Case Report

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Pathology Section

Primary Mucinous Eccrine Adenocarcinoma – A Rare Malignant Cutaneous Adnexal Neoplasm at an Unconventional Site

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ABSTRACT

Primary mucinous eccrine adenocarcinoma, a rare malignant neoplasm of the skin adnexa usually occurs in the head and neck region. The most common sites for its occurrence are the eyelid, the peri-orbital region and the scalp. We report the rare occurrence of primary mucinous eccrine adenocarcinoma of the inguinal region which has been reported in only 1% of cases according to literature search. Since the differentiation from metastatic mucinous carcinomas is difficult, a careful search for primary in the breast, gastrointestinal tract and prostate is mandatory.

This case report highlights the importance of precise diagnosis and accurate histological typing of mucinous carcinomas with an emphasis on their role in appropriate patient management along with a brief review of literature.

Keywords: Eccrine, Mucinous, Skin

CASE REPORT

A 49-year-old male patient presented with a painless swelling of one month's duration in the iliac region.

On local examination, a palpable, non-tender nodule measuring 3x2 cm which was mobile in all directions and firm in consistency was felt. There was no localized or generalized lymphadenopathy. With these findings, the clinician offered a provisional diagnosis of Desmoid tumour.

FNAC of the swelling showed inflammatory cells admixed with large cells exhibiting atypical features against myxoid/mucinous background material [Table/Fig-1]. A cytological diagnosis of a malignant mucinous neoplasm was arrived at. Histopathological examination was suggested for confirmation of diagnosis.

A detailed systemic examination coupled with radiological and biochemical work-up was undertaken, which ruled out the involvement of visceral organs. Thereafter, a wide local excision was done and the specimen was submitted for histopathological examination.

HISTOPATHOLOGICAL EXAMINATION

Gross examination of the specimen showed multiple, gelatinous tissue bits. The largest bit measured 2 x 1.5 cm. The cut surface was gelatinous [Table/Fig-2].

Microscopic examination showed large pools of mucin arranged in lobules, separated by collagenous septae [Table/Fig-3a]. Floating in the pools of mucin were epithelial tumour cells arranged in small clusters, tubules and cribriform patterns [Table/Fig-3b-d]. Individual tumour cells showed scant cytoplasm and hyperchromatic nuclei. No in-situ elements were detected.

Periodic acid Schiff (PAS) stain was positive, thereby suggesting the mucinous nature of the tumour [Table/Fig-4].

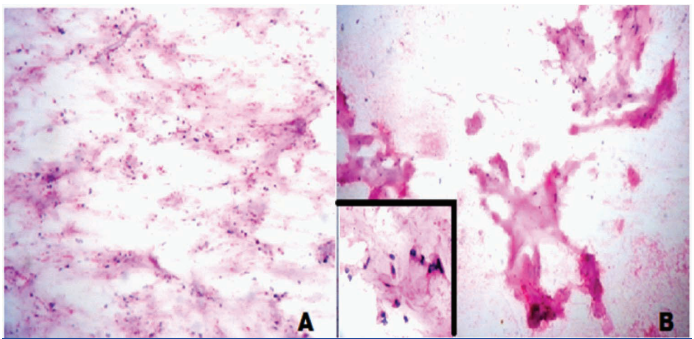
Histopathologic diagnosis of Mucinous Adenocarcinoma was made. The final diagnosis of Primary mucinous eccrine adenocarcinoma was signed out and the patient was referred to an oncology centre for further management and was lost to follow-up.

DISCUSSION

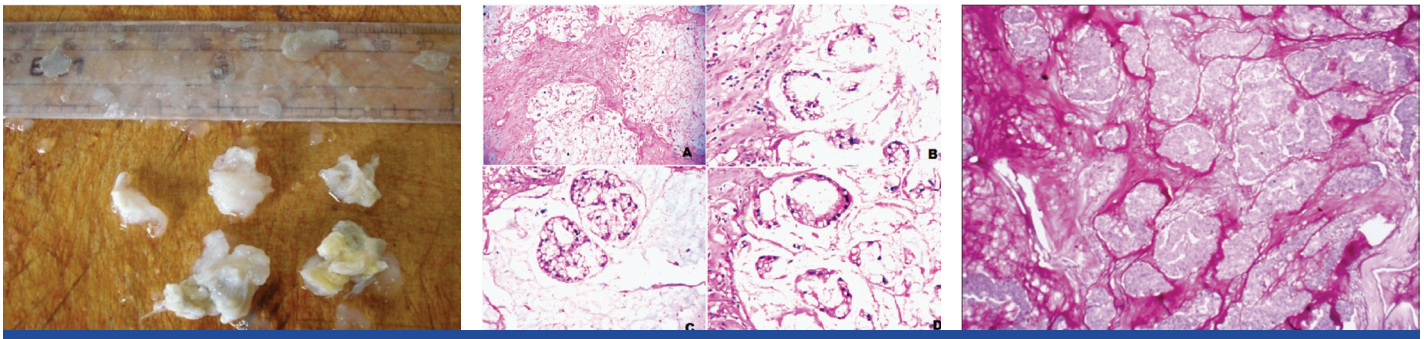
Primary mucinous eccrine adenocarcinoma is a rare malignant cutaneous adnexal tumour which is morphologically indistinguishable from metastatic mucinous carcinoma of non- cutaneous origin with primaries in breast, colon, ovaries and lung. Many other mucin producing mesenchymal and epithelial lesions may mimic this entity thereby adding to the diagnostic difficulties as encountered in the present case [1].

