A Rare Presentation of Linear Verrucous Epidermal Nevus: Case Report

Lineer Verrüköz Epidermal Nevüsün Nadir Bir Görünümü

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ABSTRACT Epidermal nevi are rare organoid nevi that originate from pluri-potent germinative cells at the basal layer of the embryonic epidermis. There are subtypes of verrucous epidermal nevus as localized, systematized, nevus unius lateris, ichthyosis hystrix and inflammatory linear verrucous nevus. Epidermal nevus is a mosaic disease that can be seen in various clinical and histopathological forms. It involves unusual areas like the palmoplantar or genital regions and may be a part of epidermal nevus syndrome. Palmar or plantar involvement may be alone or in concordance with other common involvement areas. In this report, a case of bilateral, palmoplantar linear verrucous epidermal nevus without trunkal involvement is presented. To the best of our knowledge, this is the first case distinctly described in the literature.

Key Words: Keratoderma, palmoplantar; nevus

ÖZET Epidermal nevüsler, embriyonik epidermisin bazal tabakasındaki pluripotent germinatif hücrelerden köken alan organoid nevüslerdir. Verrüköz epidermal nevüsün, lokalize, sistematize, nevus unius lateris, ihtiyosis histriks ve inflamatuar lineer verrüköz nevüs gibi alt tipleri vardır. Epidermal nevüs, çeşitli klinik ve histopatolojik formlarda görülebilen mozaik bir hastalıktır. Palmoplantar veya genital alanlar gibi alışılmadık bölgeleri tutabilir ya da epidermal nevüs sendromunun bir parçası olabilir. Palmar ya da plantar tutulum tek başına veya diğer sık tutulum alanlarıyla beraber görülebilir. Burada, gövde tutulumu olmayan bilateral palmoplantar lineer verrüköz epidermal nevüs olgusu sunulmuştur. Bilgilerimiz dahilinde literatürde bu şekilde tanımlanmış ilk olgu sunumudur.

Anahtar Kelimeler: Keratoderma,palmoplantar; nevüs

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Epidermal nevi are organoid nevi that originate from pluri-potent germinative cells at the basal layer of the embryonic epidermis. These nevi include keratinocytes and/or cutaneous appendages.[1] Linear verrucous epidermal nevus (LVEN) is a rare cutaneous hamartoma that was first described by von Baerensprung in 1863 and then reported by Unna in 1896.[2] There are subtypes of verrucous epidermal nevus as localized, systematized, nevus unius lateris, ichthyosis hystrix and inflammatory linear verrucous nevus.[3] LVEN usually settle unilaterally. They always place along Blaschko's lines. Bilateral cases are rare.[4,5] Verrucous epidermal nevi can rarely be found at the palm and sole or dorsum of the hand and the foot.[6-12] Bilateral palmoplantar LVEN without body involvement has not been reported previously. In this report, a case of bilateral, palmoplantar LVEN without trunkal involvement is presented.

CASE REPORT

A 25-year-old female suffering from palmar and plantar thickening since her birth presented with protuberance on her right hand fingers and left foot toes. On dermatological examination there were linear keratotic papules with extention to the dorsal side of the right first and second finger (Figure 1). Additionally, there were spinoid keratotic papules on a hyperemic background and punctuate dimples, settled linearly on the volar and thenar sides of the 1st finger of her right hand and 1st, 2nd and 4th fingers of her left hand (Figure 2). Keratotic papules were settled individually at the dorsal side of the 4th and 5th left toes. There were also hyperkeratotic papules forming a plaque demonstrating punctuated dimples bilaterally on the plantar sides of her feet and volar side of 4th and 5th toes of her left foot (Figure 3). Both of hand and foot lesions were following the lines of Blaschko. Her finger- and toe nails and mucous membranes were normal. There was no family history and her parents were non-consanguineous. She had diagnosed as verruca vulgaris and received salicylic acid 10 years ago.

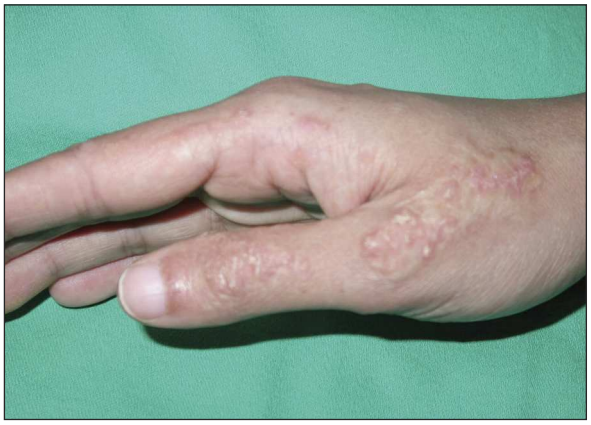

FIGURE 1: Keratotic papules settle at the dorsal side of the first and second finger of right hand linearly.

