

A Rare Case of Tumid Lupus Erythematosus with Unilateral Linear Distribution in a Young Child

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1. Abstract

Tumid Lupus Erythematosus (TLE) as a rare variant of Cutaneous Lupus Erythematosus (CLE) is characterized by edematous, urticarial-like annular papules and plaques. TLE has similar histopathologic findings to CLE such as periadnexal lymphocytic infiltration and interstitial mucin deposition. Although TLE develops on sun-exposed areas at any age, it is rarely distributed along the Blaschko lines and develops in infancy and childhood. Unlike CLE, skin lesion of TLE heals without leaving scarring or dyspigmentation. Here we report a rare case of unilateral linear TLE in a four-year-old girl which was improved by intralesional corticosteroid injection and oral antimalarial drug with leaving post-inflammatory hyperpigmentation.

2. Case Report

Tumid lupus erythematosus (TLE) is a subtype of chronic cutaneous lupus erythematosus (CLE). It was first reported by Gougerot and Burnier [1]. The word "tumid" derives from tumidus, which means "swollen" in Latin. As the name suggests, clinical manifestation of TLE can be characterized by edematous, wheal-like erythema. Like CLE, TLE has a photosensitive nature. Its skin lesions tend to develop symmetrically at sun-exposed sites. These lesions showing linear distribution are unusual. Only three cases have been reported so far. TLE affects both gender nearly equally. Even though it can occur in any ages, its occurrence in childhood is extremely rare. Treatment of TLE varies by the extent of the disease. TLE lesions tend to heal without leaving scarring or pigmentary alteration. Here we present a rare case of unilateral linear TLE developed in a four-year-old girl. Her TLE showed improvement, although she had post-inflammatory hyperpigmentation.

A 4-year-old girl presented with asymptomatic wheal-like erythematous to brownish plaques arranged linearly on the right side of her upper back, up-

per chest, and same side of forearm and hand for one year (Figure 1A-1B). Before visiting our clinic, she had been treated with topical tacrolimus without improvement. Her parents denied sun exposure of truncal lesions because she never took off her clothes when she was outside. Medical history was unremarkable. Laboratory findings showed positive results for anti-nuclear antibody (1:320), but negative results for anti-double-stranded DNA and anti-Scl-70 (anti-topoisomerase I). Histopathologic findings of biopsied skin specimen from forearm revealed perivascular and periadnexal lymphoplasmocytic infiltration and widening of spaces between dermal collagen bundles without significant changes in epidermis (Figure 2A-B). Alcian blue stains revealed interstitial mucin deposition on the dermis (Figure 2C). Direct immunofluorescence showed linear deposition of immunoglobulin M on basement membrane. Based on these clinical and histopathologic findings, the case was diagnosed as TLE. Although the lesions were treated with intralesional triamcinolone injection (2.5mg/ml) every two weeks and a topical steroid for five months, improvement was not remarkable. In the meantime, new lesions appeared. Due to insufficient effect of those treatments, we added oral hydroxychloroquine (50 mg daily) after baseline ophthalmologic examination. After four months of treatment, lesions gradually improved without developing new lesions. However, she had a brownish hyperpigmentation (Figure 1C).

TLE is diagnosed based on clinical and histopathologic findings to differentiate it from other variants of LE, polymorphous light eruption, Jessner's lymphocytic infiltration of the skin, reticular erythematous mucinosis, and pseudolymphoma, [2] although its exact diagnostic criteria have not been established yet. TLE is a subtype of CLE. TLE and CLE share some characteristic histopathologic findings, including perivascular and periadnexal infiltration of lymphocytes and plasma cells with interstitial mucin deposition as shown in our case (Figure 2A-2C). Lack of involvement of the epidermis is a usual finding of TLE as in our case (Figure



Figure 1: (A) Erythematous to brownish edematous plaques were linearly arranged along the inner forearm (B) Erythematous plaque on the same side of upper back (C) Lesions improved with brown discoloration after administration of oral hydroxychloroquine

