POROKERATOTIC ECCRINE OSTIAL AND DERMAL DUCTAL NEVUS (PEODDN)- A RARE ENTITY IN 2 CASES

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ABSTRACT

BACKGROUND

Nevi are visible, circumscribed, long-lasting lesions of the skin reflecting genetic mosaicism.[1] Porokeratotic Eccrine Ostial and Dermal Ductal Nevus (PEODDN) is believed to be a congenital hamartoma of possible eccrine origin with no malignant potential.[2] It is a rare variant of porokeratosis having characteristic feature of cornoid lamella on histopathology. It is a disorder of keratinisation involving the intraepidermal eccrine duct (acrosyringium) and is characterised by eccrine hamartoma and cornoid lamella in pathology.[3] It is usually localised at the single extremity, while wider systematised distribution has been documented rarely. Till now, a few cases have been reported. Here, we are reporting 2 cases of this rare entity with one having childhood origin and other having adult onset.

KEYWORDS

PEODDN, Porokeratosis, Coronoid, Lamella, Keratinisation Disorder, Eccrine Hamartoma.

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BACKGROUND

Case 1

A 17-year-old boy who was otherwise healthy presented in our department with multiple asymptomatic keratotic papules and plaques over dorsum of right hand extending from middle finger to wrist area since 10 years. It initially started from dorsum of hand and then gradually spread in a linear fashion. Past and family history were not relevant.

On cutaneous examination: Numerous punctate pits having comedo-like plugs were seen admixed with the papules (Figure 1). Palms and soles were spared. No extracutaneous involvement was seen.

Skin biopsy showed orthokeratosis and parakeratotic column filling invaginations in the epidermis. Epidermis at the base of column showed loss of granular cells. Eccrine ducts seen at the base of invagination were consistent with acrosyringeal origin (Figure 2).

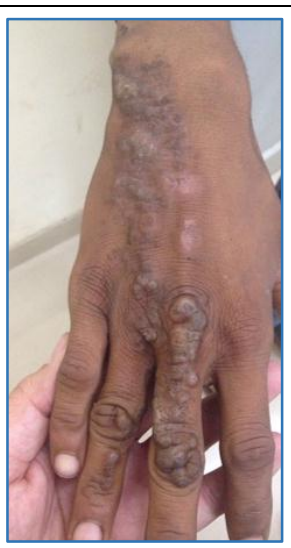

Figure 1

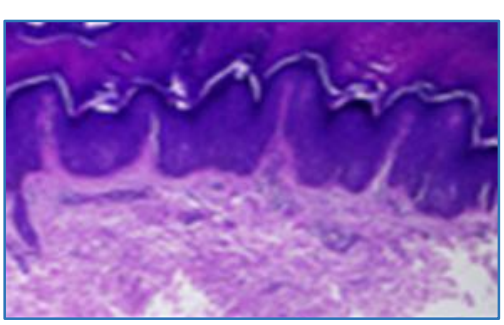

Figure 2

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Case 2

A 20-year-old female presented with multiple small asymptomatic black/dark brown papules over hyperpigmented surface of upper abdomen since 3 months with pits in the centre. Past and family history were not relevant. A few lesions had coalesced to form plaques

(Figure 3). Histopathology showed orthokeratosis filling a deep cup shaped invagination in the epidermis. Epidermis at the base of column showed loss of granular cells replaced by cluster of dyskeratotic cells. Dermis show fibrosis, pigmentary incontinence and mild interstitial and perivascular lymphocytic infiltrate (Figure 4).

