



A Case of Linear Exacerbation of Atopic Dermatitis with Secondary Prurigo Nodularis

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Inflammatory acquired Blaschko-linear dermatoses (IABLD) are a continuous concept involving diseases such as lichen striatus, blaschkitis, and atopic dermatitis. However, atopic dermatitis that showed increase in severity along Blaschko lines is rarely reported on its own. Herein, we report a rare case of atopic dermatitis with secondary prurigo nodularis along Blaschko lines, which may be valuable in broadening the concept of IABLD. A 28-year-old male presented with multiple, pruritic, brownish nodules on the left lower extremity along Blaschko lines for 3 to 4 years. The patient had atopic dermatitis since childhood. Histopathologic findings revealed compact orthohyperkeratosis, hypergranulosis, spongiosis, and irregular acanthosis in the epidermis. Fibrosis with vertically arranged collagen fibers and perivascular lymphohistiocytic infiltration were shown in the upper dermis. We diagnosed the case as secondary prurigo nodularis along Blaschko lines, accompanied by the preceding atopic dermatitis. We hypothesized that the patient's underlying atopic dermatitis increased in severity along Blaschko lines, and prurigo nodularis occurred due to frequent scratching. The lesions improved with topical methylprednisolone cream, oral antihistamines and intralesional triamcinolone injection.

Keywords: Atopic dermatitis, Blaschko lines, Inflammatory acquired Blaschko-linear dermatoses, Prurigo nodularis

INTRODUCTION

Inflammatory acquired blaschko-linear dermatoses (IABLD) are a continuous concept involving various diseases rather than individual diseases. This can be classified into subgroups of lichenoid, spongiotic, and psoriasiform dermatitis based on clinical appearance, past history, and histologic findings¹. We report a case of aggravation of atopic dermatitis along Blaschko lines, which showed secondary change of prurigo nodularis. Progression of our case may be helpful in broadening the concept of IABLD. Also, there have been only a few cases reported as linear atopic dermatitis². We hope that our case can facilitate in understanding the mechanism of linear atopic dermatitis.

CASE REPORT

A 28-year-old male presented with multiple, pruritic, brownish nodules and plaques on the left lower extremity for 3 to 4 years. The lesions were distributed along Blaschko lines, from the posterior heel to the buttock, and they had become elevated and hardened over the years (Fig. 1). We received the patient's consent form about publishing all photographic materials. The patient had atopic dermatitis and had been treated with topical desonide lotion and oral antihistamines intermittently. The general physical examination was not remarkable other than the above-described skin lesions. According to the clinical manifestation, prurigo nodularis, blaschkitis, lichen striatus (LS) were assumed to be diagnosed. On laboratory examination, immunoglobulin E was checked as 205.9 IU/ml (normal range:0~158 IU/ml), multiple allergen simultaneous



test showed class 6 in cats, class 4 in dogs and *Dermatophagoides pteronyssinus*, and class 3 in *Dermatophagoides farinae*. Punch biopsy from a nodule on the left leg was performed (Fig. 2A). The epidermis revealed compact orthohyperkeratosis, hy-

pergranulosis, irregular acanthosis, and spongiosis (Fig. 2B). Fibrosis with vertically arranged collagen fibers (Fig. 2C) and perivascular lymphohistiocytic infiltration were shown in the upper dermis (Fig. 2D). These pathologic findings of chronic



Fig. 1. (A) The general physical examination of the patient was not remarkable except for the Blaschko-linear lesions on the left lower extremity. (B) The general physical examination of the patient was not remarkable except for the Blaschko-linear lesions on the left lower extremity. (C) Multiple, pruritic, brownish nodules and plaques on the left lower extremity for 3 to 4 years.

