



Primary cutaneous eccrine mucinous carcinoma: A rare case

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Abstract

Primary mucinous carcinoma of the skin is a rare skin adnexal neoplasm with sweat gland differentiation. There have been only few cases of it reported in literature. It is a low grade malignant tumor arising preferentially in the head and neck and periorbital area. We report a case of primary cutaneous eccrine mucinous carcinoma in a 75-year-old male presenting with a swelling in the lateral angle of his right eye.

Keywords: mucinous carcinoma, malignant tumour, periorbital, skin adnexal tumour

Introduction

Primary mucinous carcinoma of the skin is a rare adnexal neoplasm with sweat gland differentiation [1]. Primary cutaneous eccrine mucinous carcinoma was first described by Lennexet alin 1952 [2], and later designated by Mendoza and Helwig, in 1971 [3]. There have been only about hundred cases of it reported in English literature [4]. It is a rare, low grade malignant tumor arising preferentially on the head and neck, typically arises in the periorbital area [5, 6].

Case Report

A 75-year-old male came with a history of nodular skin lesion near the outer angle of right eye since six months. The swelling was insidious in onset and gradually increasing in its size. On examination, a firm skin coloured nodule of approximately 2 × 2 cm was noted on the lateral canthal region of right eye with some tiny telangiectatic blood vessels over the surface. The case was clinically suspected as basal cell carcinoma. Excision biopsy was done and the specimen was sent in 10% formalin for histopathological examination to rule out the possibility of basal cell carcinoma.

Gross examination revealed a nodular circumscribed soft tissue mass, partially covered by skin, measuring 2x1.75 cm in diameter. The cut surface of the tumor had a greyish brown mucoid appearance [Figure 1]. Microscopically, the sections revealed a tumor in the dermis composed of tumor cells arranged in nests, acini, and cribriform pattern floating in a mucoid background, separated by fibrocollagenous septae. The individual tumor cells were round with hyperchromatic nucleus [Figure 2]. The mucin was periodic acid Schiff (PAS) positive [Figure 3]. Based on these histological findings final diagnosis of primary cutaneous mucinous carcinoma was rendered. There was no clinical evidence of any local lymph

node enlargement. Thorough evaluation of respiratory, gastrointestinal, genitourinary system had not revealed any evidence of primary tumours.

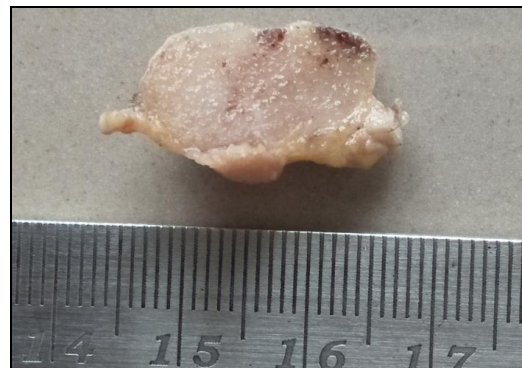


Fig 1: Gross: spherical, well-circumscribed tumor with a greyish brown mucoid cut surface.

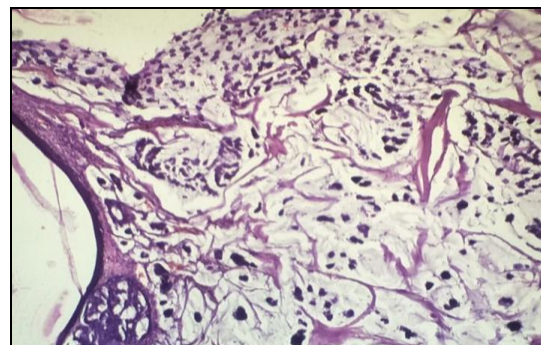


Fig 2: Tumour in the dermis composed of tumor cells arranged in nests, acini, and cribriform pattern floating in a mucoid background.