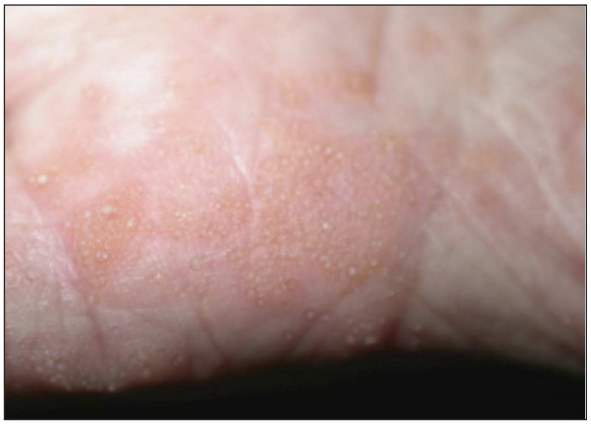

FIGURE 2: Spinoid keratotic papules and punctuate dimples on palmar region.

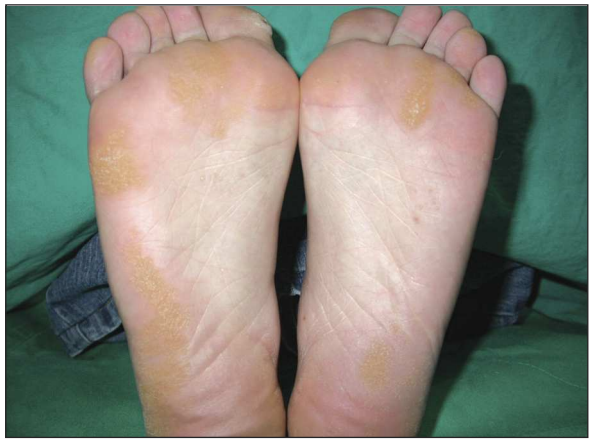

FIGURE 3: Hyperkeratotic plaques with punctuate dimples following the lines of Blaschko on plantar region.

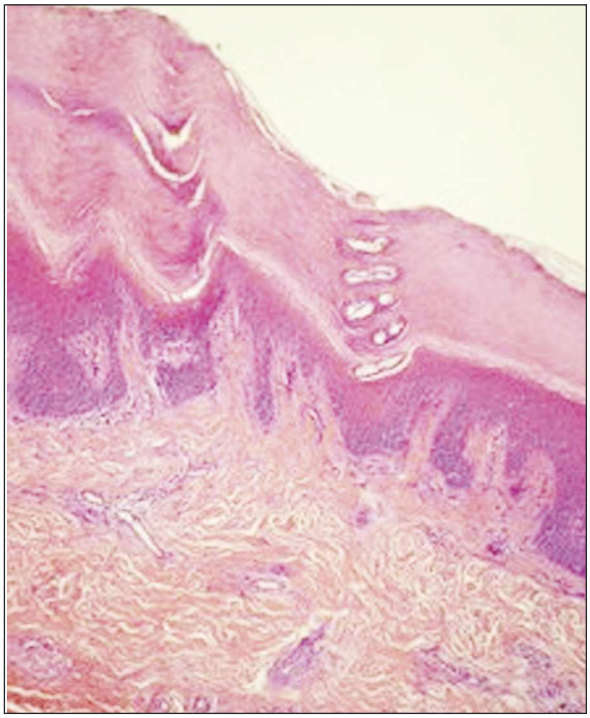

FIGURE 4: Acanthosis, papillomatosis with a lamellar hyperkeratosis and mild inflammatory infiltrate (H-E X 40).

A punch biopsy specimen from thenar sides of the left hand revealed acanthosis, papillomatosis, hyperkeratosis and mildly hypergranulosis in the epidermis, with sparsely chronic inflammatory cells in the dermoepidermal junction (Figure 4).

Hemogram, biochemical analysis, erythrocyte sedimentation rate, urogram, abdominal sonographic examination, cranial and extremities radiographics were found normal. Ophthalmologic and neurologic examinations revealed no abnormality. The patient refused treatment.

DISCUSSION

Epidermal nevus is a mosaic disease that can be seen in various clinical and histopathological forms. It can be localized or widespread. It involves unusual areas like the palmoplantar or genital regions and may be a part of epidermal nevus syndrome. Palmar or plantar involvement may be alone or in concordance with other common involvement areas.[6-13]

LVEN is an epidermal nevus that demonstrates keratinocytic differentiation. Although these nevi usually appear at birth or in early childhood, they can first become obvious in adolescence and adulthood.[14] In our case, the lesions have been existed since birth.

