Mucinous eccrine adenocarcinoma of the eyelid: report of 6 cases

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ABSTRACT  •  RÉSUMÉ

Objective: To report on patients with mucinous eccrine adenocarcinoma of the eyelid.
Design: Retrospective case series.
Participants: Biopsy specimens of 6 patients with eyelid neoplasm were obtained. The pathologic diagnoses were mucinous eccrine adenocarcinomas.
Methods: The medical records, including demographic, clinical, histopathologic, and follow-up information, were reviewed.
Results: Five patients underwent surgical removal of the lesion by means of Mohs micrographic surgery. Four of these patients were doing well; 1 of them, with positive margins of resection, had 2 recurrences. One case was lost to follow-up.
Conclusions: Mucinous eccrine adenocarcinoma is an uncommon adnexal tumour that can involve the eyelid, has low metastasis and mortality, but can be invasive or locally recur. Mohs micrographic surgery is a recommended treatment of mucinous eccrine adenocarcinoma of the eyelid.

Objet : Compte-rendu sur des patients atteints d’andénocarcinome eccrine mucineux de la paupière.
Nature : Rétrospective d’une série de cas.
Participants : Spécimens de biopsie des néoplasmes de 6 patients dont le diagnostic pathologique était un andénocarcinome eccrine mucineux.
Méthodes : Examen des dossiers médicaux, comprenant des données démographiques, cliniques, histopathologiques et les suivis.
Résultats : Cinq patients ont subi une opération pour retirer la lésion par la chirurgie micrographique de Mohs. Quatre d’entre eux s’en sont bien tirés; un, avec des marges de résection positives, a eu 2 récurrences. Un cas n’a pu être suivi.
Conclusions : L’andénocarcinome eccrine mucineux est une tumeur annexielle rare qui peut toucher la paupière et a peu de métastase et mortalité, mais qui peut être invasive et se reproduire localement. La chirurgie micrographique de Mohs est recommandée pour le traitement de l’andénocarcinome eccrine mucineux de la paupière.

Mucinous eccrine adenocarcinoma may occur in the periocular region. Since the initial description of this tumour in 1952,1 several individual case reports have been described. The lesion may be locally invasive and recur after incomplete excision, although it rarely metastasizes. The tumour is often clinically mistaken for other cutaneous lesions because of its variable appearance.

METHODS

After obtaining approval from the Emory University Institutional Review Board, we identified 6 cases with the pathologic diagnosis of mucinous eccrine carcinoma of the eyelid in the L.F. Montgomery Ophthalmic Pathology Laboratory, Emory University, Atlanta, Ga. All cases had been diagnosed by an experienced ophthalmic pathologist (Hans E. Grossniklaus). Demographic information, clinical manifestations, pathologic features, and follow-up data of the patients were reviewed. Hematoxylin and eosin, and periodic acid-Schiff stained sections were examined in all cases. Immunohistochemical staining for cytokeratin 7 (CK7), epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA), estrogen receptor (ER), and progesterone receptor (PR), and electron microscopic examination were performed in 1 representative case.

RESULTS

A summary of the clinical findings is shown in Table 1. The tumour was more common in women than men, and the mean age at presentation was 50 years (range 30–78). Three cases occurred in the upper eyelid and 3 in the lower eyelid. The clinical description in most cases was that of a slow-growing lesion that arose over months to years. The lesions measured from 3 × 2 to 8 × 5 mm in diameter, and most appeared as a solitary elevated nodule. Five patients underwent surgical removal of the tumour. Examination of the surgical margins of resection in 4 of the patients undergoing Mohs micrographic surgery showed no evidence of tumour, and all of them have remained tumour free after...
3 months to 4 years of follow-up. One of the 5 patients, with positive margins of resection, had 2 recurrences, which were surgically excised. Information on the treatment and follow-up of 1 patient (case 2) was unavailable.

