Linear cutaneous lupus erythematosus/discoid lupus erythematosus in an adult

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Linear cutaneous lupus erythematosus (LCLE) is rare. The LCLE was proposed by Abe *et al.* [1] for discoid lupus erythematosus (DLE) with a linear configuration in 1988. It occurs mainly in children and young adults. Lesions appear as linear unilateral erythematous plaques following the Blaschko lines and are observed most frequently on the face, although the neck, trunk and extremities may also be affected [2]. Neither photosensitivity nor progression to systemic LE is observed [3]. To our knowledge, only 9 cases in adults have been reported in the literature. We report here the tenth case in an adult.

A 32-year-old woman presented with a 1-month history of slightly pruritic plaque on her left jaw and neck. These lesions first appeared on her jaw and spread to the left side of the neck in a linear arrangement. There was no history of trauma around the lesions, nor of intensive exposure to sunlight. The patient reported no fever, chills or photosensitivity. Her past medical history was unremarkable and there was no similar condition reported in her family members.

On physical examination, she had linear, slightly atrophic, reddish-brown plaque on her left jaw and neck (Figure 1 A) which did not follow the lines of Blaschko strictly (Figure 1 B). Laboratory investigations revealed positive antinuclear antibodies (ANA) with a titer of 1 : 80 (normal: < 1 : 40) and a granular fluorescence pattern, decreased complement C4 of 0.13 g/l (normal: 0.17–4 g/l) and decreased leukocyte of 3.62 × 10^9/l (normal: 4–10 × 10^9/l). Liver function tests, blood urea, creatinine and urine analysis were within normal limits. Antibodies against double-stranded DNA and Sm were negative. Skin biopsy from the lesion on the jaw revealed epidermal atrophy, follicular plugging and liquefaction degeneration in the basal layer of the epidermis. In the underlying dermis, inflammatory infiltrates of lymphocytes around the adnexal and vascular structures were observed (Figures 2 A, B). Alcian blue staining did not reveal obvious mucin depositions in the dermis (Figure 2 C). Direct immunofluorescence was not performed. Based on the clinical, laboratory, and histological findings, a diagnosis of LCLE was made. The patient

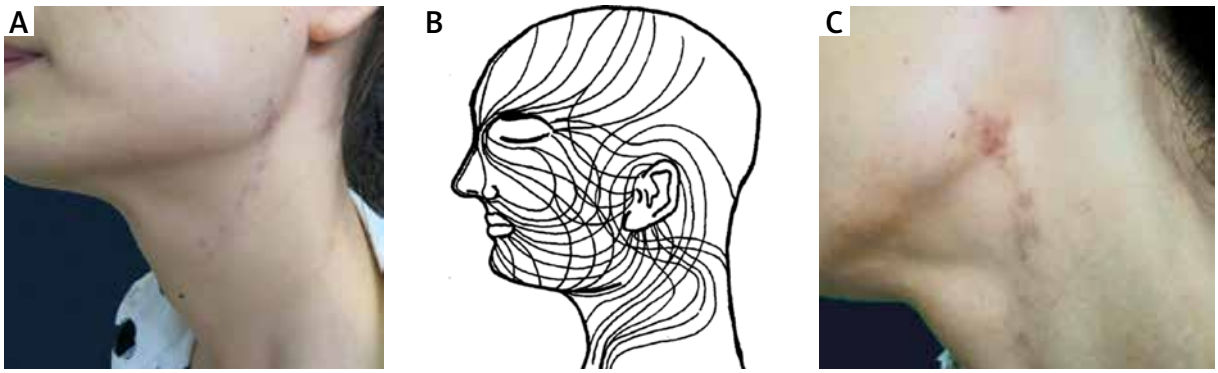

Figure 1. A – The linear, slightly atrophic, reddish-brown plaque on the left jaw and neck following Blaschko lines generally; **B** – distribution of the Blaschko lines on the plaque on the head and neck (pattern elaborated by Happle and Assim [11]); **C** – the lesions stopped spreading and became darker after treatment

