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Case Report

Porokeratotic eccrine ostial and dermal duct nevus: A case report with brief review of literature

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Abstract Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare benign cutaneous disorder. It has clinical resemblance with comedo nevus, usually presents in palms and soles. The disease is characterized histologically by well-formed cornoid lamellae occurring in close association with or overlying dilated eccrine ducts and acrosyringia. The lesions follow the Blaschko's lines and may be systematized. We describe a 20-year-old female with PEODDN.

Key words
Porokeratotic eccrine ostial and dermal duct nevus, Blaschko's lines, linear.

Introduction

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare benign cutaneous disorder. It has clinical resemblance with comedo nevus, usually present in palm and sole, where pilosebaceous follicles are absent. PEODDN was first reported by Marsden *et al*.[1] in 1979 as a "comedo nevus of the palm". It was later given its name by Abell and Read in 1980,[2] when they described a linear epidermal nevus localized on the inner foot. The disease is characterized histologically by well-formed cornoid lamellae occurring in close association with or overlying dilated eccrine ducts and acrosyringia.[3] The lesions usually present at birth or in childhood, although cases of late-onset adult PEODDN have been described.[3,4]

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Case Report

A 20-year-old female presented with asymptomatic linear pits with comedo like plugs on left sole extending from heel along the medial border of foot upto tip of left great toe along the Blaschko's lines **(Figure 1)**. Lesions were persistent from childhood, since 3 years of age and growing as the patient grows up. Her family history was unremarkable. Mucocutaneous examination shows normal oral mucosa, hair and nail. Her routine laboratory tests, serum chemistry panel and HIV serology were within normal limits.

Histopathology showed epidermal acanthosis, deep epidermal invagination of a parakeratotic column (cornoid lamella), which is distinctive feature of PEODDN with absent granular layer . There was an eccrine unit in the dermis, in close approximation to the epidermal invagination **(Figure 2)**.

The patient presented for follow-up after being placed on tretinoin 0.1% cream, 10% salicylic acid ointment and clobetasol propionate. After 2

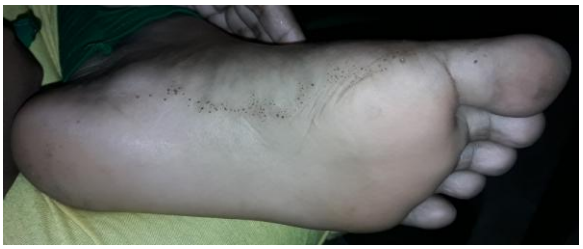

Figure 1 Linear pits with comedo like plugs on left sole.

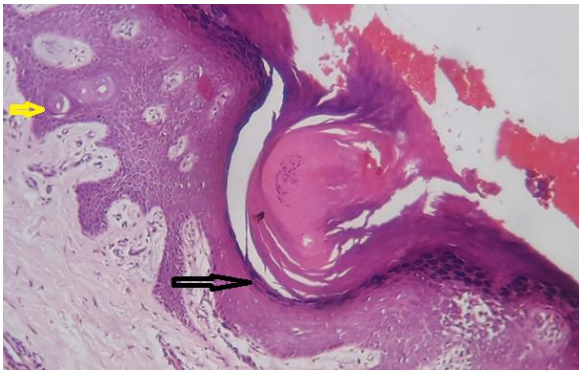

Figure 2 Parakeratotic cornoid lamellation above deep epidermal invagination with loss of granular layer. Underlying, slightly tortuous, eccrine duct nearby the epidermis (H&E X 100).

months of treatment with these topical therapies, there was no observable improvement. The patient was then treated with radiofrequency cautery with notable improvement after two treatment sessions.

Discussion

Porokeratotic eccrine ostial and dermal duct nevus is thought to be an eccrine hamartoma that presents at birth and persists into adulthood. But adult-onset POEDDN case have been described, and its frequency may be as high as 26%.[5] Lesions of POEDDN are usually asymptomatic but mild pruritus may present. It predominantly occurs on the palms and soles as linear punctate pits or pitted papules. The condition is very rare and has a relatively equal occurrence in both genders. PEODDN clinically presents as either multiple linear punctuate pits with comedo-like plugs on palms and soles or keratotic plaques and papules that resemble linear VEN on other areas.[6,7] But it may consist of multiple verrucous, keratotic, brown to flesh-colored papules often coalescing into linear plaques. The lesions follow the Blaschko's lines and may be systematized.[8] The etiology of PEODDN is currently unknown, however, studies have alluded to a genetic contribution through genetic mosaicism and a possible eccrine or circumscribed epidermal keratinization abnormality.[9,10] PEODDN is also associated with many conditions including scoliosis, anhidrosis, seizure disorder, sensory polyneuropathy, deafness, left hemiparesis, developmental delay, breast hypoplasia, alopecia, onychodysplasia, Bowen's disease, and squamous cell carcinoma.[11,12]

Our patient was clinically healthy, without any of the aforementioned associated disorders. The differential diagnoses include nevus comedonicus, punctate keratoderma, linear verrucous epidermal nevus, inflammatory linear verrucous epidermal nevus, and linear porokeratosis.[12] PEODDN is usually differentiated from these entities by its characteristic presentation and histological presence of porokeratosis-like features in close association with the eccrine unit. POEDDN is a benign condition, usually stationary or sometimes progressive but no malignant transformation has been reported. Topical management with topical corticosteroids, tar, psoralen ultraviolet A, ultraviolet B, anthralin, keratolytics, and retinoids may try but unsatisfactory. Surgical management with laser therapy, in the form of ultrapulse CO_2 laser and combined erbium/CO_2 laser have been reported.[13] Lesions in our patient was deeply cauterized with radiofrequency cautery and the lesions subsided significantly.

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