Linear lesions on the arm of a child: a diagnostic challenge

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Abstract

There are a number of conditions that follow the lines of Blaschko. Linear discoid lupus erythematosus is a rare variant of chronic lupus erythematosus with less than 20 cases reported in children. It can be misdiagnosed as lichen striatus or linear morphea. We describe a 15-year-old boy with a confirmed histologic diagnosis of linear chronic discoid lupus erythematosus following the lines of Blaschko, with no signs of systemic involvement.

Keywords: lupus erythematosus, discoid, mosaicism, child

Introduction

Discoid lupus erythematosus is a chronic cutaneous lupus variant [1]. It was first described by Moritz Kaposi in 1872 [2]. In 1988, Abe et al. [3] reported a linear configuration as one of the uncommon variants of discoid lupus erythematosus. However, it has rarely been documented in children, with less than 2% of all cases appearing before the age of 10 years [4]. Herein, we present the case of a 15-year-old boy with an 8-year history of linear chronic discoid lupus erythematosus following the lines of Blaschko.

Case Synopsis

A 15-year-old boy presented with an 8-year history of asymptomatic linear lesions on the left upper extremity. He had been previously diagnosed with lichen striatus and he had been applying desonide cream 0.05% and mometasone cream 0.1% almost daily for the last 8 years, without any improvement.

The physical examination revealed atrophic, pigmented plaques with violaceous borders and central hypopigmentation following a linear pattern, extending from the mid left forearm to the arm and shoulder. Apparently normal skin was preserved between the lesions (**Figure 1**). There were no motor or sensation disturbances. Skin atrophy related to chronic topical steroids, linear morphea, linear atrophic lichen planus, and chronic discoid linear erythematous lupus were considered as potential diagnoses and a skin biopsy was taken.

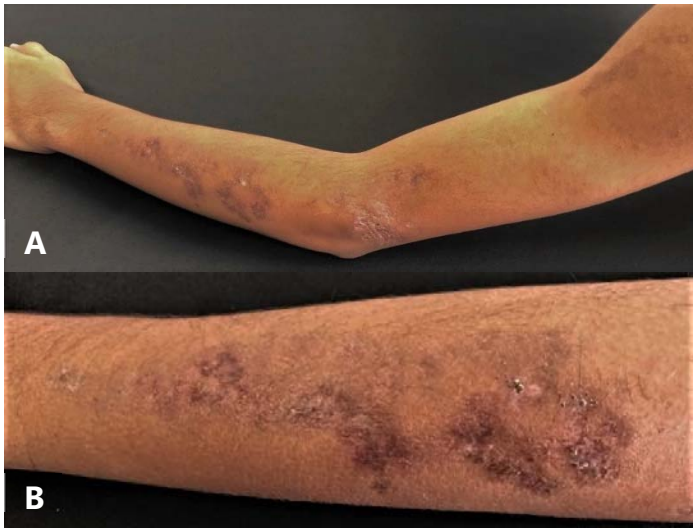

Figure 1. ***A)*** *Blaschkoid pigmented plaques extending from the left forearm to the shoulder.* ***B)*** *Discoid plaque with central atrophy and peripheral scales.*

The skin biopsy revealed atrophic epidermis, hyperkeratosis, and a lympho-histiocytic multifocal band infiltrate, with hydropic degeneration of the basal layer accompanied by multiple necrotic keratinocytes, pigmentary incontinence, and erythrocyte extravasation. The dermis showed an intense lympho-histioplasmocytic infiltrate around the adnexal structures and vessels. Increased interstitial mucin deposits were confirmed with the Alcian blue stain. Periodic acid-Schiff staining highlighted the basement membrane thickening

