Bilateral Systematized Porokeratotic Eccrine Ostial and Dermal Duct Nevus

Sir,

There are about 82 reported cases of porokeratotic eccrine ostial and dermal duct nevus (PEODDN) reported in the literature, however, most of them are acral and few are localized to a single limb. Cases with extensive distribution either unilateral or bilateral are reported rarely in the literature. We could find less than 20 cases of bilateral systematized PEODDN with none of them reported from India.

A 16-year-old boy presented with chief complaints of multiple asymptomatic skin-colored raised lesions all over the body since 3 months of age. Lesions started from thighs and gradually involved trunk, face, upper, and lower limbs in a symmetrical pattern. There was no history of decreased hearing, eye redness or discharge, photophobia, seizures, heat intolerance, hyperhidrosis, or anhidrosis. There was no family history of similar disease. On dermatological examination, multiple skin-colored papules coalescing to form plaques with mild scaling were present symmetrically on the bilateral aspect of the face, upper chest, upper back, abdomen, arms, flexor aspect of forearms, flexor aspect of thighs and legs and extensor aspect of legs in a linear pattern along the lines of Blaschko. [Figures 1-3] Multiple papules were present on the scrotum and few on penile shaft. Well-defined linear plaque with pits was present on bilateral palms. Soles were also involved. Hair, nails, mucosae, and teeth were within normal limits. The ophthalmological and auditory examination was unremarkable. Differential diagnosis of PEODDN, verrucous epidermal nevus, inflammatory linear verrucous epidermal nevus (ILVEN), linear psoriasis, linear lichen planus, and linear Darier's disease were kept. On histopathological examination of skin biopsy, epidermis revealed hyperkeratosis and discrete parakeratotic columns overlying a small vertical zone of dyskeratosis. There was the focal loss of the granular cell layer at the site from which the parakeratotic column rises [Figures 4a, b and 5]. A mild lymphocytic infiltrate was seen around capillaries in the dermis. On clinico-histopathological correlation the diagnosis of PEODDN was made. The patient was planned for oral and topical retinoids along with urea-based emollient.

PEODDN is a rare nonhereditary eccrine hamartoma.[1] The condition was first reported by Marsden *et al*. in 1979 as "comedo nevus of the palm." Abell and Reed redefined this rare entity in 1980 and named it "linear eccrine nevus with comedones".[2] Goddard *et al*. proposed "porokeratotic adnexal ostial nevus" a new unifying name for PEODDN and porokeratotic eccrine and hair follicle nevus because some patients have involvement of both acrosyringia and acrotrichia.[3]

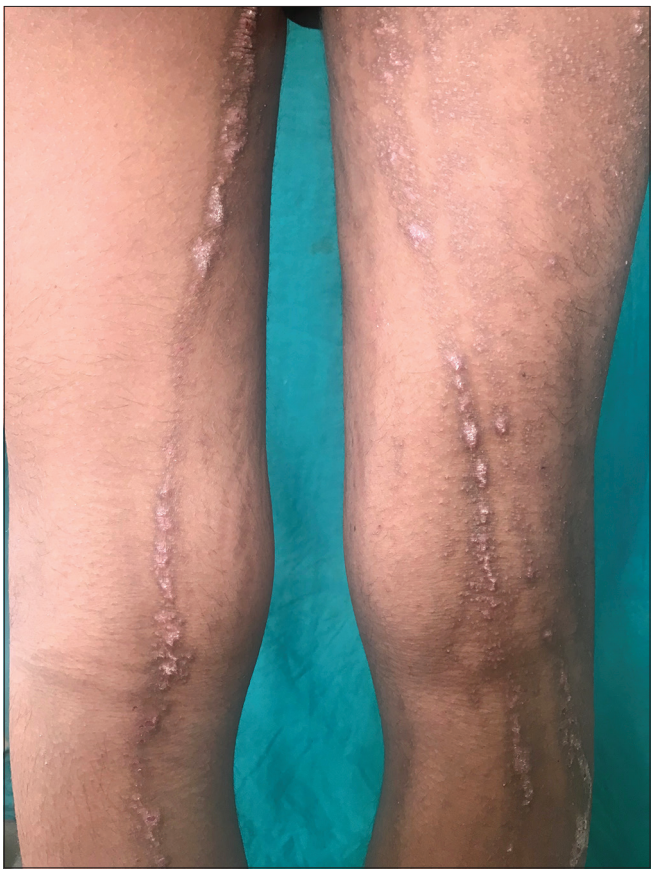

Figure 1: Multiple skin-colored papules coalescing to form plaque present symmetrically on the flexor aspect of bilateral legs in a linear pattern along lines of Blaschko

