ORIGINAL RESEARCH PAPER

Pathology

PRIMARY CUTANEOUS MUCINOUS CARCINOMA OF EYELID (PCMC) -A RARE CASE REPORT.

KEY WORDS: Mucinous carcinoma, eyelid , Ck7

Dr Shushruta Mohanty* Senior Resident, Department of Pathology, M.K.C.G Medical college, Berhampur, Odisha, India *Corresponding Author

Dr Pranati Mohanty Associate Professor, Department of Pathology, P.R.M.Medical college, Baripada, Odisha, India.

Dr Bijaya Kumar Pattnaik Assistant Professor, Department of Opthalmology,S.C.B.Medical college, Cuttack, Odisha, India.

Dr Shilpa Dash Post graduate, Department of Pathology, S.C.B.Medical college, Cuttack, Odisha, India.

ABSTRACT

Primary cutaneous mucinous carcinoma (PCMC) of the eyelid is an adenocarcinoma of the eccrine glands. It is a rare tumor with indolent growth. While there is still controversy regarding its origin as to being apocrine or eccrine , most authors favor eccrine differentiation based on evidence obtained from immunohistochemical studies and electron microscopic ultra structural analysis. However as per WHO PCMC have both eccrine and apocrine differentiation. We report a case of PCMC of eyelid in a 45 yr male who was earlier diagnosed as benign adnexal tumor on FNAC. Histopathology revealed basaloid tumor epithelial cells embedded in mucinous stroma and positive CK7 helped in arriving at correct diagnosis. This case report further deals with the differential diagnosis and management of this rare entity.

CASE REPORT:

A 35-year old male presented to the ophthalmology department with a painless, superficial nodular lesion over his right upper eyelid since 18 months, measuring 2cm x1cm. The lesion appeared free from the underlying orbital ridge .There was no regional lymphadenopathy.. Fine needle aspiration cytology from this lesion done outside was reported as benign adenexal tumor. Prelimnary investigations before surgery was unremarkable. The lesion was excised with 5 mm margins under general anesthesia. Gross pathology revealed a subcutaneous nodule of tan white to pink, gelatinous tissue measuring 2×1.0cm. Microscopic examination showed a dermal tumor composed of epithelial cell islands surrounded by lakes of mucin consistent with the diagnosis of mucinous carcinoma. A thorough search for other possible sources of any primary mucinous adenocarcinoma was done. 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:

PCMC was first described by Lenox *et al* [1] in 1951.It most commonly arises in the head or neck, with the eyelid being the most common site. Men are mostly 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)[2] .

Because of the lack of unique morphologic characteristics, the diagnosis is made on histologic grounds .Primary CMC has distinctive histochemical and ultrastructural features. The tumor is composed of small, irregular clusters of basaloid tumor cells in mucinous stroma. The tumor cells have a centrally placed, large hyperchromatic nuclei and eosinophilic cytoplasm with no mitosis.

Local recurrence occurs frequently (29.4%) following excision. mucinous carcinomas are typically slow growing in nature and avascular, a factor that helps explain their low rate of metastasis (9.6%)[3] .Metastases if occurs seen mostly to regional lymph nodes.[4] .

Mucin production is consistent with retained cellular function and an indication that the tumor is well-differentiated. large amount of mucin secretion inhibits cellular nutrition that results in decreased growth and differentiation of cancer cells.[5] The mucin is diastase-resistant, periodic acid Schiff-positive, hyaluronidase-resistant and alcian blue-positive (pH 2.5). Pathologically PCMC in comparison with other sweat gland tumours contains more mucin, epithelial cell clusters and few ductal structures.[6]

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[7]. It is extremely difficult and challenging to distinguish primary adnexal malignancy from visceral adenocarcinoma metastasising to the skin on histological grounds alone, however positivity for tumour protein p63 helps in resolving this issue as it is more commonly associated with adnexal origin and rules out metastasis.[8] PCMC is ER+/PR+/GCDFP-15/CK7+/CK20–, which poses diagnostic similarity to mammary adenocarcinomas but different from gastrointestinal adenocarcinomas, which are CK7–/CK20+[9]. Kazakov *et al* demonstrated that mammary neoplasms have a strong tendency to manifest at the chest wall, breast and axilla and rarely involve the head and neck.[9] They similarly demonstrated that mucinous carcinoma of intestinal origin is more likely to be detected after local extension to internal structures or the abdominal wall and overlying skin rather than distant metastasis. Another clue that the cells may have come from intestinal origin is the presence of dirty necrosis and epithelial cells with goblet cell differentiation which was not seen in our case[7].IHC in our case was CK 7 positive and P63 negative.

