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Case Report

Porokeratotic eccrine and ostial dermal duct nevus

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ABSTRACT

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare benign eccrine nevoid hamartoma with only a few cases reported till date. It is characterized by asymptomatic or mildly pruritic pitted comedone-like papules on palms or soles and keratotic papules and plaques in a linear pattern on the extremities, usually present in childhood. We present here three interesting cases of the same. Diagnosis was confirmed by histopathology which showed parakeratotic columns overlying mature eccrine sweat glands in dermis and dyskeratotic cells of lower epidermis.

Keywords: Porokeratotic eccrine ostial, Dermal duct nevus, Cornoid lamella

INTRODUCTION

Porokeratotic eccrine and ostial dermal duct nevus (PEODDN) is a rare nevoid cutaneous disorder, characterised by multiple, asymptomatic or mild pruritic pits or comedone-like papules on palms and soles and keratotic, whitish to brown papules in linear distribution on extremities, which may become warty over time and persists despite treatment.[1] Most commonly it is localised to palm and sole but can rarely involve trunk along the Blaschko's lines.[2] Lesion can be present at birth, appear early childhood or can have late onset.[1,2] Histopathology is the main stay of diagnosis of this rare disorder of keratinisation. Histopathology reveals cornoid lamellae overlying dilated acrosyringia/hair follicle ostia.[2]

CASE REPORTS

Case 1

A 23-year-old male patient presented with multiple, asymptomatic, hyperkeratotic scaly papulo-follicular lesions over the extensor aspect of left arm, elbow, and forearm extending up to left ring finger in a linear fashion with comedone-like lesions on left palm. Lesions were present since early childhood and were gradually increasing in extent (Figure 1A).

Case 2

A 6-year-old male patient presented with hyperkeratotic comedone-like scaly papules in a linear fashion on left palm since birth which were gradually progressive over dorsum of hand and forearm, associated with mild pruritus (Figure 1B).

Case 3

A 12-year-old male patient presented with plantar dyskeratosis on the right sole. On close observation asymptomatic, non-progressive hyperkeratotic scaly papules and tiny/comedone-like pits were observed on the right great toe and sole in linear fashion associated with diffuse dyskeratosis on sole (Figure 1C) KOH scrapping was negative for fungal hyphae.

A 4 mm punch biopsy was taken in each patient with differentials of nevus comedonicus, inflammatory linear verrucous epidermal nevus, linear psoriasis, verrucous epidermal nevus.

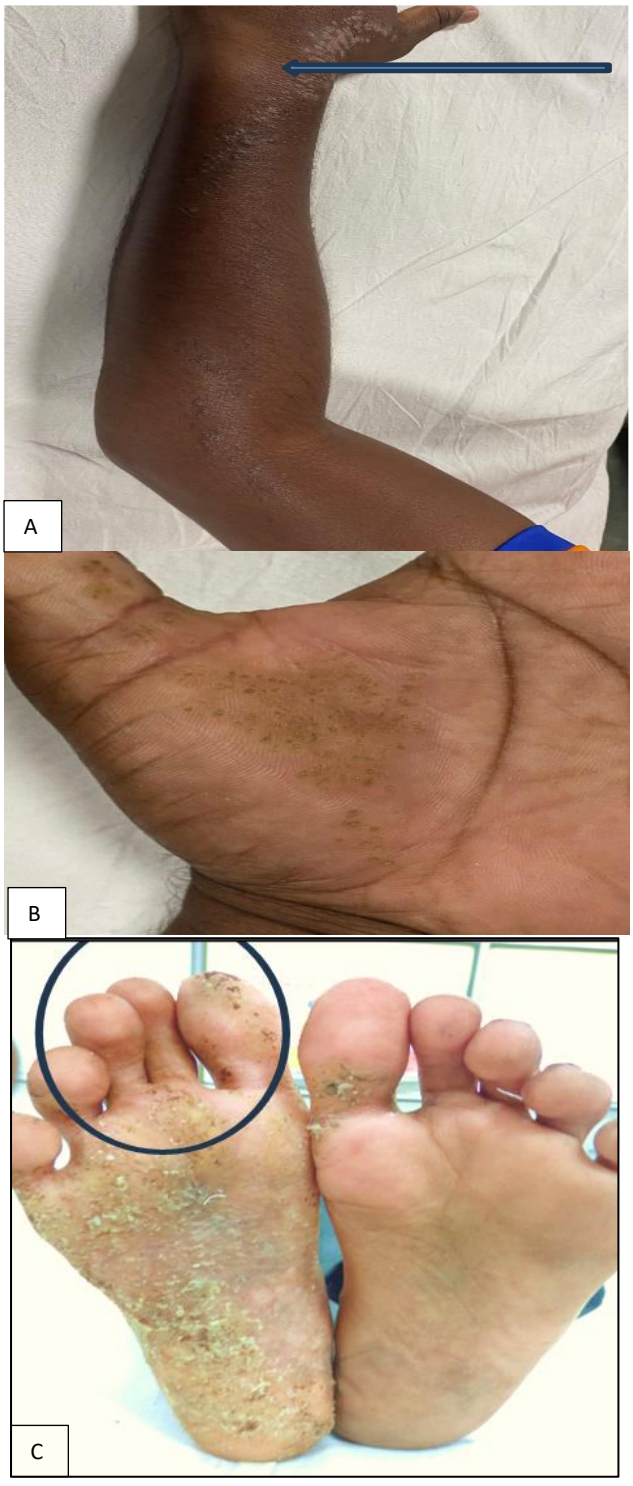

Figure 1: (A) Case 1 is a 23-year-old male patient presented with multiple, papulo-follicular lesions over extensor aspect of left forearm; (B) case 2 is a 6-year-old male patient presented with comedone-like pits on left palm and (C) case 3 is a 12-year-old male patient presented with plantar dyskeratosis and tiny comedone-like pits on right sole.