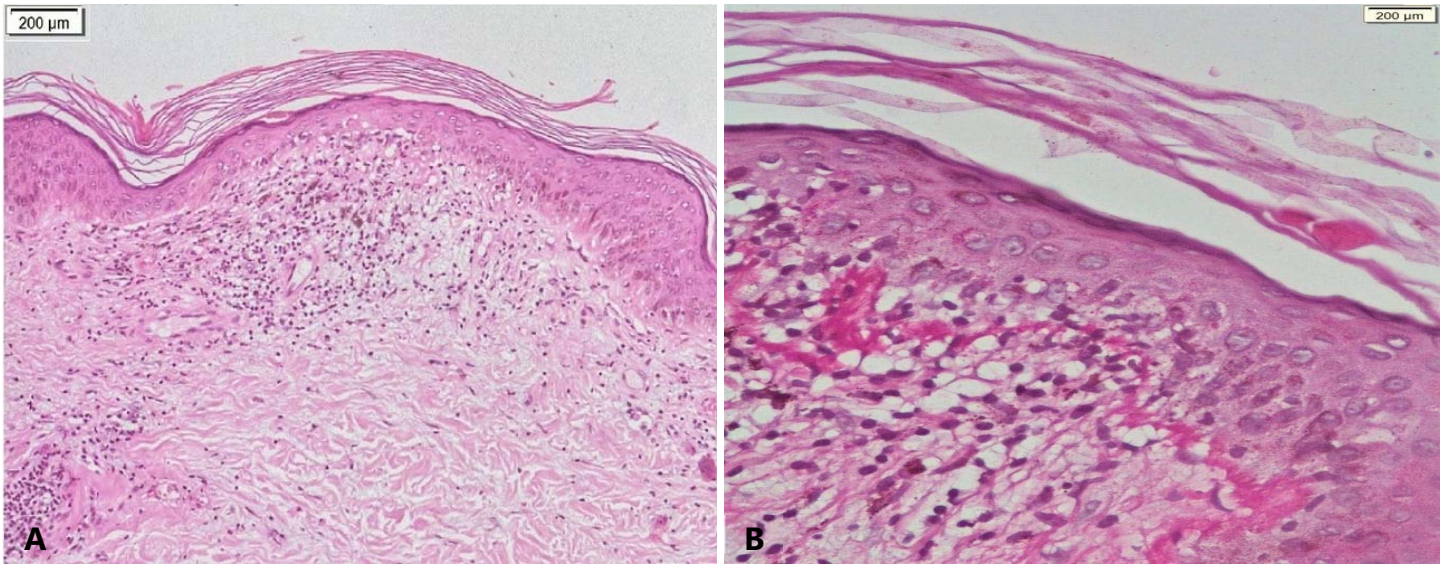

Figure 2. ***A)*** *Skin biopsy of the pigmented atrophic plaque on the left arm shows hyperkeratotic and atrophic epidermis, basement membrane thickening and hydropic degeneration. In the dermis, mucin deposits and a chronic inflammatory infiltrate.* ***B)*** *A thick basement membrane highlighted with the periodic acid-Schiff stain.*

(**Figure 2**). With those findings a diagnosis of discoid lupus erythematosus was confirmed.

A 200mg daily dose of oral hydroxychloroquine and topical tacrolimus ointment were prescribed, leading to improvement. Blood count and kidney function panel showed no abnormalities, anti-nuclear antibody and anti-DNA antibodies were negative; complement levels were normal. There were no signs of systemic involvement.

Case Discussion

Discoid lupus erythematosus has rarely been reported in children [5]. Until 2017, only 19 cases of linear discoid lupus erythematosus (LDLE) have been described in children [6].

The pathogenesis remains unknown. Immunological abnormalities play an important role in the actual understanding of the disease [7]. Abe et al. described the linear blaschkoid variants of cutaneous lupus [3]. Blaschkoid lesions reflect an abnormal cell migration of embryonic skin tissue during embryogenesis related to post-zygotic mutation in keratinocytes [1]. The cells arising from these mosaicisms express neoantigens eliciting an immune response [8]. Other inflammatory diseases such as lichen striatus, lichen planus, and psoriasis may follow a Blaschkoid pattern [1].

The majority of the LDLE cases begin during the first decade of life [9], without sex or ethnic preference [7]. Clinical manifestations include the classic features of discoid lupus erythematosus with well-demarcated, erythematous plaques that are devoid of hair, centrally hypopigmented, with characteristic atrophy, scarring, and telangiectasia. Normal-appearing skin is noted between lesions, following a Blaschkoid pattern. The most common affected areas are the face, neck, and trunk [3].

Anti-nuclear antibodies are negative or slightly positive. Typically there is no photosensitivity or progression to systemic lupus erythematosus [10]. Histologic features show epidermal atrophy, follicular plugging, hyperkeratosis, basement membrane thickening, basal layer hydropic degeneration, telangiectasia, pigmentary incontinence, superficial, medium, and deep perivascular and periadnexal chronic inflammatory infiltrate, and dermal mucin deposit [5].

Segmental inflammatory skin diseases are generally known to have a chronic course and often require long-term treatment. Therapeutic options of LDLE include hydroxychloroquine, topical corticosteroids, tacrolimus, and sun avoidance, leading to significantly improvement or even a complete resolution of the plaques [10].

Our case was initially misdiagnosed as linear lichen striatus and was chronically treated with topical steroids with subsequent skin atrophy, but owing to a lack of response to treatment a skin biopsy was taken and it confirmed the diagnosis of LDLE, an unusual variant of the disease in a child.

Conclusion

To our knowledge, this is one of a few cases of LDLE in a child. Our patient had all the clinical features and histological findings described for the disease. The linear variant of chronic cutaneous lupus erythematosus can be easily confused with other Blaschkoid diseases and it represents a diagnostic and therapeutic challenge. We prompt the scientific community to be aware of this unusual variant of cutaneous lupus in the pediatric population, even as a limited skin disease it would be important to rule out systemic involvement.

Potential conflicts of interest

The authors declare no conflicts of interests.

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