Primary cutaneous mucinous carcinoma of the eyelid treated with Mohs surgery

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INTRODUCTION

Primary mucinous carcinoma (PCMC) is a rare, slow-growing mucinous variant of sweat gland tumors. Clinically, PCMC presents with a variety of morphologies and most commonly affects the eyelid or brow followed by the neck and scalp.[1] According to a recent meta-analysis by Kamalpour et al,[1] approximately 215 cases of PCMC have been reported from 1952 to 2010, with an increased prevalence in whites, elderly, and men. Most of these cases (136 of 159) were treated with traditional surgical excision with 1-cm minimum margins, and only a limited number (15 of 159) were treated with MMS. Of the patients treated with Mohs micrographic surgery (MMS), 2 (13%) had recurring disease with no reported metastases. In contrast, 46 patients (34%) either had recurrence of disease or disease that metastasized when treated with wide local excision. Overall, the rate of PCMC metastasis was low (6.1%), and the rate of local recurrence was high (19.6%). Mean follow-up time was 30.1 (standard deviation, 39.4) months for patients treated with excision and 23.1 (standard deviation, 17.5) months for patients treated with MMS.[1] We describe a patient with PCMC of the eyelid treated with MMS who has shown no evidence of recurrence for a longer follow-up period than currently reported in the literature.

CASE REPORT

The patient was a 50-year-old white man with no personal or family history of skin cancer who presented with a 5-year history of a slow-growing, asymptomatic lesion on the right upper eyelid. The lesion was removed twice in the past and recurred both times.

Abbreviations used:

MMS: Mohs micrographic surgery
PCMC: Primary mucinous carcinoma

Clinical examination found a 1.0- × 0.5-cm pink papule located on the upper portion of the right eyelid (Fig 1, *A*). There was no appreciable lymphadenopathy. Histopathologic examination found a dermal tumor consisting of basaloid islands floating in large pools of mucin separated by fibrous septae (Fig 2, *A* and *B*). The tumor cells were cuboidal and formed tubular spaces. The nuclei showed minimal pleomorphism. These findings were consistent with an invasive mucinous carcinoma.

To investigate the possibility that this lesion represented a metastasis to the skin, the patient had a complete physical examination and a computed tomography of the head, neck, chest, abdomen, and pelvis. Colonoscopy results from the prior year were reviewed; no abnormalities were detected. Orbital magnetic resonance imaging showed no evidence of orbital extension. The patient was treated with MMS requiring 4 stages resulting in 2- × 1-cm full-thickness eyelid defect (Fig 1, *B*), which was repaired with a reverse Tenzel and an upper lid sliding tarsoconjunctival flap with full-thickness skin graft from the left upper lid by plastic surgery. He is 36 months postsurgery with no evidence of recurrence.

DISCUSSION

PCMC is a rare, slow-growing mucinous sweat gland tumor of controversial origination, with authors historically favoring eccrine differentiation.[2] However, recent opinions shift toward apocrine differentiation.[2,3] Although the role of ultraviolet

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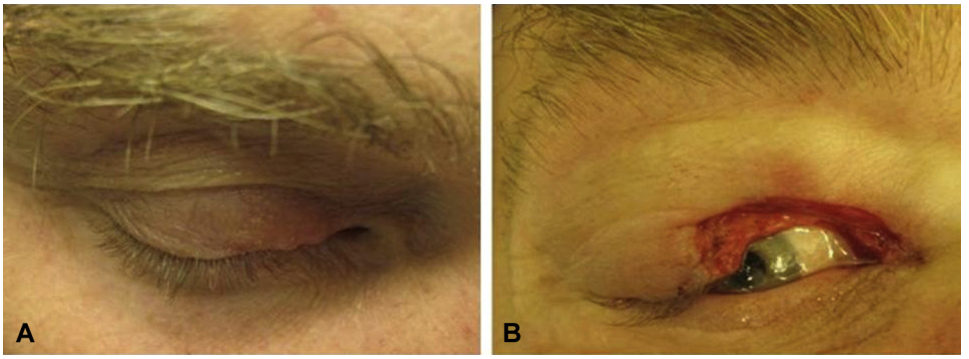

Fig 1. Clinical presentation of primary cutaneous mucinous carcinoma of the right upper eyelid before (**A**) and after (**B**) treatment with 4 stages of Mohs micrographic surgery.

