

Case Report

Primary Mucinous Eccrine Adenocarcinoma of Skin: Study of Two Cases

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Abstract: Primary mucinous adenocarcinoma (PMC) of skin is a rare adnexal epithelial neoplasm with eccrine differentiation. It is a rare tumour most commonly affecting the face and scalp region. We should be cautious before making its diagnoses as it closely mimics metastatic mucinous carcinoma from internal organs and a thorough search for any primary is warranted. Diagnosis can be only confirmed after excluding a primary mucinous carcinoma of other organs like breast, gastrointestinal tract, lung, salivary and lacrimal glands, urinary tract, prostate or paranasal sinuses metastatic to skin. Here, we report two cases of primary mucinous adenocarcinoma, one of axilla and other arising in periorbital region of face. Both the cases shows presence of PAS positive, diastase resistant mucin and are negative for CK-20 expression. Patients were undergone through an extensive search for any other neoplastic lesion by doing CT scan of thorax and abdomen, upper GI tract endoscopy, colonoscopy, USG lower abdomen and pelvis but reveals nothing. Both the patients are doing well on one year follow up period.

Keywords: Primary mucinous adenocarcinoma; orbit; axilla; PAS

INTRODUCTION

Primary mucinous adenocarcinoma (PMC) of skin is a rare adnexal epithelial neoplasm with eccrine differentiation. Since its first description by Lennex *et al* [1] in 1952, there has been about a hundred cases of it reported in English literature [2]. It is a rare tumour most commonly affecting face and scalp region. Diagnosis can be only confirmed after excluding a primary mucinous carcinoma of other organs like breast, gastrointestinal tract, lung, salivary and lacrimal glands, urinary tract, prostate or paranasal sinuses metastatic to skin. Thus, the rarity and differential diagnostic concerns makes it an important pathological entity. Here, we report two cases of primary mucinous adenocarcinoma, one of axilla and other in periorbital region.

CASE 1

A 74 year old man presented with two year history of slow growing swelling in the right axilla. Surgical excision of tumour was done. On gross

examination, it was a globular lump measuring 5x4x2 cms with ulceration on the surface. The cut surface showed a variegated appearance with multiple white nodules [Figure-1a]. Microscopic examination revealed a large lobulated tumour showing sheets of atypical anaplastic epithelial cells with hyperchromatic pleomorphic nuclei and prominent nucleoli. The cells had moderate cytoplasm and formed tubular lumen at places. Areas having abundant extracellular mucin pools with floating tumour cells noted. Infiltration into subcutaneous fat was present [Figure-1b&1c]. Periodic acid Schiff (PAS) stain shows presence of PAS positive, diastase resistant mucin [Figure-1d]. Patient was undergone an extensive search for any other neoplastic lesion by doing CT scan of thorax and abdomen, upper GI tract endoscopy, colonoscopy, USG lower abdomen and pelvis but nothing found. His serum PSA level was low. Immunostain to detect CK-20 expression was negative. A final diagnosis of Mucinous eccrine adenocarcinoma of skin was given.

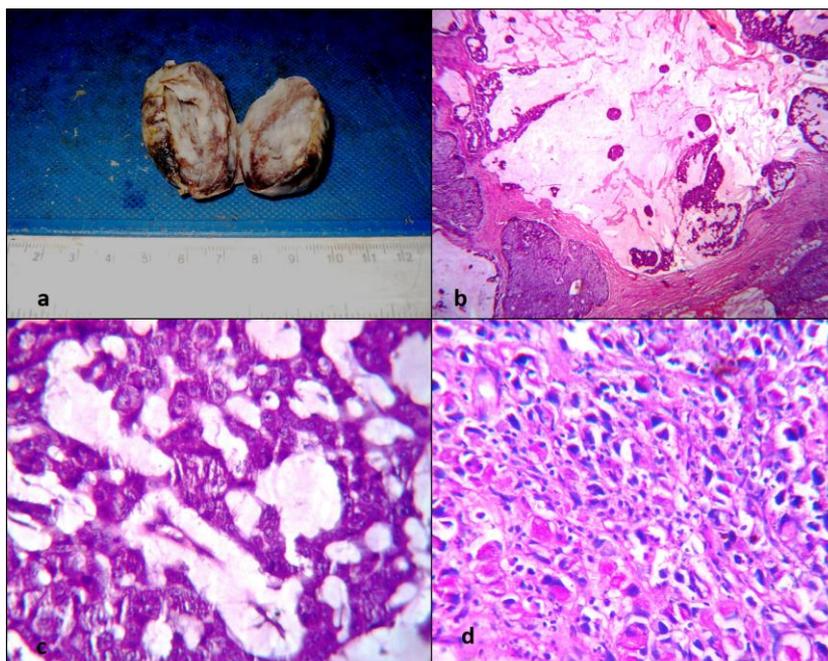


Fig-1: A) Gross photograph case 1 showing a globular lump measuring 5x4x2 centimetres; B&C) Photomicrograph showing PMC case 1(H&E stain 40x & 400x); D) PAS stain showing intracellular mucin in case 1 (PAS stain 400x)

CASE 2

A 52 year old man presented with asymptomatic swelling in right outer canthus since one year. Clinical examination revealed one centimetre firm nodular swelling with smooth surface. After resection gross pathology revealed well circumscribed skin covered subcutaneous nodule with gelatinous cut

surface. Microscopical examination showed features of mucinous eccrine adenocarcinoma. The mucin in this case was also PAS positive, diastase resistant. Tumor was negative for CK-20. An extensive search for any other primary tumor revealed nothing. Both the patients are doing well on one year follow up period.

