Sebaceous Gland Carcinoma with Misleading Clinical Appearance: A Case Report of an Eyelid Lesion

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Conflict of interest: None declared

Patient: Female, 53-year-old
Final Diagnosis: Sebaceous gland carcinoma
Symptoms: Mass in right lower lid
Medication: —
Clinical Procedure: Full-thickness wedge resection with frozen section control
Specialty: Oncology • Ophthalmology • Pathology

Objective: Challenging differential diagnosis
Background: Sebaceous gland carcinoma (SGC) is a rare malignant lesion that occurs on the eyelids. It is known to mimic other benign or malignant lesions in clinical presentation, such as a chalazion, basal cell carcinoma, and squamous cell carcinoma. The histopathological diagnosis is the mainstay for diagnosis and is often challenging.

Case Report: We describe a case of SGC in a 53-year-old woman who presented with a cauliflower-appearing lesion with pearly telangiectatic vessels and raised margins at the lower eyelid margin. Clinically, we suspected a diagnosis of basal cell carcinoma. Upon complete resection of the lesion, the final diagnosis was SGC based on the histopathological features and immunohistochemical staining characteristics of the tissue.

Conclusions: Due to the possibility of SGC presenting similarly to other lesions, it is essential for ophthalmologists to have a high index of suspicion in its diagnosis. The early and accurate diagnosis of such lesions is important for appropriate management to prevent metastasis or recurrence related to advanced tumors.

MeSH Keywords: Carcinoma, Basal Cell • Carcinoma, Squamous Cell • Immunohistochemistry • Pathology • Sebaceous Gland Neoplasms

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Background

Sebaceous gland carcinoma (SGC) is a relatively rare malignant lesion. It represents 1% to 3% of all malignant tumors [1]. However, given the high density of sebaceous glands in the eyelid, the reported frequencies among all reported cases of malignant eyelid lesions vary from 1% to 5.5% in the United States to as high as 28% to 60% in Asian-Indian populations [2,3]. Early diagnosis of SGC is crucial to prevent metastasis and extraocular spread. The clinical presentation of SGC may vary widely and may mimic other lesions of the eyelid; therefore, reaching a definitive diagnosis prior to histopathological examination can be challenging. We present a case in which SGC was not clinically suspected.

Case Report

A 53-year-old woman presented to our tertiary care eye center in Riyadh, Saudi Arabia, with a 9-month history of a right lower lid lesion. The patient stated that the lesion had started as a ruptured cyst that was removed at a private clinic 9 months previously. The lesion recurred 1 week later. She denied any history of pain, bleeding, discharge, or recurrent chalazion. She had no medical problems, with no prior ocular or nonocular surgery, apart from the removal of the ruptured eyelid cyst, and no history of periocular irradiation.

On examination, her best corrected visual acuity was found to be 20/30 and 20/20 in her right and left eyes, respectively, and both eyes had normal intraocular pressure. Slit lamp examination of the right eye revealed a 1×0.5 cm lesion with a cauliflower appearance on the lower eyelid margin. The lesion had pearly telangiectatic vessels on the raised and rolled-in anterior margin, while the posterior part of the lesion had fragile flesh with central ulceration (Figure 1). The lesion spared the normal appearance of the surrounding tissues. Note the preservation of lashes and the normal appearance of the surrounding tissues.

The patient underwent excisional biopsy of the cauliflower lesion, with the base being shaved off at the eyelid margin. The histopathologic examination of the excised tissue revealed a mass with papillary configuration lined with nonkeratinizing stratified squamous epithelium that had variable thickness and showed a focal area of ulceration. The subepithelial area contained several islands of moderately differentiated proliferating sebaceous-looking cells with a mixed lobular and infiltrative patterns and intervening stroma showing a mild degree of chronic infiltration by inflammatory cells. In one focal area, similar abnormal cells were found to infiltrate the overlying epithelium. The tumor cells expressed reactivity with p16 and p63 as well as epithelial membrane antigen (EMA; Figure 2). The final diagnosis was SGC, which unfortunately extended to the margins of excision, specifically the deep surgical margin where the lesion was shaved off at the bed of the mass.

The patient underwent complete thickness wedge resection of the lower eyelid with wide surgical margins of excision (2 mm medially and 4 mm laterally) and a frozen section control of the margins of excision. The conjunctiva of the inferior fornix was also excised for frozen section control of the deep surgical margin. Finally, lateral canthotomy and cantholysis were done, and the defect was closed with direct closure. The frozen sections revealed clear margins with no deep fornical conjunctival extension of the SGC. Two weeks later, sutures were removed. The aesthetic outcome was good, with no ectropion, entropion, or lid retraction, and head and neck lymph nodes were normal with unremarkable systemic workup.

Figure 1. A cauliflower-like pearly telangiectatic lesion arising from the posterior lamella at the eyelid margin with central ulceration.
Discussion

SGC is a great masquerader, and ophthalmologists should therefore be aware of the many presentations it can have. In a study on 60 patients with SGC, SGC was initially suspected in only 32% [4]. Hence, we believe unusual clinical and pathological presentations of SGC should be reported to raise clinical suspicion for such an aggressive eyelid tumor.

