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Case Presentation

Porokeratotic eccrine ostial and dermal duct nevus - revisited

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Abstract

We hereby report a rare case of a 14-year-old girl presenting with asymptomatic pitted papules over the flexor aspect of her right 4th and 5th digits. This was histopathologically proven to be porokeratotic eccrine ostial and dermal duct nevus (PEODDN).

Keywords: Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) , porokeratosis, eccrine glands.

Introduction

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a benign skin condition characterized by multiple keratotic papules and plaques with plugged pits having a comedo-like appearance, predominantly occurring on the acral extremities. The characteristic histological features are that of a cornoid lamella with an absent or decreased underlying granular layer and dyskeratotic cells involving the acrosyringia [1, 2]. The name was first coined in 1980 by Abell and Read [3]. However, the condition was first described by Marsden in 1979 as “comedo nevus of palm [4].” Although various treatment options can be tried, no conservative management has been reported to be fully effective in the literature [2]. Herein we report a patient with PEODDN because of its rarity and its therapeutic response to topical therapy.

Case synopsis

A 14-year-old girl presented with asymptomatic pitted papules on the 4th and 5th digits of her right hand since infancy. The lesions were very slowly progressive. There was no history of similar skin findings in any member of her family. On examination, there were multiple punctate pits with comedo-like keratinous plugs on the antero-medial aspect of the 4th digit

and on the lateral side of the adjacent 5th digit of right hand (Figure 1). The keratinous plugs were not easily removable. Thorough systemic evaluation and routine laboratory investigations did not reveal any abnormality. A 4mm punch biopsy was performed including a keratotic pit. On histopathology there was a dilated comedo-like epidermal invagination, which was filled with keratin. Parakeratosis was evident at the bottom of the invagination, beneath which, there was absence of the granular cell layer (Figure 2). The keratinocytes in that region had vacuolated cytoplasm and pyknotic nuclei. There were a few eccrine units in the dermis, in close approximation to the epidermal invagination (Figure 3). Although there was parakeratosis in the bottom of invagination, we could not demonstrate a well-formed column of parakeratosis in our case, even after repeated sectioning of the tissue. On the basis of clinical and histopathological findings, we made the diagnosis of PEODDN. We prescribed her topical tretinoin cream (0.05%) once daily and topical urea cream (10%) twice daily. After 6 months of continuous application, the patient showed marked improvement clinically (Figure 4).



Figure 1. Plugged papules on the antero-medial aspect of the 4th digit and lateral side of the 5th digit of right hand