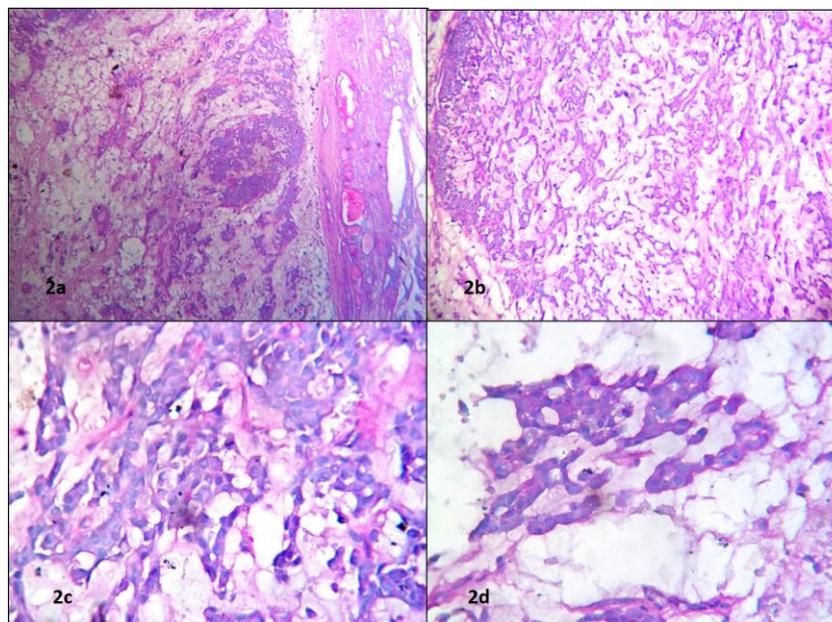


Fig-2: A) Photomicrograph showing PMC case 2. Normal adnexal glands present in right side of the field (H&E stain 40x); B&C) Photomicrograph showing the neoplastic cells of case 2 (H&E stain-100x & 400x respectively); D) Photomicrograph showing PAS positive mucin in case2 (PAS stain, 400x)

DISCUSSION

PMC of the skin is a low grade adnexal carcinoma of eccrine differentiation. Histochemical and electron microscopic studies have proven the origin of the tumour to be the secretory coil of eccrine duct [3]. Older individuals between 50-70 years are commonly affected though it is known to affect a wide age range of 8-87 years with a predilection for males. Most cases occur on the head particularly around the eyelids. Other affected sites include the scalp (17%), face (14%), axilla (9%), vulva (4%), chest/abdominal wall (7%), neck (2%), extremity (2%), canthus (2%), groin (1%) and ear (1%) [2].

The tumour occurs in dermis as an asymmetrical unencapsulated mass characterised by islands and clusters of epithelial cells floating in pools of basophilic mucin compartmentalized by delicate fibrous septa creating a honeycomb pattern. The neoplastic cells are cuboidal, round or oval with little nuclear atypia. The main differential diagnosis are metastatic mucinous carcinoma from breast, salivary and lacrimal glands, gastrointestinal tract, nose and paranasal sinuses, bronchi and ovary. The mucin in PMC is PAS positive, diastase resistant, hyaluronidase and sialinase labile and consisting of non-sulphated acid mucopolysaccharides [4]. Immunohistochemically expression of low molecular weight cytokeratins, CEA, EMA, GCDFP-15, alpha-lactalbumin, salivary amylase, beta-2-microglobulin is seen. Negative CK20 expression helps differentiating it from metastatic colorectal mucinous carcinoma [5]. Differentiation from breast carcinoma is a challenge as both show positive expression of estrogen receptor, progesterone receptor and GCDFP-15. However, demonstration of in situ component that stains positive for myoepithelial cell markers (p63 or CK 5/6) goes in favour of PMC [6].

Other differential diagnostic entities include malignant mixed tumour of skin and adenoid cystic basal cell carcinoma. Malignant mixed tumour is characterised by polygonal and plasmacytoid neoplastic epithelial cells forming tubular structures and embedded in a myxoid or chondroid stroma. There is lack of characteristic honeycomb pattern of PMC [7]. Mucin in adenoid cystic basal cell carcinoma is PAS negative and hyaluronidase sensitive and sialidase resistant [8].

It is a low grade carcinoma following an indolent course. Treatment consists of wide local excision of the tumour. Follow up is recommended as PMC are known to recur locally. Regional lymph node involvement is seen in 33% of cases involving the axilla [2], hence prophylactic lymph nodes dissection should be considered in these cases.

CONCLUSION

PMC of skin is a rare sweat gland neoplasm with a slow clinical course. Though multiple

recurrences can occur, death from the tumour is unusual. We should be cautious before making its diagnoses as it closely mimics metastatic mucinous carcinoma from internal organs and search for any primary is warranted.

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