Primary cutaneous mucinous carcinoma: A rare entity

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ABSTRACT

Primary mucinous carcinoma of the skin is a rare adnexal tumor of sweat gland origin. A case report is presented of a 70-year-old male, who presented with a slowly growing mass near the lateral canthus of his left eye. The case was clinically diagnosed as a fibroma. An excisional biopsy of the lesion revealed mucinous carcinoma of the skin. Investigations excluded the possibility of metastatic mucinous carcinoma. Thus, the lesion in the lateral canthus region was diagnosed as Primary Mucinous Carcinoma of the skin, a rare site of occurrence.

Key words: Primary mucinous carcinoma, skin, sweat gland

INTRODUCTION

Primary cutaneous mucinous carcinoma is a rare pathological entity. It is an adenocarcinoma of the eccrine glands. There have been sporadic cases reported since it was first described by Lennox et al., in 1952. We report an additional case of PMC of the skin in a 70-year-old male presenting with a swelling in the lateral canthal region of his left eye, and review the pertinent literature.

CASE REPORT

A 70-year-old male noticed a swelling near the left canthus since one-and-a-half years. The swelling was gradually increasing in size. On examination, a raised, freely mobile, firm skin mass of approximately 3.0 x 2.5 cm was present on the lateral canthal region of the left eye. The patient’s extraocular eye movements were unaffected. Examination revealed no other suspicious skin lesions or palpable lymph nodes. The case was clinically diagnosed as fibroma. Excision biopsy was done and the specimen was sent for histopathological examination. Gross examination revealed a well-circumscribed, spherical soft tissue mass, partially skin covered, measuring 2 cm in diameter. The cut surface of the tumor had a gray-brown gelatinous appearance [Figure 1]. Microscopically, the sections revealed a tumor in the dermis composed of tumor cells arranged in nests, glands, and cribriform patterns [Figure 2]. The individual tumor cells were columnar and had a hyperchromatic nucleus. The cells were seen floating in large pools of mucin separated by thin fibrovascular septa [Figure 3]. The mucin was periodic acid schiff (PAS) positive and diastase-resistant. [Figure 4] Further investigations, including a colonoscopy, ultrasonographic examination of the abdomen, and computerized tomography (CT) scans of the chest, abdomen, and pelvis were found to be normal. Thus, the lesion in the skin was reported to be PMC of the skin.

DISCUSSION

Primary mucinous carcinoma of the skin (PMCS) is a rare adnexal neoplasm with sweat gland differentiation. This tumor was first described by Lennox et al., in 1952, and later designated by Mendoza and Helwig, in 1971. Breiting et al. recently studied approximately 200 cases of PMCS documented in the literature and found that the mean annual age standardized incidence of PMCS during the period 1978 to 2003 was 0.07 per million person-years.

Although some debate exists as to the apocrine or eccrine origin of this tumor, most authors favor the eccrine differentiation based on the evidence obtained from the immunohistochemical studies and electron microscopic ultrastructural analysis. It has been suggested that PMCS may develop as a progression from abnormal apocrine or
eccrine ducts, analogous to the progression seen in the mucinous carcinoma of the breast. A ductal in-situ component can occasionally be seen ranging from ductal hyperplasia to frank carcinoma in-situ. Rosai originally suggested that the ductal proliferation continues until the overproduction of mucin results in islands of tumor cells breaking off and ‘floating’ in the mucinous pools.\(^6\)

Primary mucinous carcinoma of the skin is slightly more common in men than women and typically affects people in the age range of 50 to 70 years. The large majority of specimens of PMCS arise on the face, especially on the eyelids and scalp. The primary lesion of Mesenchymal chondrosarcoma (MCS) is often solitary, and the history is often of a small cutaneous lesion that grows slowly for many months or years and suddenly enlarges. By the time it is first brought to medical attention, the neoplasm is a nodule that varies in diameter from about 0.7 – 8.0 cm.

Mucinous carcinoma rarely originates in the skin; the majority of examples in the skin are actually metastatic to it.\(^6\) Common sites of origin of mucinous carcinomas are the breast, gastrointestinal tract, salivary glands, lacrimal glands, nose, paranasal sinuses, bronchi, renal pelvis, and ovary.\(^6\) Metastatic lesions from the breast or colon are most likely to mimic mucinous carcinoma of the skin.\(^7\) Differentiating primary mucinous carcinoma from metastatic tumors, particularly from these sites, can be difficult. The absence of expression of CK20 by immunohistochemical staining may exclude the diagnosis of metastatic colorectal mucinous carcinoma.\(^7\) Differentiating from breast carcinoma can be more difficult. It is important to recognize, however, that the likelihood of mucinous carcinoma

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**Figure 1:** Gross specimen revealing a spherical, well-circumscribed tumor with a gelatinous cut surface.

