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ECCRINE POROCARCINOMA – A RARE MALIGNANT SKIN LESION

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ABSTRACT

Eccrine porocarcinoma (EPC) is a rare type of skin cancer arising from the intra epidermal portion of eccrine sweat glands, representing 0.005-0.01% of all cutaneous tumors. About 20% of EPC recur and about 20% will metastasize to regional lymph nodes. These cancers have a mortality rate of 67% in patients with lymph node metastases. Due to its nonspecific clinical presentation, it can be misdiagnosed with Basal Cell Carcinoma (BCC) and Trichoepitheliomas. Hence the diagnosis of EPC must be based on histopathological features and IHC markers because of its potential to metastasize to distant organs. The main line of management is gross total surgical excision with a wide safety margin. Adjuvant chemotherapy and radiotherapy are used in cases of metastasis and to reduce local recurrence. Here we present a case of a 50 year old female who presented with a swelling with an overlying ulceration on the back of her left thigh. The swelling was intermittently painful. On examination the swelling was 2.2 x 2 x 1cm on left thigh region and the clinical diagnosis was given as papilloma. An excision biopsy was done. Histopathological and immunohistochemical examination showed features of Eccrine porocarcinoma. This case is reported here due to its extreme rarity.

KEYWORDS: *Eccrine porocarcinoma; Malignant skin adnexal tumors; Sweat gland tumors; Adnexal tumours.*

INTRODUCTION

Eccrine porocarcinoma (EPC) is an extremely rare malignant tumor of skin cancer arising from the intraepidermal portion of eccrine sweat glands. It accounts for 0.005-0.01% of all skin tumors with a potential for local destruction and metastasis.[1,2] It is the malignant form

of eccrine poroma, a common benign skin tumour of sweat glands.[3] Thigh is a rare site for EPC with a very few cases reported in the literature.[1,3] EPC may develop *de novo* or as a result of malignant transformation of eccrine poroma.[6] About 20% of EPC show recurrence and about 20% tend to metastasize to regional lymph nodes. There is a mortality rate of 67% in patient with lymph node metastases. The main line of management is gross total surgical excision with safety margin.[11] Adjuvant chemotherapy and radiotherapy are used in cases of metastasis and to reduce local recurrence.

CASE REPORT

We present a case of a 50 year old female who came to general surgery out patient department with chief complaints of swelling on back of the left thigh region for 2 years with on and off pain. On examination, there was an ulceration over the swelling measuring about 0.8 cm and the swelling was 2 x 1.3 x 1cm on left thigh region. An excision biopsy was done and on histopathological examination followed by immunohistochemical examination showed features of Eccrine porocarcinoma. A single fragment of skin which was congested and gray white in color with underlying soft tissue mass measuring 1.7x1x0.7cm was received for histopathological examination. On microscopic examination, the section studies showed a stratified squamous epithelium with focal erosion and proliferation of non pigmented small keratinocytes. (Figure 1) The tumor cells extending from the epidermis to the dermis, were arranged in nests, ducts and few squamous island. (Figure 2) Few scattered lymphocytes, neutrophils, fibrinoid material and congested blood vessels and few mitotic figures were also seen. (Figure 3) A probable diagnosis of eccrine porocarcinoma was given. On further immunohistochemistry for confirmation, EMA showed 60 to 70 % with moderate to strong cytoplasmic membrane positivity and Ki 67 showed more than 50% of strong nuclear positivity with high proliferation index. CK 20 showed negative staining. All these findings helped in the confirmation of the diagnosis of eccrine porocarcinoma.

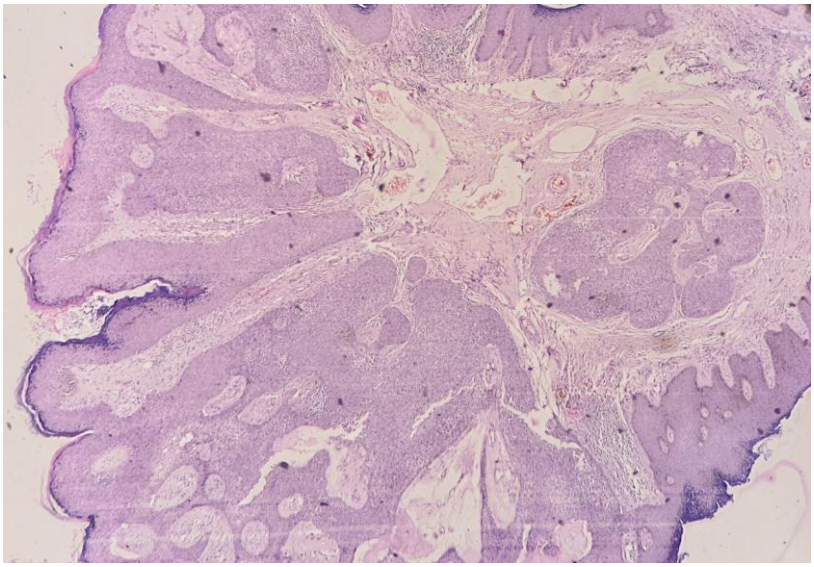

Fig. 1: Tumor nests extending from epidermis to dermis (H & E; 4X).

