Case Report

Adenoid Cystic Carcinoma of the Upper Eyelid: A Rare Case Report

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Abstract

Adenoid cystic carcinoma is a sporadic, locally aggressive, and highly recurrent malignant epithelial tumor accounting for 1% of all head and neck cancers. It primarily affects minor and major salivary glands and has a high tendency of local invasion and distant metastasis. It uncommonly originates from the palpebral portion of the lacrimal gland in the orbit. In this case report, we present a 30-year-old female patient who complained of a painless nodule in the left upper eyelid, which initially was of pea size; over time, the swelling increased to the present size. The swelling was then excised and sent for histopathological examination. Histopathological examination revealed tumor cells arranged in cribriform and tubular patterns, and finally, the diagnosis of Adenoid cystic carcinoma was made.

Keywords: Adenoid cystic carcinoma, Eyelid, Lacrimal gland, Cribriform

Introduction

Adenoid cystic carcinoma is a rare, aggressive, invasive, and recurrent malignant epithelial tumor originating most commonly from major salivary glands and rarely from minor salivary glands, esophagus, bronchial glands, skin, lung, vulva, cervix, and prostate. It follows a sluggish growth rate [1]. Primary cutaneous adenoid cystic carcinoma is a rare type of adenoid cystic carcinoma traditionally considered to originate from eccrine glands. It may arise from the primary accessory glands of the conjunctiva, ectopic lacrimal glands, and adrenal glands. It usually originates from the palpebral part of the lacrimal gland; eyelid involvement is rarely observed and comprises only 1.0% of head and neck malignancies [2]. Primary cutaneous adenoid cystic carcinomas are often misdiagnosed as chalazions because the accurate diagnosis of primary cutaneous adenoid cystic carcinoma is typically made either through histopathological evaluation or due to postoperative recurrence.

Case Report

A 30-year-old female patient was admitted to the Department of Ophthalmology, Jawaharlal Nehru Medical College and Hospital, AMU, Aligarh, due to a complaint of swelling in the left upper eyelid for the last 3 months. The swelling gradually

increased in size, was non-mobile, tender, and fixed to the bone, with no history of discharge. The swelling was associated with loss of vision, fever. On clinical examination, the swelling was 3.5×3.0 cm on the left upper eyelid, just above the medial canthus of the left eye, extending up to the upper eyebrow over the orbital rim. The swelling was non-mobile and firm in consistency, and fixed to bone. The patient underwent exenteration, and the specimen was sent for histopathological examination.

Upon gross examination, we received a specimen of the exenterated Left eye with the attached eyelid; the entire specimen measured $6.0 \times 5.7 \times 3.5$ cm. Eyelid measured 5.3×4.0 cm. Tumor measured $4.0 \times 3.8 \times 1.6$ cm. The eyeball plus optic nerve measured $2.8 \times 2.0 \times 1.5$ cm. The optic nerve measured 1.3×0.5 cm. The cut surface of the tumor showed homogeneous white areas of hemorrhage, and the eye appeared distorted (Figure 1). Representative sections were taken from the specimen and examined histopathologically.

Figure 1: Gross specimen showing exenterated eye with tumor mass in the upper eyelid.

On microscopic examination, H&E-stained sections revealed nests and clusters of biphasic tumor cells, composed of ductal and myoepithelial cells, separated by fibrous septa arranged in a cribriform and tubular pattern, with myxoid globules present in the lumen. Myoepithelial cells showed a basaloid appearance, characterized by hyperchromatic, angulated nuclei and scant cytoplasm. Sections from the eyeball and optic nerve were unremarkable (Figure 2). No immunohistochemistry was performed in our cases, as the tumor morphology was classical.

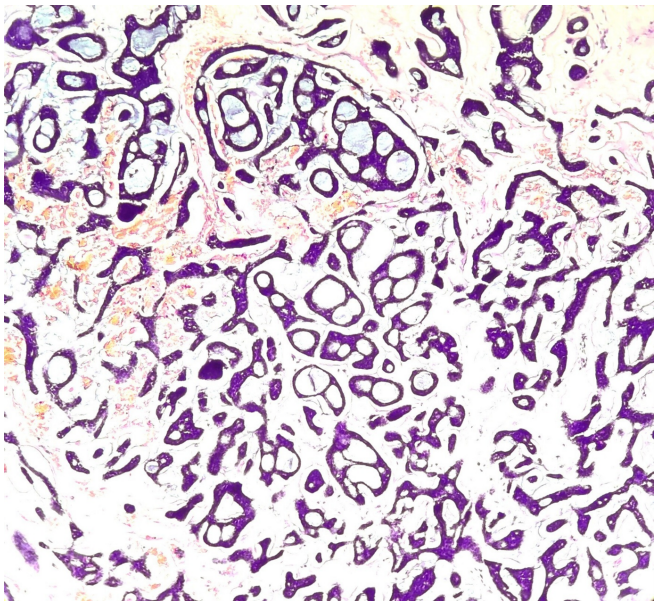

Figure 2: H & E-stained section showing tumor cells arranged in cribriform and tubular patterns, having myxoid globules in the lumen (100x)

