

# Primary Cutaneous Mucinous Carcinoma: Case Report of a Rare Adnexal Tumour

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## ABSTRACT

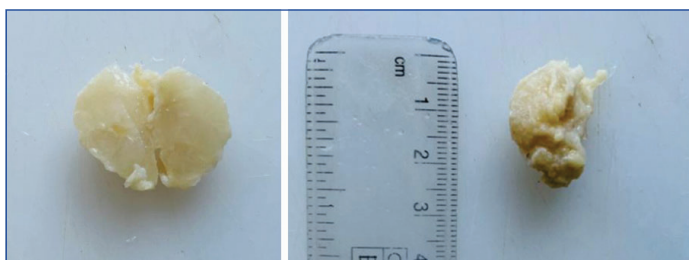
Primary cutaneous mucinous carcinoma is a rare malignant adnexal tumour of the skin, with the most common location being the periorbital region. Differentiating this tumour from cutaneous metastases and mucinous adenocarcinomas from other sites can be challenging, and due to its rarity, this tumour merits a more detailed study. The authors present a case of a 55-year-old male who presented with a slow-growing tumour located below the right earlobe. The lesion was clinically differentially diagnosed as either a lymph node or a soft tissue mass. An excisional biopsy of the mass revealed features of cutaneous mucinous carcinoma. The patient was clinically assessed, and the possibility of metastasis from other sites was ruled out. Hence, the present case lesion was diagnosed as primary mucinous carcinoma of the skin. The patient is doing well on follow-up, with no history of recurrence or other relevant issues.

**Keywords:** Adenocarcinomas, Excisional biopsy, Metastases

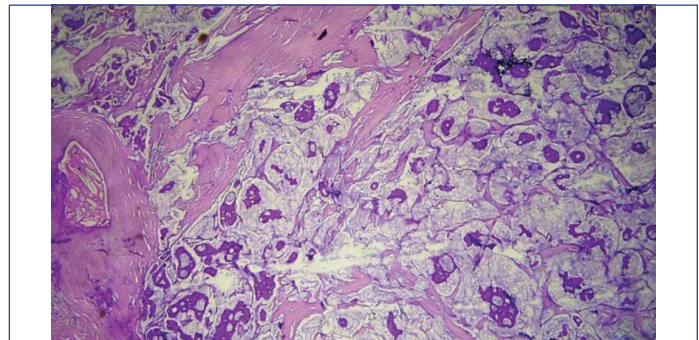
## CASE REPORT

A 55-year-old male, a known case of hypertension not on any medication and with no other co-morbidities, presented with complaints of a slow-growing tumour below the right earlobe for the past year. Clinically, the lesion was given a differential diagnosis of either a lymph node or a soft-tissue mass. Ultrasound findings suggested a benign cystic lesion, possibly an epidermal inclusion cyst. Upon clinical examination, the lesion revealed a 2×2 cm subcutaneous swelling with firm consistency. The patient underwent excision biopsy, and the specimen was sent to the Histopathology Department of the Institution.

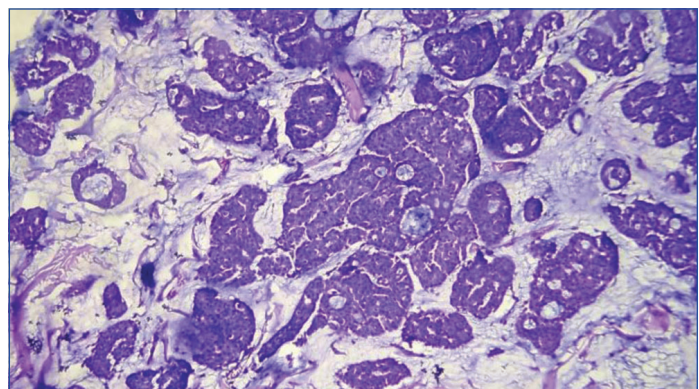
The specimen received in the department showed a pale white to translucent firm nodular mass measuring 2.5×2×1 cm. No attached skin was seen on gross examination. Cut sections revealed a solid, pale white gelatinous appearance [Table/Fig-1,2]. Formalin-fixed paraffin-embedded sections stained with Haematoxylin and Eosin (H&E) showed a fairly circumscribed neoplasm with tumour cells arranged in solid nests and glands floating in pools of extracellular mucin, separated by fibrocollagenous septae of varying thickness [Table/Fig-3]. Some areas in the periphery exhibited neoplastic cells arranged in closely packed glandular and cribriform clusters [Table/Fig-4]. No mitotic figures or necrosis were observed. The possible histopathological differential diagnoses included primary cutaneous mucinous carcinoma or a metastatic deposit from an unknown primary carcinoma. The extracellular mucin showed Periodic Acid-Schiff (PAS) positivity [Table/Fig-5]. Most cutaneous mucinous carcinomas are positive for oestrogen receptors and exhibit neuroendocrine differentiation. In the present case, Immunohistochemical (IHC) analysis revealed that tumour cells were



