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Porokeratotic Eccrine Ostial and Dermal Duct Nevus: Report of a Rare Case

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ABSTRACT

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare hamartoma of eccrine duct. It usually appears at birth or in early childhood and unilateral in distribution. Histologically, well-formed cornoid lamellae in close association with a dilated eccrine duct is the diagnostic feature. Here we are presenting one such case of PEODDN with bilateral distribution in multiple sites.

Key words: Porokeratotic eccrine ostial and dermal duct nevus (PEODDN), Comedo nevus, Porokeratotic adnexal ostial naevus (PAON).

INTRODUCTION

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare benign disorder of keratinisation involving the intra epidermal part of the eccrine duct (acrosyringium). It is characterized by linearly arranged grouped keratotic papules and plaques in palm and soles mainly, but may occur at other areas of the extremities and even on multiple locations. In case of extra palmer locations, PEODDNs usually present like a verrucous lesion [1]. Here we present a case of bilateral PEODDN with lesions in multiple locations.

CASE REPORT

We report an otherwise healthy, 16 year-old young girl developing numbers of asymptomatic skin lesions over 10 years. She had applied various topical medications without any benefit. There was no family history of similar condition.

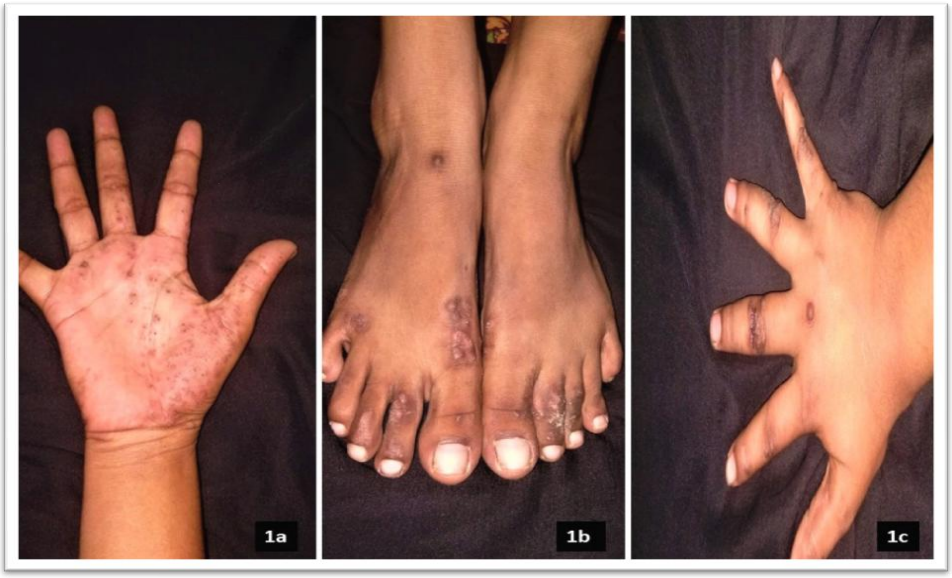

Figure 1 (a) Pits with keratinous plug in the right palmer surface (b) Verrucous fused plaques over dorsal aspect of both foot, few containing pores (c) Similar skin lesion over dorsum of right hand

Clinical examination revealed grouped linear brown keratotic papules on her right and left foot and right hand. There were associated pits with keratinous plugs in the right palmer surface. The lesions in dorsal aspect of left, right foot and right hand were mainly verrucous fused plaques due to coalescence of papules. Some of the fused plaques contain central pits within them (Fig 1a-c). Initial clinical differential diagnosis included linear verrucous epidermal nevus (VEN), inflammatory VEN, porokeratosis, lichen planus, blaschkitis. General physical examination was otherwise normal. A punch biopsy from one such lesion was performed.

Microscopical examination showed orthokeratotic hyperkeratosis, acanthosis and a cup shaped invagination of parakeratotic column (cornoid lamella). The epidermis at the base of column showed loss of granular layer. A dilated eccrine duct was observed at the base of invagination, suggesting acrosyringeal origin (Fig 2a, b). Based on the clinical and histopathological findings, diagnosis of PEODDN was made.