Various proposed pathogenetic mechanisms of PEODDN include (a) abnormally dilated parakeratotically plugged acrosyringium, (b) lack of carcinoembryonic antigen expression, (c) increased proliferation of basal keratinocytes, (d) genetic mosaicism, and (e) somatic *GJB2* gene mutation coding for connexin 26 protein.[1] A person with PEODDN having a somatic mutation in *GJB2* should be counseled regarding the risk of having a child with syndromes such as keratitis-ichthyosis-deafness syndrome, Vohwinkel syndrome, Bart–Pumphrey syndrome, and non-epidermolytic palmoplantar keratoderma with deafness that are associated with *GJB2* mutations.[4]

Clinically, PEODDN is asymptomatic or mildly pruritic and predominantly occurs on the palms and soles as linear punctate pits or pitted papules. Elsewhere, they consist of multiple verrucous,

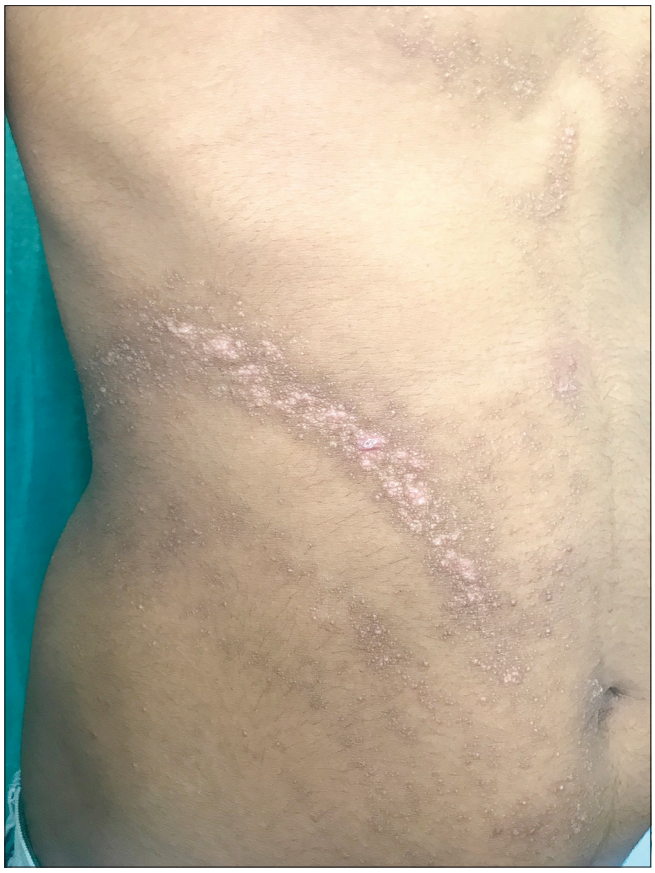

Figure 2: Multiple skin-colored papules coalescing to form plaque along lines of Blaschko on the trunk (close-up view)

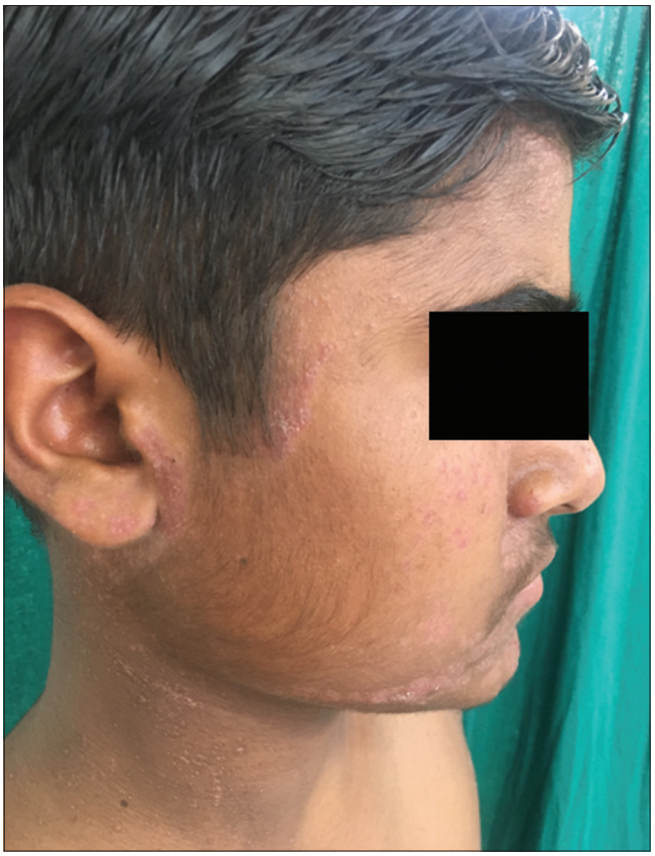

Figure 3: Multiple erythematous plaques with fine scaling on the face and few skin-colored papules over the neck

