A Case of Acquired Blaschko Dermatitis in a Child; A Variant of Lichen Striatus, or Not?

Eun Ah Cho, M.D., Woo Sun Lee, M.D., Yoon Young Kim, M.D.,
Kyung Moon Kim, M.D., Si-Yong Kim, M.D.

Department of Dermatology, St. Vincent Hospital, College of Medicine, The Catholic University of Korea

Acquired Blaschko dermatitis is a rare inflammatory linear eruption that presents as multiple lines of itching papules and vesicles following the Blaschko's lines. Only ten cases have been reported in literatures since Grosshans' first description in 1990 and all cases have been in adults.

A 2-year-old girl presented with a 4-month history of a slightly pruritic papulovesicular eruption, which was limited to the left side of her body, along the Blaschko's line. A skin biopsy revealed interface dermatitis. The eruption initially failed to respond to treatment with topical steroids but responded to systemic steroids. We present the first case of acquired Blaschko dermatitis in a child with a review of the relevant literature.
(Ann Dermatol (Seoul) 19(4) 181～184, 2007)

Key Words: Blaschko dermatitis, Blaschko's line

INTRODUCTION

In 1990, Grosshans et al. described a man with acquired unilateral relapsing inflammatory linear lesions along the Blaschko's line[1]. They considered the disease to be a new entity and named it 'Blashkite de l'adulte'. Megahead et al. subsequently proposed the term 'acquired relapsing self-healing Blaschko dermatitis'[2]. Since then, Lee et al. proposed the term 'acquired Blaschko dermatitis'[3].

Although the etiology and pathogenesis of this condition is unclear, previously reported cases share common features. In all cases, the lesions are distributed linearly along the Blaschko's lines in adults, ranging in age from 24 to 65 years old (mean age 40 years), and have a tendency to relapse over time. Histopathologically, they show spongiotic or interface dermatitis. All published cases were in adults[1-7]. We describe the first case of acquired Blaschko dermatitis in a child.

Received February 16, 2007
Accepted for publication September 13, 2007

Reprint request to: Si-Yong Kim, M.D., Department of Dermatology, St. Vincent Hospital, College of Medicine, The Catholic University of Korea, Tel: 82-31-249-7465, Fax: 82-31-253-8927, E-mail: dervint@catholic.ac.kr

CASE REPORT

A 2-year-old girl presented with a 4-month history of a slightly pruritic eruption, which was limited to the left side of her body including the leg and trunk. The lesions were erythematous papules and vesicles that were distributed linearly along the Blaschko's lines (Fig. 1). Except for the skin lesions, there were no remarkable findings on physical examination. There was no personal or family history of atopic disorders, psoriasis or any other type of dermatitis or viral infection. She had been treated with topical corticosteroid therapy at a local clinic for three months with little improvement.

A biopsy specimen from a lesion on her left leg demonstrated mild hyperkeratosis of the epidermis with perivascular lymphohistiocytes infiltration and mild telangiectasis in the papillary dermis. A closer view revealed exocytosis, spongiosis and focal vacuolar degeneration of the basal cell layer in the epidermis (Fig. 2).

The eruption did not respond to methylprednisolone acetonate 0.1% ointment that had been