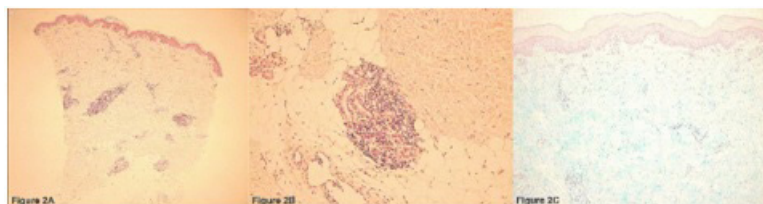


Figure 2: (A) There was no significant pathologic change in epidermis. (Hematoxylin & Eosin [H&E], original magnification ×40). (B) Lymphoplasmacytic infiltration around the skin appendage (H&E, original magnification ×200). (C) Alcian blue stain showed mucin deposition between dermal collagen bundles (original magnification ×100).

2A), although there might be minor epidermal changes such as vacuolar degeneration in the basal layer or epidermal atrophy in some cases [2]. Therefore, our case could be compatible with TLE.

Clinical findings of smooth-surfaced, erythematous edematous papules and plaques in our case (Figure 1A and 1B) were in accord with TLE [1], supporting that diagnosis. However, there were unique findings in our case. Ultraviolet light is a well-known triggering factor of TLE, and most of the skin lesions develop on sun-exposed areas [1]. However, the skin lesions in our case developed on non-exposed areas such as the inner wrist, forearm, and arm, anterior chest, and back (Figure 1A and 1B). In addition, the lesions were distributed linearly along lines of Blaschko (Figure 1A and 1B). Blaschko-linear distribution is a very rare clinical presentation of TLE. Only very few cases have been reported in English written literature [3-5]. These cases along with our case are summarized in (Table 1). Including our case, two patients had skin lesions on the arm and same side of the back [5]. Although the other two patients had lesions on their foreheads, they denied sun-exposure. These findings suggest a po-

tential role of somatic mosaicism in its pathogenesis. Differing from other three cases, only our case developed in childhood. TLE usually occurs in the fourth decade [2]. Childhood TLE in itself is exceedingly rare with only a few reported cases in the literature [6].

TLE has been treated with topical corticosteroids or intralesional corticosteroid injection, although these treatments were insufficient to control skin lesions in our patient. The outcome of topical therapy showed various success rates, ranging from 12 to 80%. [7,8] When TLE is refractory to topical therapies, systemic antimalarial agents for four to six weeks may show a good response [2]. Treatment with hydroxychloroquine in our case also brought significant flattening of lesion without developing new lesions. However, complete resolution by hydroxychloroquine for four months was not examined (Figure 1C). Lower doses (approximately 2 mg/kg per day) than recommendation (up to 6 mg/kg per day) [2] might be responsible for the less effective outcome. TLE usually spontaneously disappears after several years, although it may last for 30 years [2]. TLE is known to heal without scarring, atrophy, or dyspigmentation. However, our patient had hyperpigmentation after her lesions improved. Consid-

Table 1: Characteristic findings of linear tumid lupus erythematosus: reported cases vs this case

No.	Age	Gender	Location	Photosensitivity	Immunofluorescence test	Laboratory tests	Phototest	
1	32	F	Forehead	No history of Sun exposure	IF : IgM	Normal	Not performed	Bouzit et al. ³
2	43	M	Forehead	No history of Sun exposure	IF : IgM, C3	Normal	Not performed	Hinz et al. ⁴
3	27	M	Arm	No history of photosensitivity	IF : C3	Normal	Not performed	Pacheco et al. ⁵
			Back					
4	4	F	Arm	Unclear	IF : IgM	ANA(+)	Not performed	This case
			Back					

ering that there are reports about TLE with hypopigmentation in black patients [9] and hyperpigmentation in a Korean patient [10], TLE may show pigmentary alteration depending on their skin color.

Our case showed Blaschko-linear distribution on non-exposed area, onset in early childhood, and improvement leaving post-inflammatory hyperpigmentation as rare findings in TLE.

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