BRIEF ARTICLES

Primary Mucinous Carcinoma of the Nasal Bridge

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ABSTRACT

Mucinous carcinoma is a rare appendage tumor that is resistant to surgery, chemotherapy, and radiotherapy. It has an unremarkable appearance and must be distinguished as a primary tumor or as a metastasis from distant sites. Detailed imaging studies and immunohistochemical investigation are often essential. Here, we report a case of surgery-refractory primary mucinous carcinoma on the left nasal bridge in a 62-year-old female that ultimately required removal by Mohs surgery.

INTRODUCTION

Primary cutaneous mucinous carcinoma (PCMC) is an extremely rare appendage tumor with an incidence rate of less than 0.1 cases per million individuals.[1] It is often clinically missed due to its unremarkable presentation as a small indolent, asymptomatic nodule that may be indurated or ulcerated. Mucinous carcinoma is a malignant tumor with a low rate of metastasis, but high rate of recurrence. It can rarely arise as a primary lesion from the skin or as a metastatic lesion from distant sites, most commonly the breast or gastrointestinal tract.[2] It is clinically impossible to differentiate between PCMC or metastatic cutaneous mucinous carcinoma (MCMC) and detailed imaging studies and immunohistochemical investigations are usually required to definitively diagnose PCMC.[1]

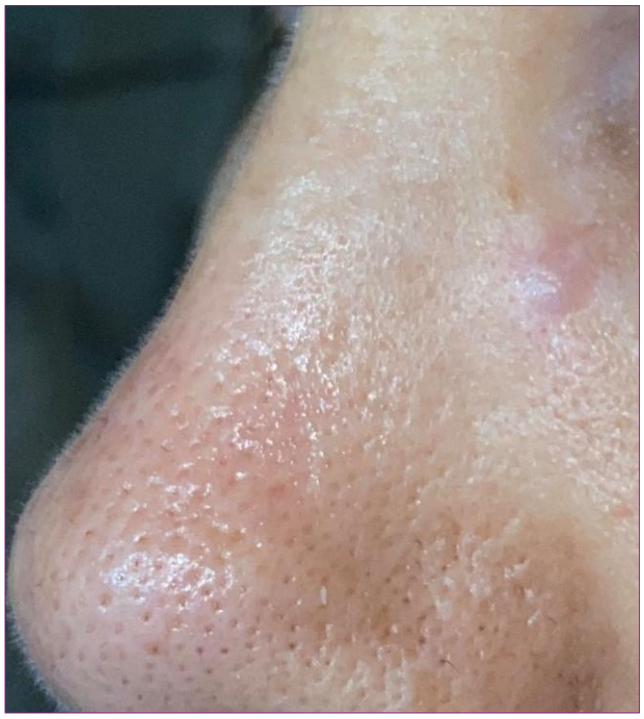
There is no standard of care established for treatment of PCMC and although rare, distant metastasis is highly resistant to both chemotherapy and radiotherapy.[3] In this report, we highlight a case of PCMC on the nasal bridge that was refractory to two previous surgical interventions and ultimately treated successfully with Mohs Surgery.

CASE PRESENTATION

A 62-year-old Iranian female presented with complaint of a cystic lesion on her left nasal bridge. The lesion was previously removed by two outside dermatologists but has since recurred. Previous pathology reports and medical records were unavailable for review. Past medical history if significant for breast cancer treated with chemotherapy and radiotherapy. Physical exam showed a well-demarcated nodule on the left nasal bridge (Figure 1). Excisional biopsy of the lesion was performed.

The lesion was described microscopically as neoplasm in the dermis comprised of islands

Figure 1. Mucinous Carcinoma Presenting as a Well-Demarcated Nodule on the Left Nasal Bridge.



of epithelioid cells with abundant mucin around and inside the islands (Figure 2). The preliminary diagnosis was probable primary cutaneous mucinous carcinoma.

Since the patient was already established with an oncologist and monitored for breast cancer treatment and signs of metastasis, other sources of malignancy were excluded. Diagnosis of PCMC was established based on history and histologic findings. Due to the lesion's resistance to surgical excision and extension beyond the deep surgical margin, the patient was referred to a Mohs surgeon for removal of the remaining lesion.

