

Eccrine Porocarcinoma of Thigh : A Rare Adnexal Tumour

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ABSTRACT

We report a rare case of eccrine porocarcinoma (EPC) in a 65 year old male patient. EPC arises from the intraepidermal portion of the eccrinesweat glands (acrosyringium). Previously the tumour was known as eccrine adenocarcinoma or malignant eccrine poroma with incidence predominantly in extreme of old age. The tumour arises most commonly on hands and feet. Rare sites like scalp, face, penis and abdomen are also reported in the literature. The distinction of porocarcinoma and basal cell carcinoma can be a diagnostic dilemma on a superficial biopsy. The histological features of high mitotic rate, tumour depth and lymphovascular invasion are bad prognostic markers. Immunohistochemistry for CK15, CK7, epithelial membrane antigen(EMA) and estrogen confirmed the diagnosis of EPC. This tumour is preferably treated with Moh's surgery though few cases of metastasis are reported.

Keywords: Basal Cell Carcinoma, CK15, CK 7, Eccrine Porocarcinoma, EMA, Immunohistochemistry.

Introduction

Eccrine porocarcinoma (EPC) is a cutaneous adnexal neoplasm. Cutaneous adnexal neoplasms are varied group of neoplasm with most challenging areas of dermatopathology. EPC is very rare tumour with incidence ranging from 0.005% to 0.01% of all cutaneous neoplasm.^[1,2] It is seen in the age group 60-80 years with female predominance.^[1,2] The tumour commonly occurs on hands and feet. We report a 65 year old male patient with EPC that developed on right thigh.

Case History

A 65 year old patient presented with complains of non healing ulcer on antero-lateral aspect of right thigh since past 6 months. The lesion was progressively increasing in size. On examination the lesion was ulceroproliferative mass, hard in consistency. The surgeons sent in a superficial biopsy to our department with a clinical diagnosis of basal cell carcinoma. On histopathological examination the biopsy revealed a palisading arrangement in a focal area with cells originating from the epidermis. The cells also showed high mitotic activity. With the atypical appearance of the malignant cells the patient was advised for a resection of the tumour in suspicion of a variant of basal cell carcinoma. Moh's resection of tumour with clear margin was performed by the operating surgeon. Surgical resection of 16x6x2 cm was received with tumour mass measuring 3 cm in diameter. Grossly all the surgical margins appeared free from tumour (Fig. 1A, 1B).

On close examination few cystic areas were noted in the superficial region along with infiltration of the tumour in the

subcutaneous region (Fig. 1C, 1D). On histopathological examination the tumour revealed atypical poroid cells infiltrating into dermis as anatomizing bands and dermal tumour infiltrate (Fig. 2A). Multiple spiraling ductules confirmed the diagnosis of EPC (Fig. 2B). Individual cells of the tumour showed marked pleomorphism with hyperchromatic nucleus showing prominent 1-2 nuclei with scant to moderate cytoplasm (Fig. 2C). Mitotic activity in few areas was > 14 mitotic figures/hpf (Fig. 2D). On immunohistochemistry (IHC) epithelial markers CK15, CK7 and EMA along with focal estrogen receptor came positive confirming EPC (Fig. 3). The initial suspicion of basal cell carcinoma was ruled out on histopathology by typical appearance of EPC with confirmation on IHC.

Discussion

In 1963 the first case of EPC, previously known as malignant eccrine poroma or eccrine adenocarcinoma, was reported by Pinkus and Mehregan using term epidermotrophic eccrine carcinoma.^[3] Since then many advances in histopathology of similar tumours and their differentiation with help of recent IHC techniques has been done. Mehregan AH et al,^[1] Robson et al,^[2] Luz MA et al,^[4] studied subsequently large group of patients, with 35, 69 and 8 cases respectively. The incidence of 0.005% to 0.01% highlights the rarity of this tumour.^[1,2] The guidelines for high grade EPC of positive lymphovascular invasion, depth of > 7mm and mitotic activity > 14 cells/hpf were seen in present case. Subsequently negative lymphovascular invasion, depth of < 7 mm and mitotic activity of < 14 mitotic cells/hpf were highlighted as features of low grade EPC. Luz Muerilo de A et al^[4] also evaluated additional two factors i.e. perineural

