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CASE REPORT

Metastatic squamous cell carcinoma arising from a serious widespread porokeratotic eccrine ostial and dermal duct nevus: Case report and literature review

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ABSTRACT

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare benign skin disease. Widespread involvement of PEODDN is more uncommon and its association with squamous cell carcinoma (SCC) is also rare. In this case report, we present the case of a 50-year-old woman with serious widespread PEODDN along Blaschko's lines and concomitant invasive SCC with lymph node metastasis. This was the first reported case of metastatic SCC arising from PEODDN.

Introduction

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a congenital hamartoma of possible eccrine origin with no malignant potential. It is usually localized at the extremity, while extensive distribution involving the trunk and extremities has rarely been documented and association with squamous cell carcinoma (SCC) has also rarely been reported. A woman with a serious widespread PEODDN with a concomitant metastatic SCC is reported herein.

Case Report

A 50-year-old woman presented with many asymptomatic grouped comedo-like papules on her trunk and extremities, which were present since birth, in addition to an ulcer on her left lower leg, which was present for 2 months. With age the lesions had become larger and some of them showed confluence, especially on her extremities. She denied any history of systemic diseases. No one else in her family had similar complaints.

Dermatological examination revealed multiple hyperkeratotic papules and filiform spine over her trunk and extremities where lesions presented along Blaschko's line in the form of a systematized linear epidermal nevus (Figures 1A–1F). On closer examination, the hyperkeratotic lines were found to be composed of small filiform keratotic plugs. At their base, the plugs had a pitted keratotic papule similar to a comedo nevus. On her left lower extremity, the more serious lesions had arisen in a thornlike fashion vertically (Figures 1D–1F). The thornlike projections with larger plugs were very closely grouped and some of them coalesced into hyperkeratotic verrucous plaques. Those plugs could be extracted and some pits remained. Both palms and soles had similar lesions and the soles were highly hyperkeratotic. At the left pretibial site, a demarcated circular ulcer 2.5 cm in diameter was found. The head and face were spared. The systemic examination, including chest X-

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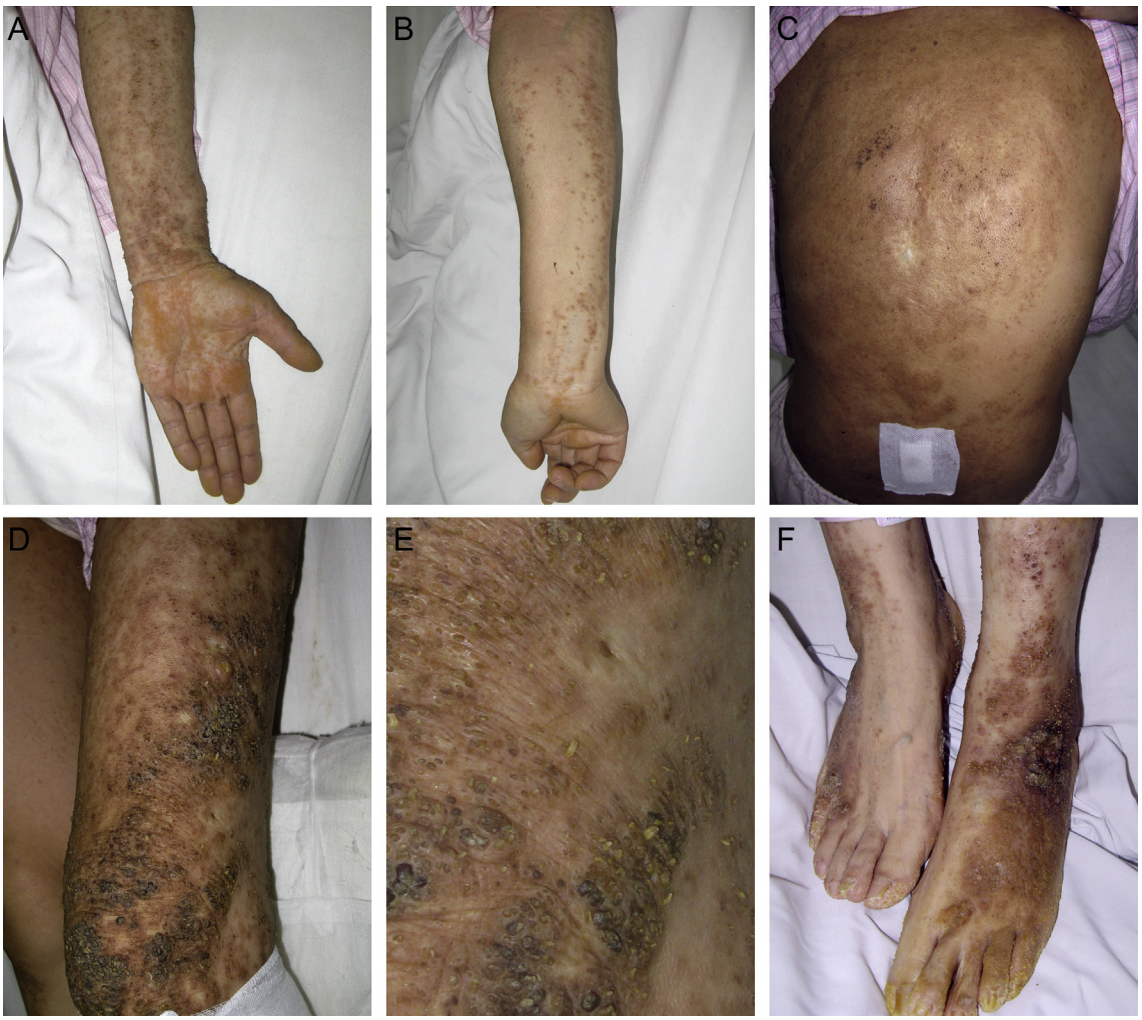

