Psoriasis in a linear or unilateral distribution is rare. We report a 1-year-old boy who had a few erythematous scaly plaques at birth that gradually progressed to widespread unilateral plaques along the lines of Blaschko. The term congenital blaschkoid psoriasis was proposed by Lehners-Weber C et al. in 1996 to describe congenital linear psoriatic lesions following the lines of Blaschko. The differentiation of linear psoriasis from other linear dermatoses is not easy. The combination of a complete history, careful skin examination, and histopathology are essential for making the correct diagnosis and insuring proper treatment. (Dermatol Sinica 24: 256-259, 2006)

Key words: Congenital Blaschkoid psoriasis, Linear psoriasis, Nevoid psoriasis, ILVEN, Blaschko’s lines
INTRODUCTION

Linear or nevoid psoriasis are terms for psoriasis appearing in a linear or nevoid distribution. There have been a few reports of psoriasis occurring in early childhood in an unusual nevoid distribution following the lines of Blaschko. Lehners-Weber et al. described a girl with congenital linear psoriatic lesions following the lines of Blaschko and proposed the term congenital blaschkoid psoriasis for this unusual clinical expression. We present a similar case.

CASE REPORT

A one-year-old boy presented with unilateral linear and whirled erythematous plaques on the left trunk and limbs. He had only a few erythematous lesions with scales at birth gradually progressing to widespread unilateral lesions. The child did not appear to have pruritus or other symptoms related to the skin lesions. On examination, scaling erythematous plaques were found exclusively on the left side of the body in lines running from the chest over the axillary folds to the dorsum of the hand (Fig. 1A-C), from the sole to the thigh and genital area (Fig. 1D), from the chest to the back (Fig. 1A-C), and from the abdomen to the back. The lesions were linear streaks or bands along the lines of Blaschko. There were also a few erythematous plaques on left scalp. The patient was treated with topical hydrocortisone and urea cream, but the response was poor. Our differential diagnosis at that point was linear verrucous epidermal nevus (ILVEN) or lichen striatus. A skin biopsy was taken from the left arm.

A low-power microscopic view of the specimen showed regular psoriasiform hyperplasia with a superficial perivascular lymphocytic infiltrate (Fig. 2A), along with confluent parakeratosis, hypogranulosis and papillary dermal edema (Fig. 2B). Neutrophilic microabscesses were present in the superficial epidermis and within the stratum corneum (Fig. 2C). These findings were most consistent with a diagnosis of psoriasis.

No other family members had psoriasis or similar skin lesions. When the patient returned for removal of the sutures, it was noted that the skin lesion at the biopsy site had worsened, consistent with the Koebner phenomenon. Topical desoximetasone ointment was prescribed for the trunk and limb lesions and betamethasone valerate cream for the scalp, both resulting in an excellent response by the end of 2 weeks. The
plaques on the scalp, trunk and limbs had either disappeared completely or had thinned and faded (Fig. 3).

**DISCUSSION**

Psoriasis rarely occurs in a linear or unilateral distribution. When it does, it may very closely resemble other linear dermatoses such as epidermal nevi and lichen striatus. However, histopathologic examination alone often fails to reliably differentiate linear psoriasis from dermatitic epidermal nevi. Consequently, many authors have questioned the existence of true linear psoriasis. They have assumed such disorders to be linear epidermal nevi with psoriatic features or epidermal nevi occurring in patient with psoriasis. There certainly have been many reports of the invasion of linear epidermal nevi or systematized epidermal nevi by psoriasis as a manifestation of the Koebner phenomenon. However, many investigators still believe that linear or nevoid psoriasis is a distinct entity. This belief is supported by the fact that quite a few individual cases have been reported that have unmistakable clinical and histopathologic evidence of psoriasis.

The main differential diagnosis is ILVEN, clinical and pathological criteria for which were described by Altman and Mehregan in 1971. In a study of 25 patients they defined the diagnostic criteria as: (i) early age of onset, (ii) female predominance, (iii) frequent involvement of the left lower extremity, (iv) substantial pruritus, (v) persistence of the lesions, and (vi) refractoriness to treatment. They described an inflammatory and psoriasiform appearance histopathologically, including areas of parakeratosis with loss of the granular cell layer and orthokeratosis with a prominent granular layer with a sharp demarcation. This finding was absent in our patient.

Lesions of any particular disorder that happen to be distributed along the lines of Blaschko still tend to exhibit the same clinical and histologic picture as similar lesions with a more common distribution. Linear psoriasis has the same pathologic features as classic psoriasis. However, ILVEN and psoriasis share certain histologic features, making the differentiation difficult by pathology alone. A definitive diagnosis requires not only a detailed history and careful visual and microscopic examination of the skin lesions but also on close monitoring of the disease course and response to treatment.

Another possible diagnosis to consider is the invasion of a linear epidermal nevus by psoriasis. In such a case, typical plaque or guttate psoriasis lesions are present in addition to the linear skin lesions. A careful history may reveal that non-inflammatory, minimally erythematous lesions preceded the appearance of plaques. Our patient had no preceding linear lesions, and all of his psoriatic lesions developed in linear zones following the lines of Blaschko. Further clues supporting the diagnosis of psoriasis for this patient included the lack of pruritus, a good response to topical steroids, and the histopathology which was consistent with psoriasis. Even the relatively rapid progression and widespread distribution of the lesions was more consistent with psoriasis rather than ILVEN.

The issue of treatment response is important. A poor response to antipsoriatic therapy may indicate inadequate treatment rather than misdiagnosis. It's better not to jump to a conclusion that a patient has ILVEN. Rather, ade-
Adequate antipsoriatic treatment should be given and the patient followed for a reasonable period of time before determining that the lesions are unresponsive.

Blaschko’s lines were described and drawn in 1901 by Alfred Blaschko. These lines are to be distinguished from other linear patterns such as Voight’s lines, Langer’s lines, and the lines of innervation of the spinal nerves. They do not follow any known nerves, vascular or lymphatic structure in the skin. It is now well-established that the lines of Blaschko are a result of genetic mosaicism, where chromosomal differences cause specific cells or groups of cells react differently from other cells in the same anatomic area. Migration along lines of Blaschko of a population of mutant cells during embryogenesis lead to streaks of genetically abnormal skin contrasting with genetically normal skin.

In conclusion, we believe that linear nevoid psoriasis is indeed a distinct entity that can be diagnosed based on a combination of historical, gross, and histopathological features. The lesions in our case were identical to those reported for congenital blaschkoid psoriasis by Lehners-Weber et al. A precise descriptive term for this unique form of congenital psoriasis.

REFERENCES