LVEN is characterized by rough, gray-brown or skin-like in color, verrucous or velvety papules that tend to be coalescent. Sometimes these papules may form well-circumscribed plaques. Although the trunk and the extremities are the main areas of settlement, the nevi can be found on the head and even on the face. Diffuse and bilateral lesions of epidermal nevus are usually appeared in wavy transverse band form in the trunk. However, the lesions tend to be spiral and longitudinal form in the extremities according to Blaschko's lines as in our patient.[14]

LVEN usually settle on the trunk, lower and upper extremities in three large series of epidermal nevus and/or epidermal nevus syndrome. In these series, there were no LVEN with palmar or plantar localization.[4,5]

Generalize body involvement can accompany to palmoplantar LVEN.[8] The other clinical aspects of palmoplantar LVEN are unilateral hand or foot involvement alone and unilateral both hand and foot involvement.[6,7,9,10] Bilateral verrucous epidermal nevi have been reported as a generalized form or a form with extremity involvement without palmoplantar involvement in a few cases.[8,15] In our case, lesions were localized to palmoplantar areas and dorsal sides of the toes and fingers bilaterally without trunkal or extremity involvement. Medline search revealed 5 previously reported cases of palmar or plantar LVEN. The main features of these cases together with our case are summarized in Table 1.[6-10]

Despite the fact that histopathology of LVEN shows lengthening in rete with hyperkeratosis, acanthosis and papillomatosis, histopathological appearances similar to that of epidermolytic hyperkeratosis, Darier's disease, psoriasis, Hailey–Hailey disease, seborrheic keratosis, porokeratosis, acrokeratosis verruciformis and acanthosis nigricans have also been observed. Histopathological exami-

TABLO 1: Clinical and histopathological data on 5 patients with palmar or plantar LVEN.

Case no	Reference	Localization of body lesions	Unilateral PP lesions	Bilateral PP lesions	Histopathological findings
1	(6)	(-)	Left plantar	(-)	Hyperkeratosis, hypergranulosis, acanthosis, papillomatosis
2	(7)	(-)	Left palmar	(-)	Hyperkeratosis, papillomatosis, epidermal hyperplasia
3	(8)	Bilateral trunk, extremites	(-)	(+)	Epidermolytic hyperkeratosis
4	(9)	(-)	Left plantar	(-)	Acanthosis, orthokeratotic hyperkeratosis, suprabasal acantholysis, dyskeratosis
5	(10)	(-)	Right PP	(-)	Hyperkeratosis, vacuolar degeneration of granular layer, hypergranulosis
6	Present case	Dorsum of right hand and left foot	(-)	(+)	Lamellar hyperkeratosis, papillomatosis, acanthosis

PP palmoplantar, (+) with involvement, (-) without involvement

nation of our case demonstrated a seborrheic keratosis-like pattern.[4,14]

The lesions of our patient follow the Blaschko's lines. The linear distribution of the Blaschko's lines possibly suggests genetic mosaicism. Lichen striatus, linear porokeratosis, linear Darier's disease, linear lichen planus and linear psoriasis should be included in the differential diagnosis of LVEN. Especially, inflammatory linear verrucous epidermal nevus, a subtype of LVEN, is proposed to be a form of psoriasis. However, there are no clear-cut detectable differences between inflammatory LVEN and linear psoriasis.[3,16-18]

In the differential diagnosis of LVEN cases with palmoplantar involvement, porokeratotic eccrine ostial and dermal duct nevus, keratosis punctuate palmo-plantaris Buschke–Fischer–Brauer, porokeratosis punctuate palmaris et plantaris, calluses, viral warts, acrokeratoelastoidosis and focal acral hyperkeratosis should be considered.[19-22]

Porokeratotic eccrine ostial and dermal duct nevus is a congenital, asymptomatic dermatosis that follows Blaschko lines. It is characterized by multiple punctuate papules and dimples with comedone-like keratotic plugs settled at the center of the lesion. They can be differentiated from LVEN by the absence of a granular layer and by demonstrating parakeratotic columns in epidermal invaginations.[19]

Keratosis punctuate palmo-plantaris Buschke–Fischer-Brauer is characterized by symmetrical, pin-like in size, depressed, hyperkeratotic papules or hyperkeratosis areas like a water drop or a pearl in areas that are usually not exposed to mechanical irritation. On the other hand, in areas that are exposed to pressure, such as the lateral sole, it demonstrates hyperkeratotic papule plaques. Absence of the lesions at the dorsal sides of the hands and the feet and demonstration of nonspecific acanthosis, orthohyperkeratosis, hypergranulosis and depressed epidermis under the hyperkeratotic plug are main features that make it distinguished from LVEN.[20]

Porokeratosis punctuate palmaris et plantaris is characterized by multiple spindle-like keratosis with keratotic plugs over the palmoplantar area. It can be differentiated from LVEN by its cornoid lamellae and parakeratosis.[20]

Acrokeratoelastoidosis is characterized by small, yellowish white, oval-polygonal, rhomboid, wart-like, hard papules and plaques. Its histopathology demonstrates hyperkeratosis, epidermal hypertrophy and decrease in dermal elastic fibers and these can be used to differentiate them from LVEN. Fokal acral hyperkeratosis resembles acrokeratoelastoidosis clinically. These are more common among blacks and demonstrate absence of elastorrhexis and presence of intact elastic tissue in the dermis.[21]

Consequently, this report has demonstrated an unusual case of LVEN that involved the palmoplantar regions bilateral and the dorsal side of the fingers along the Blaschko's lines, without body involvement, demonstrating seborrheic keratosis-like pattern histopathologically. To the best of our knowledge, this is the first case distinctly described in the literature. This report contributes to the wide clinical spectrum of epidermal nevus.

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