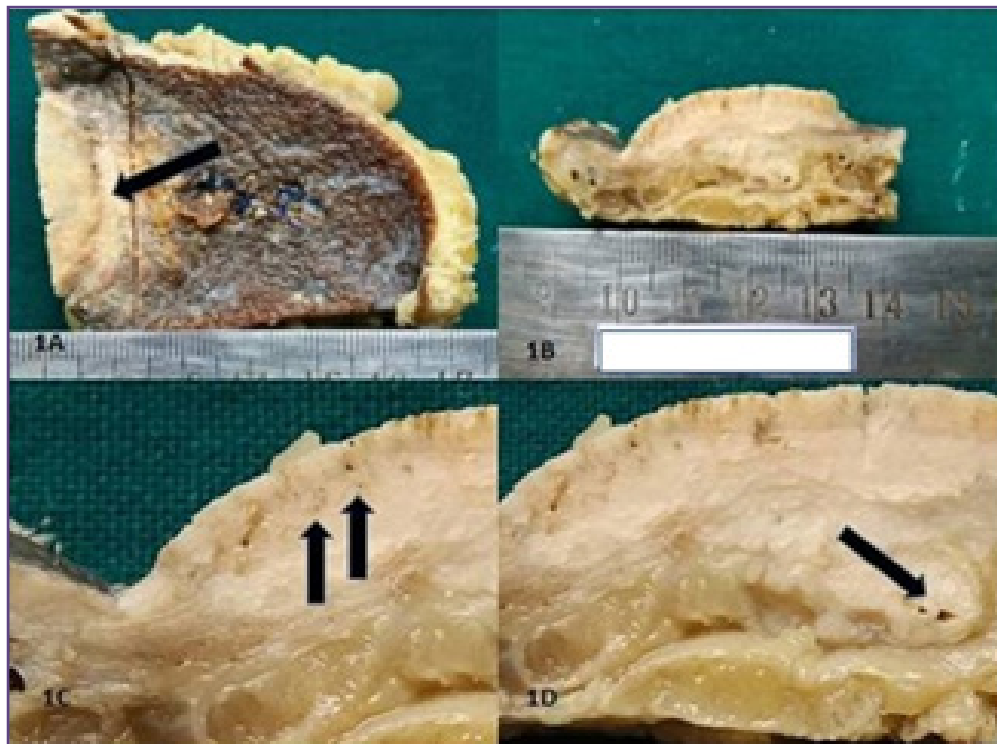


Fig. 1A Gross appearance of ulceroproliferative mass with arrow. 1B Cut section of the tumour showing mass whitish in colour. 1C arrow pointing to cystic lumina. 1D Tumour invading in subcutaneous tissue.

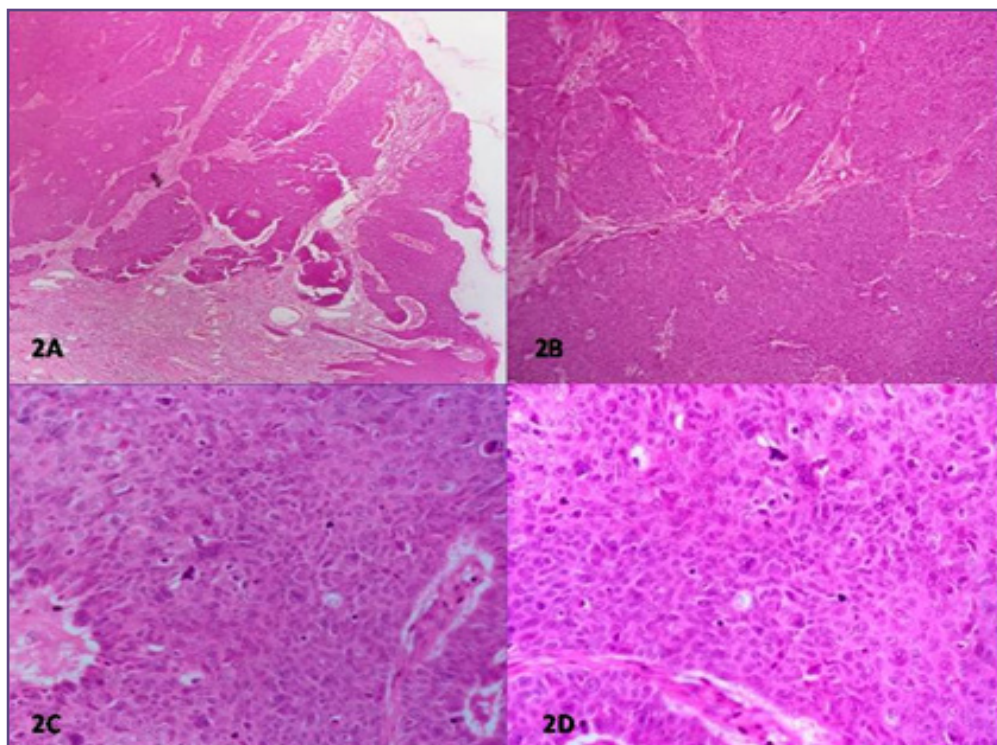


Fig. 2A photomicrograph showing tumour infiltrating into deeper dermis. H&Ex100. Fig. 2B Multiple spiraling ductules. H&Ex100. Fig. 2C & 2D Marked pleomorphism of tumor cells with high mitotic activity H&Ex400.

