

Primary Cutaneous Mucinous Carcinoma

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ABSTRACT

Primary mucinous carcinoma of the skin is an uncommon malignancy of sweat gland origin. It is usually misdiagnosed clinically because of its benign appearance. On histological examination it should be differentiated from metastatic mucinous carcinomas arising elsewhere in the body. We present a case of a 77 years old male presenting with a swelling on face which was diagnosed clinically as a benign lesion but on histological examination it was diagnosed as mucinous carcinoma and after thorough investigations it was labelled as primary mucinous carcinoma of skin.

Keywords: Carcinoma, Cutaneous carcinoma, Mucinous carcinoma.

Authors' Contribution:

All authors contributed equally to the conception, literature search, manuscript drafting, editing and review

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Article info:

Received: January 11, 2024
Accepted: March 01, 2024

Cite this article: Ishaq Q, Nabi N, Mehmood U, Ahmed N. Primary Cutaneous Mucinous Carcinoma. J Islamabad Med Dental Coll. 2024; 13(1): 183-186.
DOI: <https://doi.org/10.35787/jimdc.v13i1.1088>

Funding Source: Nil
Conflict of interest: Nil

Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare malignant tumor of skin adnexa. Lennox et al., was the first one to discover this rare entity in 1952.¹ Till now in literature, approximately 200 cases have been reported.² This is a locally aggressive neoplasm with a tendency to recur but metastasis is rare. Most commonly mucinous carcinomas arise in the viscera and metastasize to the skin. Therefore, metastatic mucinous carcinoma should be excluded before the diagnosis of primary mucinous carcinoma.³ Wide surgical excision with or without regional lymph node dissection is considered as the treatment of choice. Distant occurs then it becomes highly resistant to both radio and chemotherapy.⁴

Case Report

A 77 years old male was presented in a private clinic for the treatment of a boil on cheek below left eye. As it resembles a benign lesion so it was excised and

no microscopic examination was done, it reappeared after some time and re excision was done not followed by histopathological examination and after some period of time it again appeared and on 3rd consecutive re excision it was sent to excel labs where its gross and histopathological examination was done. On gross examination it was a single brown soft tissue fragment measuring 0.4x0.4x0.3cm. It was entirely submitted in one cassette. On microscopic examination, the biopsy was composed of epidermis and dermis. It showed a tumor in the dermis. The tumor cells were in small clusters immersed in mucin lakes. The tumor cells had vesicular nuclei and prominent nucleoli. The lesion was involving the deep margin (Figure 1). On immuno-histochemical panel, AE/AE3 was positive, CK 20 was negative, CK7 was positive, p63 was negative, Estrogen and progesterone receptors were also positive.

DISCUSSION

Primary mucinous carcinoma of the skin is an uncommon malignant tumor of eccrine glands. Although it can occur at a wide age range (8-84 years), it affects individuals in 5-7th decade with approximately 2:1 male to female ratio. Increased incidence is observed in white race as compared to African American, East Asian or Indian population.⁵ It usually occurs in head and neck region, especially eyelids but can be found on other regions as axillae, thorax, abdomen, groin, foot, hand, and vulva. The lesion is un-encapsulated but well circumscribed, nodular, ranging in size from 0.7 to 120 cm, with the average size of around 1.8 cm. Its clinical appearance can be confused with many inflammatory, benign and malignant lesions like lipoma, epidermoid cyst, nevus, hemangioma, pyogenic granuloma, sebaceous cyst or carcinomas, metastasis from other sites, which is the most important and challenging differential because mucinous carcinomas usually metastasize to the skin from other sites.⁶ The most common sites from where it metastasizes are the breast, salivary glands, lacrimal glands, nose and paranasal sinuses, bronchi, renal pelvis, and ovary. This definitive diagnosis can be made by thorough clinical and radiological examination. Moreover, there are some microscopic and immunohistochemical parameters which can help differentiate primary and secondary mucinous carcinomas. In mucinous carcinomas the microscopic examination shows invasive nests of neoplastic cells arranged in glandular, cribriform or micro-papillary patterns surrounded by lakes of mucin. The individual tumor cells are usually uniform with scanty eosinophilic cytoplasm. The mucin is Periodic acid Schiff positive and diastase resistant. This mucin is considered to be responsible for the slow growth of the tumor by depriving tumor cells from nutrients and thus rare metastasis. Primary and metastatic mucinous carcinoma shows same morphology but metastatic tumors usually show more dysplastic features which include poorly

