



## Case Report

# Adenoid cystic carcinoma of eyelid: A rare case presentation

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## ABSTRACT

Adenoid cystic carcinoma (ACC) is a rare tumor identified for its severe aggressiveness and tendency for local invasion and distant metastases. It primarily affects the salivary glands, although it can also arise in other glandular tissues, such as the lacrimal glands of the eyelid. Primary cutaneous adenoid cystic carcinoma (ACC) is an extremely rare type of malignant tumor of the eyelids, accounting for just around 1% of all head and neck cancers. We present here a rare case of primary ACC in a 75-year-old woman who came with a painless, progressive nodule in her left upper eyelid. Following a preliminary diagnosis of a lymphangioma, a thorough excision was performed and the eyelid defect corrected. Histopathology showed a solid tumor predominantly of tumor cells arranged in a cribriform architecture. The PAS stain revealed granular myxoid material in the lumina, and the tumor cells stained positive for the CD117 IHC marker. Based on the correlation between immunohistochemistry and histology, adenoid cystic carcinoma of the eyelid was diagnosed.

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## 1. Introduction

Adenoid cystic carcinoma (ACC) is a type of basaloid tumor that consists of both myoepithelial and epithelial cells with a range of morphologic patterns, such as solid, cribriform, and tubular. It usually has a deadly outcome and a relentless clinical course. This rare epithelial tumor has a sluggish growth rate, and cutaneous adenoid cystic carcinoma is most commonly reported as affecting the scalp; eyelid involvement is extremely uncommon. The biggest recorded series from the USA contained 152 instances of PCACC over three decades, with an incidence rate of 0.23 per million person years.<sup>1</sup> Usually the majority of primary cutaneous ACCs of the eyelid are mistaken for chalazion; the diagnosis was made either through pathological examination or as a result of postoperative recurrence. Due to its localized aggressiveness and potential for recurrence

even after a successful surgical excision, adenoid cystic carcinoma needs long-term observation. Because of its rarity, PCACC is frequently misdiagnosed or ignored in initial examinations, emphasizing the significance of histological evaluation for appropriate diagnosis.

## 2. Case Report

A 75-year-old woman with no prior known medical history consulted an ophthalmology department with a complaint of painless nodule in the left upper that had been bothering her for the previous five years associated with difficulty in opening and shutting the eyelid. On examination, a pale pink-colored nodule measuring approximately 15x4 mm with lost cilia was found on the medial half of the left upper eyelid, and no skin ulcers were seen in relation to the indurated lump of the eyelid (Figure 1). An initial clinical diagnosis of ? lymphangioma was made since the tumor was pale pink in color, followed by excision of the tumor

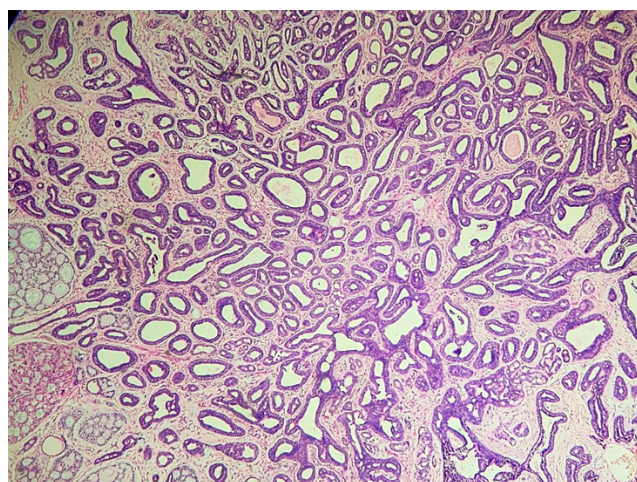
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and sent for histopathological examination. Microscopic examination showed a solid tumor consisting primarily of tumor cells grouped in bands that interacted with lumina (tubular), and a few cells are arranged in a cribriform pattern with hyalinized material inside (Figures 2 and 3).