Histopathological examination in all three cases showed well-formed cornoid lamellae which is a parakeratotic column overlying a small vertical zone of dyskeratotic and vacuolated epidermis, hypogranulosis below cornoid lamella. Dermis showed mature eccrine sweat glands in mid and lower dermis in two patients and dilated hair follicle ostia in one patient just underlying the parakeratotic columns (Figure 2A and B).

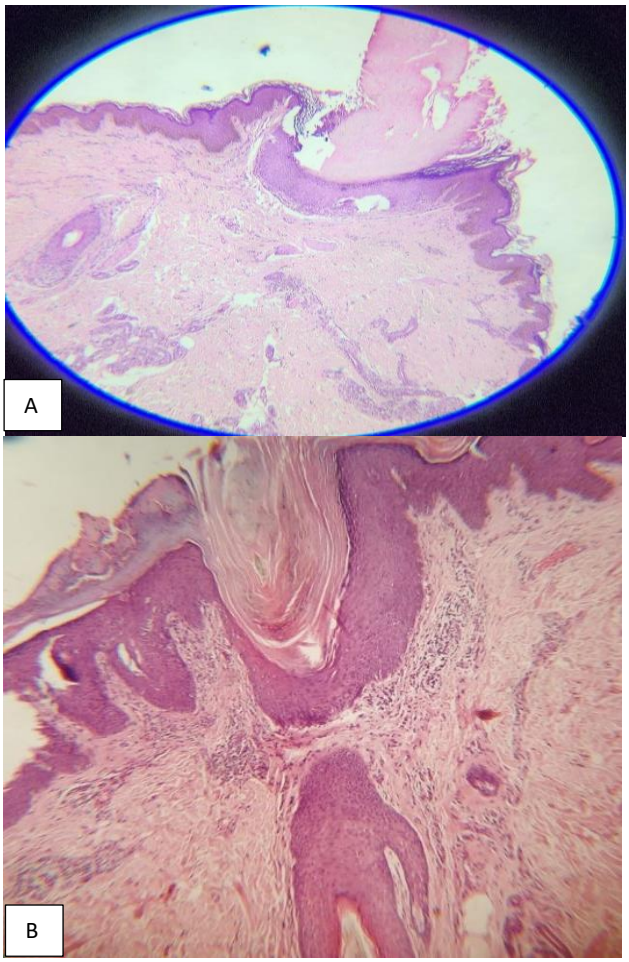

Figure 2: (A) Mature eccrine glands in dermis with overlying parakeratotic coloumn as seen in histopathology of lesion on palm (H and E stain, 10× magnification) and (B) parakeratotic coloumn overlying dilated follicular ostia from lesion on forearm (H and E stain, 10× magnification.

On the basis of clinical findings and classical histopathological findings all our patients were confirmed to have PEODDN.

None of the patient had any family history, systemic association, comorbid condition or mucocutaneous involvement on detailed assessment.

DISCUSSION

PEODDN is a rare benign eccrine nevoid hamartoma with only a few cases reported till date in the literature. Indian literature reports only about 10 cases. The condition was first described by Marsden et al in 1979 as Comedo nevus of palm, and showed keratotic plugs

occluding eccrine ostia by scanning electron microscopy and later it was redefined by Abell and Reed. In 1980 Goddard et al suggested a new name as porokeratotic ecrrine and hair follicle nevus as involvement of both acrosyringia and acrotrichia may be seen.[3]

Pathogenesis of PEODDN is not known exactly but recently somatic GJB2 gene mutation coding for connexin 26 protein has been found.[3]

Rarely a systematized PEODDN is reported associated with various CNS anomalies, endocrine anomalies, scoliosis, breast hypoplasia, alopecia etc. Rarely long-standing lesions are reported to be converted in Bowen's disease or squamous cell carcinoma.[2-4]

All our patients were male and two were in pre-pubertal age though all 3-patient had early age of onset of lesions. In a study by Alorman et al five out of eight cases were male.[4] Case report by us in 2015 was also a male and had congenital lesions but due to asymptomatic nature presented in adulthood.[5]

None of our patients had any family history of similar condition or any systemic association. The case series by Alomran et al have shown nail changes in one patient and systemic endocrine disorders in two of their eight patients.[4]

PEODDN may be treated by emollients, keratolytic and topical retinoids, the study by Alomran et al has shown the efficacy of topical Tazoretene gel.[4]

CONCLUSION

PEODDN is a rare nevoid condition, we have seen three such histopathology proven cases of the same in our department over a period of five years. Many such cases might be missed as these cases are asymptomatic and due to lack of histopathology at peripheral centres.

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