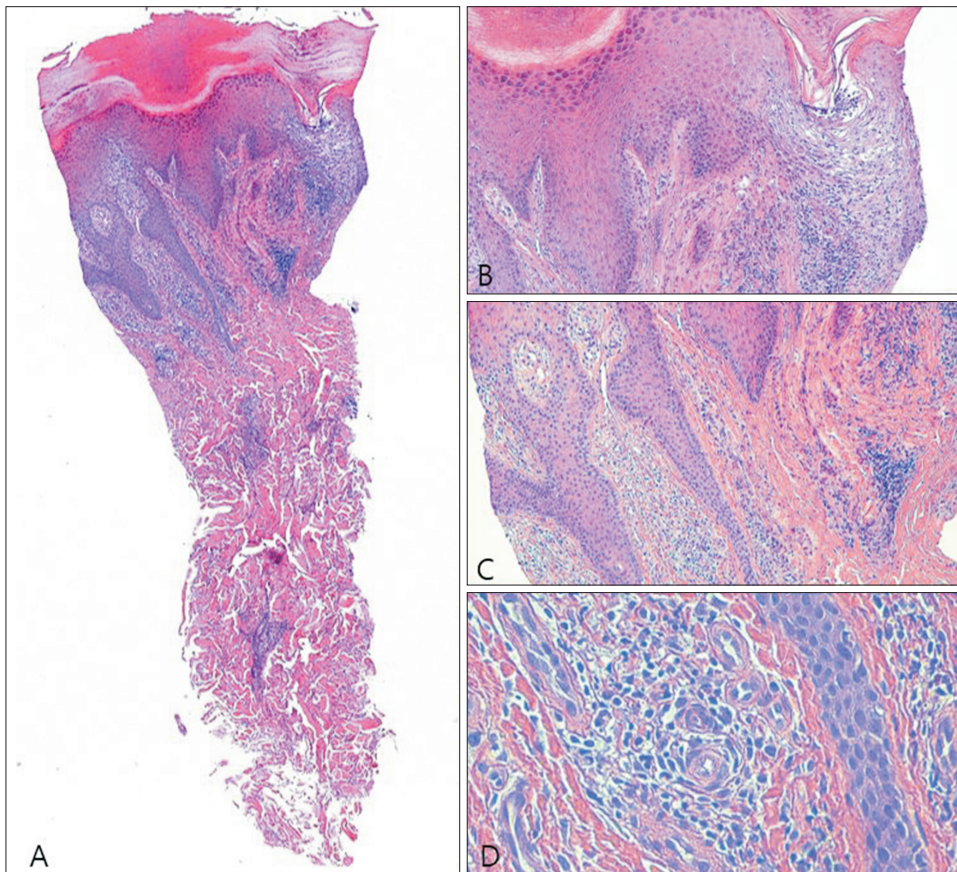


Fig. 2. (A) Photomicrograph from the lower leg portion of the lesion (H&E, $\times 40$). (B) Hyperkeratosis, orthokeratosis, hypergranulosis, irregular acanthosis, and spongiosis in the epidermis (H&E, $\times 100$). (C) Fibrosis with vertically arranged collagen fibers (H&E, $\times 100$). (D) Perivascular lymphohistiocytic infiltration in upper dermis (H&E, $\times 400$).

eczematous dermatitis were consistent with prurigo nodularis and chronic atopic dermatitis. In addition, the pathologic features could explain the clinical hyperkeratotic nodular lesion of the patient. Based on the clinical, laboratory and pathologic findings, we diagnosed this case as secondary prurigo nodularis along Blaschko lines, accompanied by preceding atopic dermatitis. The patient is undergoing follow up with topical methylprednisolone cream, oral antihistamines and intral-lesional triamcinolone injection with 2 weeks interval. He is showing slight improvement with treatment.

DISCUSSION

Taieb et al.³ suggested that LS and blaschkitis exist on a spectrum of Blaschko-linear acquired inflammatory skin eruptions. Keegan et al.¹ regarded LS, blaschkitis, and atopic dermatitis as continuous diseases that are classified within the spectrum of IABLD rather than as individual diseases. At first, our case was considered as LS or blaschkitis because of the typical distribution of the cutaneous lesion. However, LS is characterized by its clinical feature of flat-topped papules with smooth surface, early age onset, absence of symptom and spontaneous resolution. Conversely, blaschkitis commonly has papulovesicular morphology with extensive involvement, late age onset, severe pruritus and frequent recurrences. Our case did not belong to these characteristics of the two diseases. Rather, the hyperkeratotic firm nodules with a chronic course implied chronic dermatitis with a secondary hyperkeratotic change.

Considering the patient's history of atopic dermatitis, we hypothesized that atopic dermatitis increased in severity along Blaschko lines. Prurigo nodularis was presumed to be a secondary change from frequent scratching caused by itching of atopic dermatitis. Hladik et al.² reported that clinical and pathologic features of linear atopic dermatitis and blaschkitis are very similar, and we could differentiate them with past history of atopic dermatitis and typical distribution of atopic dermatitis. Hladik et al.² and López-Cedeño et al.⁴ reported total four cases of linear atopic dermatitis which showed increase in severity along the lines of Blaschko, which initially showed similar clinical course shown in our case. While segmental mosaicism is common in monogenic skin diseases, linear distribution is a rare manifestation in polygenic diseases such as atopic dermatitis. Blaschko line arrangement may reflect

clonal loss of heterozygosity in a subpopulation of early skin stem cells. Van Gysel and Grimalt⁵ concluded that a linear manifestation of atopic dermatitis shows the clonal outgrowth of cells harboring a postzygotic mutation which increased the predisposition to atopic dermatitis.

Previous reported cases of linear atopic dermatitis and our case both shared clinical features of past history of atopic dermatitis and pathologic features of spongiotic dermatitis. However, our case showed a secondary hyperkeratotic change with a pathologic sign of prurigo nodularis, which were not observed in previous reports. The association between prurigo nodularis and atopic dermatitis is well known. Miyachi et al.⁶ elucidated the relationship between prurigo nodularis and atopic dermatitis. He speculated that atopic dermatitis patients are sensitive to insect bites or other scratch-provoking factors which lead to intensive scratching resulting in verrucous appearance of nodular lesions. Napolitano et al.⁷ considered prurigo nodularis as a clinical pattern of adult atopic dermatitis. Considering the clinical manifestation of prurigo nodularis which can be explained by aggravation of atopic dermatitis along Blaschko lines and pathologic findings showing both spongiotic dermatitis and chronic eczematous dermatitis, our case was compatible with atopic-type prurigo nodularis clinicopathologically.

This case is worth reporting in two points. First of all, it shared features of both spongiotic dermatitis and chronic eczematous dermatitis, which may broaden the concept of continuous spectrum of IABLD. In other words, having the clinicopathologic characteristics of both atopic dermatitis and prurigo nodularis in an individual support the idea that IABLD consists of diseases on a continuous spectrum. Secondly, atopic dermatitis along Blaschko lines is rarely reported on its own. We hope that this additional case of Blaschko-linear atopic dermatitis supports other case reports which suggest that clonal loss of heterozygosity affects atopic dermatitis.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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