Linear Pitted Plaque Over the Foot

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A 22-year-old female presented with a 10-year history of a gradually progressive, mildly pruritic, ill-defined linear plaque of size 7-cm over the medial surface of the right foot. The plaque had multiple well-defined pits with elevated keratotic margins of size 1–2 mm over the surface [Figure 1]. A skin biopsy from the margin of the pit showed epidermal invaginations containing parakeratotic plugs with thinned-out to absent granular layer with few dyskeratotic epidermal cells. There were multiple eccrine units in upper dermis underneath this. Moderately dense infiltrate of lymphocytes and histiocytes was focally present at the dermoepidermal junction without any interface change [Figures 2 and 3].

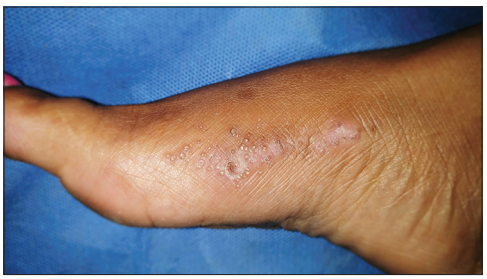

Figure 1: Linearly arranged plaque with multiple well-defined pits having elevated keratotic margins over the right foot

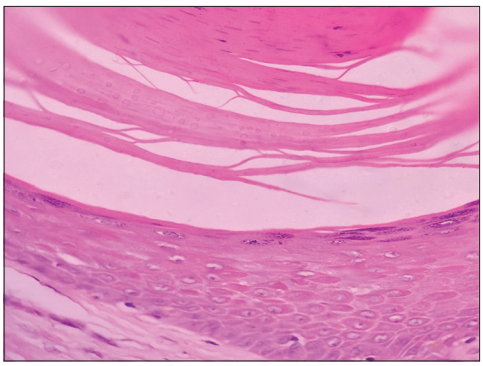

Figure 3: A parakeratotic plug invaginating the underlying epidermis with thinned-out granular layer and few dyskeratotic cells (H and E, ×400)

Question

What is the Diagnosis?

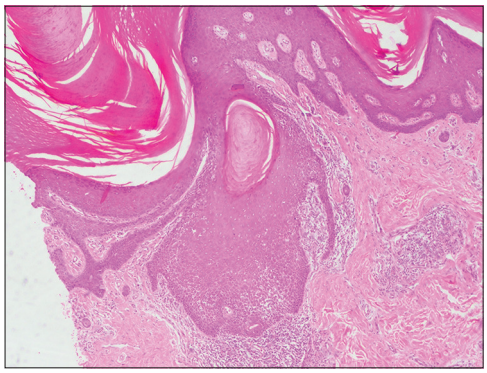

Figure 2: Epidermal invaginations containing parakeratotic plugs with thinned-out to absent granular layer. Multiple eccrine units are seen in upper dermis with focal moderately dense infiltrate of lymphocytes and histiocytes at the dermoepidermal junction (H and E, ×100)

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Answer

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN).

Discussion

PEODDN is believed to be an eccrine hamartoma with possible keratinization defect. The pathogenesis of the disease is still unclear. The proposed mechanism includes genetic mosaicism and circumscribed epidermal or eccrine keratinization abnormality. Recent literature suggests that PEODDN might be a mosaic form of keratitis ichthyosis deafness (KID) syndrome and is caused by somatic mutation in *GJB2*, which encodes a gap junction protein connexin-26.[1]

It usually appears at birth or during childhood as linearly arranged multiple punctate or keratotic papules and plaques.[2] There is a relatively equal occurrence in both genders.[2] The lesions are usually asymptomatic but occasionally it may be accompanied by moderate pruritus and anhidrosis.[3-5] The commonly affected sites are palms and soles. Lesions on other parts of the body usually manifest as multiple verrucous, keratotic, brown-to-flesh–colored papules which may coalesce into linear plaques in a Blaschkoid distribution.[2,3] Most of the lesions are persistent, however, occasionally spontaneous flattening of lesions have been reported.[6] Lesions are thought to be benign; however, malignant transformation has been reported.[7,8] Rare associations are seizure disorder, deafness and development delay, hemiparesis, sensory polyneuropathy, hyperthyroidism, scoliosis, breast hypoplasia, alopecia, and onychodysplasia.[2,3,8]

Classic histopathology of PEODDN includes a parakeratotic column (cornoid lamellae) occupying as invagination in the epidermis associated with loss of granular cells at the base of the column. Multiple eccrine ducts are seen in the upper dermis in relation to these cornoid lamellae. A moderately dense, lymphocytic infiltrate may be present in the papillary dermis.[8,9]

Clinically, it may be difficult to differentiate PEODDN from various linear and segmental dermatoses, i.e., linear porokeratosis, linear lichen planus, linear verrucous epidermal nevus (VEN), inflammatory linear verrucous epidermal nevus (ILVEN), linear and nevoid psoriasis, and linear Darier's disease. In such circumstances, diagnosis mainly depends on clinicopathological correlation. Linear porokeratosis, the closest clinical differential, usually presents as flat atrophic lesions with thready hyperkeratotic elevated margins. Pitting is usually not seen, and there is absence of eccrine units in close association with overlying parakeratotic columns invaginating the underlying epidermis. Linear lichen planus lesions are less keratotic, flat-topped, violaceous papules and plaques with marked lichenoid interface pattern on histopathology. VEN lesions are usually asymptomatic, linear verrucous plaques. Histopathology is characterized by constellation of three findings, i.e., acanthosis, papillomatosis, and hyperkeratosis. ILVEN lesions are inflammatory and pruritic compared to VEN and histopathology shows sharply demarcated alternating areas of hypergranulosis with overlying orthokeratotic hyperkeratosis and hypogranulosis with overlying parakeratosis. An upper dermal perivascular inflammatory infiltrate composed of lymphocytes and histiocytes is a regular feature. Linear and naevoid psoriasis is a rare form of psoriasis, which is characterized by linear distribution of the erythematous, scaly lesions along Blaschko's lines. The classic histological features of psoriasis are hyperkeratosis, parakeratosis, absence of granular cell layer, elongated rete ridges, suprapapillary thinning, and Munro's microabscesses. Discrete tiny keratotic papules that exacerbates with light, heat, sweating, and friction with histology showing suprabasal acantholysis and dyskeratotic cells in upper layers of epidermis are distinctive features of linear Darier disease.

Surgical excision and ablative lasers are the first line therapy for PEODDN.[9] Smaller and localized lesions can be treated with surgical excision; however, recurrence is possible.[10] Large lesions are relatively better managed with ablative lasers such as CO_2 laser and combined Erbium/CO_2 laser. Various other treatment options, i.e., topical corticosteroids and calcipotriol, retinoids, keratolytics, phototherapy, and cryotherapy have been tried with disappointing results.[9,10]

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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