

Primary Cutaneous Mucinous Carcinoma Treated with Micrographic Surgery

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Introduction

Primary mucinous carcinoma of the skin (PMCS) is a rare, slow growing tumor with a high recurrence rate and occasional metastasis. It most commonly presents as an asymptomatic nodule on the face, particularly the eyelids.¹ PMCS is resistant to radiation and chemotherapy. Historically, a wide local excision has been the treatment of choice, but it has high recurrence rates. While no current definitive treatment guidelines are available, limited data suggests that Mohs micrographic surgery (MMS) may result in significantly lower recurrence rates than excision.² We report a case of recurrent PMCS treated with MMS.

Case Report

A 48-year-old African American male presented with a two-year history of a subcutaneous, asymptomatic growth below his right eye. On clinical exam, a 3x2x1 cm non-tender, mobile, subcutaneous nodule was noted on the right lower eyelid and the zygomatic arch. He underwent excisional biopsy by plastic surgery two months later. Pathology showed an island of CK7 positive and CK 20 negative basaloid cells in the mucinous stroma consistent with primary mucinous carcinoma (figures 1, 2 & 3). The

tumor was approaching the surgical margins. The patient was lost to follow up and presented fifteen months later with a recurrent growth in the right infraorbital area.

CT imaging showed a soft tissue mass without bony involvement. An MRI of the orbits, face and neck showed a 3.8x7x1.1 cm right lower eyelid lesion with no bony involvement, as well as prominent bilateral neck lymph nodes and a thyroid mass. Chest X-ray and metastatic workup were unremarkable. A fine needle aspiration of the thyroid mass and cervical lymph node biopsy were both negative for metastasis. After a tumor board discussion MMS was recommended for complete resection of the tumor with adjuvant radiation therapy.

MMS was performed in 2 stages in an attempt to clear the deep margins (first stage shown in figure 4). The second stage showed clear margins without any residual tumor (the final MMS defect shown in figure 5). The orbital septum was violated medially. The patient underwent reconstruction by plastic surgery two

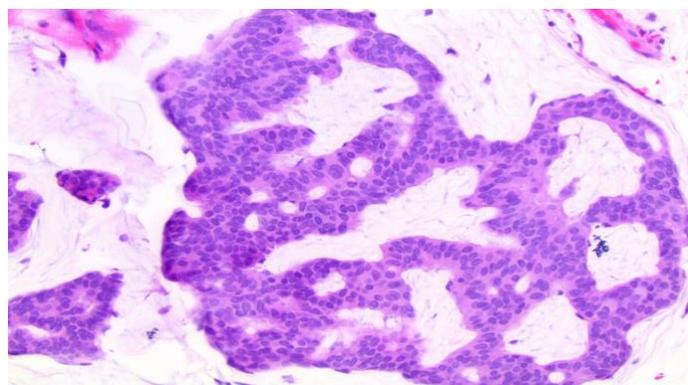


Figure 1. H&E stain at 10x showing islands of basaloid cells surrounded by mucinous stroma;

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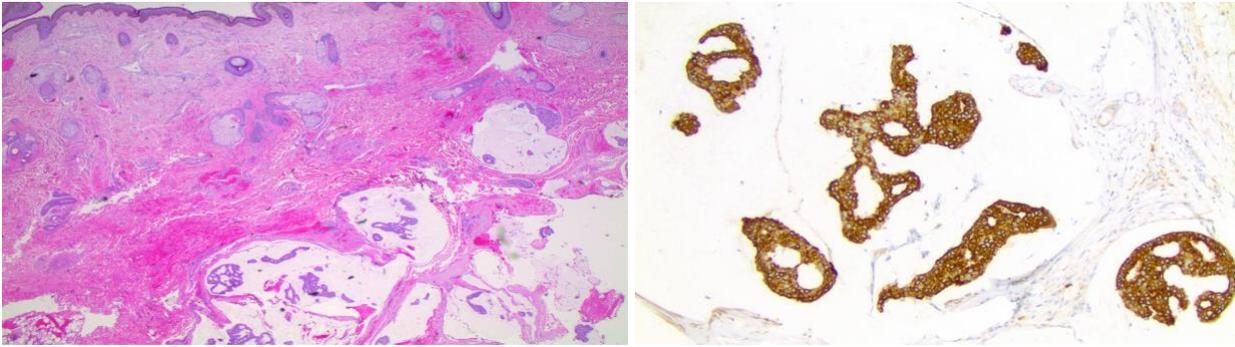


Figure 2. Deep dermal component at 2x magnification; **Figure 3.** CK7 positive tumor cells at 20x

days later with right cheek fasciculocutaneous flaps, right inferior orbicularis oculi muscle flap, medial canthopexy and lateral canthoplasty. In addition, the patient received 24 radiation treatments as per radiation oncology recommendations to prevent recurrence. Eight month follow up showed no recurrence.

