Neutrophilic Dermatosis with an Erythema Gyratum Repens-like Pattern in Systemic Lupus Erythematosus

Benjamin Khan Durani[1], Konrad Andrassy[2] and Wolfgang Hartschuh[1]*
[1]*Department of Dermatology and* [2]*Department of Internal Medicine, University of Heidelberg, Bergheimerstr. 56a, DE-69115 Heidelberg, Germany. *E-mail: wolfgang_hartschuh@med.uni-heidelberg.de*
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Sir,

In 1952, Gammel reported on a 56-year-old woman presenting with erythema in irregular wavy bands with a collarette-type or marginal desquamation. The lesions moved constantly at a rate of 1 cm/day. Further examinations revealed a poorly differentiated adenocarcinoma of the breast. Ten days after resection of the carcinoma the eruption cleared almost completely (1). Other variations of figurate erythemas have been observed in multiple skin diseases, for example, erythema annulare centrifugum, neutrophilic figurate erythema of infancy and necrolytic migratory erythema. An association with different internal illnesses such as malignant tumours, autoimmune diseases and infections is well known. We describe here a patient with systemic lupus erythematosus (SLE) and figurate erythemas, histologically characterized by a neutrophilic dermatosis.

CASE REPORT

A 69-year-old man presented with multiple reddish, infiltrated plaques on the trunk. He had neither itching nor pain, but had suffered from joint pain for many years. Treatment of the arthritis included harpagophytum extract, celecoxib and prednisone (5 mg). Concerned about possible side effects from the medication, the general practitioner stopped all drugs. The skin lesions persisted in spite of this intervention. One month later, gyrate erythema appeared in addition on the legs. The bizarre lesions with a varying reddish tinge were arranged in a wood grain-like crescent pattern, changing formation at short intervals, sometimes within a few hours, leaving slight post-inflammatory hyperpigmentation (Fig. 1). Neither scaling nor elevation/oedema of the lesions was observed.

Laboratory abnormalities included antinuclear antibodies, 1/20 480 (normal value, 1/80); anti-dsDNA, 44 IU/ml (normal value, $<$40 IU/ml); positive histone antibodies; anticardiolipin antibodies IgG, 21 GPL IU/ml (normal value, $<$10 GPL IU/ml); pANCA 1/32 (normal value, $<$1/10); low C3c, 0.34 g/l (normal value, 0.9–1.8 g/l); low C4, 0.03 g/l (normal value 0.1–0.4 g/l), leucocytopenia, 2.88/nl (normal value 4–10/nl) and a nephritic sediment. An extensive search showed no malignant growth, but a biopsy of the kidneys revealed a diffuse proliferative lupus nephritis (WHO IVa).

Considering the American Rheumatism Association (ARA) criteria (2), SLE was diagnosed. Skin biopsies from the red plaques and gyrate erythema showed a diffuse dermal neutrophilic infiltrate without signs of vasculitis or epidermal involvement (Fig. 2). Immunohistology revealed a dermo-epidermal lupus band.

The gyrate erythema and infiltrated plaques disappeared after 2 weeks of treatment with local steroids. Thereafter, the nephrologist started therapy with a cyclophosphamide bolus (500 mg every 4 weeks) because of the proliferative lupus nephritis. No relapse of the skin lesions was observed during the following disease course.

DISCUSSION

We report a patient with SLE presenting reddish crescent gyrate skin lesions on the legs, which resemble erythema gyratum repens of Gammel (1), a rare disorder that usually appears as a paraneoplastic condition. The histopathological findings of erythema gyratum repens are not specific; however, the neutrophilic dermatosis found in our patient was not described. Furthermore, Gammel reported slow movement of the erythema at a rate of 1 cm/day instead of rapid changing of lesions and