The pathologic features of the tumour are illustrated by case 5 (Figs. 1–3). All cases included dermis infiltrated with nests of tumour cells floating in a sea of mucin, as demonstrated by alcian blue and colloidal iron stains. The tumour cells exhibited mildly pleomorphic, vesiculated nuclei, occasional prominent nucleoli, and scanty, eosinophilic cytoplasm. Several tumours formed cysts lined by a proliferation of neoplastic cells exhibiting a papillary configuration or tubules with central lumens. Immunohistochemical stains in case 5 were positive for CK7, EMA, CEA, ER, and PR, and ultrastructural examination showed a tumour composed of stratified cells with centrally placed, round to oval nuclei, margined chromatin, and variable amounts of cytoplasm, forming glandular structures with lumens. These cells exhibited intercellular junctions, scattered mitochondria, glycogen granules, and microvilli projecting toward the lumens of the glandular structures.

**Conclusions**

There is controversy regarding whether mucinous eccrine carcinoma arises in a sweat gland or apocrine gland. This tumour has been described as arising in the eyelid, mostly in individual case reports.2–7 When the tumour arises in the eyelid, it is usually a unilateral lesion, although bilateral cases have been reported.4 The tumour usually appears as a solitary, asymptomatic, slow-growing nodule, cyst or ulcer. The contour of the lesion may be elevated, pedunculated, or papillomatous, and the surface may be smooth, irregular, or crusted. The colour of the lesion may be tan, grey, blue, or brown. Usually the lesion is less than 3 mm in diameter, although a 20 mm diameter has been previously reported.4 Because of the variable clinical appearance, clinical diagnoses have included chalazion, epidermoid cyst, hemangioma, myxoma, lipoma, papilloma, keratoacanthoma, pyogenic granuloma, sebaceous cyst, sebaceous carcinoma, squamous cell carcinoma, basal cell carcinoma, adenoid cystic carcinoma, malignant melanoma, and Kaposi sarcoma.3–7

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age, yr</th>
<th>Sex</th>
<th>Location</th>
<th>Size</th>
<th>Clinical appearance</th>
<th>Clinical diagnosis</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>78</td>
<td>F</td>
<td>Left upper eyelid</td>
<td>5 × 5 mm</td>
<td>Round elevated nodule</td>
<td>Papilloma</td>
<td>MMS</td>
<td>8 years, 2 recurrences, no metastasis</td>
</tr>
<tr>
<td>2</td>
<td>61</td>
<td>F</td>
<td>Right lower eyelid</td>
<td>5 × 5 mm</td>
<td>Elevated nodule</td>
<td>Neoplasm</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>3</td>
<td>69</td>
<td>F</td>
<td>Left lower eyelid</td>
<td>3 × 3 mm</td>
<td>Elevated nodule</td>
<td>Chalazion</td>
<td>MMS</td>
<td>4 years, no recurrence or metastasis</td>
</tr>
<tr>
<td>4</td>
<td>30</td>
<td>M</td>
<td>Left upper eyelid</td>
<td>4 × 3 mm</td>
<td>Elevated nodule</td>
<td>Pyogenic granuloma</td>
<td>MMS</td>
<td>2 years, no recurrence or metastasis</td>
</tr>
<tr>
<td>5</td>
<td>34</td>
<td>M</td>
<td>Right lower eyelid</td>
<td>8 × 5 mm</td>
<td>Red nodule</td>
<td>Hemangioma</td>
<td>MMS</td>
<td>9 months, no recurrence or metastasis</td>
</tr>
<tr>
<td>6</td>
<td>64</td>
<td>F</td>
<td>Left upper eyelid</td>
<td>3 × 2 mm</td>
<td>Lump</td>
<td>Basal cell carcinoma</td>
<td>MMS</td>
<td>3 months, no recurrence or metastasis</td>
</tr>
</tbody>
</table>

Note: y, year; F, female; M, male; MMS, Mohs micrographic surgery; N/A, not available.
Immunohistochemical stains for mucinous eccrine carcinoma may be positive for cytokeratins (CK7, CAM5.2), CEA, EMA, mucus-associated peptides of the trefoil factor family (TFF1 and 3), and tumour-associated glycoprotein. Characteristic electron microscopic features include dark cells at the periphery with mucin production and pale cells with little or no mucin production centrally placed in each nest of cells. Complete excision of this tumour is recommended, varying from excision with clinically visible margins to wide local excision. We recommend complete surgical excision with histologic confirmation of negative surgical margins of resection. This may be accomplished by Mohs micrographic surgery.

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REFERENCES

Keywords: adenocarcinoma, mucinous, eyelid, Mohs surgery