Primary mucinous carcinoma of eyelid: A rare clinical entity

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Primary cutaneous mucinous carcinoma of the eyelid, a rare pathologic entity, is an adenocarcinoma of the eccrine glands. Though it has low metastatic potential, it does have a significant recurrence rate. We present the occurrence, clinical and histological features, and management of this tumor in a 62-year-old male who presented with a recurrent, firm, nodular left lower lid lesion. He underwent excision with a 5 mm margin and the defect was repaired with a Mustarde's cheek rotation flap. A full oncological screening, including whole-body Positron Emission Tomography scan, excluded the presence of primary mucinous carcinoma elsewhere and any metastatic spread. This case underscores the importance of considering this tumor in recalcitrant eyelid lesions and highlights the pathology of this tumor.

Key words: Eyelid, primary mucinous adenocarcinoma

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Primary cutaneous mucinous carcinoma (MC) of the eyelid is an adenocarcinoma of the eccrine glands. It is a rare tumor with indolent growth.[1] About 120 cases have been reported in literature so far, mostly as case reports.[2] We report a case of primary MC of the eyelid and present a brief review of literature on its pathology and treatment options.

Case Report

A 62-year old male presented to the oncology center for evaluation of a painless, superficial nodular lesion over his left lower eyelid that had slowly grown over the course of approximately 18 months, to measure 4.0 x 2.0 cm. Patient gave history of a similar swelling at the same site which had appeared in July 2005 and after a similar slow, painless progressive increase in size, it had been excised in February 2006. The histopathology report from the previous surgery was not available for review. The swelling recurred within six months and displayed a painless, gradual, progressive increase in size till he presented at our center. On examination, he had a well-defined, irregularly marginated nodular lesion over the left lower eyelid [Fig. 1]. The overlying skin was normal in appearance and freely mobile over the underlying nodular lesion except for an area of 1 x 1 cm at the lateral end, where the scar of the previous surgery was tethered to the mass. There was no regional lymphadenopathy. The lesion appeared free from the underlying orbital ridge. Fine needle aspiration cytology from this lesion reported a benign adenexal tumor. The lesion was excised with 5 mm margins under general anesthesia. The resultant defect involved the entire lower eyelid and was reconstructed by a Mustarde's cheek rotation flap. This was a large skin flap which was rotated from the cheek. Incision began at the lateral canthal angle, extending upward onto the temple, and swinging posteriorly just anterior to the ear and then inferiorly across the mandible [Fig. 2]. Eight weeks postoperatively, there was marginal ectropion and lateral tissue sag [Fig. 3]. Gross pathology revealed a subcutaneous nodule of tan, gelatinous tissue measuring 4 x 2 x 1.0 cm. Microscopic examination revealed a dermal tumor composed of epithelial cell islands surrounded by lakes of mucin consistent with the diagnosis of mucinous carcinoma [Fig. 4]. The lateral margins were tumor cell-free but the deep resected margin was involved. A thorough search for other possible sources of mucinous adenocarcinoma was made. Upper and lower gastrointestinal study, contrast enhanced computer tomography study of the chest and abdomen, as well as a whole body Positron Emission Tomography scan were negative for any other primary.

Discussion

First described by Lenox *et al*.,[3] in 1951, primary mucinous

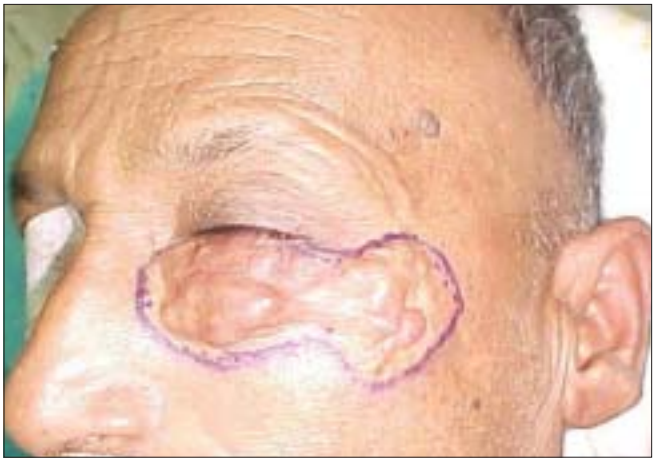

Figure 1: Tumor involving entire lower eyelid (left) with resection margins inked out

Figure 2: Reconstruction with Mustarde's Flap

Figure 3: Postoperative Week 8

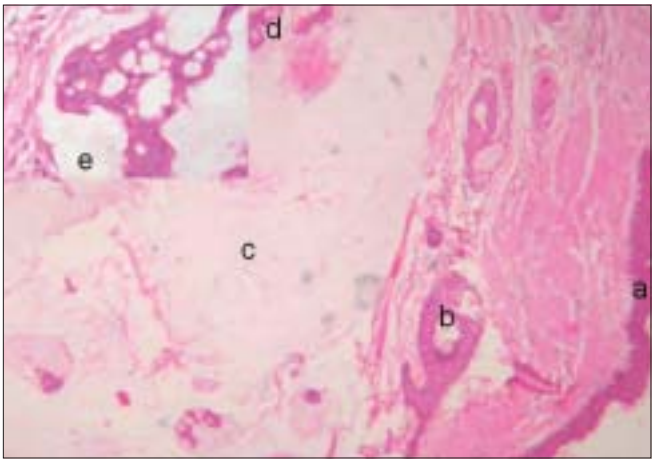

Figure 4: Histopathology: H and E (x100) (a) Epidermis (b) Dermal adnexa (c) Mucin matrix (d) carcinoma cells. Inset (e) x400 view of carcinoma cells within mucin matrix

carcinoma of the skin (MCS) is a rare subtype of sweat gland tumor. While some debate exists as to the apocrine or eccrine origins of this tumor, most authors favor eccrine differentiation based on evidence obtained from immunohistochemical studies and electron microscopic ultra structural analysis.[4] MC most commonly arises in the head or neck, with the eyelid being the most common site. Men are more affected than women in a 2:1 ratio and it tends to occur in more elderly individuals (average age 62 years, range 34-84 years).[5]