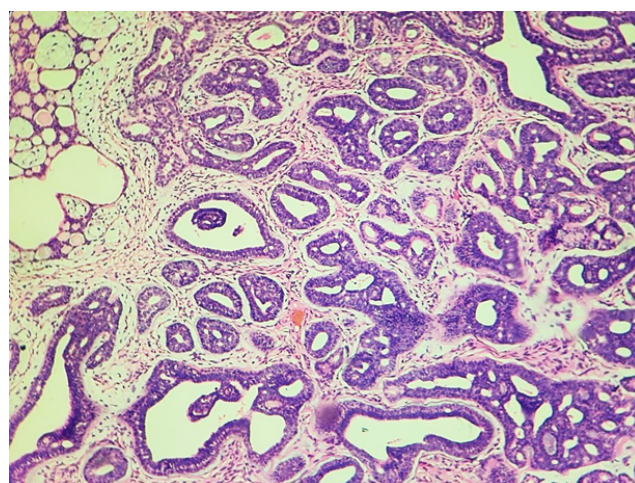


**Figure 1:** Photomicrograph showing nodule in medial half of left upper eyelid with loss of cilia measuring 15x4mm



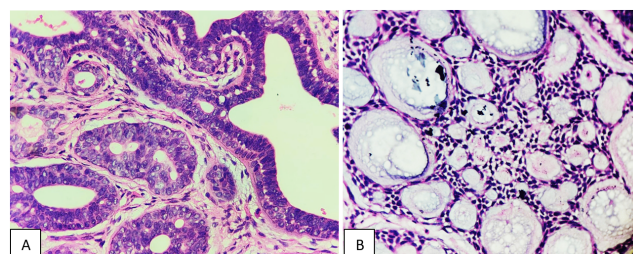
**Figure 2:** Photomicrograph showing tumour cells arranged in interlacing band with lumina H&E 4x

The cells are deep basophilic with round to oval nuclei and some with round to elongated nuclei, prominent nucleoli and moderately pale eosinophilic cytoplasm (Figure 4 A,B). The tumor also show presence of granular myxoid material in the lumina which stained positive with Periodic Acid Schiff (PAS). These cells are tested positive for CD117 immunohistochemically (Figure 5). Both histopathological and immunohistochemical evaluation confirms diagnosis of adenoid cystic carcinoma of the eyelid. The postoperative phase was uneventful, and the patient received a thorough systemic assessment



**Figure 3:** Photomicrograph showing tumour cells arranged in both tubular and cribriform pattern H&E-10x

following this diagnosis. No metastases or similar lesions were discovered throughout the systemic examination. At present, 6 months following surgery, the patient is asymptomatic but is being closely monitored due to the high chance of recurrence.

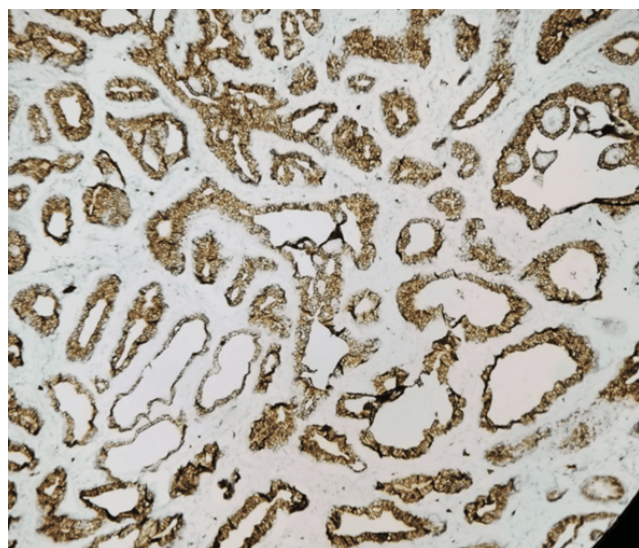


**Figure 4:** A): Photomicrograph showing tubular pattern contain simple tubules composed of inner ductal and outer myoepithelial cells H&E- 40x; B): Photomicrograph showing cribriform pattern composed of myoepithelial cells with myxoid or hyalinized globules H&E- 40x

### 3. Discussion

Adenoid cystic carcinoma (ACC) accounts for 1-2% of head and neck tumors and 10% to 15% of malignant tumors of the salivary gland.<sup>2</sup> This tumor is extremely rare to involve the skin; if present, most commonly reported at the scalp, Palpebral or eyelid localization is still unique; in this location, it can arise from the Moll's glands or lacrimal gland of the palpebral lobe, the conjunctiva's supplemental lacrimal glands, or an ectopic lacrimal glandular tissue.<sup>3</sup> When an adenoid cystic carcinoma of the lacrimal gland develops, the eye is typically pulled downward, toward the nose, and forward. It may cause eye bulging (also known as proptosis). Another characteristic of adenoid