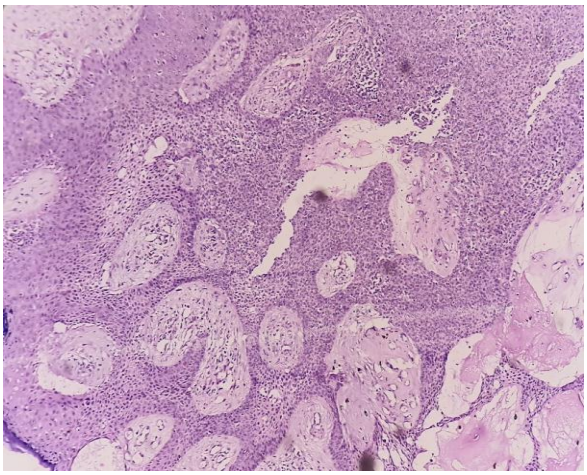

Figure 2: Tumour nests (H & E; 20X).

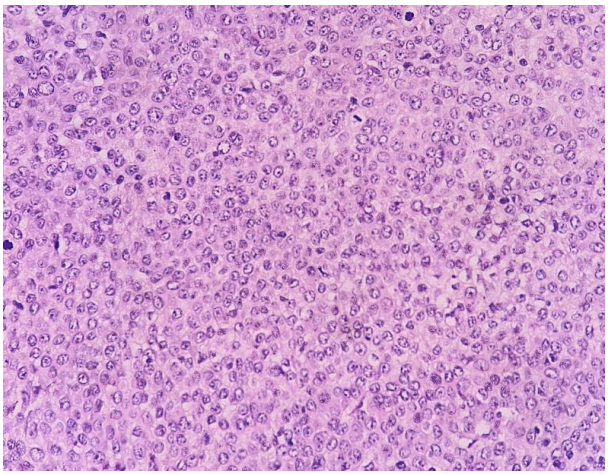

Figure 3: Tumour cells with mitoses (H & E; 40X).

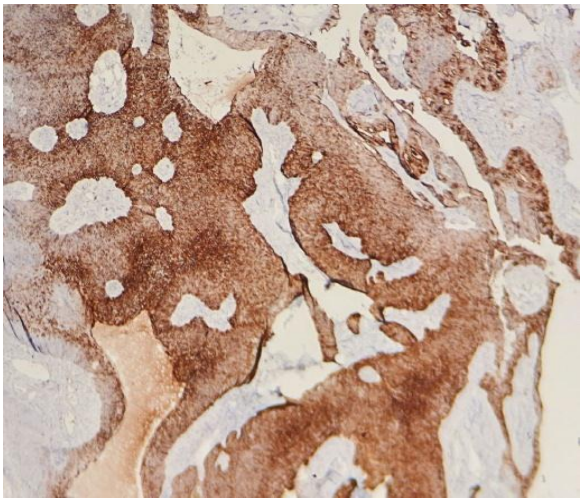

Figure 4a: EMA positive

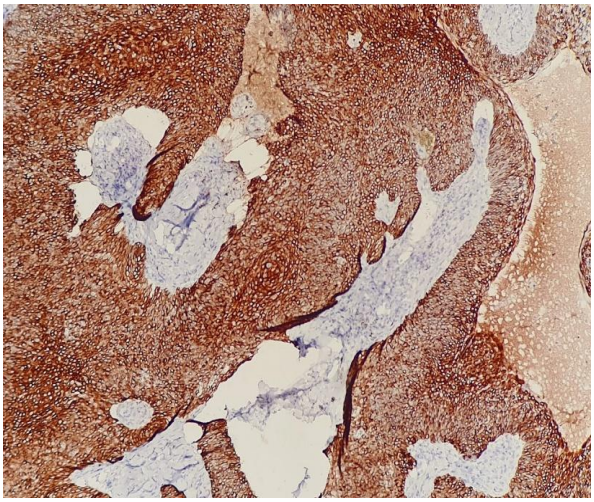

Figure 4b: EMA positive.

