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Inflammatory Linear Verrucous Epidermal Nevus Co-Existing with Nevoid Psoriasis Associated with Arthropathy: A Rare Association

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Abstract

Inflammatory linear verrucous epidermal nevus (ILVEN) is a keratinocytic nevus characterized by erythematous pruritic plaque along the lines of Blaschko. Epidermal nevi are associated with primarily skeletal abnormalities and less commonly central nervous system (CNS) manifestations. There have been only few case reports with the involvement of the musculoskeletal system. We present a case of ILVEN associated with a deformity of hand secondary to arthropathy.

Keywords: Nevoid psoriasis; Inflammatory verrucous epidermal nevus; Arthropathy; Deformity; Blashckoid

Introduction

Segmental mosaicism leads to variable presentation of nevoid condition leading to Blashckoid distribution of lesions [1]. ILVEN and nevoid psoriasis are two such conditions having similarities clinically and histologically which may lead to diagnostic difficulties. Arthritis has been described as a new association in the literature, which if detected early and managed promptly may prevent complications. We came across case of ILVEN associated with arthropathy which was initially thought to be nevoid psoriasis with arthropathy leading to a diagnostic dilemma.

Case Discussion

An 11-year-old female had itchy pruritic lesions predominantly involving the left side of her body since 6 years (Figure 1,2 and 4). There were episodes of exacerbation in winter. There was no involvement by the disease in any of the family members. There was a progressive loss of movement of the left little and ring finger at the distal interphalangeal joint (Figure 3) during the past 2 years. There was no preceding history of joint pain or swelling. On examination, there were multiple erythematous scaly plaques along the lines of Blaschko involving the left side of the chest, back, arm, forearm and palm intermingled with atrophic hypopigmented macules and patches. Similar erythematous scaly plaques were present on the right palm (Figure 3). A fixed flexion deformity of the left little and ring finger was observed. The scalp and nails were normal.

Clinically a diagnosis of naevoid psoriasis with arthritis was made based on the morphology and distribution of the lesions and the patient was treated with topical Flucinolone acetonide cream. On further investigation, radiological examination of the deformity revealed bony ankylosis of the left ring and little finger distal interphalangeal joint and a thinning of the middle phalanx (Figure 5). On histology, there was stratified squamous epithelium with moderate acanthosis, papillomatosis, foci of parakeratosis, elongation of rete ridges with slight spongiosis and exocytosis of lymphocytes in the epidermis and mild-to-moderate perivascular chronic inflammatory infiltrate of lymphocytes and histiocytes in the dermis (Figure 6-8). These were the features suggestive of ILVEN. It was difficult to arrive at a diagnosis in our case as there were features of both naevoid psoriasis and ILVEN. After the literature review, we considered the possibility of ILVEN and naevoid psoriasis being present concomitantly. She showed partial response to topical steroid at 6 weeks and was subsequently lost to follow-up.

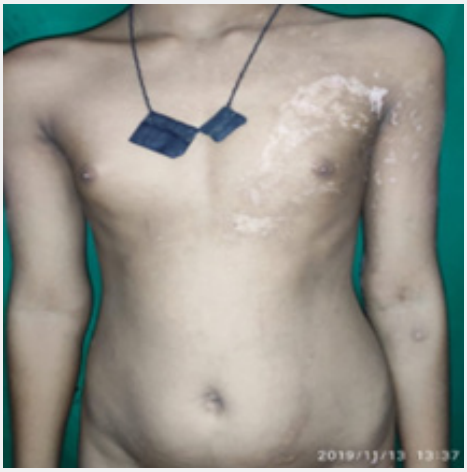

Figure 1: Atrophic macules and scaly plaques along the lines of Blashcko.

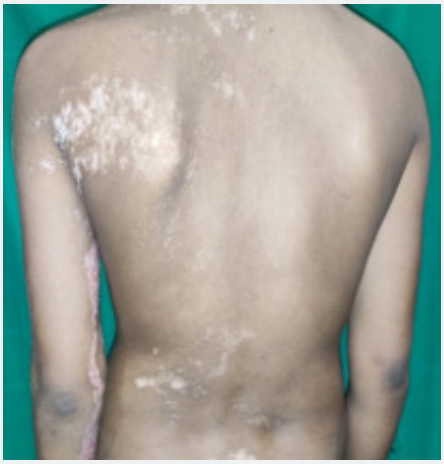

Figure 2: Atrophic macules, patches and erythematous scaly plaques in a Blaschkoid distribution.

