Linear Lupus Erythematosus Profundus:  
−Report of a Case and Review of the Literature−

Jeng-Wei Tjiu Chia-Yu Chu

Lupus erythematosus profundus (LEP) is a rare entity characterized by erythematous or flesh-colored deep nodules and plaques, with a predilection for the buttocks, arms, thighs and face. There are only 4 reported cases of linear variant of LEP in the English literature. We herein report another case of linear LEP. The patient was a 26-year-old female who presented with a linear indurated erythematous plaque along the left arm to her left palm. Skin biopsy taken from the lesion showed lymphocytic panniculitis with hyaline fat necrosis. Direct immunofluorescence study revealed granular depositions of IgM, C3 and C1q along the basement membrane. She was treated with oral hydroxychloroquine, topical corticosteroids and topical tacrolimus ointment. Control of the disease activity was achieved after treatment for nine months. (Dermatol Sinica 22 : 221-226, 2004)

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From the Department of Dermatology, National Taiwan University Hospital, Taipei, Taiwan
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Reprint requests: Chia-Yu Chu, M.D., Department of Dermatology, National Taiwan University Hospital, No. 7, Chung-Shan South Road, Taipei 100, Taiwan
TEL: 886-2-23562141 FAX: 886-2-23934177
INTRODUCTION

Lupus erythematosus profundus (LEP) is characterized by erythematous or flesh-colored deep nodules and plaques, with a predilection for the buttocks, arms, thighs and face.\(^1\) LEP occurs mostly in middle-aged women and is generally accompanied by other clinical or serologic evidence of lupus erythematosus (LE). A linear variant of LEP had been rarely reported (only 4 cases had been reported) in the English literature.\(^2\)\(^-\)\(^5\) We report another case of linear LEP (LLEP) in a young female.

CASE REPORT

A 26-year-old female presented with a 16-year-history of a linear skin lesion from her left upper arm extended to the left forearm and left palm gradually. She had no past history of any congenital or systemic disease. There was no family history of any autoimmune rheumatologic disease. She also denied an operation or trauma history. Some asymptomatic, erythematous to violaceous, pea-sized to coin-sized nodules and plaques developed on the inner aspect of the left upper arm when she was 10 years old. The nodules and plaques increased in number and gradually extended to the flexor side of the left forearm and to the left palm. The lesions became confluent to a long linear, indurated, erythematous to violaceous plaque from her left upper arm to left palm one year after she noticed it. No alopecia or photosensitivity was associated with the skin lesions. She had been treated with topical steroid ointment for a few months prior to her visit to our hospital, but the topical therapy was ineffective.

Physical examination revealed a linear indurated erythematous to violaceous plaque extending from the left upper arm and descending to the flexor side of the left forearm and the ulnar side of the left palm (Fig. 1). The overlying skin was slightly atrophic, depressed and covered with a few adherent scales. Laboratory studies showed that the blood cell counts, erythrocyte sedimentation rate, urinalysis, anti-DNA, anti-ENA, anti-nuclear antibody, and the complement levels were all within normal lim-
its. A biopsy taken from the lesion on the upper arm showed focal hydropic degeneration of the epidermal basal layer, thickening of the basement membrane and a predominantly lymphocytic infiltrate with plasma cells arranged around appendages and blood vessels in the deep dermis (Fig. 2A). Lymphoplasmacytic infiltration around the fat lobules with fibrous septal thickening and mild hyalinized change was found (Fig. 2B, 2C). Special stains of alcian blue demonstrated the presence of mucin in the dermis and subcutis. Direct immunofluorescence (DIF) study revealed granular deposits of IgM, C1q (Fig. 3) and C3 at the dermoepidermal junction. Linear LEP was diagnosed. She was treated with hydroxychloroquine 200 mg/day and topical clobetasol propionate ointment at the initial 3 months, but the topical agent was changed to 0.1% tacrolimus ointment due to steroid acne in recent 6 months. The disease activity was under controlled now with absence of erythema, partially resolution of the indurated plaques and no new lesion developed.

DISCUSSION

Lupus erythematosus profundus (LEP) is a chronic panniculitis that affects about 1 to 3% patients with cutaneous lupus erythematosus. LEP usually affects patient aged from 20 to 50 years old with female predominance. A recent study by Ng et al. showed a 2:1 female to male
ratio and younger age of onset (mean age 31.3 years, range 16–51 years). A Japanese study in 1996 also showed similar results with mean age 31.9 years and 94% female in a series of 16 LEP cases. The frequency of involvement of various sites were: face (50%), upper limbs (33%), scalp (25%), lower limbs (17%), and trunk (8.3%). The lesions of LEP are usually subcutaneous nodules or plaques but some patients may develop chronic scarring plaques or transient erythema. Trauma to subcutaneous fat seems to be a precipitating factor for lesions of LEP. LEP was initially thought as a manifestation of DLE, but later LEP was reported to occur roughly frequent in the setting of SLE. Clinical DLE lesions were reported from 13% to 33% among LEP patients, while as SLE were found in 4% to 25% of them in recent series.