Mucinous carcinomas most commonly arise in the head or neck, with the eyelid being the most common site [2]. Men are more affected than women in a 2:1 ratio. They tend to occur more frequently in elderly individuals (average age 62y, range 34-84 y) [3]. Cutaneous mucinous carcinomas may present with a variety of clinical presentations. They are known to present as painless, papular or nodular lesions ranging from 5cm to 120cm in size [4]. They are usually single as in the present case. They may have a smooth surface as in the present case, or may have an ulcerated or crusted surface.

The differential diagnosis includes many benign and malignant cutaneous lesions, but the most important one to rule out is the mucinous carcinoma of skin metastatic from another site. An associated in- situ component may help in establishing the diagnosis



[Table/Fig-1]: A. Cytological smear showing abundant myxoid/mucinous material and atypical cells (H & E, X 40)
B- Cytological smear showing atypical cells embedded within the mucinous material (H & E, X 40). Inset shows high power view of the same (H& E, × 400)



[Table/Fig-2]: Gross examination of the specimen showing gelatinous tissue bits
[Table/Fig-3]: a- .Histopathologic section showing pools of mucin, divided into compartments by collagenous septae and islands of tumour cells floating within it (H & E, X 40)
b- Tumour cells arranged in tubular patterns, floating in pools of mucin (H & E, X 100)
c- Section showing tumour cells arranged in cribriform patterns (H&E, X 400)
d- Islands of tumour cells showing mild cytological atypia and focal duct formation (H & E, X 400)
[Table/Fig-4]: Photomicrograph showing pools of PAS positive, diastase resistant mucin between the tumour nests (PAS stain, X 20)

of a primary tumour. Unfortunately, the tumour in the present case did not show any in- situ component and we had to rely on the other parameters to rule out a primary in other sites. Further, the histopathologic clue to an intestinal origin is a combination of dirty necrosis and the presence of epithelial cells with goblet cell differentiation [5]. The present case did not show any of these features which helped in ruling out a primary in the gastrointestinal tract.

Primary cutaneous mucinous carcinomas are slow-growing tumours with a local recurrence rate of 29.4% following excision and a low metastatic rate of 9.6% [6,7]. Majority of these tumours metastasize to regional lymph nodes. Rarely, skeletal metastasis has been reported in about 7% cases [8]. Local recurrences are known to occur frequently in these tumours, but they rarely metastasize. Death following metastatic spread has been reported in three cases as per literature review [7,9]. No metastasis was detected in the present case.

The slow local growth and relatively low rate of metastasis in mucinous carcinomas are supposedly due to the copious mucin secretion which interferes with cellular nutrition, thereby hindering growth and differentiation of cancer cells.

The mucin produced by the tumour cells in mucinous carcinomas is Periodic acid Schiff (PAS) positive. The tumoural PAS positivity, as seen in our case, is a regular feature in all the mucinous carcinomas of the skin.

A wide local excision with at least 1 cm margin as was done in the present case which is the recommended treatment modality in primary mucinous carcinomas of the skin.

The patient needs to be followed up regularly, because of the risk of recurrences and secondly, because of the possibility that the tumour which lacks in- situ elements might actually represent a metastasis. Accordingly, our patient was advised regular follow-up at half-yearly intervals.

CONCLUSION

A high index of suspicion is required to diagnose a rare entity like primary cutaneous mucinous eccrine adenocarcinoma especially in a rare site like iliac region as in the present case. Following diagnosis histologically, it warrants an extensive search for the primary site as in this case where a thorough search ruled out a primary elsewhere.

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