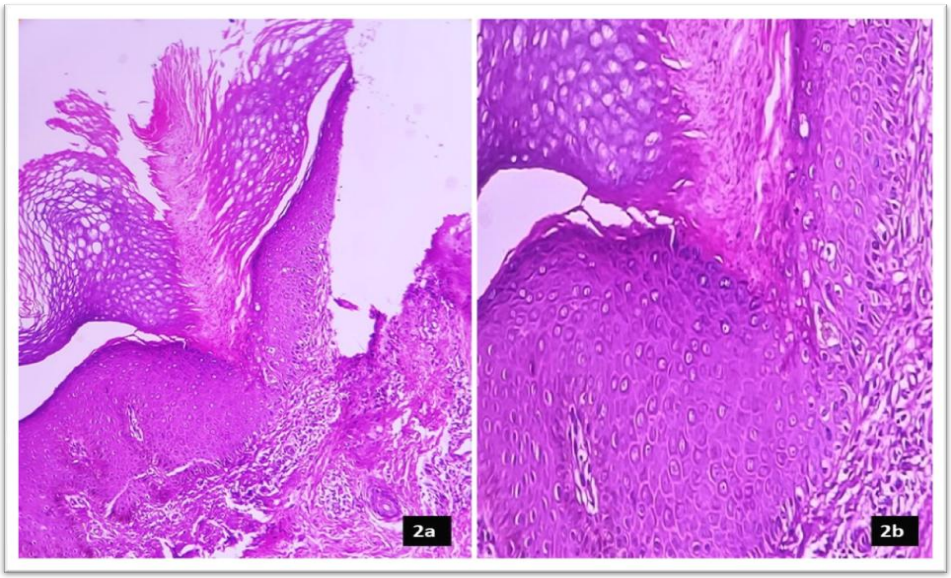

Figure 2 (a) Low magnification showing orthokeratotic hyperkeratosis and cornoid lamella. The granular layer beneath the cornoid lamella is absent (100x, H& E) (b) High magnification view showing one dilated eccrine duct beneath the invagination (400x, H& E)

DISCUSSION

Porokeratotic eccrine ostial and dermal duct nevus is a very rare skin condition characterized by porokeratotic dermatosis. PEODDN was first described by Marsden et al. in 1979 [2] as "comedo nevus of the palm" but the term was coined by Abell & Read in 1980[3]. It is composed of small keratotic papules or comedo like pits filled with keratotic plugs in linear distribution. PEODDN usually has unilateral distribution on the distal portion of a limb such as on the palm or sole. A wider distribution of the disease can be found rarely along Blaschko lines[1].

The cause of the disease is still unclear. Out of the two hypothesis, Stoof et al. postulated that the epidermal invagination is widely dilated keratin plugged acrosyringial duct which is continuous with the dermal duct at base[4] .Bergman et al. proposed that the invagination represents an abnormal clonal proliferation of epidermal cells that produce the cornoid lamella like column[5] .Recently somatic mutation in GJB2 encoding a gap junction protein connexin 26 has been documented in PEODDN and it may be a mosaic form of keratitis ichthyosis deafness (KID) syndrome[6].

Differential diagnoses include linear verrucous epidermal nevus (VEN), inflammatory linear VEN, linear porokeratosis, nevus comedonicus,

punctuate keratoderma. All the above mentioned diseases lack characteristic histological features of cornoid lamella occurring in association with dilated eccrine duct and acrosyringia. PEODDN like features involving the hair follicle of non acral areas are known as porokeratotic eccrine and hair follicle nevus (PEHFN). Recently Goddard et al. [7] suggested that PEODDN and PEHFN may be two different spectrum of same disease and coined a common term "porokeratotic adnexal ostial nevus" (PAON).

PEODDN is a benign condition and the clinical course is usually stationary. Some cases with progressive disease also noted[8]. Most of the cases are asymptomatic, however severely pruritic lesions are documented in past. The lesions are commonly congenital or present in childhood, although cases of adult onset PEODDN have been described[9]. Association with other condition is rare and includes neurological disorders, scoliosis, palmoplantar keratoderma, breast hypoplasia, onychodysplasia, alopecia and hyperthyroidism. Recently a diffuse squamous cell carcinoma and Bowen disease have been described to arise from PEODDN, therefore periodical prolonged follow up is mandatory in every case[10,11].

Our case presented with asymptomatic skin lesion since childhood and it was bilateral involving multiple sites of acral region. Histological features showed characteristic cornoid lamella involving dilated eccrine duct, thus ruling out other differentials. There was no associated anomaly of the dermoepidermal interface.

Various treatment modalities are tried in PEODDN but the results are not satisfactory[12]. There is little success with topical corticosteroids, anthralin, keratinolytics, psoralen, UVA, UVB and retinoids. Nowadays, laser therapy in the form of ultrapulse CO2 laser and combined erbium/CO2 laser have produced good results with little scarring[13].

CONCLUSION

PEODDN is very rare in India and only 5 cases have been reported so far. [12,14,15,16,17] Our case is unique because of its bilaterality and multiple acral parts involvement. We conclude that, PEODDN must be considered in differential diagnosis of linear keratotic papules.

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