The histopathology of LEP comprises lob-
ular panniculitis with lymphocytes, histiocytes and plasma cells infiltrations of subcutaneous fat lobules.\textsuperscript{1, 6-8} Hyaline change of subcutaneous collagen bundles and interstitial lymphocytic or plasma infiltrations could be seen. Lymphid follicle in subcutaneous fat can be observed in some cases.\textsuperscript{1} In more than half of the LEP cases, there are also epidermal and dermal changes of DLE.\textsuperscript{1} These include atrophy of the epidermis, vacuolar change at the dermoepidermal junction, thickened basement membrane, interstitial mucin deposition, and perivascular infiltrations.\textsuperscript{8} DIF studies in lesions of lupus panniculitis usually showed granular depositions of IgM and C3 and linear fibrin deposits at the dermal-epidermal junction and the basement membrane of hair follicles.\textsuperscript{8}

The clinical diagnosis of LLEP is difficult, but the histopathological and DIF findings favored the diagnosis in this patient. The cause for linearity remained unknown and trauma or Kobner phenomenon had been suggested as possible causes.\textsuperscript{2-5} This variant of LEP is extremely rare and to our knowledge, only four cases had been reported. Tada et al. reported a 9-year-old boy having a LLEP along his left lower extremity;\textsuperscript{2} Innocenzi et al. reported a 17-year-old Italian boy with LLEP along the inner aspect of his right arm;\textsuperscript{3} Tamada et al. reported another 19-year-old man with LLEP on the left arm;\textsuperscript{4} Nagai et al. reported a 10-year-old girl with LLEP on the scalp following the lines of Blaschko.\textsuperscript{5} Our case served as the fifth reported case of LLEP. The clinical, histopathological and laboratory features of the five reported cases are summarized in Table I.

The differential diagnosis of patients with LLEP includes linear morphea, linear lichen sclerosus et atrophicus (LSA),\textsuperscript{10} linear DLE,\textsuperscript{11} linear lichen planus (LP),\textsuperscript{12} LE-LP overlap syndrome, inflammatory linear verrucous epidermal nevus (ILVEN),\textsuperscript{13} lichen stiatus\textsuperscript{14} and linear psoriasis. Linear morphea is characterized by sclerotic change of the skin, homogenization of collagen bundles in the dermis without significant interface change. LSA usually have atrophic whitish macules clinically and atrophy of the epidermis with plugs, hydropic degeneration of basal cells histologically.\textsuperscript{10} DLE is characterized by red-purple macules or papules that evolve into larger, well-demarcated, erythematous plaques. With maturation and resolution of DLE lesions, the plaques become centrally hypopigmented, atrophy, scarring and telangiectasia.\textsuperscript{11} DLE did not have firm, sharply defined small nodules and histopathological features of lymphocytic panniculitis with hyalinization changes. In this case, some of the superficial lesions might be DLE lesions, but most of the indurated lesions are LEP, which is evidenced by histopathological findings. Linear LP usually had concurrent classic LP-like lesions that are pruritic, polygonal, violaceous papules and plaques with some nail changes or mucous membrane involvement.\textsuperscript{12} LE-LP overlap syndrome usually has both clinical and histological features of LE and LP. ILVEN lesions are usually scaly and verrucous plaques characterized by psoriasiform proliferation of epidermis in histology.\textsuperscript{15} Lichen striatus had histological features including band-like infiltrations in the papillary dermis, or superficial and deep infiltrations in the dermis, parakeratosis and spongiosis.\textsuperscript{16} Based on the clinical features, histological and DIF findings, all the above mentioned differential diagnoses were unlikely except LLEP.

Topical corticosteroid\textsuperscript{2} and intralesional injection of steroid\textsuperscript{4} were used to treat LLEP in previously reported case. Our case was treated with oral hydroxychloroquine and topical high potency corticosteroid ointment with partial improvement. The topical agent was changed to topical 0.1% tacrolimus ointment due to steroid acne. The beneficial effects of tacrolimus ointment probable rely on its immunomodulatory effects in the inflammatory disorder.

In conclusion, we reported here a rare case of LLEP, which is the fourth reported case in the literature. The clinical diagnosis of LLEP would be difficult and it must be differentiated from other disease entities presenting as localized linear cutaneous eruptions. In our case, the diagnosis of LLEP was established based on the histopathological and DIF findings. The cause

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of LLEP remains unknown but an immune mechanism is certainly involved. With the use of corticosteroid and tacrolimus ointments, the LLEP lesions were clinically improved.

REFERENCES