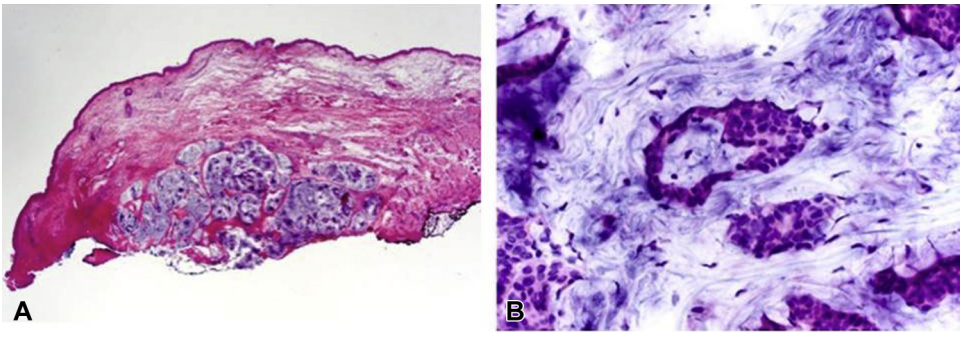

Fig 2. Primary cutaneous mucinous carcinoma with islands of bland basaloid cells arranged around ducts, floating in a pool of mucin surrounded by fibrous septae seen with hematoxylin-eosin stain on frozen section under low power (**A**) and high power (**B**).

radiation exposure has yet to be elucidated for cutaneous appendageal tumors, the anatomic predilection of PCMC and higher prevalence in elderly and in those with less skin pigmentation are consistent with the hypothesis that ultraviolet radiation may be a risk factor for its development.[4] Clinically, PCMC primarily affects the head and neck. The lesions have been described as nontender, soft, round, well-circumscribed red, pink, gray, blue, purple, or flesh colored nodules with or without telangiectasias or ulceration.[2,5] Because of the lack of unique morphologic characteristics, the diagnosis is made on histologic grounds. The differential diagnosis of PCMC includes epidermal inclusion cyst, hemangioma, chalazion, basal cell carcinoma, squamous cell carcinoma, melanoma, sebaceous carcinoma, and, most importantly, cutaneous metastasis from an alternative primary adenocarcinoma.[5] Because PCMC shares many of clinical, histologic, and immunohistochemical findings of the adenocarcinomas arising from more common sites such as the breast, gastrointestinal tract, lung, ovary, and prostate,[2,3,5] a complete history and physical examination with emphasis on lymph nodes and breast examination is suggested. Colonoscopy or barium enema, mammography, and computed tomography scan of the head, neck, chest, abdomen, and pelvis should also be considered.

Histologically, PCMC is typically avascular and composed of small, irregular clusters of epithelial cells with few mitotic figures, round to cuboidal nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm. The cells are arranged in small glands or islands surrounded by abundant mucin.[2,5] Careful analyses of primary mucinous carcinomas of the skin have found a spectrum of microscopic heterogeneity analogous to that observed in the breast. There are pure types (mucinous carcinoma without an invasive ductal component), mixed types (mucinous carcinoma plus invasive ductal component), and mucocelelike configurations.[3] Most pure type tumors appear to emanate from in situ lesions. As the neoplasm develops, accumulation of mucin leads to the distention of the ducts and subsequent disruption of the myoepithelial layer, its misplacement, and, finally, its complete disappearance. The result is a pure mucinous carcinoma with typical histology of epithelioid nests in a pool of mucin with no identifiable in situ component.[3]

PCMC consists of a diastase-resistant, hyaluronidase-resistant, periodic acid Schiff–positive sialomucin.[5] Alcian blue is positive at pH 2.5 but negative at pH 0.4.[2] PCMC is ER+/PR+/CK7+/CK20−, which is similar to mammary adenocarcinomas but different from gastrointestinal adenocarcinomas, which are CK7−/CK20+.[3]

Most PCMC cases are managed conventionally with wide local excision, and although PCMC metastasis is rare, local recurrence is quite common. Interestingly, lower rates of metastasis and recurrence have been reported among younger patients and Asians.[1] To date, PCMC has been resistant to both chemotherapy and radiation.[5] MMS has been suggested as a more favorable treatment option than wide excision given the complete margin control, cosmetic considerations, and lower reported rates of metastasis and recurrence. Our findings support this claim, as our patient with PCMC of the eyelid treated with MMS has shown no evidence of recurrence or metastasis for a longer follow-up period (36 months) than currently reported in the literature.

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