PCMC has been resistant to both chemotherapy and radiation.[7] Most PCMC cases are managed conventionally with wide local excision with at least 1 cm margins. However some authors have documented that the use of Mohs micrographic surgical technique seems promising in managing these rare entity [10]

CONCLUSION:

Primary mucinous carcinoma of the skin adnexa is rare, and should be included in the differential diagnosis of subcutaneous swellings in the head and neck, especially in the periorbital region .Mucinous carcinomas are more common in other sites and may metastasise to eye. Although there are multiple markers to differentiate primary mucinous carcinoma from metastatic adenocarcinomas, and histologic and immunohistochemical findings of the two forms tend to overlap, therefore, a careful workup to rule out metastatic tumors is necessary in all cases of PCMC for better management of the patients.

Figure 1-Nodular lesion in right upper eyelid (arrow)

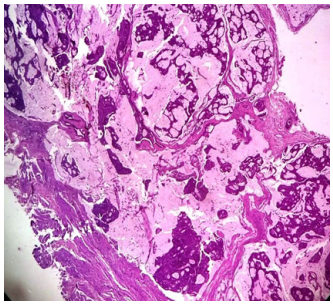

Figure 2(a)- Photomicrograph (scanner 40x)- showing epithelial cell islands in mucin pools.

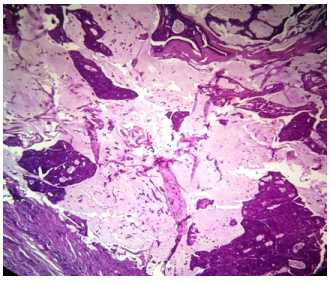

Figure 2 (b)- Photomicrograph (low power 100x)-showing epithelial cells in mucin pools.

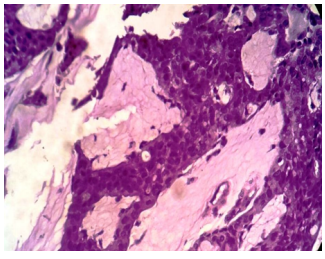

Figure 2 (c) -Photomicrograph (High power 400x) showing basaloid tumor cells in mucinous stroma

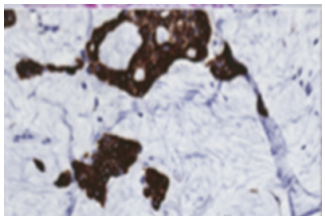

Figure 3- Positive CK7 in tumour epithelial cells.

REFERENCES

1. Lennox B, Pearse AGE, Richards HG. 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. [PubMed].
2. 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. [PubMed].
3. Carson HJ, Gattuso P, Raslan WF, Reddy V. Mucinous carcinoma of the eyelid: An immunohistochemical study. Am J Dermatopathol. 2005;17:494–8. [PubMed]
4. Sudesh R, Siddique S, Pace L. Primary eyelid mucinous adenocarcinoma of eccrine origin. Ophthalmic Surg Lasers. 1999;30:394–5. [PubMed].
5. A.L.K.S.K.Hemlatha,K.Amita,M.Sanja,M.S.Lavanya,Primary mucinous eccrine adenocarcinoma-a rare malignant cutaneous adnexal neoplasm at an unconventional site,J.Clin.Diagn.Res.8(8)(2014) FD14-FD15.
6. I.M.Scholz, W.Hartschuc,Primary mucinous eccrine carcinoma of the skin – a rare clinical tumor with many differential diagnosis J.Dtsch.Dermatol .Ges. 8(6)(2010) 446-448.
7. Breiting L., Christensen L., Dahlstrom K., Breiting V., Winther J.F. Primary mucinous carcinoma of the skin: a population based study. Int J Dermatol. 2008;47:242–245. [PubMed].
8.. Ivan D, Nash JW, Prieto VG, et al. Use of p63 expression in distinguishing primary and metastatic cutaneous adnexal neoplasms from metastatic adenocarcinoma to skin. J Cutan Pathol 2007;34:474–80[PubMed].
9. Kazakov D.V., Suster S., LeBoit P.E. 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;29:764–782. [PubMed].
10. Durairaj VD, Hink , Eric M, Kahook MY. Mucinous eccrine adenocarcinoma of the periocular region. Ophthal Plast Reconstr Surg. 2006;22:2230–5. [PubMed].