Our patient was a 53-year-old woman. Her age was slightly less than the average age at SGC diagnosis, which is 70 years [5], but it was close to the average age at diagnosis in the same region reported from India and Oman (57 and 59 years, respectively) [2,3]. For unknown reasons, SGC has a female predominance [6]. Other predisposing factors include immunosuppression, history of periocular radiation, familial retinoblastoma, and Muir-Torre syndrome [7,8].

SGC may have a wide variability in initial clinical presentation, mimicking other lesions such as chalazion, chronic conjunctivitis, posterior blepharitis, SCC, BCC, and Merkel cell tumors [8]. However, it classically presents as a firm, painless, and indurated thickening of the eyelid with a yellow hue; upon eyelid eversion, it appears as a multinodular mass [8]. Intra-epithelial spread is suggested by the presence of blepharoconjunctivitis unilaterally in the affected eye and fornical shortening [8]. In a recent study on 30 patients with SGC, the most common clinical presentation was either diffused lid thickening or localized nodules. However, the authors did not describe any patient who had a pearly telangiectatic lesion similar to the one in our case [6]. Madarosis, destruction of the lid margin, diffuse lid thickening, and fornical shrinkage may provide clinical hints towards the underlying diagnosis. However, none of these signs were present in our case. Our case was clinically unique in various aspects. The lesion had a raised cauliflower shape at the lid margin, sparing the overlying skin, orbicularis, and eyelid lashes despite 9 months duration. The nearby lid

Figure 2. (A) Histopathologic appearance of the proliferating sebaceous-looking cells in a lobular pattern within the subepithelial stroma (original magnification ×200, hematoxylin and eosin). (B) The tumor cells reacting to epithelial membrane antigen (original magnification ×200). (C) Lower power showing the nuclear staining of SGC cells using p63 immunohistochemistry marker (original magnification ×50). (D) The SGC tumor cells reacting to p16 (original magnification ×200).
margins looked normal without any palpable mass or induration. The lesion had central ulceration and a pearly telangiectatic anterior part that was suggestive of BCC, yet a lack of skin involvement deferred the clinical decision of BCC. The fleshy and fragile posterior architecture raised the possibility of SCC. The diagnosis of SGC was not expected, especially given the lack of established clinical signs of SGC.

Histopathological examination of SGC lesions reveals various morphological growth patterns, including trabecular, lobular, and papillary [7]. The most common is the lobular pattern, which was present in our case [2]. However, different morphological types may overlap in a lesion [9]. Many immunohistochemical stains and proteins have been recommended to aid in the diagnosis of SGC, of which EMA, Ber-Ep4, androgen receptor, and adipophilin appear to be useful [10]. Cytokeratin stains (with the exception of CK19) are not very helpful in differentiating SGC from SCC and clear cell variants of BCC, with a rate of misdiagnosis that may reach 77% [11]. It is even more challenging to distinguish SGC from BCC that has sebaceous differentiation, in which case, BCC cells would lack expression of EMA [11,12]. The combination of p16 and p53 is particularly helpful in detecting intra-epithelial SGC and determining the extent of tumor spread [13]. In our case, the morphology was not basaloid, and the tumor cells expressed strong reactivity with p16, which was considered to be diagnostic. In addition, EMA demonstrated membranous and cytoplasmic reactivity, but this was only supportive because it could also be positive in SCC. Unfortunately, adipophilin and p53 stains were not available in our laboratory. Of note, the reported cell origin of SGC cannot be identified in 50% to 60% of cases [3]. The exact origin of the SGC was not determined histopathologically because the lesion was confined to the lid margin, but it was speculated to possibly arise from glands of Zeis.

The mainstay treatment modality of SGC is surgical resection with wide local excision and frozen section followed by eyelid reconstruction [6]. It is recommended that patients undergo sentinel lymph node biopsy or strict regional nodal surveillance in tumors that are 10 mm or larger in dimension. Patients with regional lymph node metastasis should undergo radical neck dissection followed by radiation therapy [14].

Fortunately, our patient had complete excision with no evidence of deep invasion or metastasis. This good outcome is attributed to the relatively early presentation of the patient. In one study, the average delay of SGC diagnosis was found to be 23 months [4]. In a review published in 2002, Snow et al. [15] reported a metastasis rate of 8%, which was mostly attributed to advanced tumors. Metastasis sites most commonly include regional lymph nodes, lung, liver, brain, and bone [14]. The mortality rate associated with SGC is 5% to 10%. However, recent studies have found that mortality rates have been reduced by appropriate treatment [6,16].

Conclusions

SGC remains a challenging diagnosis even for expert ophthalmic surgeons with long experience in eyelid lesions. SGC is a great masquerader; therefore, a high index of suspicion is essential in the diagnosis of this aggressive eyelid tumor. Tissue diagnosis is the key diagnostic method for SGC, and it can be aided by a panel of immunohistochemical stains. Reaching an accurate diagnosis is important for planning complete surgical excision as well as preventing metastasis due to advanced tumor stages.

Conflict of interest

None.

References: