An Extensive Cutaneous Erythema associated with Vasculitis in a Patient with Systemic Lupus Erythematosus

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A 42-year-old woman with systemic lupus erythematosus (SLE) has had wide-spread erythemas with some purpuric patches on her trunk and upper extremities during exacerbation of her disease. Biopsy findings from erythematous lesions and purpuric areas revealed the pathology of lupus erythematosus and leukocytoclastic vasculitis (LCV) with immune deposits, respectively. A feature of wide-spread symmetrical erythemas with foci of LCV occurring in this patient is considered to be an unusual presentation.
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Key Words : Extensive cutaneous erythema, Leukocytoclastic vasculitis, Systemic lupus erythematosus

The type of skin manifestations in systemic lupus erythematosus (SLE) may be regarded as a marker of the disease activities, and sometimes it can be used as a predictor of prognosis or organ involvement[1,2]. Extensive erythemas or leukocytoclastic vasculitis (LCV), respectively, is not an infrequent feature in patients with SLE, however, the coexistence of these two cutaneous features is rare;they may be associated with an active phase of disease[1-3]. In review of literatures, we could not find any case resembling ours.

CASE REPORT

A 42-year-old woman, who had been diagnosed as SLE 4 years ago, was admitted for further evaluation of intermittent fever, multiple arthralgia and general malaise. At the initial presentation, her disease manifested as malar rash, photosensitivity, leukopenia, arthritis, and glomerulonephritis. For one and a half months, she has experienced some aggravating symptoms of arthritis with recurrent fever and skin rashes. The rashes were noted as wide-spread erythemas, pink to dull red in color, with scattered purpuric-erythematous areas, which appeared almost symmetrically on her anterior chest, shoulders, arms, upper back, and abdomen, with several foci of ulcerative lesions on the arms and anterior chest (Fig. 1). These erythematous lesions with variable intensities of inflammation had persisted for more than 3 weeks prior to the admission. Initially these lesions had occurred on the upper arms and shoulders as erythematous patches with a few purpuric lesions, then further developed over the trunk for a period of 10 days. Mild itching and prickling sensations were the only cutaneous symptoms. She did not have any relevant history of trauma, infection, or any other suspected precipitating episode for these lesions. Besides her general weakness and fever, malar erythemas, photosensitivity, and oral erosions were also recognized on physical examination. Her past medical history and family history had nothing to be noted.

Biopsy specimens of the skin were taken from erythematous lesions on the flank and upper arm, and from purpuric-erythematous areas on the upper

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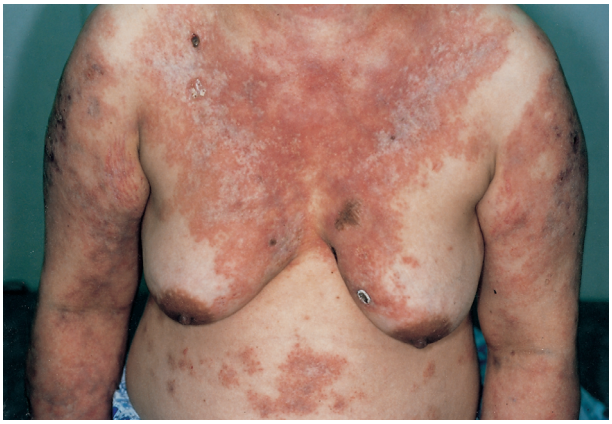

Fig. 1. Wide-spread erythematous lesions with some purpuric patches on the trunk and arms.

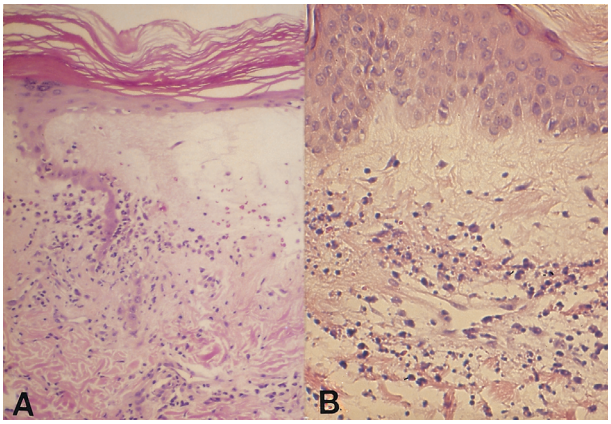

Fig. 2A. Epidermal atrophy, focal hydropic degeneration of basal cells, marked edema of upper dermis and a moderate degree of lymphocytic infiltration(perivascular/periappendageal) are seen (H&E, × 100).
2B. Neutrophil-dominant inflammatory infiltrates with leukocytoclasia, endothelial swellings, vasodilation with some extravasation of red blood cells are observed in the upper dermis (H&E, × 200).

arm and clavicular areas. Histopathologic findings revealed the features of LE (epidermal atrophy, focal hydropic degeneration of basal cells, marked edema of upper dermis, perivascular/periappendageal lymphocytic infiltrations) on the erythematous/edematous lesions on her flank and arm (Fig. 2A) and the features of LCV (neutrophil-dominant inflammatory infiltrates with leukocytoclasia, endothelial swellings, vasodilation with extravasation of red blood cells, and foci of fibrinoid degenerations) on the purpuric/erythematous areas (Fig. 2B).

Laboratory findings showed leukopenia (2400/mm^3), thrombocytopenia (90,000/mm^3), and an elevated erythrocyte sedimentation rate (55 mm/hr). The creatinine clearance rate was 30 ml/min. Urinalysis revealed proteinuria. The chest roentgenogram showed slight pleural effusions. The titer of anti-nuclear antibodies was 1:2560 (speckled pattern), and anti-nDNA antibody was seen as strong-positive on the *Crithidia luciliae* substrates. The serum complement concentrations revealed low levels of C3 (43 mg/dL) and C4 (6 mg/dL). Anti-neutrophil cytoplasmic antibodies, anti-phospholipid antibodies, and cryoglobulins were not detected.

Noticing the presence of some exacerbation of systemic symptoms, the dose of oral prednisolone was increased to 60 mg/day, with hydroxychloroquine (400 mg/day), and within the next 3 weeks a subsequent improvement of the activities of cutaneous erythemas and of her constitutional symptoms were observed.

DISCUSSION

The wide-spread erythema is one of the LE-specific skin lesions, and LCV can be detected in the active phase of SLE[4,5]. A feature of wide-spread symmetrical erythemas and cutaneous vasculitis occurring simultaneously, as have observed in this patient, is considered to be an unusual and rarely encountering presentation; the patient was recognized to be in the stage of flare-up of SLE.

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