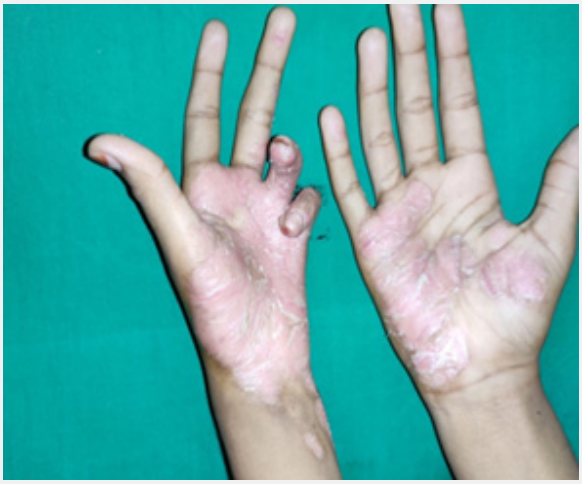

Figure 3: Erythematous scaly plaques involving both palms with a flexion deformity of left ring and little Finger.

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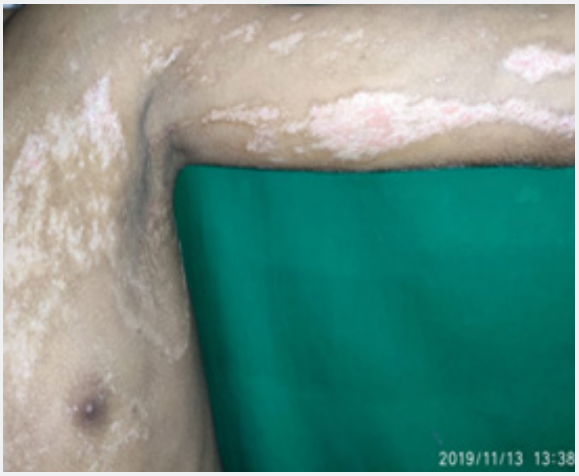

Figure 4: Scaly erythematous plaques with atrophic macules coalescing to form a patch over left upper limb and trunk.

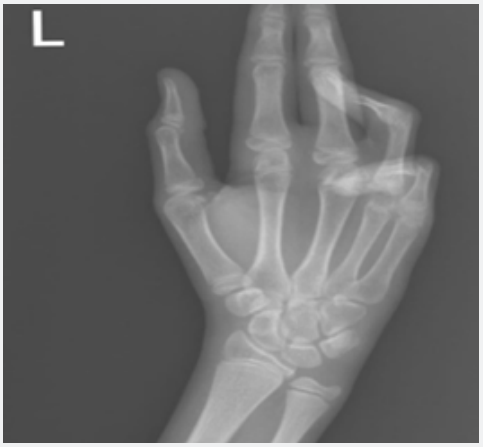

Figure 5: Radiological image of left hand showing bony ankylosis of left interphalangeal joint and thinning of middle phalanges of left ring and little finger.

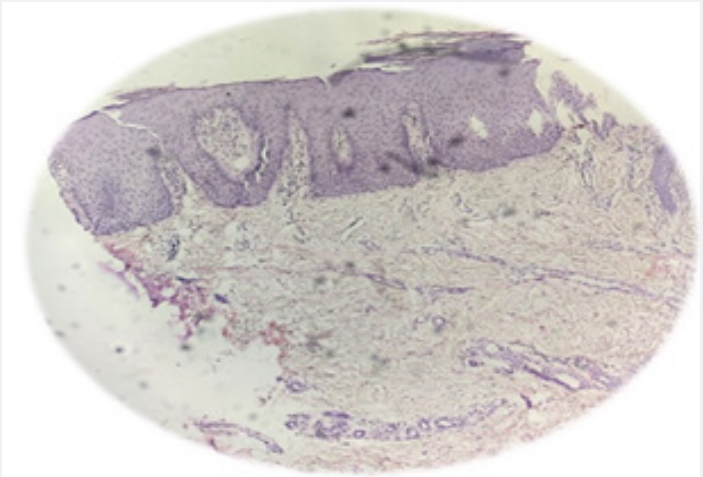

Figure 6: Skin biopsy showing parakeratosis, acanthosis, elongated rete ridges, spongiosis.

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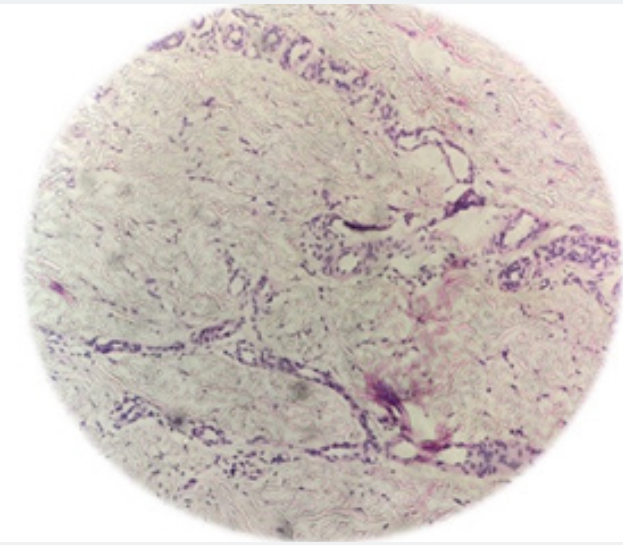

Figure 7: Dermal findings on histopathology showing inflammatory lymphocytic infiltrate in the dermis.

