous / meibomian gland carcinoma. She

received post-operative radiotherapy in view of margin positivity. Patient is kept on follow up and leading a good quality of life for six months.

Case 2: A 30 year old male presented with swelling in the lateral aspect of right lower eyelid since last four years. It was gradually progressive and was associated with mild pain. Wide local excision of mass was done four years back. Previous report was not available with the patient, but it recurred since last three months again with the same complaint. CECT orbit showed evidence of hypodense ill-defined mass showing uniform enhancement seen predominantly at the lateral canthus, inferior part of preseptal compartment of eye showing ill-defined fat planes. He was again operated and surgical pathology report showed infiltrating tumor composed of sheets and nest of tumor cells separated by fibrous tissue. The tumor cells show moderate nuclear pleomorphism and have moderate to abundant clear vacuolated cytoplasm. Histopathological features were of meibomian gland carcinoma. The disease again recurred after one year and underwent salvage surgery under GA, in the form of right orbito-zygomatico maxillectomy with superficial parotidectomy and right neck node dissection. Histopathology examination revealed poorly differentiated sebaceous carcinoma of right eyelid involving lateral canthus, part of upper and lower conjunctiva, lacrimal gland and lateral rectus muscle. He received both radiotherapy and chemotherapy after surgery. After 6 months of follow up a repeat CECT head was done which revealed only bony defect in right frontal and zygomatic bone, right side maxillary sinus and greater wing of sphenoid with no evidence of growth. The patient is tumor free since last four months of follow up.



Fig. 1: Clinical presentation showing large mass of the left lower eyelid

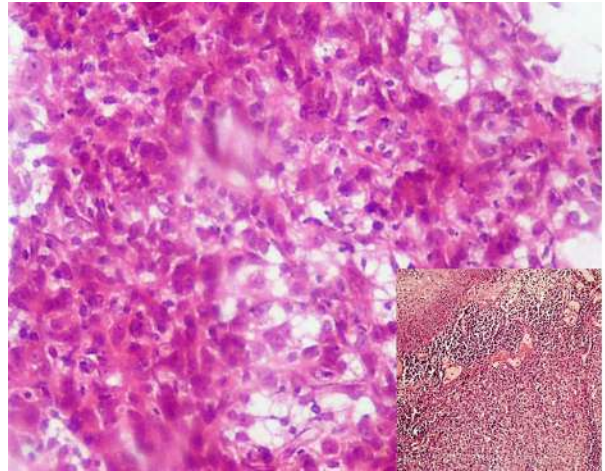


Fig. 2: Malignant cells with vacuolated cytoplasm and areas of necrosis (H & E, $\times 40$). Inset shows lobular arrangement with dense inflammatory infiltrate

Discussion

Meibomian glands are modified sebaceous glands present in the stroma of tarsal plate arranged vertically. They are about 30-40 in the upper lid and about 20-30 in the lower eyelid [2]. The clinical presentations of ocular sebaceous carcinomas are diverse and may delay an accurate diagnosis for months to years. It is a very slow growing tumor and commonly found in elderly population with female predisposition [1,3,4]. Mean age at diagnosis is mid-sixties, however the tumor has been reported in children as young as 3-5 years old. Two important features differentiate meibomian carcinoma from other periocular malignancies. First, unlike single origin of other tumors, meibomian carcinomas appear to arise from multifocal origins. Second, unlike radial spread of basal cell and squamous cell carcinomas, meibomian gland carcinoma tends to spread superficially in a pattern known as pagetoid spread, which is a hallmark of this tumour [1,2,4].

There are five areas in the periorbital region that contain sebaceous cells [5-8]:

1. Regular sebaceous glands associated with the pilosebaceous units of the facial and eyelid skin,
2. Meibomian glands located deep in the tarsus which secrete the oily outer constituent of the tear film
3. The Glands of Zeiss which nourish the eyelash hair follicles with oily secretions
4. The pilosebaceous units of the eyebrow hairs
5. The sebaceous glands within the caruncle.

