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CASE REPORT

Eccrine Porocarcinoma in Linear Epidermal Nevus

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Abstract: Linear epidermal nevus is a congenital focal epidermal dysplasia common at birth or during childhood. Linear epidermal nevus followed by cutaneous malignancy is extremely rare. Here, a case of linear epidermal nevus followed by eccrine porocarcinoma (EPC) is reported.

Keywords: linear epidermal nevus, eccrine porocarcinoma

Introduction

Generally, linear epidermal nevus presents as linear distributions of skin-colored, light red, or yellow-brown keratotic papules and plaques.[1] Linear epidermal nevus followed by malignancy is very uncommon. Currently, most reported malignancies following linear epidermal nevus are epithelial-derived tumors, such as basal cell carcinoma (BCC),[2] squamous cell carcinoma,[3,4] and keratoacanthoma.[5] However, skin adnexal malignancies have rarely been reported.[6,7] Here, we describe one case of eccrine porocarcinoma arising from linear epidermal nevus.

Case Report

A 77-year-old man presented with linear yellow-brown keratotic papules and plaques on the left side of the abdomen for over 70 years and erythematous plaques on the lesion for 8 months. The yellow-brown papules and plaques have occurred since childhood. Eight months before the presentation, erythematous plaques with erosion developed on the existing lesion, which was hard to cure. There was no subjective symptoms. Dermatologic examination showed linear, dense, yellow-brown papules and plaques on the left side of the abdomen, which were well-circumscribed and had a rough surface. In the meantime, an erythematous plaque (approximately 1 cm in diameter) with mild surface erosion and yellow overlying eschar was found on the lesion (Figure 1). The histopathology of the erythematous plaque showed hyperkeratosis and a large number of tumor masses in the epidermis and mid dermis. The masses were composed of basaloid cells, which had marked atypia and formed ductal structures (Figure 2a and b). Linear epidermal nevus followed by eccrine porocarcinoma (EPC) was diagnosed. Extended tumor resection was performed. There was no evidence of recurrence at 1 year of follow-up, and the patient continues to be followed up regularly.

Discussion

Eccrine porocarcinoma (EPC), first reported in 1963, is a rare skin adnexal malignancy derived from eccrine ducts. The pathogenesis of EPC is unknown, and the EPC following linear epidermal nevus is likely to develop from the epidermal nevus because of the immature eccrine structures in the diseased tissues.[7] The current study contends that some specific oncogenic drivers, including epidermal growth factor receptor (EGFR), tumor protein p53 (TP53), cyclin-dependent kinase inhibitor 2A (CDKN2A), and mitogen-activated protein kinase (MAPK), play a role in the pathogenesis of EPC.[8]

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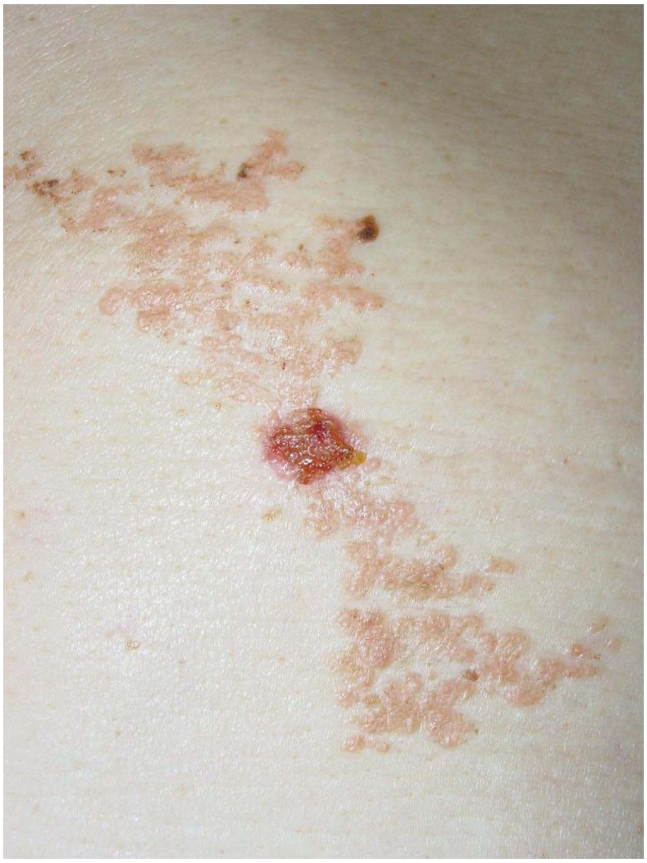