Figure 1 (A) Multiple comedo-like papules on the left upper extremity. (B) Multiple linear comedo-like papules on the right upper extremity. (C) Some small pitted keratotic papules on the back. (D) On the left lower extremity, the more serious lesions had arisen in a thornlike fashion and coalesced into hyperkeratotic verrucous plaques. (E) Many thornlike and comedo-like lesions on the left leg in a close-up view. (F) Some comedo-like papules coalesced into verrucous plaques on the dorsal feet.

ray, electrocardiogram, echocardiogram, and ultrasonogram of the abdomen, was normal.

The biopsy of papules from the left leg showed a parakeratotic column associated with eccrine ducts, appearing as cornoid lamella similar to porokeratosis and standing within an epidermal invagination. The cornoid lamella-like columns were composed of compact hyperkeratosis with underlying hypokeratosis. The granular layer was absent in this invagination, which contained vacuolated keratinocytes. The eccrine ducts in the dermis were significantly tortuous, dilated, hyperplastic, and some were full of sweat (Figure 2A). In addition, significant papillomatosis with irregular acanthosis and some inflammatory cells around the invaginated epidermis were observed. No involved follicles were found. All these histopathological features were consistent with PEODDN. The histology of the ulcer showed poorly differentiated tumor nests with some atypical keratinocytes and necrosis in the dermis (Figure 2B). Atypical cells exhibited glassy eosinophilic cytoplasm, hyperchromatic nuclei, atypical mitoses, and disorganized architecture (Figure 2C). These features of the ulcer conformed to invasive SCC. The SCC lesion was surgically excised and no management of PEODDN was initiated because of the patient's refusal. Unfortunately, lymphatic metastasis of her left groin was found 8 months later (Figure 2D).

Discussion

PEODDN is a rare hamartoma of the eccrine sweat glands first termed by Abell and Read in 1980.[1] Recently, the term *porokeratotic adnexal ostial nevus*, which includes PEODDN and porokeratotic eccrine and hair follicle nevus (PEHFN), was proposed because some cases had involvement of both acrosyringia and acrotrichia histologically.[2] The pathogenesis of PEODDN is still unclear. It has been suggested that PEODDN could be a congenital hamartoma of the acrosyringium and dermal duct.[2] In a recent report, it was considered that PEODDN is a mosaic form of keratitis–ichthyosis–deafness syndrome with a gene mutation in *GJB2* encoding a gap junction protein connexin 26 (Cx26).[3]

The lesions of PEODDN are usually present at birth. However, rare cases of late onset in adults had also been reported.[4] The usual distribution of these lesions is the distal portion of a limb. A widespread distribution of PEODDN along Blaschko's line is a rare condition.[5,6] The lesions are composed of small keratotic papules or comedo-like pits filled with keratotic plugs. The plugs usually arise vertically from the skin surface as multiple tiny filiform spines. In serious cases, the thornlike projections with larger plugs can be closely grouped and some of them can coalesce into hyperkeratotic verrucous plaques as in our case. It is usually asymptomatic, but

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