

## Case report

# Nevoid psoriasis: an uncommon blaschkolinear dermatosis

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**Abstract** Nevoid or linear psoriasis is an uncommon form of psoriasis characterized by the linear or zosteriform distribution of the psoriatic lesions. It usually follows the lines of Blaschko with unilateral involvement. It is commonly confused clinically and histopathologically with verrucous epidermal nevus. We report a young male, who had linear psoriasis in a unilateral distribution over his shoulder and arm. He was diagnosed on clinical as well as histological ground and was managed on the lines of psoriasis.

**Key words**

Linear psoriasis, nevoid psoriasis, lines of Blaschko, inflammatory linear verrucous epidermal naevi.

### Introduction

Psoriasis is a common chronic, genetic, noncontagious inflammatory skin disorder that appears in many clinico-morphological forms and can affect any part of the body. Nevoid or linear psoriasis is a rare variant of this common disorder and is characterized by a linear distribution of the psoriatic lesions. It follows the lines of Blaschko with unilateral involvement and usually occurs in young adults but has also been reported in childhood.<sup>1,2</sup> The pathogenesis is unclear, but it could be explained as a result of the migration of cells harboring a somatic mutation following the lines of Blaschko during early embryogenesis.<sup>3,4</sup> It was first described in 1951 by Leslie<sup>5</sup> and Sobel.<sup>6</sup> In psoriasis lesions in a

blaschkolinear distribution mostly occur together with scattered lesions, but occasionally they may be isolated and may present in a purely nevoid form following Blaschko's lines, may koebnerize, or may superimpose an epidermal nevus.<sup>7,8</sup> Very rarely it may coexist with another rare dermatosis that follows Blaschko's line, e.g. porokeratotic eccrine ostial and dermal duct nevus.<sup>9</sup> Several clinical entities resemble nevoid psoriasis, and differential diagnosis is important. It is the inflammatory linear verrucous nevus (ILVEN) that is most commonly confused with linear psoriasis and a clear distinction between linear psoriasis vulgaris and (ILVEN) can be difficult because of the clinical and histopathologic similarities of the two conditions.<sup>10</sup> An association of ILVEN with psoriasis vulgaris has also been reported.<sup>7</sup> Lesions of the ILVEN are pruritic, most commonly occur on the legs, pelvis, and buttock, have early age of onset and female predominance. The congenital hemidysplasia with ichthyosiform nevus and limb deficiency

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(CHILD) syndrome presents with verrucous plaques in a segmental distribution but other associated features clearly differentiate it. Lichen striatus and linear lichen planus also resemble nevoid psoriasis or ILVEN, however, their histology is quite distinct.<sup>10-12</sup> Above clinical and histopathologic features clearly differentiate these disorders from linear psoriasis. In difficult to diagnose cases, involucrin immunohistochemistry can be a useful diagnostic tool to confirm diagnosis of nevoid psoriasis.<sup>13</sup> Treatment with keratolytics and topical calcipotriol, topical steroids and other topical anti-psoriatic regimens lead to a significant improvement.<sup>1,2</sup> The purpose of reporting the case was to highlight the existence of nevoid or linear psoriasis as an entity.

### Case report

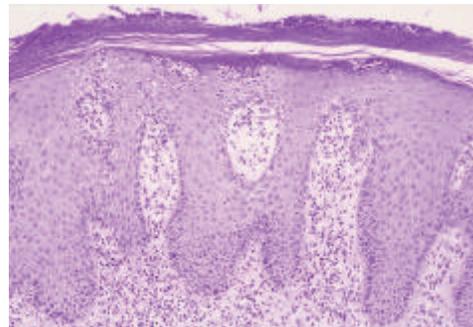
A 25-year-old man, presented with a slightly pruritic, linear plaques over his left shoulder and adjacent arm. The lesions started appearing three years ago as small discrete papules and plaques roughly in a linear fashion on back of his right shoulder and arm and gradually progressed and coalesced to form large linear scaly plaques. He had no relevant personal or family history. On clinical examination, large erythematous scaly verrucous lesions were seen that were linearly distributed over back of his left shoulder axilla and outer aspect of the left upper arm (**Figure 1 and 2**). There were no lesions suggestive of psoriasis else where over his skin nails or scalp. Skin biopsy revealed classical histological features of psoriasis showing acanthosis with regular elongation of rete ridges, parakeratosis, Munro's microabscesses, spongiform pustules (**Figure 3**). There were no columns of hypergranulosis with orthokeratosis



**Figure 1** Linear erythematous, scaly eruption affecting left axilla, shoulder and upper arm



**Figure 2** Another view of linear hyperkeratotic, erythematous and scaly eruption.



**Figure 3** Histopathology of the lesion, psoriasiform hyperplasia of epidermis along with Munro's microabscesses.

alternating with columns of agranulosis with parakeratosis as we see in ILVEN.

Clinical examination and results of histopathology confirmed the diagnosis of unilateral nevoid or linear psoriasis. He

was managed with topical anti-psoriatic ointment (containing tar, salicylic acid, topical steroid dispensed in emulsifying ointment).

## **Discussion**

Psoriasis is one of a number of autoimmune diseases that display significant HLA associations (HLA-Cw6). However, only about 10% of Cw6-positive individuals develop disease, suggesting that other genetic and/or environmental factors must be involved. Several compelling lines of epidemiologic evidence indicate that psoriasis susceptibility is inherited, albeit not in a simple monogenic fashion, and that genetic, rather than environmental, factors are primarily responsible for the variability in inheritance of psoriasis. Taken together, these observations suggest that one or more loci in addition to HLA are necessary for the development of psoriasis. The number of additional loci is likely to be small, because i) the disease is very common ii) substantial excess risk of psoriasis is observed in first degree relatives, and iii) nevoid variants of psoriasis have been reported, suggestive of somatic mutation of a single gene during development.<sup>3,4</sup> The existence of a linear psoriasis has frequently been a subject of debate. Several authors have disputed the existence of true linear psoriasis and contend that many reports of linear psoriasis represent either inflammatory linear verrucous epidermal nevus (ILVEN) in a patient with psoriasis, a nevus, or even an invasion of ILVEN by psoriasis as a manifestation of the isomorphic reaction.<sup>10,11</sup> In contrast, some authors have a strong opinion that linear psoriasis is a separate entity. Therefore, before diagnosis, it should be differentiated clinically and histopathologically from

ILVEN. The criteria for the diagnosis of ILVEN established by Altman and Mehregan<sup>14</sup> include early age of onset, 4 times more common among women than men, frequent involvement of the left leg, pruritus, psoriasiform appearance, persistence, and resistance to treatment. Among these criteria, a differential point is that the lesions of ILVEN take the form of intensely pruritic linear groups of excoriated eczematous papules, which proves extremely refractory to therapy. The histopathologic aspect of ILVEN is very similar to psoriasis, but is characterized by columns of hypergranulosis with orthokeratosis alternating with columns of agranulosis with parakeratosis. The case reported by us was clinically more suggestive of nevoid psoriasis rather than ILVEN, as the patient was male, the disease had late onset, and the lesions were not much pruritic and were distributed over upper half of the body (these features were against ILVEN). Moreover, the histology of the lesion revealed classical features of psoriasis and there were no alternating columns of hypergranulosis with orthokeratosis and agranulosis with parakeratosis. The response to anti-psoriatic treatment was satisfactory.

## **Conclusion**

Psoriasis may koebnerize in a linear fashion, may superimpose or invade an epidermal nevus but a nevoid form following Blaschko's lines should be taken a separate entity.

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