Case Report

Porokeratotic eccrine ostial and dermal duct nevus: A rare case report

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Abstract

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is an uncommon benign disorder that clinically mimicks comedo nevus but usually favors the palms and soles, where pilosebaceous follicles are absent. It also can present with widespread involvement along Blaschko's lines. It is a disorder of keratinisation involving the intraepidermal eccrine duct (acrosyringium) with classical histopathological examination findings (eccrine hamartoma and cornoid lamellation). The patient is a 32-year-old woman with a 12-year history of pruritic skin lesions on her right palm and index finger. Histopathological examination revealed multiple small epidermal invaginations with overlying parakeratotic cornoid lamellation and loss of granular layer. Few dyskeratotic cells are seen at the base of epidermal invagination. After clinic-pathologic correlation, the diagnosis of PEODDN was made. Late-onset and rare clinical presentation as pruritic lesion are the characteristic features that make this patient an extraordinary presentation of PEODDN.

Keywords: Coronoid lamellae, eccrine hamartoma, keratinisation disorder, porokeratotic eccrine ostial and dermal duct nevus

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Submitted: 14-Sep-2020 **Revised:** 06-Apr-2021 **Accepted:** 02-Jun-2021 **Published:** 25-Oct-2021

INTRODUCTION

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) uncommon variant of porokeratosis, is a disorder of keratinisation which clinically presents as asymptomatic localised area of plugged pits some times in a linear distribution on palms and soles or a generalised form mimicking linear verrucous epidermal nevus in blashkoid distribution, having histological features of thin column of parakeratotic cells (coronoid lamellae) with complete absence or decreased underlying granular layer and few dyskeratotic cells that involve the acrosyringia characteristically.[1]

CASE REPORT

A 32-year-old female presented with pruritic keratotic papules and plaques on the right palm and index finger of right hand which had been since 12 years. No history of a similar condition in the family members. Clinical

Access this article online	
Quick Response Code:	Website: www.jcsr.co.in
	DOI: 10.4103/JCSR.JCSR_83_20

How to cite this article: Kumar SS, Kumar KR, Chenchaih V, Usha G, Ramana PV. Porokeratotic eccrine ostial and dermal duct nevus: A rare case report. J Clin Sci Res 2021;10:246-8.

examination showed keratotic papules (some discrete whilst others coalescing to form plaques in a linear configuration), distributed over right palm, extending from thenar eminence to first web space (Figure 1) and the outer aspect of index finger (Figure 2). Few pits had comedo-like plugs were seen in between the papules. Our differential diagnosis based on clinical picture, was blaschkoid porokeratosis, linear verrucous epidermal nevus (VEN), punctate palmoplantar keratoderma and darriers disease. General physical examination showed no abnormality. The patient was advised punch biopsy. The histopathological features were consistent with PEODDN (Figure 3), showing orthokeratosis and parakeratotic column (Coronoid lamellae). Epidermis at the base of parakeratotic column showed loss of granular cells with few clusters of dyskeratotic cells. A dilated eccrine duct was seen at the base of invagination). The patient was advised to undergo ablative laser surgery.

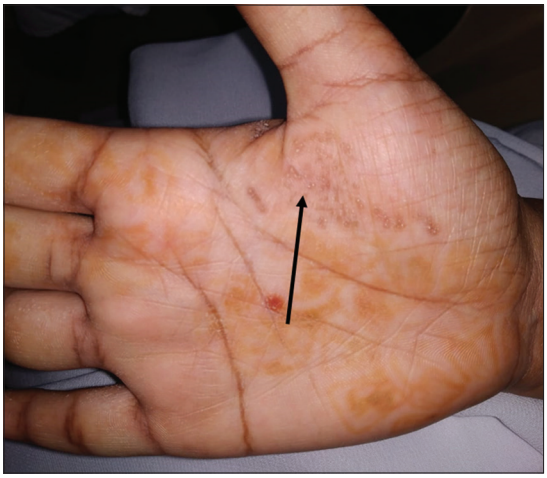

Figure 1: Clinical photograph showing keratotic papules over thenar eminence (arrow)

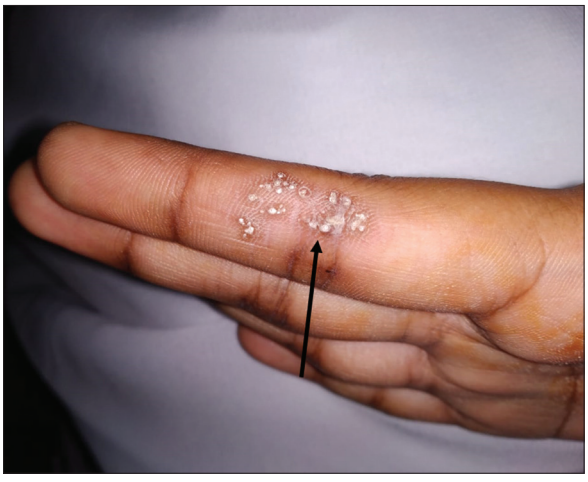

Figure 2: Clinical photograph showing keratotic papules over index finger (arrow)

