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Mucinous Eccrine Adenocarcinoma: Report of Six Cases

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Abstract

Six patients with mucinous eccrine carcinoma of the eyelid were identified in the case records of the L.F. Montgomery Laboratory. Five of these patients had undergone Mohs micrographic surgery after their initial diagnosis. Follow-up ranged from three months to eight years. Four patients with negative margins of excision had no evidence of recurrence, one patient was lost to follow-up and one patient with positive margins of excision had two recurrences after incomplete excision of the tumor. We recommend surgical excision of mucinous eccrine carcinoma of the eyelid with histologic monitoring of the surgical margins of resection.

Mucinous eccrine adenocarcinoma may occur in the periocular region. Since the initial description of this tumor in 1952^{1,2}, there have been several individual case reports described. The lesion may be locally invasive and recur after incomplete excision, although it rarely metastasizes. The tumor is often clinically mistaken for other cutaneous lesions due to its variable appearance.

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

A summary of the clinical findings is shown in Table 1. The tumor 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, two in the lower eyelid and one in the periocular tissue. 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 Mohs micrographic surgical removal of the tumor. Examination of the surgical margins of resection in four of those patients showed no evidence of tumor and all of those patients have remained tumor free with 3 months to 2 years follow-up. One of those patients had positive surgical margins of resection and had

two recurrences which were surgically excised. The treatment and follow-up for one patient (case 2) was unavailable.

The pathologic features of the tumor are illustrated by case 5 (Figs 1–3). In all cases included dermis infiltrated with nests of tumor cells floating in a sea of mucin demonstrated by alcian blue and colloidal iron stains. The tumor cells exhibited mildly pleomorphic, vesiculated nuclei, occasional prominent nucleoli and scanty, eosinophilic cytoplasm. Several tumors formed cysts lined by a proliferation of neoplastic cells exhibiting a papillary configurations or tubules with central lumens. Immunohistochemical stains were positive for CK7, EMA, CEA, ER, and PR in ?cases. Ultrastructural examination of case 5 showed a tumor 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 microvillae projecting toward the lumens of the glandular structures.

There is controversy regarding whether mucinous eccrine carcinoma arises in a sweat gland or apocrine gland.³ This tumor has been described to arise in the eyelid, mostly in individual case reports.^{4–9} When this tumor arises in the eyelid, it is usually a unilateral lesion, although bilateral cases have been reported.⁸ The tumor usually appears as a solitary, asymptomatic, slow-growing nodule, cyst or ulcer.^{6–8} The contour of the lesion may be elevated, pedunculated, or papillomatous and the surface may be smooth, irregular or crusted. The color of the lesion may be tan, grey, blue or brown. The lesion is usually less than 3mm in diameter, although a 20mm diameter tumor has been previously reported.⁸ Due to 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.⁹

Immunohistochemical stains for mucinous eccrine carcinoma may be positive for cytokeratins (CK7, CAM5.2), carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), mucous-associated peptides of the trefoil factor family (TFF1 and 3), and tumor-associated glycoprotein (TAG-72).^{10,11} 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.^{12,13} Complete excision of this tumor is recommended, varying from excision with clinically visible margins to wide local excision.^{6,7} We recommend complete surgical excision with histologic confirmation of negative surgical margins of resection. This may be accomplished by Mohs micrographic surgery.¹⁴ There is no evidence that a sentinel lymph node biopsy is indicated for patients with mucinous eccrine carcinoma of the eyelid.

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Figure 1.
The right lower eyelid of a patient with mucinous eccrine adenocarcinoma shows a slow-growing elevated nodule.

Figure 2.

A photomicrograph shows a section of mucinous eccrine adenocarcinoma of eyelid with nests of tumor cells with mildly pleomorphic nuclei, occasional prominent nucleoli and scanty, eosinophilic cytoplasm floating in a sea of mucin (hematoxylin-eosin, original magnification $\times 10$).

Figure 3.

A photomicrograph shows a section of mucinous eccrine adenocarcinoma of eyelid stained with colloidal iron demonstrates the presence of mucin (original magnification $\times 25$).

Figure 4. Electron microscopic examination shows the tumor cells with centrally placed, round to oval nuclei, marginated chromatin and variable amounts of cytoplasm, exhibiting intercellular junctions, scattered mitochondria, glycogen granules and microvilli projecting toward the lumens of the glandular structures.

Table 1

Clinical Characteristics of Mucinous Eccrine Carcinoma of the Eyelid

case	age (yrs)	sex	location	size (mm)	clinical appearance	clinical diagnosis	treatment	follow-up
1	78	f	upper eyelid	5×5	round elevated nodule	papilloma	Mohs	8 years, two recurrences, no metastasis
2	61	f	eyelid	5×5	elevated nodule	neoplasm	n/a	n/a
3	69	f	lower eyelid	3×3 mm	elevated nodule	chalazion	Mohs	4 years, no recurrence or metastasis
4	30	m	upper eyelid	4×3 mm	elevated nodule	pyogenic granuloma	Mohs	2 years, no recurrence or metastasis
5	34	m	lower eyelid	8×5 mm	red nodule	hemangioma	Mohs	9 months, no recurrence or metastasis
6	64	f	upper eyelid	3×2 mm	lump	basal cell carcinoma	Mohs	3 months, no recurrence or metastasis

m=male, f=female, Mohs=Mohs micrographic surgery, n/a=not available