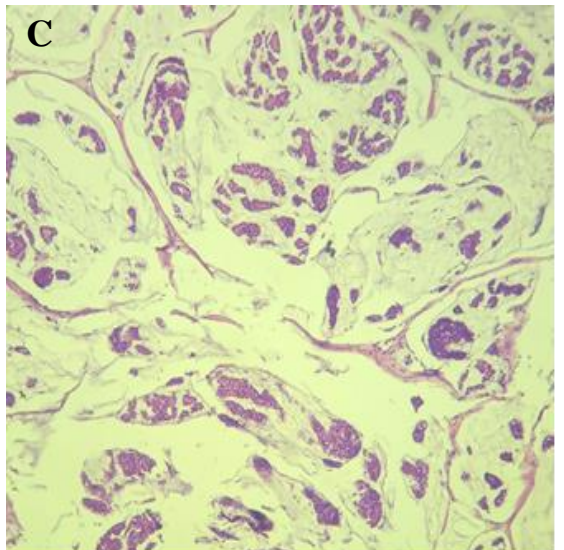
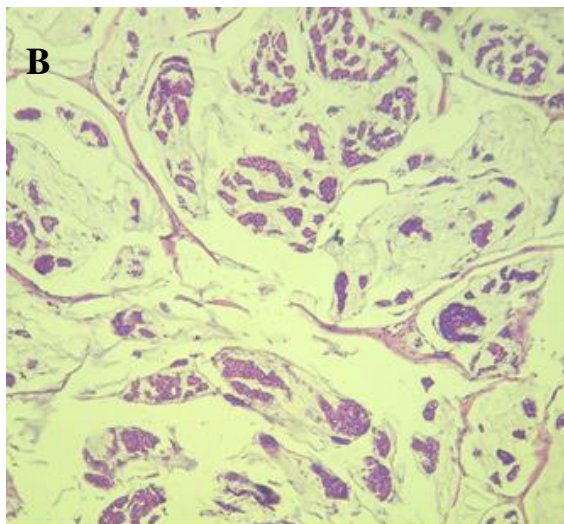
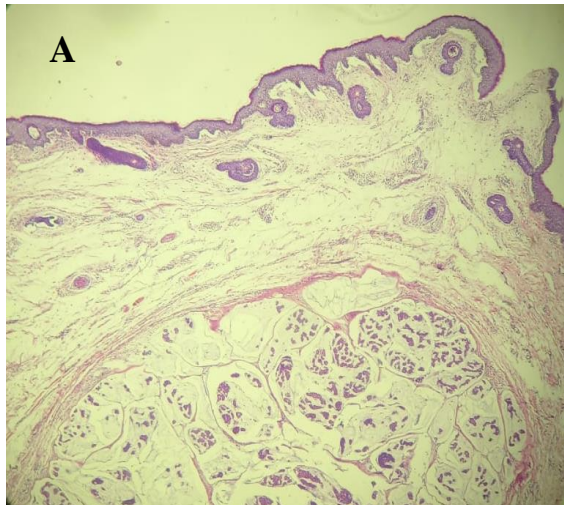


Figure 1: Morphological features of primary mucinous carcinoma of skin; A: the dermis showed nests of tumor cells in lakes of mucin (H&E X10), B: Fibrous septa around the neoplastic cells and the surrounding mucin (H&E X 20), C: The tumor cells are round to cuboid

organized epithelial cells, prominent hyperchromasia and more mitotic figures than what is observed in primary mucinous carcinoma of skin.