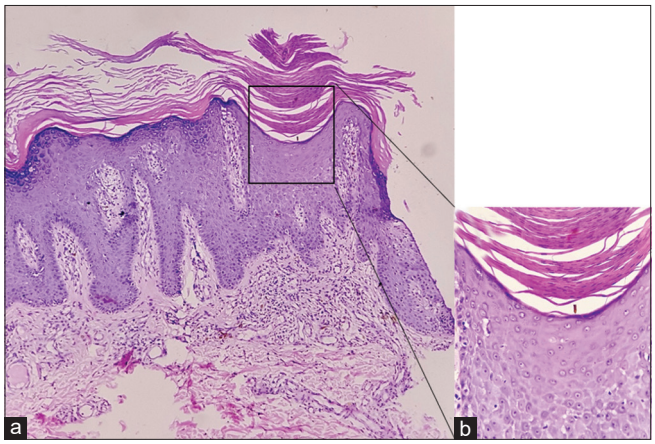

Figure 4: (a) Hyperkeratosis and discrete parakeratotic columns overlying a small vertical zone of dyskeratosis with focal loss of granular cell layer at the site from which parakeratotic column rises (H and E, 10×), (b) Hyperkeratosis and discrete parakeratotic column with focal loss of granular layer at the site (H and E, 40×)

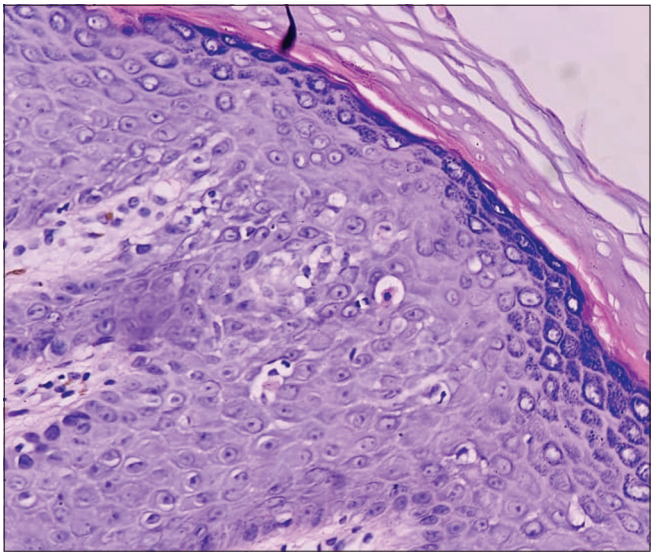

Figure 5: Epidermis showing a small vertical zone of dyskeratotic cells underlying parakeratotic column (H and E, 40×)

keratotic, brown-to-flesh-colored papules often coalescing into linear plaques.[5] The lesions follow the Blaschko lines and maybe systematized as in our case.[6] PEODDN usually occurs at birth or in early childhood but adult onset has been reported in the literature.[1] Rare associations of PEODDN include hyperthyroidism and sensory polyneuropathy, breast hypoplasia, Bowen disease, deafness and development delay, seizure disorder, hemiparesis, scoliosis, alopecia, onychodysplasia, and squamous cell carcinoma.[1]

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.[5] However, sometimes this connection may be barely visible as in our case.[6]

PEODDN was differentiated from ILVEN by the lack of pruritus and the presence of pits on palms and soles. Early-onset of lesions, lack of pruritus, and silvery-white scales with negative Grattage and Auspitz sign ruled out linear psoriasis. Early-onset of lesions, lack of violaceous hue, Wickham's striae, and absence of nail and mucosal findings ruled out linear lichen planus. Lesions in our patient simulate linear VEN and linear Darier's disease, however, the presence of fine scales in few lesions and the absence of nail findings were against the above diagnosis. Histopathology of PEODDN is characteristic and distinct from all the above-mentioned differential diagnoses.

The treatment options are limited. Few cases with spontaneous involution have been described.[7] Small and localized lesions are suitable candidates for surgery. Ultra-pulse CO_2 laser is an acceptable therapy because the chances of scarring and pigmentary changes are minimal. Other treatment modalities include topical steroids, retinoids, keratolytics, phototherapy, electrocautery, and cryotherapy but with limited benefits.[1] PEODDN is largely a benign disorder with no malignant potential, however, few reports of the association of extensive PEODDN with malignancy warrants close monitoring of all diagnosed cases.[8]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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