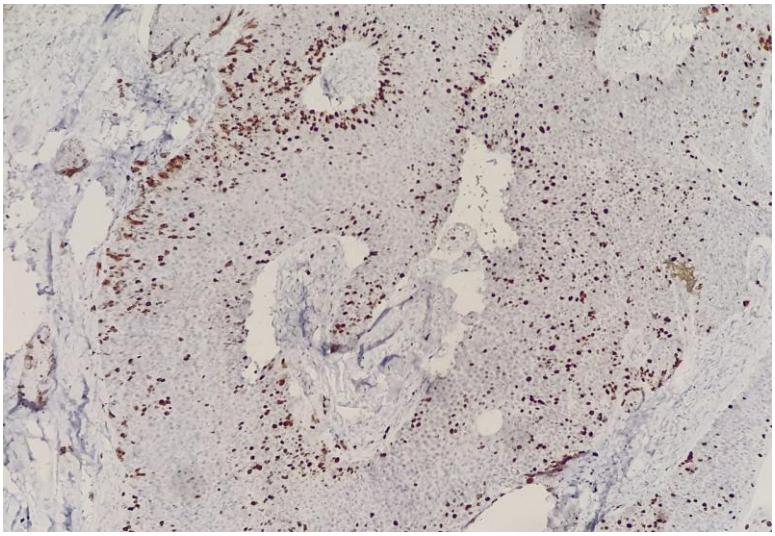

Figure 5: Ki67 showing more than 50% positivity.

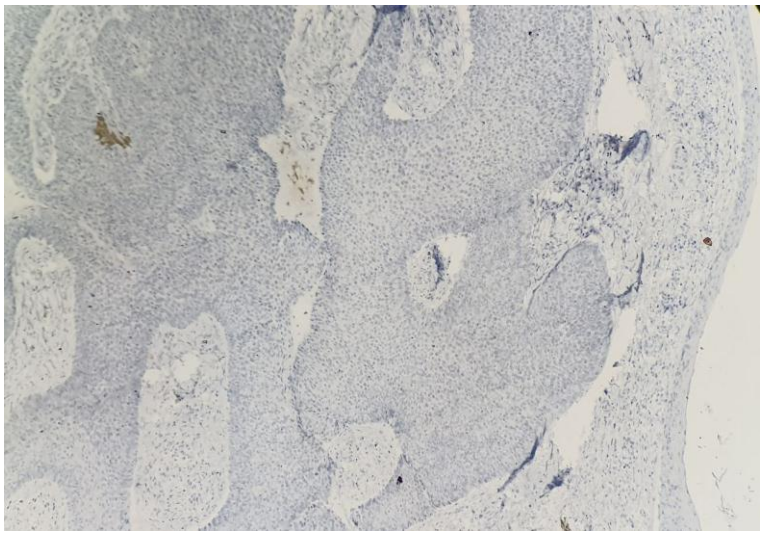

Figure 6: CK 20 Negative.

DISCUSSION

Eccrine porocarcinoma (EPC) is a potential neoplasm of the skin that arises from eccrine sweat glands. It was previously known as an eccrine adenocarcinoma or malignant eccrine poroma.[1] The first case of eccrine porocarcinoma was reported by Pinkus and Mehregan in 1963.[12] It commonly occurs between 60 to 80 years of age. Usually arising from hands and feet commonly in lower extremities. In the primary tumor, the malignant cells arise from the intraepidermal portion of the eccrine sweat glands and may be limited to the epidermis or may extend into the dermis.The tumour is asymmetrical with an infiltrative growth pattern.[11] Infiltrating tumour shows multiple attachments to the epidermis.Two types of atypical cells: eosinophilic and clear cells are present in these tumours.[4] The eosinophilic cells appear as polyhedral or fusiform with round to oval hyperchromatic nuclei, distinct

nucleoli, indistinct cell boundaries, and a variable amount of cytoplasm. The clear cells appear as large and polyhedral with round to oval nuclei, abundant clear cytoplasm, and distinct cell borders. Our case showed both types of cells.

The two main differential diagnoses are Basal cell carcinoma and Trichoepithelioma. For basal cell carcinoma, clinical appearance often parallels to eccrine porocarcinoma. Most common appearance is a papule or nodule with eroded or ulcerated. The presence of myxoid stroma and peripheral clefting has been suggested to be most helpful to distinguish BCC from porocarcinoma[10] BCC stains positive for p53, BCL2 and p63 and negative for EMA, CEA and CK20 IHC markers.

Trichoepitheliomas are benign adnexal tumors which may display some features that mimic like eccrine porocarcinoma. Most common tumor types of trichoepitheliomas have an association with nevus sebaceus. Papillary mesenchymal bodies and calcifications is present in trichoepitheliomas.[7] These tumours may have epidermal connection but ulceration is rare. The IHC marker CK 20 shows positive staining.

CONCLUSIONS

The present case is being reported because of its rarity and it is challenging to diagnose eccrine porocarcinoma based on clinical presentation alone. Hence histopathology along with immunohistochemistry are always the mainstay for early definitive diagnosis and treatment of these tumours. Local excision with negative margins is treatment regimen and if regional lymphnodes are involved then lymphadenectomy should be done. Perilesional injection of interferon alpha and interleukin 2 has been reported to produce a partial response.

Conflict of interest

There is no conflict of interest.

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Nil

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published.

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