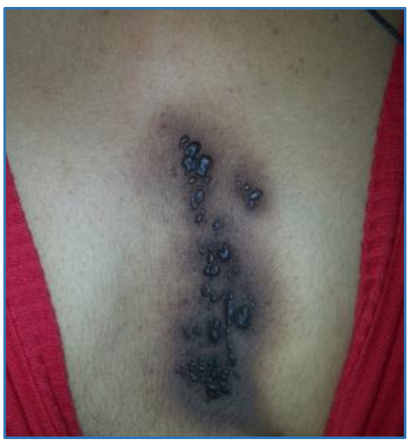

Figure 3

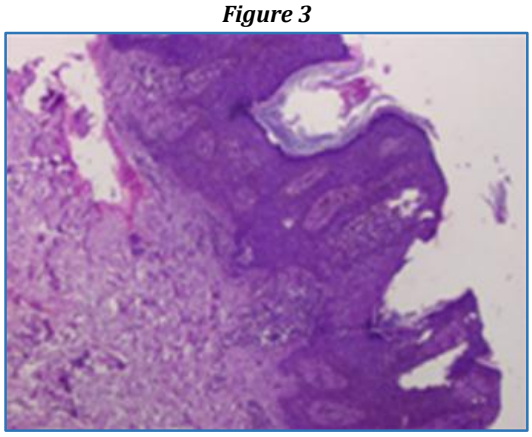

Figure 4

DISCUSSION

The term "PEODDN" was first given by Abell and Read in 1980.[4] However, it was described as comedo nevus of the palm by Marsden et al in 1979 and on scanning electron microscopy keratotic plugs occluding the eccrine ostia were present.[5] Other synonymous terms used for PEODDN are comedo nevus of the palm, linear eccrine nevus with comedones, palmar and plantar pseudocomedo porokeratotic sweat nevus and porokeratotic eccrine ostial and hair follicle nevus.[6] It usually appears at birth or early childhood.[7] Although, PEODDN is considered to be congenital, but a review of the literature showed us that the frequency of a late-onset variant may be as high as 26%.[8] It has a linear distribution following the Blaschko lines. Most common sites of presentation are palms and soles, but can involve other areas like trunk, neck and forehead. Duration between onset and diagnosis may vary from several months to years. Out of the 23 cases of PEODDN described in the review by Sassmannshausen et al, the ratio of M:F was 12:10 suggesting the gender ratio is almost equal.[6] It is usually asymptomatic, but may be associated with pruritus, hyperhidrosis or anhydrosis. Pathogenesis of PEODDN is still not clear. Hypothesis suggests that it might be caused by mutations in the GJB2 gene coding for the gap junction protein connexin 26 (Cx26) associated with KID or similar gap junction disorders.[9] Bergman et al demonstrated that it stained positively for carcinoembryonic antigen (CEA) in its lower course, but negatively in the inner borders of the invagination from which the parakeratotic column arose which suggests that the epithelial structure in PEODDN is an abnormally keratinising epidermal invagination through which an acrosyringium-like duct traverses.[10]

There are rare case reports of PEODDN associated with conditions like hyperthyroidism, sensory polyneuropathy, breast hypoplasia, Bowen's disease, deafness, developmental delay, seizure disorder, hemiparesis, scoliosis, alopecia, onychodysplasia and squamous cell carcinoma.[11] Co-occurrence with linear psoriasis has also been described.[12]

The mainstay of diagnosis is histopathology and its pathognomonic feature is multiple cornoid lamella-like parakeratotic columns arising over eccrine sweat ducts with dilated acrosyringium. Our diagnosis was made on the basis of clinical and histopathological correlation. Differential diagnoses include porokeratosis plantaris discreta, inflammatory linear verrucous epidermal nevus, nevus comedonicus, linear epidermal nevus, linear psoriasis, spiny keratoderma and linear porokeratosis. Nevus comedonicus was excluded by the absence of grouped undeveloped hair follicles filled with cornified debris devoid of hair shafts.[13]

Usually, the treatment is not effective. Various treatment options that have been tried in the past are topical agents like keratolytics, retinoids, steroids, calcipotriol, phototherapy and cryotherapy. Ultrapulse CO2 laser is comparatively effective and has better cosmetic results.[11]

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