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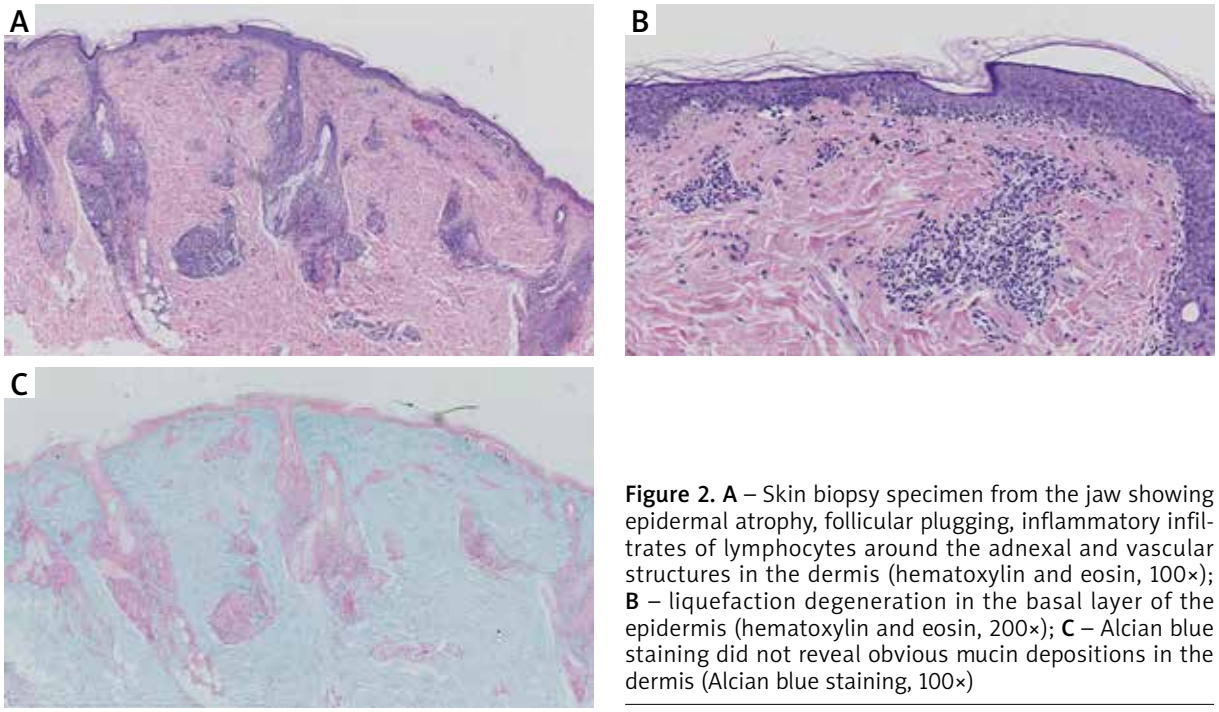

Figure 2. A – Skin biopsy specimen from the jaw showing epidermal atrophy, follicular plugging, inflammatory infiltrates of lymphocytes around the adnexal and vascular structures in the dermis (hematoxylin and eosin, 100×); **B** – liquefaction degeneration in the basal layer of the epidermis (hematoxylin and eosin, 200×); **C** – Alcian blue staining did not reveal obvious mucin depositions in the dermis (Alcian blue staining, 100×)

was injected with 1 ml of a betamethasone injection and was also treated with desonide cream and tacrolimus 0.1% ointment for 1 month, the lesions stopped spreading and became darker (Figure 1 C).

The LCLE is a highly unusual variation of discoid lupus erythematosus (DLE) [4]. In 1998, when reporting 2 cases of linear childhood CLE following the lines of Blaschko, Abe *et al*. [1] proposed the term 'linear cutaneous lupus erythematosus (LCLE)' for DLE with a linear configuration. Nevertheless, other subtypes of LE that follow a linear pattern have already been described, including deep LE, subacute LE and tumid LE. The LCLE occurs mainly in children and young adults, with a similar incidence in both sexes, without ethnic preference. Lesions appear as linear unilateral erythematous plaques and are observed most frequently on the face, although the neck, trunk and extremities may also be affected [2]. Most commonly, anti-nuclear antibodies are negative or slightly positive. Neither photosensitivity nor progression to systemic LE is observed. The histological findings in LCLE include hyperkeratosis, atrophy of the epidermis, hydropic degeneration of the basal cell layer in the epidermis, perivascular and periadnexal dense infiltrates of lymphocytes, and mucinous deposition in the dermis. These findings are compatible with DLE [3]. Differential diagnosis must be established with other conditions with Blaschko linear distribution such as linear lichen planus, lichen striatus, linear granuloma annulare, linear psoriasis, morphea or inflammatory linear verrucous epidermal nevus [2]. The treatment of linear cutaneous LE includes potent topical corticosteroids or calcineurin inhibitors. In more widespread disease, systemic corticosteroids, methotrexate, chloroquine or hydroxychloroquine or other immunosuppressants may be necessary [5].