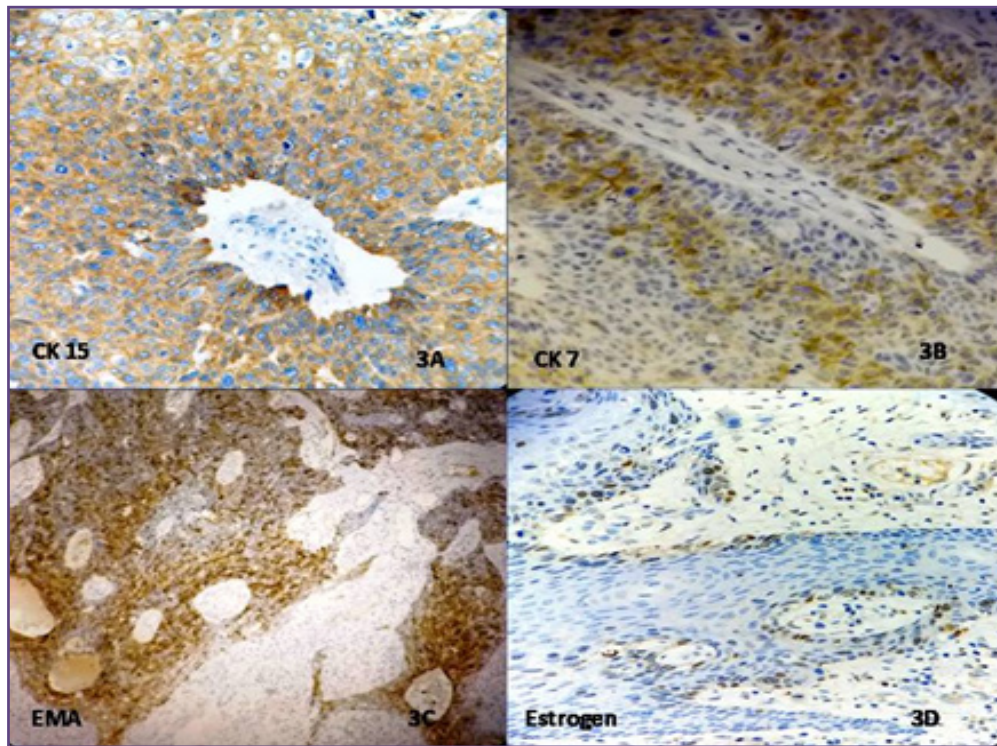


Fig. 3 Immunohistochemistry shows tumour cells positive for CK15 (3A), CK7 (3B), EMA (3C) and focal positivity for estrogen (3D)..

invasion and necrosis. Clinically the differential diagnosis of patients with age more than fifty years and lesion over the limbs include basal cell carcinoma (BCC), Paget's disease, melanoma, metastatic cancer.^[5] Histopathology with adequate resection of tumour provides a better approach to rule out these differential diagnoses.

Luz MA et al^[4] in his study of eight cases found one case of EPC mimicking basal cell carcinoma. Basaloid pattern was seen with absence of ductal differentiation as in our case initially. Similarly Klenzner T et al^[6] encountered EPC of the ear mimicking basaloid squamous cell carcinoma. These close encounters of EPCs with other adnexal neoplasm makes identification of the origin of tumour an important factor for appropriate therapy and prognosis. IHC has emerged as a very useful technique in identification of cells of eccrine sweat glands in EPCs. Low molecular weight cytokeratin are positive for excretory coil of eccrine sweat glands, epithelial membrane antigen (EMA), carcino-embryonic antigen (CEA), and S100 protein are positive in basal layer only. CEA is negative in most of EPC as in our case, it is positive in well formed ducts.^[7] Neoplastic cells with myoepithelial differentiation exhibits immunoreactivity for S-100 protein in eccrine spiradenoma, eccrine acrospiroma, and dermal mixed tumour, the stain

is useful for the differential diagnosis of these tumours with EPC.^[8] The acrosyringial cells stain for high molecular weight keratin. Some EPCs show positivity for estrogen and progesterone receptors which have a important clinical implication, as affected patients may be treated with hormonal therapy along with the main line treatment.^[9] The EPCs has a tendency to arise on lower limbs (44%), trunk (24%) and head and neck region (18%).^[2] Rare cases of EPCs infiltrating in cranium and involvement of penis are also reported.^[10,11] Regional lymph node metastases are found in about 20 % of EPCs and distant metastases in 10 % which have not been observed in the present case.^[4]

Conclusion

In this paper we report a rare case of EPC which mimicked basal cell carcinoma on a superficial biopsy. The suspicion highlighted the importance of adequate surgical excision for histopathological diagnosis and treatment. In spite of the clear resection, an accurate prediction of the tumour outcome is challenging. Early detection with complete resection provides better prognosis.

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