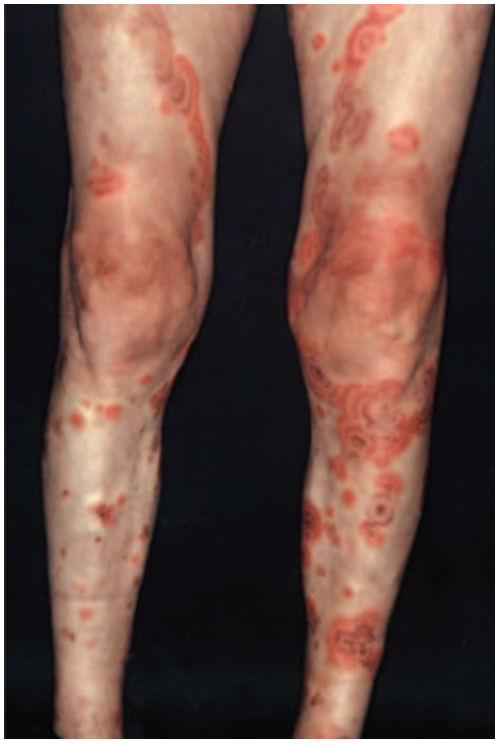

Fig. 1. Curious lesions with a varying reddish tinge forming wood grain-like crescent patterns on the legs.

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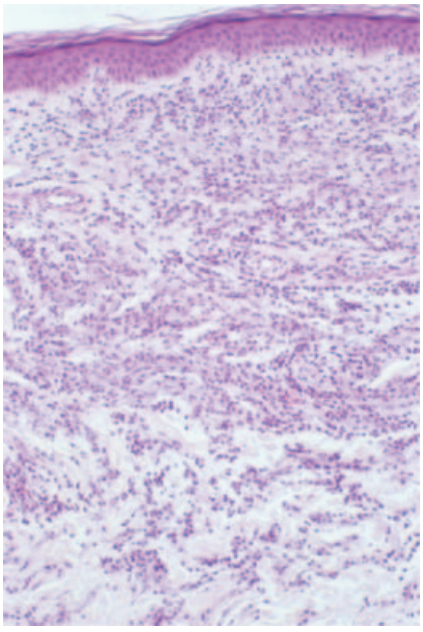

Fig. 2. Histology (H&E stain) reveals a diffuse dermal neutrophilic infiltrate (original magnification ×30).

observation of linear desquamation, which was missing in our case (1).

In Japanese patients with Sjögren syndrome, annular erythema mimicking Sweets syndrome or erythema gyratum repens have been reported (3). Lesions were characterized by wide, elevated borders and central pallor. Histologically, there was a coat sleeve-like infiltration of lymphocytes around blood vessels throughout the dermis, nuclear dust around the blood vessels or between collagen bundles, marked oedema of the papillary dermis and slight epidermal changes. However, no neutrophilic infiltrate was observed.

In 1975, Helwitt et al. (4) introduced the term 'lupus erythematosus gyratum repens' for another form of cutaneous lupus erythematosus. However, in skin biopsies dyskeratotic keratinocytes were prominent while neutrophilic dermatosis was absent, and the clinical features were clearly different from our case.

Furthermore, urticarial vasculitis can be considered as a differential diagnosis in view of the clinical features. However, histological findings in urticarial vasculitis with papillary dermal oedema, slight perivascular infiltrates with variable amounts of neutrophils, sometimes slight leukocytoclasia, slight focal fibrin deposits of superficial venules and haemorrhage are conspicuously distinct from the histological features found in our patient.

The crescent erythematous rings of our case resemble the lesions in a patient presented by Pique et al. in 2002 (5). They reported on a 38-year-old white woman with SLE developing lesions arranged in a zebra-like pattern. Histopathological study showed leucocytoclastic vasculitis involving the small blood vessels of the superficial dermis. A perivascular infiltrate of neutrophils, nuclear dust and fibrinoid degeneration of the vessel walls were found (5). However, in contrast to our case no distinct diffuse dermal neutrophilic infiltrate was reported.

From a histological viewpoint Sweets syndrome should be considered as a differential diagnosis (6). However, following the diagnostic criteria initiated by Su & Liu (7) for the diagnosis of acute febrile neutrophilic dermatosis, our patient does not fulfil all of the required conditions, which excludes classical Sweets syndrome.

Further histological differential diagnoses presenting with neutrophilic dermatosis, such as bowel-associated dermatosis-arthritis syndrome (8), pyoderma gangrenosum (9), rheumatoid neutrophilic dermatosis (10), Behcet's syndrome and acute generalized pustulosis (11), can easily be excluded by the clinical features.

This case seems to be the first report on a characteristic neutrophilic dermatosis in SLE resembling erythema gyratum repens of Gammel.

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