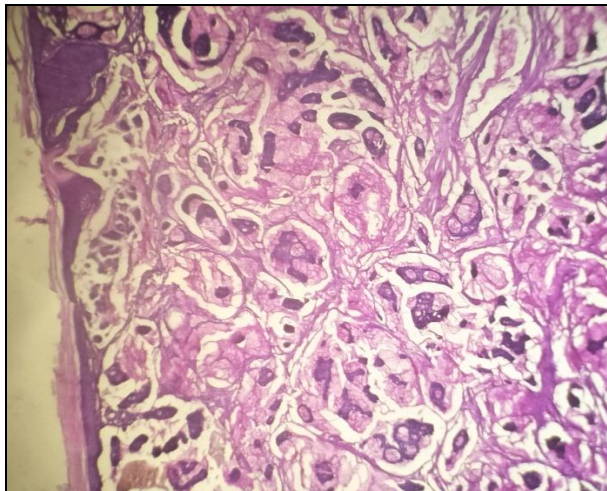


Fig 3: PAS Stain-the mucin was periodic acid Schiff (PAS) positive.

Discussion

Primary cutaneous mucinous carcinoma is a rare epithelial neoplasm occurring mostly, but not exclusively, in middle aged and older patients [5]. This tumor is slightly more common in men than in women and commonly seen in people of age group between 50 to 70 years. The majority of primary mucinous carcinoma of skin arise on the face, especially on the eyelids and scalp [7]. Although this is characterized by destructive local growth and the potential of metastasizing to regional lymph nodes, it generally follows an indolent course [5].

This is a slow-growing type of tumor with a high local recurrence rate. However, incidence of distant metastases is rare [7, 8]. Primary mucinous carcinoma of the eyelid skin has historically been regarded as having high rates of recurrence (up to 40%) and low metastatic potential [6, 9]. In current case there was no evidence of any local tissue destruction or lymph node involvement or metastasis.

Microscopically this tumours presents as an un-encapsulated asymmetric dermal tumour that may extend into the sub cutis and even deeper tissue planes, characterized by large pools of basophilic mucin, which are compartmentalized by delicate fibrous septa, thereby creating a honeycomb pattern. Within the lakes of mucin are small "floating" islands and bizarre clusters of neoplastic epithelial cells, sometimes exhibiting a cribriform arrangement. In our case tumor had focal cribriform pattern. The small neoplastic cells are cuboidal, round or oval with abundant cytoplasm. Nuclei are small with very little atypia. Mitoses are rare. The epithelial mucin is PAS-positive [5] these features were also appreciated well in our case.

Before a diagnosis of mucinous carcinoma is established, a primary carcinoma in a breast or other organs like salivary and lacrimal glands, gastrointestinal tract, nose and paranasal sinuses, bronchi, ovary and renal pelvis should be specifically sought and excluded as most cases of mucinous carcinoma in the skin are metastatic. In our case thorough investigations have failed to show any trace of these primary tumours. Histological differentiation between primary cutaneous mucinous and metastatic mucinous carcinoma to the skin may be impossible [5].

Histochemically, these lesions contain a nonsulfated mucoprotein, probably sialomucin. This substance is thought to be elaborated by the dark cells. Sialomucin is found in both apocrine and eccrine sweat glands [6, 10].

Immunohistochemically, neoplastic cells express low molecular weight cytokeratins, CEA, EMA, GCDFP- 15, alpha-lactalbumin, salivary amylase, beta-2-microglobulin. Cytokeratin 20 expression allows differentiation of mucinous gastrointestinal carcinoma metastatic to the skin from primary cytokeratin 20-negative cutaneous mucinous carcinoma [7].

Conclusion

Primary mucinous carcinoma of the skin is a rare skin adnexal neoplasm; it generally follows an indolent course with frequent local recurrences. Microscopically neoplastic cells are cuboidal, round, or oval having small nuclei with little atypia floating in abundant mucin pool. Mucinous carcinoma metastatic to skin from another organ, particularly the gastrointestinal tract is the differential diagnosis.

References

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