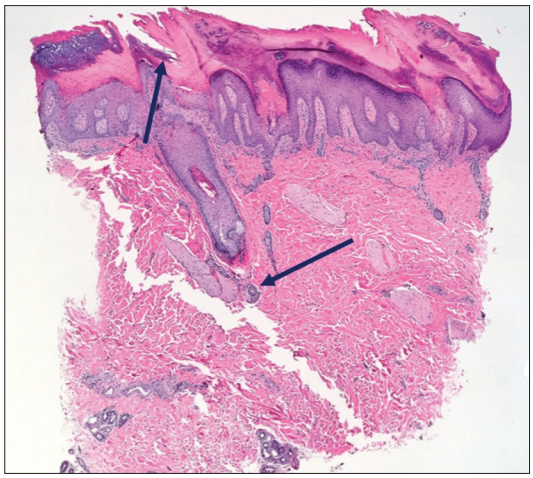

Figure 3: Histopathological examination showing orthokeratosis, coronoid lamella, dilated eccrine duct at the base of invagination (arrows) (Haematoxylin and eosin, X100)

DISCUSSION

PEODDN is a rare dermatological entity, whose pathogenesis is yet to be described. It was proposed that the epidermal invagination originates from a widely dilated acrosyringium which is continuous with dermal duct at its base while others believe that the invagination is the end result of the proliferation of abnormal clone of epidermal cells producing the cornoid lamella.[1] It was suggested that PEODDN originates from a circumscribed keratinisation abnormality based on carcinoembryonic antigen staining positivity along the ductal lumina through the parakeratotic column of cornoid lamella.[1] Recent findings indicate that PEODDN may be a mosaic form of keratitis ichthyosis deafness syndrome and is caused by somatic mutation in GJB2 encoding a gap junction protein connexin-26.[2] The condition is usually asymptomatic but is occasionally pruritic. The onset may range from birth till adulthood. PEODDN usually presents as grouped keratotic pits and is usually noted on the palms and soles as in our case. When sites other than palms and soles are involved, it presents as keratotic papules and plaques resembling linear VEN.[1] Depending on the morphology, extent and patterns of involvement, the clinical differentials of PEODDN may include nevus comedonicus, linear VEN, inflammatory linear VEN (ILVEN), linear psoriasis, linear porokeratosis, dilated pore nevus, linear lichen planus, punctate palmoplantar keratoderma and punctate porokeratosis. The condition can be differentiated from ILVEN by the absence of extremely pruritic bands of eczematous or psoriasiform lesions and the presence of pitted papules and punctate pits on palms and/or soles. Histological features, however, can discriminate the clinical differentials.[2] Characteristic histopathological features

of PEODDN include orthokeratosis, and a column of parakeratosis occupying an invagination of the epidermis, which, at the base of the column shows loss of granular cells. Dilated acrosyringia at the base of the invaginations point to an eccrine origin.

There are rare reports of associations with Bowen disease, deafness and development delay, seizure disorder, hemiparesis, scoliosis, hyperthyroidism and sensory polyneuropathy, breast hypoplasia, alopecia, onychodysplasia and squamous cell carcinoma.[1] Treatment options are limited. Some lesions may undergo spontaneous flattening with time. Small and localised lesions may be suitable candidates for surgery. Laser therapy, preferably ultra-pulse CO_2 laser, is an excellent modality because the chances of pigmentary changes and scarring are minimal.[3] Combined erbium/CO_2 laser therapy has been shown to offer significant cosmetic improvement in PEODDN patients. Modalities such as topical steroids, retinoids, keratolytics, phototherapy, electrocautery and cryotherapy have not shown any promising results.[4]

We would like to emphasise that late-onset and pruritic lesions with blaschkoid distribution could be one of the clinical presentations of PEODDN.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Easton JA, Donnelly S, Kamps MA, Steijlen PM, Martin PE, Tadini G, *et al*. Porokeratotic eccrine nevus may be caused by somatic connexin26 mutations. J Invest Dermatol 2012;132:2184-91.
2. Bandyopadhyay D, Saha A, Das D, Das A. Porokeratotic eccrine ostial and dermal duct nevus. Indian Dermatol Online J 2015;6:117-9.
3. Mondal A, Kumar P, Das A, Debbarman P, Mandal RK. Porokeratotic eccrine ostial and dermal duct nevus: A noteworthy presentation. Indian Dermatol Online J 2015;6:130-1.
4. Goddard DS, Rogers M, Frieden IJ, Krol AL, White CR Jr., Jayaraman AG, *et al*. Widespread porokeratotic adnexal ostial nevus: Clinical features and proposal of a new name unifying porokeratotic eccrine ostial and dermal duct nevus and porokeratotic eccrine and hair follicle nevus. J Am Acad Dermatol 2009;61:1060.e1-14.