Meibomian carcinoma can arise from any of these

types of sebaceous glands. However, classically it arises from the Meibomian glands of tarsal plate and from the upper eyelids, which accounts for approximately 2/3 of cases. Less commonly, it is found in the Glands of Zeiss and the lower eyelids. It is rare to find the lesion in the caruncle or eyebrow and very rare to find it in the lacrimal gland, which accounts for only a handful of poorly differentiated cases [9-11].

Although sebaceous cell carcinoma is one of the most malignant lesions of the eyelids, it may be difficult to diagnose. This is demonstrated by the fact that the average intervening time between presentation and diagnosis ranges from one to three years. The reason for this difficulty in diagnosis is due to its varied presentation both clinically and histopathologically. It tends to masquerade as more common benign conditions, mimicking other tumors as well as inflammatory conditions. So it is important to include sebaceous gland carcinoma in the differential diagnosis of most eyelid masses and recurrent inflammatory conditions [12-15].

The most common presentation is a small, rubbery, firm nodule that looks like a chalazion. This is complicated by the fact that sometimes there is true chalazion formation secondary to obstruction of the meibomian ducts by a mass. The mass may be yellow in color due to its lipid content, especially as it encroaches on the epidermal surface [3,7,9]. The caruncle is an unusual location for this tumor, but here it presents as a multi-lobulated, grey-yellow subconjunctival mass covered with epithelium. When arising from the glands of Zeis, the lesions form small yellow nodules in front of the gray line and this in turn results in entropion of the eyelid. Besides presenting as various masses, sebaceous cell carcinoma also present as various inflammatory conditions. This results from the intraepithelial spread that is typical of this lesion. Pagetoid spread, characteristic of sebaceous gland carcinoma, occurs when individual tumor cells migrate up to the epithelial surface and form patchy, skip lesions separate from the original tumor mass. This causes irritation and leads to inflammatory presentations such as blepharoconjunctivitis or kerato conjunctivitis [5-9].

Similar to its varied clinical presentation, sebaceous cell carcinoma may also mimic other disease processes histopathologically. Sebaceous carcinoma may have focal areas of squamous differentiation and is therefore commonly mistaken for squamous cell carcinoma. In fact, 50% of sebaceous cell carcinomas are misdiagnosed as squamous cell carcinoma. Basal cell carcinoma on the other hand can have areas of

sebaceous differentiation. Mucoepidermoid carcinoma has clear mucinous cells that resemble sebaceous cells. Melanoma may also simulate sebaceous cell carcinoma since a previously excised melanoma may recur as a nonpigmented lesion in the conjunctiva or cornea [15,16].

The masquerading feature of sebaceous carcinoma makes it a difficult diagnosis. The frequency of wrong clinical impressions has remained unchanged for the last few decades. So it is extremely important to maintain a high clinical suspicion. There are some clues in history and physical that help raise clinical suspicion. A chalazion that presents late in life without a prior history or a recurrent chalazion is quite suggestive. Further investigation is also warranted for inflammatory presentations such as blepharitis or keratoconjunctivitis that are unresponsive or only partially responsive to treatment. Clues on physical exam that should raise suspicion include a diffuse lesion or thickening and madarosis. Histopathologically, special stains such as oil-red-O may be helpful in the diagnosis. However, this requires fresh or wet tissue since paraffin processing washes out lipid [11-18].

Conclusion

Sebaceous cell carcinoma is an aggressive tumor seen most often on the eyelid. Since it clinically mimics other disease it is difficult to diagnose. However, accurate and prompt diagnosis is crucial since it is not only one of the most malignant lesions on the eyelid, but also one with serious associations such as Muir-Torre syndrome and high potential for regional and distant metastases. Since it is seen almost exclusively on the eyelid, the burden for maintaining high clinical suspicion and making this challenging diagnosis falls on the ophthalmologist and ocular pathologist.

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