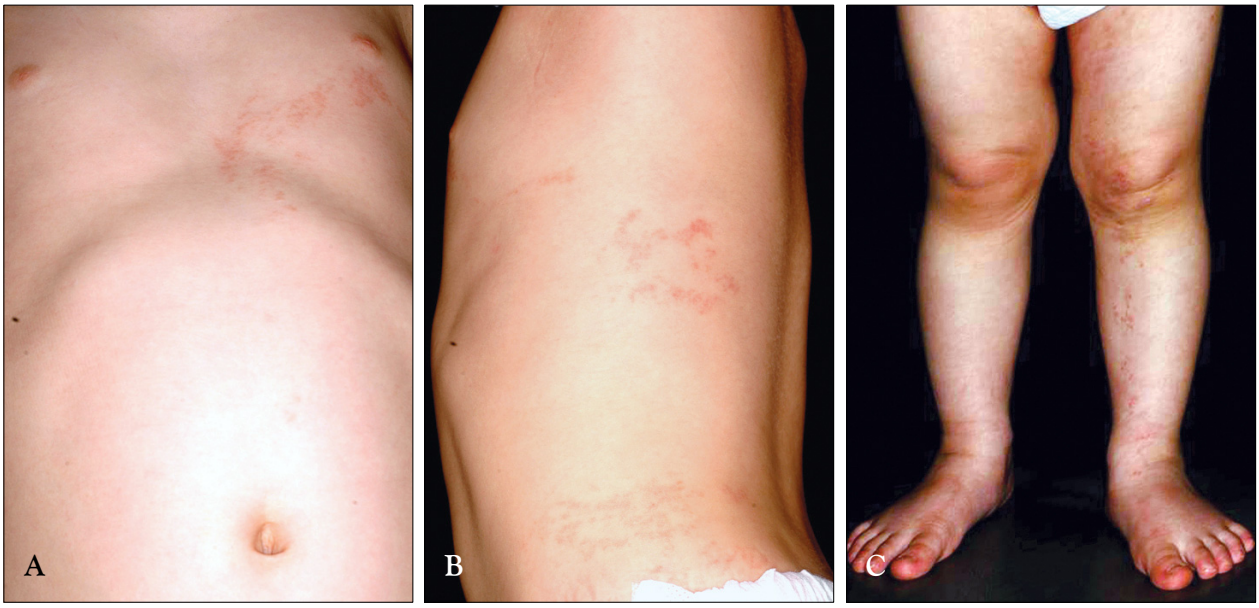

Fig. 1. A, B, C. Erythematous discrete grouped papules and vesicles on the left side of the leg and flank along the Blaschko line.

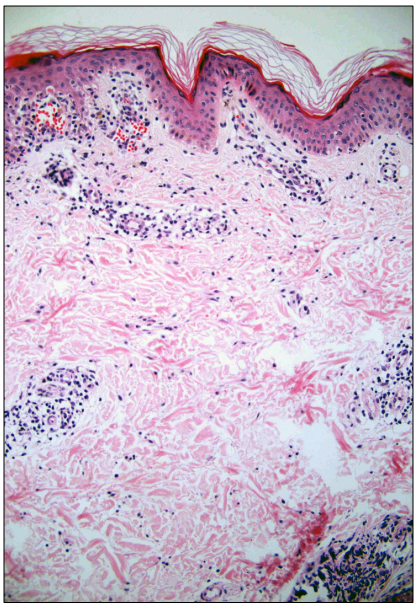

Fig. 2. Mild hyperkeratosis of the epidermis with perivascular lymphohistiocytes infiltration and mild telangiectasis in the papillary dermis. It showed exocytosis, spongiosis and vacuolar degeneration of the basal cell layer in the epidermis (H&E, × 200).

topically applied for one month. Therefore, systemic therapy was introduced with oral prednisolone 5 mg per day. Significant improvement was noted after 10 days but the lesion recurred 1 month later after the cessation of systemic therapy.

DISCUSSION

Thus far, ten cases of acquired Blaschko dermatitis have been reported in literature, and seven of them have been reviewed in this present report (Table 1)[1-7]. All seven cases were adults and there was no gender predilection. The skin lesions revealed multiple lines and uni- or bilateral lesions. An histological examination showed spongiotic or interface dermatitis. Topical steroid ointments had been applied in four cases but were only effective in one case. Systemic steroid was prescribed for one patient who showed a good response. The other three cases had no treatment but all improved within several weeks. In most cases, the lesions followed the same clinical pattern of relapsing several times for months and years.

In contrast to other reported cases, our case occurred in a child. But, the patient showed histologically non-specific interface dermatitis without lichenoid infiltration, presenting as multiple lines on the trunk and leg. More strikingly, it showed a

Table 1. Previously described cases of acquired blaschko dermatitis[1-7]