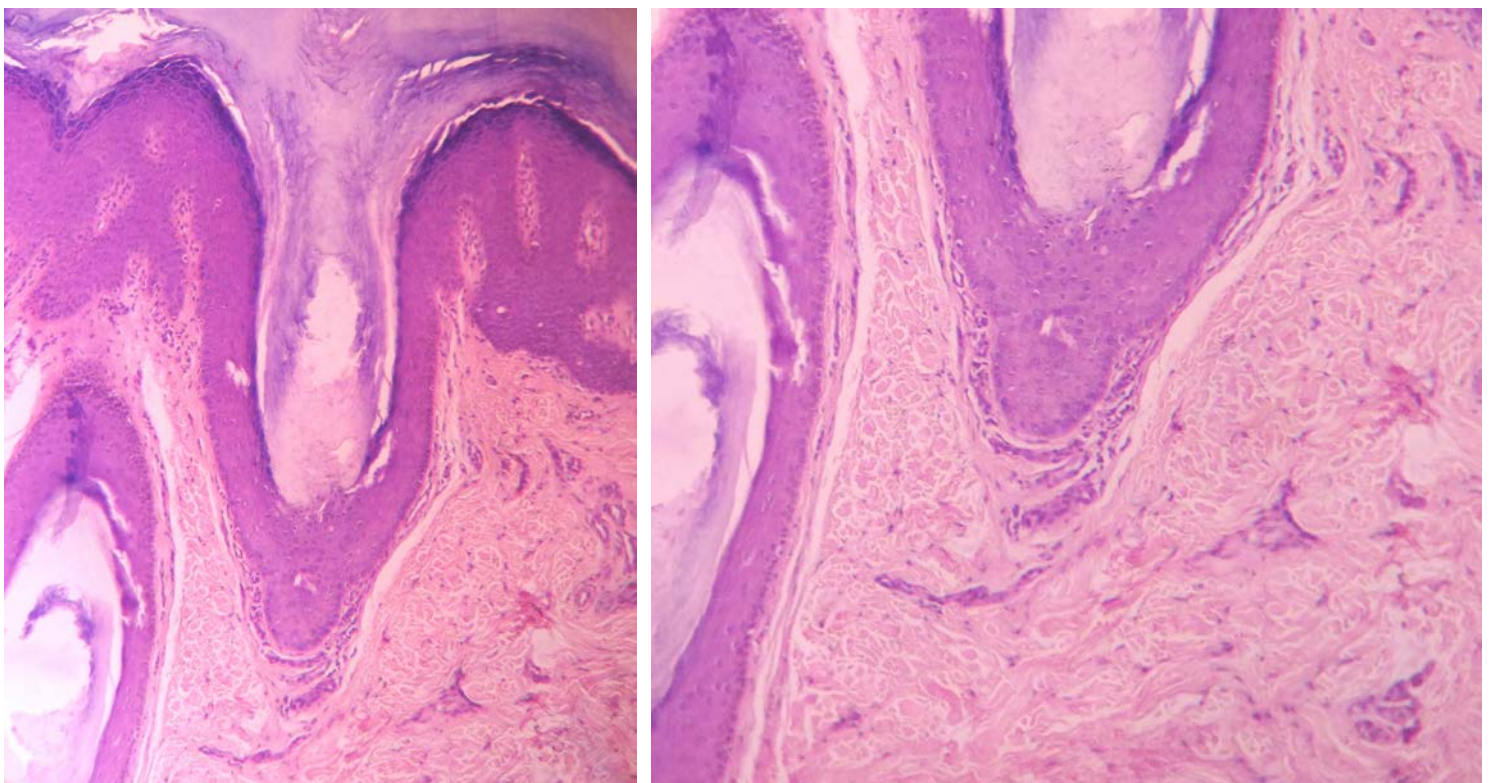


Figure 2. Photomicrograph shows hyperkeratosis, keratin filled epidermal invagination, irregular acanthosis, and focal loss of granular cell layer (H and E, $\times 40$).

Figure 3. Photomicrograph shows focal parakeratosis, absent granular cell layer, and dyskeratotic cells in epidermis with eccrine units in close approximation to epidermis (H and E, $\times 100$).



Figure 4. Clinical improvement after 6 months of topical therapy

Discussion

PEODDN is predominantly a mosaic genetic defect. Immunohistochemistry with CEA staining supports the theory that it represents an abnormally keratinizing epidermal invagination through which an acrosyringium traverses [1, 2].

Clinically it is characterized by linear or band like lesions consisting of small keratotic papules with a central plugged pit, predominantly occurring over palms and soles, though other sites may be involved [1]. It usually starts at birth or in early childhood, but late onset also has been reported [1, 5]. Lesions are mostly localized, but a few cases have been reported with extensive involvement [2]. The condition is usually asymptomatic or pruritic and may be associated with hyper or hypohidrosis [1]. Other systemic involvement is very rarely associated, although a few cases of co-incidental cutaneous disorders like Bowen's disease and linear psoriasis have been reported [6,7].

Histologically, PEODDN is characterized by dilated comedo-like epidermal invaginations filled with parakeratotic columns. The granular layer is absent at the base of these columns and in the lining of the comedo-like structures. Keratinocytes with vacuolated cytoplasm and pyknotic nuclei may also be present. In the lower portion of the invaginations, intraepidermal eccrine ducts are generally visible. It may be associated with hyperplastic intradermal eccrine ducts with comma-shaped extensions. However, many cases were also reported showing no connection between the parakeratotic columns and eccrine ducts [1].

The differential diagnoses include nevus comedonicus, punctate keratoderma, linear verrucous epidermal nevus, inflammatory linear verrucous epidermal nevus, and linear porokeratosis [5]. PEODDN is usually differentiated from these

entities by its characteristic presentation and histological presence of porokeratosis-like features in close association with the eccrine unit.

Surgical excision is the mainstay of treatment [1]. Erbium/CO₂ laser therapy was found to be effective in a report [8]. Other treatment modalities that have shown unsatisfactory results include topical and systemic retinoids, topical keratolytics, emollients, topical steroids, topical 5-fluorouracil, CO₂ laser therapy, and cryotherapy [2].

In our case, the diagnosis of PEODDN was made on the basis of the typical clinical presentation and histological features such as the presence of parakeratosis and vacuolated keratinocytes with absence of granular layer in the epidermis and the presence of eccrine units just below the epidermal invagination. Conservative therapy was helpful.

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