DISCUSSION

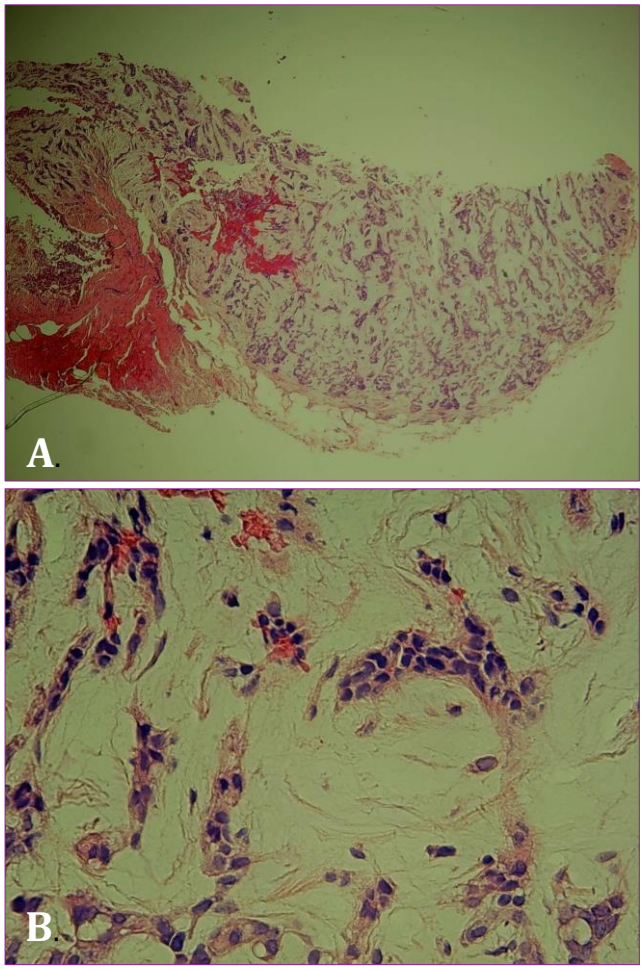
PCMC is a rare appendage tumor of sweat glands with an estimated 200 cases reported in literature, with less than 150 reported in English.[5] The abundance of mucin in PCMC interferes with nutrition of the tumor cells and is thought to be responsible for the slow growing course of the lesions.[5] Due to its indolent course, PCMC can be easily overlooked, and patients often present to the clinic after noticing its gradual enlargement.

Affected individuals usually present in their 50-70s, affecting twice as many men than women. Meta-analysis by Kamalpour *et al* found that white patients (77.2%) are disproportionally more affected when compared to Asian (12.7%) or African (10.1%) patients.[6] PCMC commonly arises in the periorbital region followed by non-periorbital regions of the face and neck, scalp, axilla and genital areas. The lesion size varies from 1.0-8.0 cm, averaging 1.5 cm in diameter.[6]

PCMC can have wide-ranging appearances. Most often, it presents as a well-circumscribed, slow-growing, painless, firm or soft nodule, but can also present as papillomatous or pedunculated lesions.[1,7] They may also be ulcerated, indurated, crusted with telangiectasias or appear as cysts.[7] The color of the lesions also varies greatly – ranging from flesh colored to grey, purple, red or blue tinged.[1] Based on the clinical exam, several lesions such as cysts, basal cell carcinoma, keratoacanthoma, nevus, apocrine hidrocystoma, or even Kaposi sarcoma can be on the differential. Since this lesion has no characteristic physical or clinical features, histological examination is required for nearly all diagnosis of PCMC.

The WHO has classified these sweat gland tumors as both apocrine and eccrine; however, the prevailing theory leans towards eccrine differentiation based on immunohistochemical and ultrastructural features.[9] Histologically, PCMC is described as nests of neoplastic epithelial cells floating

Figure 2. (**A**) Neoplasm in the dermis comprised of islands of epithelioid cells with abundant mucin around and inside the islands (H&E, 40x). (**B**) Individual clusters of epithelial cells surrounded by mucin (H&E, 400x).



in mucinous lakes.[9] It is nearly impossible to histologically differentiate PCMC from MCMC; thus, immunohistochemical and imaging studies, such as PET-CT, x-ray, CT and ultrasound are vital for determining whether the lesion is primary or secondary.[3]

MCMC most commonly originates from the breast, gastrointestinal tract, salivary glands, lacrimal glands, nose, paranasal sinuses, bronchi, renal pelvis and ovaries. MCMC from the breast and gastrointestinal tract can mimic clinical appearance of PCMC; thus, it is of special importance to distinguish PCMC from MCM from these sites.[10] Absence of expression for cytokeratin (CK) 20 can be used to exclude diagnosis of metastatic colorectal mucinous carcinoma.[11] One study of 5 PCMC samples found that positive staining for CK7, p63, and CK5/6 was indicative of PCMC, with CK7 being most indicative.[11] In our patient, immunohistochemical staining was not used, but given a history of recurrent mucinous carcinoma and no other source of metastasis, PCMC is the most likely diagnosis.

PCMC generally has a good prognosis. Most forms of metastasis are through direct invasion of local regions; however, direct invasion into lymphatics, skeletal muscle, bone, dura, and periosteum is also possible.[12] There are currently no guidelines for treatment of PCMC. One meta-analysis found that in 159 cases of PCMC with follow up, the cases treated with Mohs surgery with a mean follow up time of 23.1 months had a 13% recurrence rate and no cases of metastasis.[6] The study also found that PCMC treated with excision with a mean follow up time of 30.1 months showed that 34% recurred or metastasized. Our patient's PCMC required a total of four surgical interventions for resolution. This can be financially costly and have a negative effect on patient's self-image and cosmetic appearance. Therefore, we believe that Mohs surgery is the preferred treatment after initial diagnosis of PCMC. Radiation and chemotherapy are not advised for treatment as PCMC have been reported to be resistant to both modalities.[4]

CONCLUSION

PCMC is a rare, malignant appendage tumor with an unremarkable clinical appearance. Early recognition, proper work-

up and treatment of this highly recurrent, chemotherapy and radiotherapy-resistant lesion is essential to avoid potential metastasis. We hope that this case report contributes to the future development of diagnosis and treatment guidelines for PCMC.

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