**Figure 5:** Photomicrograph showing CD117/KIT positive in the epithelial component

cystic carcinoma is that it invades local nerves, causing pain. The most common signs of ACC of the lacrimal gland are discomfort and eye bulging. Lacrimal gland ACC (LGACC) is associated with a low future disease-free survival (DFS), and perineural invasion is considered a poor prognosis because it carries inherent risks of extending to the skull base and local recurrence.<sup>4</sup>

The involvement of the skin can be primary (as in our instance) or secondary, resulting from direct infiltration, peri-nervous extension, or cutaneous metastases of an ACC.<sup>5</sup> These tumors are characterized by a remarkable propensity for recurrence and low progression. Clinically, these tumors present as a firm painless nodule accompanied by loss of cilia or distortion.<sup>6</sup> The lesion may mimic a chalazion, palpebral sebaceous carcinoma, or basal cell carcinoma.<sup>2,7,8</sup> Even though there were no skin ulcers as in our instance, cilia loss was seen in line with the indurated nodule on the eyelid. On the other hand, in middle-aged or older patients, hard nodes that are accompanied with cilia loss or distortion or skin ulceration may indicate an underlying cancer.<sup>2</sup> Its microscopic appearance is the same as that of other localizations.<sup>9</sup> The tumor is composed of basophilic cells with cribriform, tubular, or solid architecture that reside in the mid-to-reticular dermis and are unattached to the upper epidermis or hair follicles.

The hyaline material, which is eosinophilic or basophilic mucin, fills the cystic spaces and stains positive for PAS seen in ACC. The prognosis is poorer for tumors with a primarily solid pattern among these.<sup>10</sup> In our case, tumor cells are predominantly arranged in a tubular and cribriform pattern with cystic spaces filled with hyaline material.

Histopathologically, one of the important differential diagnoses of adenoid cystic carcinoma is adenoid BCC. Clinically, they differ from one another by having uneven margins, everted edges, and a base made of a dark brown exudate with blackish incrustation. The surrounding periorbital region is swollen and edematous. Histologically, it can be distinguished by the absence of peripheral palisading of nuclei, which is observed in BCC,<sup>11</sup> and there is no continuity in ACC with the hair follicles or epidermis. There was no nuclear palisading, and neither was continuity with the epidermis or hair follicle in our instance.

Along with BCC, sweat gland carcinoma and sebaceous carcinoma are other differential diagnoses. Immunohistochemistry suggests positive expression of CD117 aids in the diagnosis of ACC, negative expression with BerEP4 rules out Basal cell carcinoma, and lack of adipophilin rules out sebaceous carcinoma.<sup>3</sup> In our instance, the tumor cells exhibited high CD117 membrane positivity, supporting the diagnosis of adenoid cystic carcinoma once more.

ACC of the eyelid has a better prognosis than ACC of the lacrimal gland, which is the next most prevalent site of ACC in the optical adnexa.<sup>6</sup> Because one of the possible explanations is that people with eyelid ACC, which usually manifests as nodules, seek medical attention sooner, and the eyelid ACC has an idle course. In our case, it was unclear where ACC originated; it might be ectopic lacrimal gland tissue, accessory lacrimal glands in the conjunctiva, or Moll's glands.<sup>12</sup>

Since ACC is an uncommon tumor of the eyelid, the best course of action is still unknown. Surgical excision is the preferred treatment for eyelid ACC, and this was done in our instance. Given the recurrent nature of ACC, long-term monitoring is necessary.<sup>12</sup> Our instance emphasizes the value of the histological examination, which was crucial to the diagnosis.

#### 4. Conclusion

This case emphasizes the need for including ACC in the differential diagnosis of eyelid malignancies, despite its rarity. Early diagnosis and a comprehensive treatment approach are essential for better outcomes. It can recur even after a successful and appropriate surgical excision, and it is marked by its local aggressiveness, so continuous monitoring in long-term follow-up is required to detect and manage possible recurrences.

#### 5. Source of Funding

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#### 6. Conflict of Interest

Nil.

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