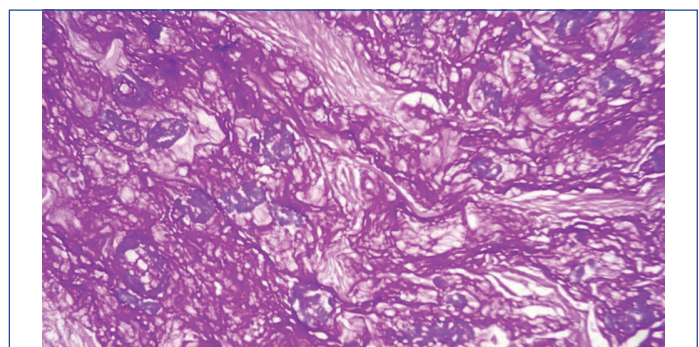
**[Table/Fig-1]:** Cut section; a solid, pale white gelatinous appearance.  
**[Table/Fig-2]:** Gross appearance showing a nodular mass. (Images from left to right)



**[Table/Fig-3]:** Tumour cells in solid nests and glands floating in pools of extracellular mucin, separated by fibrocollagenous septae (H&E, 10x).

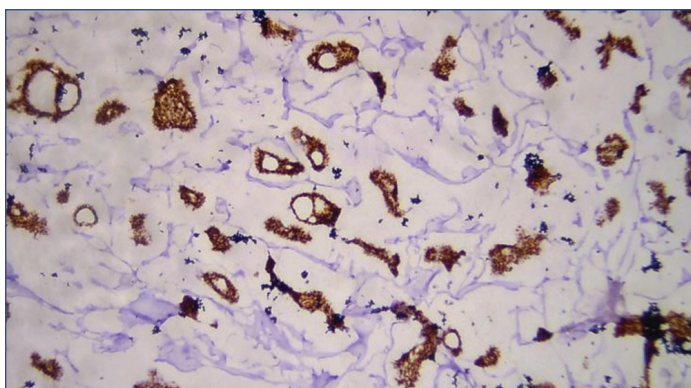


**[Table/Fig-4]:** Glands and cords of neoplastic cells floating in extracellular mucinous pools (H&E, 20x).

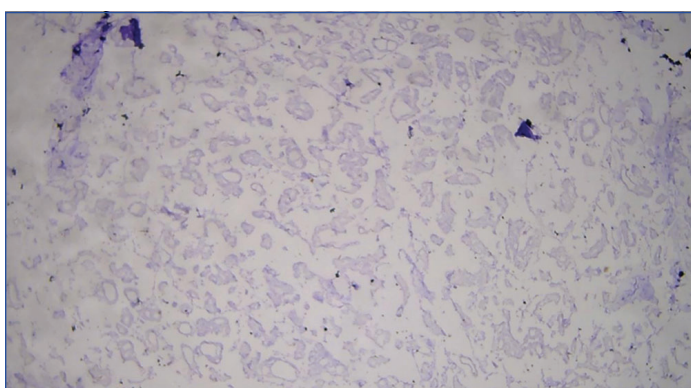


**[Table/Fig-5]:** Extracellular mucin showing positivity (PAS, 10x).

positive for Oestrogen Receptors (ER) [Table/Fig-6] and negative for synaptophysin [Table/Fig-7].



**[Table/Fig-6]:** Tumour cells showing nuclear positivity for oestrogen receptor (IHC, 20x).



**[Table/Fig-7]:** Tumour cells negative for synaptophysin (IHC, 40x).

The case was discussed with the clinician, and further investigations, including radiography, ultrasound and computed tomography, were performed, all of which were negative. Thus, the possibility of metastasis from a mucinous adenocarcinoma from other sites was ruled out. Hence, a final diagnosis of primary mucinous carcinoma of the skin was reached.