Discussion

PMCS is a rare adnexal neoplasm of the mucinous sweat gland, which is often mistaken for metastasis from extracutaneous sites, such as gastrointestinal tract or breast. Clinical presentation may vary, with the classic presentation of nontender, well-circumscribed nodules of varying color.¹ The most common location of the neoplasm is the eyelid and eyebrow (49.7%), followed by non-periorbital face & neck (19.5%) and scalp (17%).¹ Thorough physical examination must be performed with a focus on lymph node and breast exam to exclude metastasis and extracutaneous origins, respectively.

The origin of PMCS is controversial. It was historically thought to be a variant of eccrine differentiation, but is now considered a tumor of apocrine differentiation.² Histology shows dermal avascular, irregular clusters of basaloid cells with rare mitosis surrounded by abundant mucin.³ There are three types of PMCS: pure, mixed, and mucocele.⁶ Pure types originate as in-situ and lack an invasive

ductal component. As mucin builds up and distends the duct, it displaces the myoepithelial layer resulting in a classic histopathology of epithelioid nests in a pool of mucin without an identifiable in-situ component. Mixed type is similar to the pure, but with an invasive ductal component. Mucocele type displays configurations which are diastase and hyaluronidase resistant and can mimic a benign breast mucocele as well as a ductal breast adenocarcinoma.⁴

Workup for suspected PMCS includes colonoscopy, barium enema and mammography, to evaluate for extracutaneous origin. CT imaging may be necessary to exclude metastases.² Unlike PMCS, cutaneous metastases from mammary adenocarcinoma exhibit a predilection for the chest and axilla rather than the face.² Treatment has conventionally been wide local excision, but there is a high rate of recurrence, ranging from 34-36%.⁵ Metastases of the primary lesion are less common, occurring in about 14% of cases.⁶ PMCS is resistant to chemotherapy and radiation.

In a review of the literature, there are a few case reports demonstrating the success of MMS. A small case series of 6 patients with mucinous carcinoma treated with MMS showed no recurrence. The 20 to 36 month follow up indicated that long-term recurrence rates were lower (13%) than



Figure 4. First stage of MMS ; **Figure 5.** Second stage, final defect of MMS

recurrences after a wide local excision (34%).⁷

In a meta-analysis of PMCS cases reported between 1952 and 2010, a total of 59 cases were found. Caucasian race, advanced age, and male gender were demonstrated as the most likely risk factors. One hundred and thirty six of the patients in the meta-analysis were treated with wide local excision with 1 cm margins and 15 were treated with Mohs surgery. Of those treated with Mohs, 2 (13%) had recurrence with no reported metastases. In comparison, those with wide local excision, 46 (34%) had recurrence or metastases. Overall recurrence was 19.6%.²

Although there are no current official guidelines for treatment of PMCS, the data indicates that MMS may be a superior treatment with lower recurrence rates compared to wide local excision. While additional studies are needed, our experience thus far supports favorable outcomes with the use of MMS in treatment of PMCS.

References

1. Papalás JA, AD Proia. Primary mucinous carcinoma of the eyelid: a clinicopathologic and immunohistochemical study of 4 cases

and an update on recurrence rates. *Arch Ophthalmol.* 2010; 128(9):1160-1165.

2. Adefusika JA, JD Pimentel, RN Chavan, and JD Brewer. Primary mucinous carcinoma of the skin: the Mayo Clinic experience over the past 2 decades. *Dermatologic Surgery.* 2015 Feb 1; 41(2):201-8.

3. Breitling L, L Christensen, K Dahlstrøm, V Breitling, and JF Winther. Primary mucinous carcinoma of the skin: a population-based study. *Int J Derm.* 2008 Mar 1; 47(3):242-5.

4. Kazakov DV, S Suster, PE LeBoit, et al. Mucinous carcinoma of the skin, primary, and secondary: a clinicopathologic study of 63 cases with emphasis on the morphologic spectrum of primary cutaneous forms: homologies with mucinous lesions in the breast. *Am J Surg Pathol* 2005; 29 (6) 764- 782

5. Marra DE, CF Schanbacher, and A Torres. Mohs micrographic surgery of primary cutaneous mucinous carcinoma using immunohistochemistry for margin control. *Dermatologic surgery.* 2004 May 1; 30(5):799-802.

6. Snow SN, GT Reizner. Mucinous eccrine carcinoma of the eyelid. *Cancer.* 1992;70:2099-2104.

7. Chavez A, K Linos, FH Samie. Primary cutaneous mucinous carcinoma of the eyelid treated with Mohs surgery. *JAAD case reports* 2015 Mar; 1(2):85.