Based on gross and microscopic examination, the final diagnosis of well-differentiated adenoid cystic carcinoma of the left upper eyelid (cribriform and tubular pattern) was made. Patient underwent radiation therapy, and no recurrence or distant metastasis was noted after 6 months of follow-up.

Discussion

Primary cutaneous adenoid cystic carcinoma is a rare, slow-growing skin adnexal tumor, accounting for less than 1% of all head and neck tumors and 10 to 15% of the salivary gland malignant tumors. It is classified as an adnexal skin tumor in the WHO classification [3]. The precise etiology of adenoid cystic carcinoma is uncertain and likely to arise from somatic mutation. However, the specific pattern of these mutations remains largely unexplored. Primary cutaneous adenoid cystic carcinoma is a locally invasive cancer, and tumor recurrence after tumor excision is very common. The clinical diagnosis of Primary cutaneous adenoid cystic carcinoma remains challenging for clinicians due to its numerous differential diagnoses, including chalazion, adenoid cystic variant of basal cell carcinoma, mucinous carcinoma, palpebral sebaceous carcinoma, dermal cylindroma, and primary cutaneous cribriform apocrine carcinoma [4, 5, 6]. Histopathologically, all these lesions closely resemble each other and are similar to Primary cutaneous adenoid cystic carcinoma [7]. Histopathological examination remains the gold standard to arrive at a conclusive diagnosis. Microscopic examination of adenoid cystic carcinoma reveals a dermal tumor comprising basaloid cells arranged in a cribriform, tubular, or solid pattern, depending on tumor grade, with a fibrous or hyalinized stroma having pseudocysts filled with mucin [8]. Peritumoral clefting is typically absent. Primary cutaneous adenoid cystic carcinoma usually lacks a connection with the skin epithelium. This is particularly noticeable when the tumor is located on the scalp. Immunohistochemistry is typically not required for the diagnosis of adenoid cystic carcinoma; however, S-100, CD117, p63, and actin are helpful when needed. Adenoid cystic carcinoma of the eyelid is a rare and unique condition. It can arise from the meibomian gland or lacrimal gland of the palpebral lobe. These tumors may exhibit cystic intratumor spaces containing alcian blue-positive mucin [7]. Perineural invasion is present in approximately 76% of cases and is associated with an increased risk of local recurrence [2]. In our case, perineural invasion was not present. Adenoid cystic carcinoma of the eyelid has a much better prognosis than adenoid cystic carcinoma of the lacrimal gland, which is the most prevalent site of adenoid cystic carcinoma in the optical adnexa [9]. Leu et al. found that cribriform variants of adenoid cystic carcinoma are associated with better prognosis. The overall prognosis of Primary cutaneous adenoid cystic carcinoma is better than that of adenoid cystic carcinoma of other organs [10, 11]. Wide surgical excision with at least 2 cm tumor-free margins is the treatment of choice for Primary cutaneous adenoid cystic carcinoma. Perineural invasion is critical, and long-term follow-up for recurrence is recommended [12]. Mohs micrographic surgery can be helpful. The role of radiotherapy and chemotherapy is debatable because no study has advocated that radiation decreases the risk of local recurrence. Chemotherapy can be used in patients with distant metastases.

This case is reported because Primary cutaneous adenoid cystic carcinoma is a sporadic tumor of the eyelid. Wide surgical excision of the tumor is the treatment of choice for eyelid adenoid cystic carcinoma. As this tumor has a high recurrence rate, long-term monitoring is necessary. Histopathological examination of resected specimens is the gold standard for diagnosing adenoid cystic carcinoma.

Conclusion

Despite its rarity, the diagnosis of Adenoid cystic carcinoma must be considered when patients present with long-standing painless nodules in the eyelid. Early diagnosis and a comprehensive treatment approach are essential for better outcomes. The chances of recurrence are high even after adequate, wide surgical excision, and because of its local aggressiveness, continuous monitoring and long-term follow-up of the patient are highly required. A high index of suspicion must be kept to diagnose the disease, as it leads to significant morbidity, including visual impairment.

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