## DISCUSSION

Primary mucinous carcinoma of the skin was first described by Lennox B et al., in 1952 [1]. It is a rare adnexal tumour with features similar to the mucinous lesions found in the breast. Some tumours that have a solid component with immunohistochemical features of neuroendocrine tumours are diagnosed as endocrine mucin-producing sweat gland carcinoma, which is regarded as a precursor lesion [2]. A review of 159 patients found that 77%, 13% and 10% of the patients were Caucasian, Asian and Black, respectively [3]. Both males and females are affected equally, with a predominant number of cases seen in the older population [4-6]. The most common location for this carcinoma is the face, particularly the periorbital region [3]. Rare cases have been reported in the anogenital area, supposedly arising from the mammary glands in that region [7]. Most cases of primary cutaneous mucinous carcinoma present as a single, bluish, flesh-coloured nodule or cyst that grows slowly. The mean diameter is reported to be approximately 1.8 cm [8].

Classic morphology shows nests, cords and singly scattered neoplastic cells floating in extracellular mucin pools, separated by fibrous septa of variable thickness. Primary cutaneous mucinous carcinoma shows a histopathological spectrum with two variants: a pure tumour where extracellular mucin comprises over 90% and a rare mixed form with an invasive ductal component [9]. Neoplastic cells are monomorphic with scant mitotic activity. An in situ component should be searched for, as it can be a supportive feature in defining the tumour as primary cutaneous. The in situ component can range from ductal and atypical ductal hyperplasia

to ductal carcinoma in situ. Immunohistochemically, the neoplastic cells are positive for epithelial membrane antigen, low molecular weight cytokeratins, cadherins and gross cystic disease fluid protein-15. In most cases, neoplastic cells express oestrogen and progesterone receptors. Neuroendocrine differentiation may also be observed in primary cutaneous mucinous carcinoma. Although ultrastructural findings have proposed an eccrine origin for these tumours, about half of them show apocrine secretion, suggesting apocrine differentiation.

Primary cutaneous mucinous carcinoma is very rare. No morphological feature or immunohistochemical tool can clearly distinguish it from metastatic mucinous carcinoma. Primary cutaneous mucinous carcinoma has a better clinical outcome than metastatic lesions; therefore, differentiating between the two is crucial. Some features that can help differentiate primary lesions include their more organised neoplastic cells, lesser hyperchromasia, and low mitotic activity. In metastatic carcinoma, tumour cells invade between collagen bundles at the margins of the tumour.

The usual sites of origin for mucinous carcinoma include the breast, gastrointestinal tract, salivary glands, lacrimal glands, nose, paranasal sinuses, bronchi, renal pelvis and ovary [10]. Differentiating cutaneous mucinous carcinomas from breast carcinomas can be challenging. However, it is noteworthy that instances of breast mucinous carcinoma metastasising to the skin of the face or eyelid are very rare [11]. Staining for myoepithelial markers {tumour Protein 63 (p63) or Cytokeratin 5/6 (CK 5/6)} in the in situ component, if present, can help exclude metastatic mucinous breast carcinoma [12]. Metastasis from colorectal carcinoma can be excluded by immunohistochemical staining for CK20. In the present case, no primary tumour was detected in the breast, gastrointestinal tract, salivary glands, lungs, or kidneys.

The treatment for mucinous carcinoma is local excision. Due to an increased recurrence rate, excision with adequate margins is advised. Prognostically, tumours located in the head and neck have a much better outcome compared to those noted on the chest and back. A high recurrence rate and metastasis have been linked to younger age at presentation, white race and tumour size greater than 1.5 cm [3].

Since these tumours have a high recurrence rate, patients must be counselled about the need for frequent follow-ups for recurrence and regional lymphadenopathy. The present case patient is doing well nine months post-surgery during follow-up.

## CONCLUSION(S)

Primary cutaneous mucinous carcinoma is a rare malignant adnexal carcinoma. The present case patient represents one of the few rare cases. Clinical distinction from metastatic mucinous carcinoma is of utmost importance. Clinicians should consider all possibilities before reaching a diagnosis, and investigations should be thoroughly reviewed to check for any signs of rare tumours.

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