In most cases the age of onset is younger than 15 years. To our knowledge, there have been only nine previous confirmed cases of LCLE/DLE in adults, which all followed Blaschko lines [2, 5–11] (Table 1). The incidence in both sexes seemed similar. The lesions occurred mainly on the face. Testing for antinuclear antibodies was positive in 2 of the cases. Laboratory tests of blood cell count were all normal. The treatment mainly included hydroxychloroquine, topical steroid and sunblock, which led to the lesions being stable or significantly improved, even complete resolution. Our patient represents the 10th case and this is the first reported case in China in the English literature. It is unique that the lesions in our case were slightly different from the typical line patterns of Blaschko (Figure 1 B) which was elaborated by Happle and Assim [12] on the head and neck in 2001. There may be some variation among different ethnic groups or different individuals of the same race. In our case ANA was positive along with decreased complement C4 and leukocyte which had never presented in the nine previous cases. After treatment with 1 ml of betamethasone injection, topical corticosteroids and tacrolimus 0.1% ointment for 1 month, the lesions stopped spreading and became darker.

Here we report the 10th case of LCLE/DLE occurring in an adult. The clinical presentation of LCLE in children and adults seems to be similar, with a similar incidence in both sexes, essentially involving the face. The diffe-

Table 1. Summary of cases of linear cutaneous lupus erythematosus/discoid lupus erythematosus in adults

Author	Age of onset	Gender	Distribution (site)	Antibodies	DIF	Blood cell counts	TX	CT	Efficacy
Bouzit *et al.* (1999) [5]	29	F	Forehead	(–)	+	Normal	HCQ + TS	2 months	CR
Abe *et al.* (2000) [6]	23	M	Cheek	(–)	+	Normal	DDS	28 days	SI
Sàbat *et al.* (2006) [7]	19	F	Nose and epicanthal area	ANA(+)	ND	Normal	HCQ	1 year	Stable
Gaitanis *et al.* (2009) [8]	21	M	Supraorbital area, infraorbitally and mouth angle	(–)	+	Normal	HCQ + TS + SB	2 years	CR
Thind *et al.* (2009) [9]	37	F	Mouth, chin	(–)	+	Normal	TS + SB	NM	Stable
Kim *et al.* (2010) [10]	33	M	Forehead, nose	(–)	ND	Normal	HCQ + TS	NM	SI
Alcántara-González *et al.* (2011) [2]	64	M	Neck	(–)	ND	Normal	TS	2 months	CR
Alcántara-González *et al.* (2011) [2]	45	M	Head	(–)	ND	Normal	TS	3 months	SI
Verma *et al.* (2012) [4]	32	F	Face, upper back and extremities	ANA(+)	ND	Normal	TS + MTX + SB	NM	SI
Our case (2015)	32	F	Jaw and neck	ANA(+), CC4(+)	ND	Decreased leukocyte	TS + SS + TT	1 month	MI

F – female, M – male, ANA – antinuclear antibody, CC4 – complement C4, DIF – direct immunofluorescence, ND – not done, TX – treatment, HCQ – hydroxychloroquine, TS – topical steroid, DDS – diaminodiphenylsulfone, SB – sunblock, MTX – methotrexate, SS – systemic steroid, TT – topical tacrolimus, CT – course of treatment, NM – not mentioned, CR – complete resolution, SI – significantly improved, MI – mildly improved.

rential diagnosis of such linear lesions includes lichen striatus and linear lichen planus. In patients presenting with a linear inflammatory eruption on the face, a skin biopsy should be performed to rule out a linear form of cutaneous lupus erythematosus.

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Conflict of interest

The authors declare no conflict of interest.

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