Figure 1 Physical examination showed linear, dense, yellow-brown papules and plaques on the left side of the abdomen, which were well-circumscribed and had a rough surface.

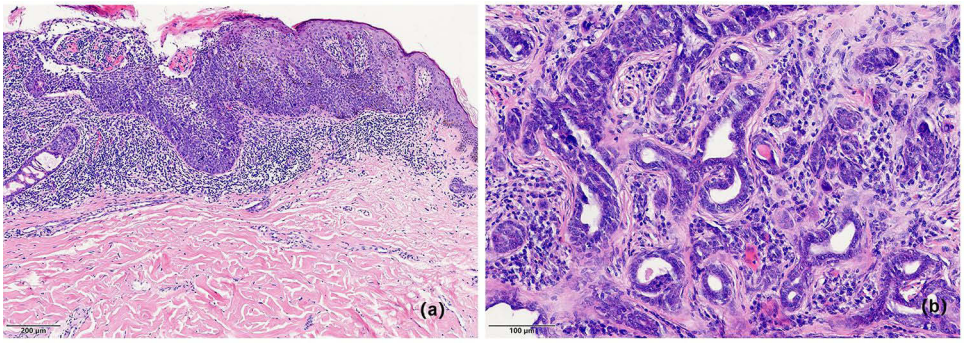

Figure 2 (**a**) Histopathological image showed hyperkeratosis and a large number of tumor masses in the epidermis and mid dermis (hematoxylin-eosin, x40). (**b**) The masses were composed of basaloid cells, which had marked atypia and formed ductal structures (hematoxylin-eosin, x100).

EPC is prevalent among those aged 60 to 80 years old, with a comparable proportion of males and females. Related skin lesions occur mostly in the lower extremities (44%), followed by the head and neck (30.6%), the trunk (24%), and some other infrequent sites.[9] There are no specific clinical manifestations, but nodules, plaques (with or without verrucous hyperplasia, ulcers, and polyps), and occasionally skin breakage and bleeding are observed. The diagnosis of EPC is mainly based on the histopathological findings presenting the appearance of invasive tumor masses confined to the epidermis or dermis. The tumor masses are mainly composed of basaloid and squamous epithelial-like cells, which have significant atypia with large hyperchromatic nuclei and common pathological

nuclear divisions. Notably, tumor cell differentiation to the ductal epithelium is an essential clue for diagnosis. Immunohistochemistry of the tumor cells generally shows positive CEA and EMA expression.[10] Nevertheless, a study reported that positive CEA expression existed in only a few cases presenting with well-differentiated ductal structure. The differential diagnoses of EPC include pyogenic granuloma, BCC, and squamous cell carcinoma. Surgical procedures for EPC can be extended tumor resection or Mohs micrographic surgery. Adjuvant radiotherapy after surgical removal of EPC could be considered in cases with positive or close margins and in cases with unfavorable histological features.[11] In the patient reported here, linear yellow-brown papules and plaques have occurred since childhood, which is the typical manifestation of linear epidermal nevus. Furthermore, the lesion's development of erythematous plaques is consistent with the clinical manifestation of EPC. Based on histopathological and immunohistochemical findings, linear epidermal nevus followed by EPC was confirmed. A long-term follow-up was performed because of the risk of recurrence and metastasis.

Conclusion

We need to pay attention to the follow-up of linear epidermal nevus based on their potential risk of developing cutaneous malignancy.

Consent Statements

Written informed consent was provided by the patient to have the case details and accompanying images published. Institutional approval was not required to publish the case details.

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Disclosure

No potential conflict of interest was reported by the author(s).

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