Primary mucinous carcinoma of the skin typically has an indolent course. Local recurrence occurs frequently (29.4%) following excision, but the rate of metastasis is low (9.6%) and most metastases are to regional lymph nodes.[2,5,6] Primary MC has distinctive histochemical and ultrastructural features. The tumor is composed of small, irregular clusters of tumor cells around a ductal lumen in mucinous stroma. The tumor cells have a centrally placed, cuboidal nucleus and eosinophilic cytoplasm with little mitosis. Mucin production is consistent with retained cellular function and an indication that the tumor is well-differentiated. Additionally, mucinous carcinomas are typically avascular, a factor that helps explain their low rate of metastasis.[4] The mucin is diastase-resistant, periodic acid Schiff-positive, hyaluronidase-resistant and alcian blue-positive (pH 2.5). This histochemical profile is consistent with the presence of a non-sulfated mucoprotein, most likely sialomucin. Indeed, in the largest series on the subject described by Kazakov *et al*., authors conclude that primary cutaneous MCs span a morphologic spectrum compatible with their mammary counterparts.[7] It is recognized that distinguishing primary cutaneous adnexal neoplasms from metastatic carcinomas can be difficult and hence organ-specific immunostaining profiles using multiple markers can be used with high sensitivity, specificity, and positive predictive value in detecting primary adenocarcinomas.[8] Although multiple markers may help to differentiate primary MC from metastatic adenocarcinomas, histologic and immunohistochemical findings of the two forms tend to overlap, and therefore, a careful workup to rule out metastatic tumors is necessary in all cases of primary cutaneous MC. To this end an extensive search for other possible primary site was done with relevant imaging procedures.

Treatment for primary MC of the skin is wide local excision and since there is a significant risk of local recurrence, it has been recommended that the excision be done with at least 1 cm margins.[1,9] Some authors have suggested Moh's micrographic technique as an alternative to wide local excision.[2] It is also recognized that these tumors have a locally invasive natural history and there is a high risk of local recurrence despite Moh's surgery.[9,10] In our case as the lower eyelid was involved into entirety, such wide margins could not be possible. Nevertheless, the 5 mm margin we could achieve was sufficient to have clear lateral margins on histopathology. Opinion is divided over the use of adjuvant radiotherapy with some favoring it,[11] while others not favoring it.[12] We have scheduled the patient for adjuvant external beam radiotherapy as the deep margin was positive for tumor deposit.

This case brings forth a rare skin adnexal tumor involving the lower eyelid. Surgeons and ophthalmologists should be aware of this tumor in the periocular region and should consider these carcinomas in the differential diagnosis of cystic/solid eyelid lesions even though they appear benign on clinical course.

References

1. Lee GA, Cominos D, Sullivan TJ Clinicopathological report: Mucinous carcinoma of the eyelid. Aust N Z J Ophthalmol 1999;27:71-3.
2. Bindra M, Keegan DJ, Guenther T, Primary cutaneous mucinous carcinoma of the eyelid in a young male. Orbit 2005;24:211-4.
3. Lennox B, Pearse AGE, 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;64:865-80.
4. Carson HJ, Gattuso P, Raslan WF, Reddy V. Mucinous carcinoma of the eyelid: An immunohistochemical study. Am J Dermatopathol 2005;17:494-8.
5. Wright JD, Font RL. Mucinous sweat gland adenocarcinoma of eyelid: A clinicopathologic study of 21 cases with histochemical and electron microscopic observations. Cancer 1979;44:1757-68.
6. Sudesh R, Siddique S, Pace L. Primary eyelid mucinous adenocarcinoma of eccrine origin. Ophthalmic Surg Lasers 1999;30:394-5.
7. Kazakov DV, Suster S, LeBoit PE, Calonje E, Bisceglia M, Kutzner H, *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;29:764-82.
8. Park SY, Kim BH, Kim JH, Lee S, Kang GH. Panels of immunohistochemical markers help determine primary sites of metastatic adenocarcinoma. Arch Pathol Lab Med 2007;131:1561-7.
9. Krishnakumar S, Rambhatla S, Subramanian N, Mahesh L, Biswas J. Recurrent mucinous carcinoma of the eyelid. Indian J Ophthalmol 2004;52:156.
10. Durairaj VD, Hink, Eric M, Kahook MY. Mucinous eccrine adenocarcinoma of the periocular region. Ophthal Plast Reconstr Surg 2006;22:2230-5.
11. Anderson PJ, Dobson CM , Berry RB. Mucinous adenocarcinoma of the face: A case report. Int J Oral Maxillofac Surg 2004;33:610-2.
12. Karimipour DJ, Johnson TM, Kang S, Wang TS, Lowe L. Mucinous carcinoma of the skin. J Am Acad Dermatol 1996;34:323-6.