

Case Report

An ulcerated exophytic tumour over the gluteal area- eccrine porocarcinoma in an immunocompetent female

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ABSTRACT

Porocarcinoma is a very rare type of skin cancer arising from the eccrine sweat glands and is poorly understood. The exact pathogenesis is unknown. Clinical features are non-specific. It may present as a painless, slow growing mass or nodule, ulcer, plaque, swelling, wart, papule or naevus that increases in size over years to decades and may ulcerate or bleed upon trauma. They occur equally in both sexes and are most commonly found in the lower extremity. Though metastasis is rare, they are considered aggressive with high mortality rate. Diagnosis requires standard histopathological studies. Immunohistochemistry is useful in difficult cases. Treated early, eccrine porocarcinoma is curable by wide excision. The author reports the case of an eccrine porocarcinoma in the lower extremity of an elderly lady and a summary of its clinical, radiological and histological findings is illustrated.

Keywords: Porocarcinoma, Appendageal tumour, Eccrine sweat gland tumour

INTRODUCTION

Eccrine porocarcinoma is an extremely rare malignancy. It arises from eccrine sweat glands and accounts for 0.005–0.01% of all cutaneous tumours.¹ It is encountered in the 6th or 7th decade of life. The tumour exhibits non-specific clinical features and can present as a plaque, nodule, and ulcer. Porocarcinoma may mimic some cutaneous lesions like seborrheic keratosis, wart, nevus, pyogenic granuloma, basal cell carcinoma, achromic melanoma, and Bowen's tumour. Although metastasis has been described in 20% cases only, it is associated with high mortality and poor response to radiation. Metastatic porocarcinoma is treated with retinoids, tamoxifen and docetaxel. Histopathology clinches the diagnosis and is based on morphology rather than immunohistochemistry, there may be an expression of carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA); PS-100 is negative. However,

these markers are also not very reliable. In view of its rarity of occurrence, we report a case of eccrine porocarcinoma presenting as an exophytic, ulcerated growth over the right gluteal region in a 62-year-old immunocompetent woman. The purpose of this report is to illustrate the features of porocarcinoma and to emphasize the need for its prompt histopathological diagnosis.

CASE REPORT

An immunocompetent 62-year-old Indian lady presented with bleeding from a skin lesion present over the lateral aspect of her right gluteal region for 3 months. The swelling was slow-growing and painless, measuring 4x5x1cm, with well-defined margins and an ulcerated surface (Figure 1). Regional lymph nodes were not involved. Differential diagnoses included ulcerated lupus

vulgaris, squamous cell carcinoma, extramammary Paget's disease, and pyoderma gangrenosum.



Figure 1: An exophytic nodular swelling over the lateral aspect of the right gluteal region with an ulcerated surface. A wide local resection was done and the surgical site was allowed to heal by secondary intention.

Biopsy from the ulcer margin showed polygonal cells in the epidermis which were infiltrating the dermis, arranged in nests, cords, and sheets. Moderate pleomorphism, round to oval hyperchromatic nucleus, inconspicuous nucleolus and moderate amount of clear to eosinophilic cytoplasm was seen. Immunohistologically, the tumour was focally positive for EMA, CEA and P63 (Figure 2). These features were consistent with a diagnosis of porocarcinoma. Computed tomography (CT) scan revealed a soft tissue tumour in the skin and subcutaneous plane of the right gluteal region measuring 43×35 mm with no signs of lymphadenopathy and metastasis. The patient underwent wide local excision of the lesion with a 2 cm margin. Grossly, the tumour was exophytic with ulceration present on the surface. All resected margins were free of tumour. The resected site was allowed to heal by secondary intention. The patient remains symptom and recurrence-free on follow up till date.

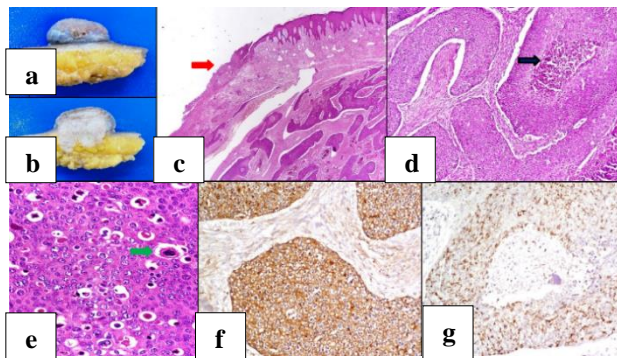


Figure 2: (a, b) Exophytic tumour with superficial erosion and crusting mimicking squamous cell carcinoma or cutaneous metastasis. Tumour having sharp, infiltrative growth pattern; (c, d) multiple sections examined from the tumour shows surface epithelial ulceration (red arrow), an infiltrative tumour with solid aggregates of invasive tumour cells growing deep into the dermis and adjacent subcutis. There are areas of comedo necrosis (black arrow) and prominent nuclear pleomorphism along with squamous differentiation; (e) atypical mitosis; (f) and (g) tumour cells focally immunopositive for CEA along with EMA.

DISCUSSION

The term “eccrine porocarcinoma” was first described by Mishima and Morika in 1969.² It arises from the intraepidermal ductal portion of eccrine sweat glands. The exact etiopathogenesis is unknown. There are two accepted tumorigenesis pathways: denovo generation and transformation from a benign precursor lesion such as a poroma. Akalin et al showed that P53 expression may have a role in carcinogenesis.³

A metanalysis of 453 patients showed 49% cases were male and 51% were female. The mean age was 67.57 years ranging from 6 months to 97 years of age.⁴ The mean duration of presentation was 5.57 years ranging from 4 days to 60 years. The most common sites of affection are the head and neck (39.9%) followed by lower extremity (33.9%). Metastasis at presentation was noted in 110 (31%) cases.

Our patient presented with a de novo tumour over the gluteal area (most commonly involves lower extremity) as a painless, nodular growth with ulceration. Both immunocompromised state (immunosuppressive therapy for renal transplantation and chronic lymphoid leukemia) and sun exposure have been reported as risk factors.⁵ However, our patient was immunocompetent and had involvement in the non-sun exposed covered site.

The growth of the tumour was rather rapid in our case and the ulcerated surface raised suspicion of malignancy which prompted us to do a biopsy from the ulcer margin which confirmed the diagnosis of porocarcinoma. Investigations revealed a circumscribed tumour with no evidence of lymph node involvement or distant metastasis. Keeping in view the localized stage of the tumor a wide local excision of the tumour with a 2 cm margin was performed. The risk of local recurrence to porocarcinoma is about 20% and hence histologically clear margins is mandatory.⁶ Some meta-data, however, favors lymph node dissection due to high rate of metastases to lymph nodes. The exact survival rate in porocarcinoma is not known. Follow up of the patient up to 1 year has not shown any recurrence or metastasis. The patient has been counselled regarding the strict follow up.

CONCLUSION

We reported a case of eccrine porocarcinoma in a 62-year-old immunocompetent Indian woman over the gluteal region. Considering its rarity, non-specific presentation and high rate of metastasis, any atypical mass/red erosive tumour presenting to dermatologists mandate high suspicion for a malignant tumor such as eccrine porocarcinoma. Early histopathology is key to success for its management.

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