**Figure 2:** Tumor in the dermis composed of cells arranged in nests, glands, and cribriform patterns, floating in mucin lakes (H and E, ×20)

**Figure 3:** Small nests of adenocarcinoma lying in pools of extracellular mucin that were separated by fibrocollagenous septae (H and E, ×40)

**Figure 4:** Photomicrograph showing pools of PAS positive, diastase-resistant mucin between the tumor nests (PAS stain, ×20)
from the breast, metastasizing to the skin (much less on the face or eyelid) is exceedingly low. Cases of PMCS have been found to be estrogen receptor, progesterone receptor, and GCDFP-15 positive. Qureshi et al., suggest that finding an in-situ component of a tumor that stains for myoepithelial cells (positive stains for p63 and CK5/6 among others) can help to exclude metastatic mucinous breast carcinoma.[8]

The distinction of MCS from a mucinous carcinoma metastatic to the skin is important because the prognosis of mucinous carcinoma metastatic to the skin is poorer than that of MCS. Primary lesions can be differentiated from metastatic lesions by their more organized epithelial cells, less hyperchromasia, and fewer mitoses in individual cells. In addition, in the case of metastatic carcinoma, tumor cells invade between the collagen bundles at the margin of the nodule. Due to the difficulty in distinguishing these histologically, a thorough workup for metastatic lesions should be completed with any new diagnosis. In our case, no primary tumor was detected in the breast, digestive tract, salivary glands, lacrimal glands, paranasal sinuses, lungs or kidneys.

Primary mucinous carcinoma of the skin is a slow-growing benign tumor with a high local recurrence rate. However, incidence of distant metastases is rare.[9] Treatment of mucinous carcinoma entails local excision. On account of the high rate of recurrence, adequate excision with generous margins (at least 1 cm) is recommended. Several reports of successful treatment using Mohs micrographic surgery have been described. Other treatments, such as chemotherapy and radiation, are generally not employed in the management of these tumors. Patients should be counseled about the importance of frequent follow-ups for evaluation of local tumor recurrence or development of regional lymphadenopathy.

REFERENCES

1. Ku BS, Kwon OE, Kim DC, Song KH, Lee CW, Kim KH. A case of primary mucinous carcinoma of the skin. Korean J Dermatol 2005;43:1228-32.
2. Lennox B, Pears AG, Richards HG. Mucin secreting tumors of the skin, with special reference to the so-called mixed salivary tumor of the skin and its relation to hidradenoma. J Pathol Bacteriol 1952;74:865-80.
3. Mendoza S, Helwig EB. Mucinous (adenocystic) carcinoma of the skin. Arch Dermatol 1971;103:68-78.
4. Breiting L, Christensen L, Dahlstrom K, Breiting V, Winther JF. Primary mucinous carcinoma of the skin: A population based study. Int J Dermatol 2008;47:242-5.
5. Carson HJ, Gattuso P, Raslan WF, Reddy V. Mucinous carcinoma of the eyelid: An immunohistochemical study. Am J Dermatopathol 1995;17:494-8.
6. Rosai J. Ackerman’s Surgical Pathology, 7th ed. United States: St Louis, MO: Mosby; 1989.p.1232-3.
7. Ohnishi T, Takizawa H, Watanabe S. Immunohistochemical analysis of cytokeratin and human milk fat globulin expression in mucinous carcinoma of the skin. J Cutan Pathol 2002;29:38-43.
8. Qureshi JS, Salama ME, Chitale D, Bansal I, Ma CK, Raju U, et al. Primary cutaneous mucinous carcinoma: Presence of Myoepithelial cells as a clue to the cutaneous origin. Am J Dermatopathol 2004;26:353-58.
9. Miyasaka M, Tanaka R, Hirabayashi K, Yamazaki A, Shinohara H, Taira H, et al. Primary mucinous carcinoma of the skin: A case of metastasis after 10 years of disease-free interval. Eur J Plast Surg 2009;32:189-93.

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