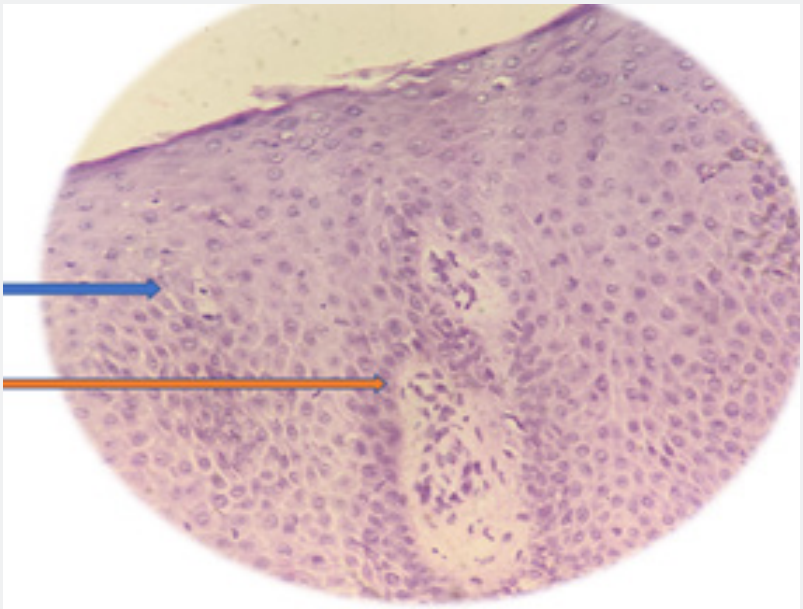

Figure 8: Exocytosis of lymphocytes (blue arrow) with dilated dermal capillaries (yellow arrow).

Discussion

There has been a considerable debate whether ILVEN and naevoid psoriasis are distinct entities or variants of each other. Though they may have a resemblance, a few differentiating features that help in arriving at a diagnosis are given in Table 1. A diagnostic criteria for ILVEN was given by Altman and Mehregan in 1971 which had the following limbs. Early age of onset, 4:1 female preponderance, frequent involvement of left lower extremity, pruritus, distinct and inflammatory linear appearance, following the lines of Blaschko and persistent lesions showing marked refractoriness to treatment [2]. In our case, the female predilection, early age of onset, left side of involvement and histopathology lead to a diagnosis of ILVEN. However, due to the deformity, a history of winter exacerbation, presence of skin lesions outside the lines of Blaschko (over right palm and lower back), bony ankylosis and thinning of the middle phalanx, which represents a late feature of psoriatic oligoarthritis [3], nevoid psoriasis with arthritis could not be ruled out.

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Table 1: Differentiating features of ILVEN and Nevoid psoriasis.

	ILVEN	Nevoid psoriasis
Clinical features	More pruritic, early age of onset, female preponderance, left side of involvement [3]	May appear later in life, slightly itchy, can involve any site
Histology	Alternate bands of hypergranulosis with overlying orthokeratosis and agranulosis with parakeratotic hypergranulosis in addition to psoriatic features [3]	Hyperkeratosis, parakeratosis, agranulosis, elongated rete ridges Munro's microabscesses
Treatment	Refractory to anti-psoriatic treatment	Responds well to anti psoriatic therapy.
Immunohistochemistry	1. Increased expression of keratin 10[3] 2. Absent involucrin, [1], 3. Lower ki 67 expressions. 4. A focal pattern of anti-keratin 16[9].	1. Keratin expression not very extensive 2. Involucrin expression is seen 3. Elastase positive cells -more significant [9] 4. A homogenous pattern of anti-keratin 16[9]

Although described as separate entities, recently, it has been proposed that ILVEN and nevoid psoriasis may co-exist in the same patient [4]. Psoriasis developing on an epidermal nevus has been described, where the subjects present with typical psoriatic lesions in other parts of the body [5]. It is possible that there was an overlap of the above two conditions in this case. ILVEN has been reported to be associated with a few musculoskeletal abnormalities like supernumerary digits, congenital bony anomalies of the ipsilateral extremities, congenital dislocation of the ipsilateral hip [6], contractures, dactylitis and bony ankylosis [7].

However, our patient had an acquired deformity of the hand with concomitant features of arthropathy. The association of arthritis with ILVEN has been described recently in a few case reports [8,9]. We hypothesize that this deformity was due to associated psoriatic arthritis secondary to the co-existent naevoid psoriasis. There have been reports where arthritis associated with ILVEN has responded to methotrexate without affecting the skin lesions. It has been recommended that ILVEN-associated arthritis should be treated on the lines of psoriatic arthritis. This association has to be kept in mind while treating a case of ILVEN as early diagnosis and proper management may help in preventing deformities.

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