Porokeratotic eccrine ostial and dermal duct nevus (PEODDN): A case report

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INTRODUCTION

The term Porokeratotic Eccrine Ostial and Dermal Duct Nevus (PEODDN) was first coined in 1980 by Abell and Read 1. However, it was first described by Marsden in 1979 2. PEODDN is a very rare skin condition which is classified as porokeratotic dermatoses. It is characterized by cornoid lamella which is a column of parakeratotic cells and is associated with dyskeratosis in the spinous layer as well as reduction in the number of granular zone cells. It is in close association with subjacent acrosyringia 3. Hereunder, we report a case with this rare condition.

CASE REPORT

A 25-year-old male came to our outpatient clinic with a ten-year history of a mildly itchy lesion on his left foot. No other symptoms were detected. Past history as well as family and drug history was unremarkable. The lesion was unilateral and limited to the lateral surface of left foot. It consisted of multiple erythematous papules which were distributed in a linear fashion and plugs were seen in the center of most lesions (Figure 1, 2). No other cutaneous lesions were noted. General physical examination showed no abnormality. A skin biopsy showed cornoid lamella which is exclusively associated with eccrine acrosyringia (Figure 3).

DISCUSSION

To date, only 42 cases (Medline search) of PEODDN have been reported. Many of them present at birth or at young ages although some may occur in adults or even in the elderly 3. Male
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predominance is seen in most of the studies, although the number of cases is too few to make a conclusion. For example, in a retrospective study on ten cases, the male/female ratio was 7:3 while in a review of 24 cases, this ratio was 12:10 (two unknown). Our case was diagnosed after 10 years, similar to a case series of ten patients in which the duration between onset and diagnosis is from several months to seventeen years.

In reported cases, the lesions are mainly located on extremities, as in our case. Trunk, forehead and neck involvement have also been reported. The lesions may also be bilateral, generalized, or occur along Blaschko lines. It is usually asymptomatic, although it may be accompanied by a mild pruritus, hyperhidrosis or anhydrosis. Association with other conditions is rare and includes neurological problems (sensory polyneuropathy, seizure, developmental delay, deafness, and hemiparesis), scoliosis, palmoplantar keratoderma, onychodysplasia, alopecia, and hyperthyroidism. There is also a report of association with Bowen disease.

Etiologically, it has been proposed that the invagination of the epidermis may result from an abnormal clone of epidermal cells which leads to the formation of cornoid lamella. Another hypothesis suggests that the invagination is a dilated acrosyringeal and dermal duct which is keratin-plugged.

Histopathology is the mainstay of diagnosis. As we mentioned earlier, cornoid lamella with the involvement of acrosyringia is pathognomonic for PEODDN. It is usually associated with the dilation of eccrine duct. Differential diagnoses include porokeratosis plantaris discreta, inflammatory linear verrucous epidermal nevus, nevus comedonicus, linear epidermal nevus, linear psoriasis, spiny keratoderma, linear porokeratosis, congenital unilateral punctate porokeratosis and porokeratosis of Mibelli.

Although the condition is benign, most treatment modalities fail to show beneficial results. Steroids,
5-FU, retinoids (both topical and oral), cryosurgery, photo and laser therapy and keratolytics all have been used but have shown limited efficacy. In conclusion, we presented a case of PEODDN and briefly reviewed the literature on the topic. Because of the rarity of this condition, each diagnosed case of PEODDN should be reported to enhance our knowledge regarding this condition.

REFERENCES