Reference	Age/Sex	Distribution of the skin lesion	Histopathology	Treatment	Prognosis
Grosshans[1] et al.	38/M	Whole body	Spongiotic dermatitis	None	Spontaneous improvement after a few days or weeks, and relapse was noted
Megahed[2] et al.	44/F	Left side of the upper and lower limb, chest and abdomen	Spongiotic dermatitis	Topical corticosteroid	After applying topical steroid for 2 weeks with little success, there was spontaneous improvement noted after 6 weeks.
Lee[3] et al.	27/M	Left side of the trunk and arm	Spongiotic dermatitis	Topical corticosteroid	Complete resolution after topical steroid treatment for a month with relapse being noted
Betti[4] et al.	24/F	Left side of the trunk	Interface dermatitis	None	Spontaneous improvement after 3 months
Lipsker[5] et al.	38/F	Whole body	Interface dermatitis	None	Spontaneous resolution within 4 weeks
Hale[6]	64/M	Left side of the chest, abdomen, back and buttock	Spongiotic dermatitis	Topical corticosteroid	Topical steroid applied for 1 month with little success
Bojanic[7] et al.	65/F	Right side of the lower limb	Interface dermatitis	Topical corticosteroid and oral corticosteroid	After applying topical steroid for 1 month with little success, complete resolution after oral corticosteroid for 1 week and relapse was noted
Our case	2/F	Left side of the chest, abdomen, back and lower limb	Interface dermatitis	Topical corticosteroid and oral corticosteroid	After applying topical steroid for 4 months with little success, complete resolution after oral corticosteroid for 10 days and relapse was noted

relapsing pattern. Therefore, this case was diagnosed as acquired Blaschko dermatitis rather than lichen striatus.

Most cases of acquired dermatoses that present in a Blaschkolinear pattern in children are common dermatoses that are usually distributed in a random manner. They show the same clinical and histological pattern as the lesions of the common presentation. This occurs in psoriasis, lichen planus, or atopic dermatitis. A few acquired dermatoses always present in a linear pattern with their own clinical and histological characteristics, such as lichen striatus or Moulin disease[8]. Acquired Blaschko dermatitis is a rare disease that has only been described in adults[1-7].

Several cases with similar clinical and histologic features, but with generalized atopic dermatitis or another atopic background were diagnosed as linear atopic dermatitis or mosaic atopic eczema[9,10]. In our case, as in other cases reported as 'acquired Blaschko dermatitis', there was no personal or family history of atopic dermatitis. Some authors suggested that these eruptions may present abortive forms of atopic dermatitis caused by mosaicism with one wild-type and one pathogenic allele in the affected Blaschko line and two wild-type alleles in the remaining tissues[2,8]. But so far the exact relationship still remains uncertain, so the term 'acquired Blaschko dermatitis' would be better in cases without systemic atopy. Definite diagnosis is only possible on the background of pronounced systemic atopy.

Linear lichen planus, affecting 0.24% to 0.62% of all patients with lichen planus, can also show the blaschkolinear pattern[11]. However, clinical histology reveals a lichenoid tissue reaction underlying an acanthotic epidermis with an accentuated granular

layer. Clinical features, such as mucosal involvement, nail changes, or concurrent classic lichen planus-like lesions also help differentiate from acquired Blaschko dermatitis.

Recently, there has been a trend to include acquired Blaschko dermatitis as an adult-onset variation of lichen striatus, as lichen striatus exhibits many variable chameleon-like features clinically and histologically[12,13]. But despite some overlaping conditions, they can be easily differentiated. Lichen striatus is a more common condition and generally affects children with an average age of 3 years. It consists of small inflammatory papules, rarely vesicles with one or few lines along Blaschko's lines unilaterally on a limb. The lesions spontaneously disappear within a few months and usually do not relapse. Non-specific spongiotic changes can be observed occasionally, but focal, band-like lymphocytic infiltration in the dermis is the most characteristic finding[12-15]. In contrast, adult Blaschkitis or acquired Blaschko dermatitis is a rare condition, generally affecting adults with a mean age of 40 years and consisting of papules and vesicles with multiple lines along Blaschko's lines uni- or bilaterally, especially on the trunk. The prognosis has a rapid resolution, less than 2 months and usually relapses are without sequelae. Histologically, a spongiotic pattern is characteristic[8]. In our case, clinical and histological features, except the unusual young age, fit acquired Blaschko dermatitis. Further cases should be collected to understand the nature of this new entity as a separate term, which should be differentiated with other dermatoses, especially lichen striatus.

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