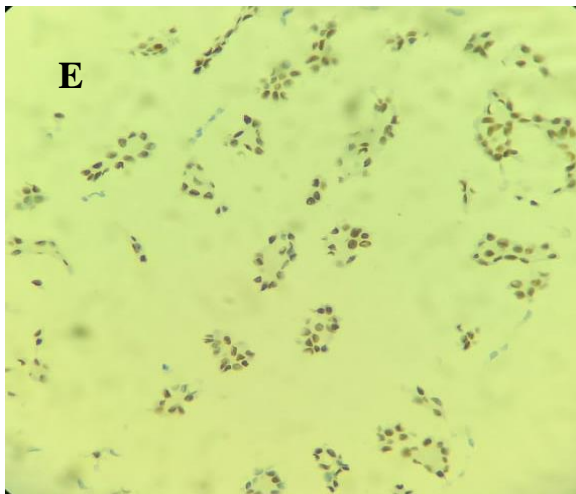
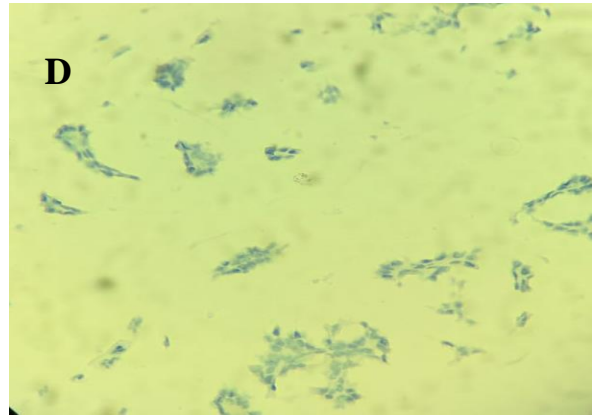
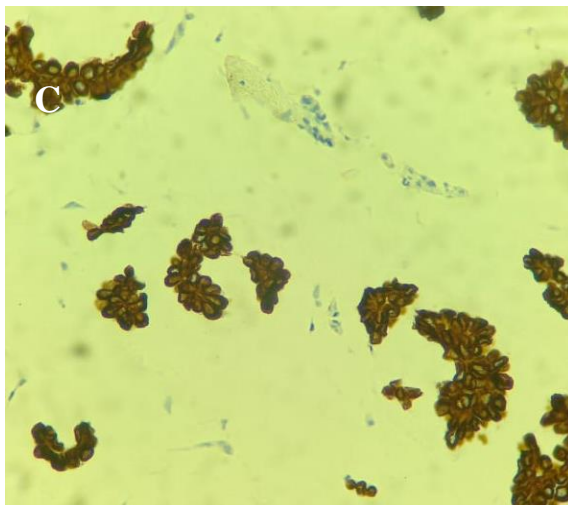
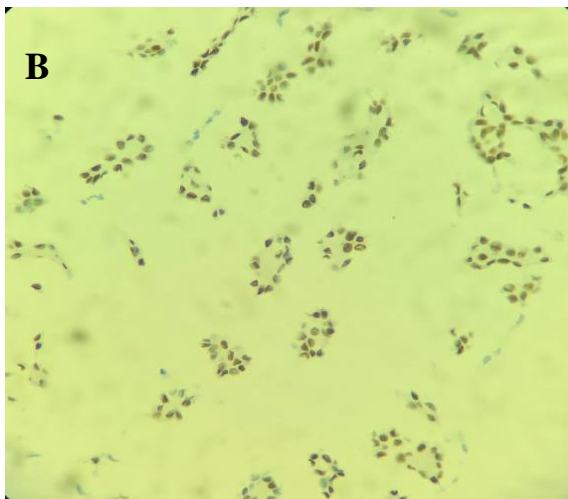
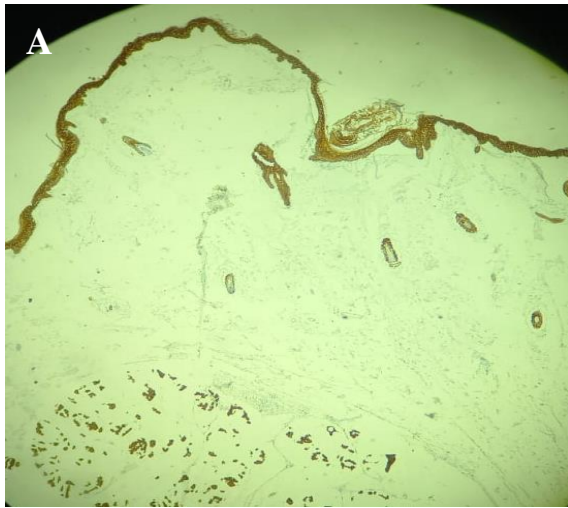


Figure 2: Immunohistochemical stains applied to PMC A: AE/AE was positive, B: CK 20 was negative, C: Ck7 was positive, D: P63 was negative, E:ER & PR was positive

Similarly, Immunohistochemical staining is also helpful in this differentiation. PMC usually express cytokeratin (CK-7), epithelial membrane antigen, ER, and PR. Mucinous carcinomas of colon and rectum express CK 20 by which these carcinomas are differentiated from PMC.⁷ According to some authors presence of an in-situ component favors PCMC, whereas the site of tumor on the chest wall or in the axilla favor a breast primary.⁸ Wide surgical excision with margin of 1cm is the treatment of choice. Chemo and radiotherapy are not effective. New treatment modalities of PMCS with aromatase inhibitors as adjuvant therapy has shown promising results.⁹

The tumor usually recurs locally which is in about 30% to 40% of cases while distant metastases is rare which is approximately in 6% of cases. The prognosis of PMC is good. Older age and Asian population showed better postsurgical outcomes. Prognosis is affected by size, duration of persistence and site.¹⁰

CONCLUSION

The tumor deserves attention not only for its rarity but also for the problems of differential diagnosis with dermal metastasis from mucinous adenocarcinoma of other sites. Diagnostic concerns include its deceptively benign appearance in some cases and the difficulty in differentiating it from secondary mucinous carcinoma of skin metastasizing from a primary source elsewhere.

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