

# An erythema gyratum repens variant of bullous lupus erythematosus



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## INTRODUCTION

Bullous lupus erythematosus is an autoantibody-mediated subepidermal blistering disease that occurs in patients with systemic lupus erythematosus (SLE).<sup>1</sup> Erythema gyratum repens (EGR) is a clinical diagnosis characterized by a wood grain pattern of centrifugally migrating erythematous plaques with fine leading scale. Although most cases of EGR are associated with underlying malignancy, several nonparaneoplastic-associated cases of EGR have been reported.<sup>2</sup> Here, we report on a patient with bullous lupus erythematosus presenting in an EGR pattern.

## REPORT OF CASE

A previously healthy 55-year-old African-American man presented for evaluation of a sudden-onset, diffuse vesiculobullous eruption affecting the trunk and extremities. The patient denied associated skin pain or pruritus. He was otherwise well, and a detailed review of systems was negative. He had a medical history of hypertension, hyperlipidemia, and diabetes mellitus type II. His medications included atorvastatin, methocarbamol, and calcium/vitamin D supplementation. He had no known drug allergies or family history of autoimmune disease.

Initial physical examination found erythematous plaques expanding centrifugally with peripheral tense vesicles and bullae (Fig 1). Over a period of days, concentric vesicular plaques developed (Fig 2). There was no mucosal involvement.

Histopathologic examination found a superficial perivascular mixed inflammatory cell infiltrate

### Abbreviations used:

EGR: erythema gyratum repens  
SLE: systemic lupus erythematosus

composed of lymphocytes, scattered eosinophils, and numerous neutrophils. Neutrophilic microabscesses were present within the papillary dermis. The neutrophilic infiltrate extended into the overlying epidermis with vacuolar alteration of the basal layer. A second specimen showed a subepidermal bulla with epidermal necrosis, lichenoid interface dermatitis, and a polymorphous dermal infiltrate.

Direct immunofluorescence was strongly positive for C3 (4+) granular deposition and shaggy linear fibrin (4+) at the dermal-epidermal junction. Granular IgG was present focally at the dermal-epidermal junction.

Serologic testing found positive antinuclear antibody, 1:1280 titer in a speckled pattern, anti-double-stranded DNA antibody (14), anti-Smith antibody (>8.0), and antiribonucleoprotein antibody (>8.0). Anti-Ro and anti-La antibodies were not present. Serum C3 was low (76 mg/dL), and C4 was within normal limits. Results of complete blood count with differential, comprehensive metabolic panel, and urinalysis were within normal limits.

Serologic, histopathologic, and direct immunofluorescence findings supported a diagnosis of bullous lupus erythematosus. Given the concentric morphology of the eruption, a diagnosis of an EGR variant of bullous lupus erythematosus was made. Because of the association of EGR with malignancy,

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**Fig 1.** Posterior shoulder and back with erythematous plaques and peripheral tense vesicles and bullae in a centrifugal pattern.



**Fig 2.** Abdomen and chest with multiple concentric vesicular plaques.

age-related cancer screenings were performed, the results of which were negative.

Initial treatment included oral prednisone monotherapy in doses of 60 mg/d without improvement. Addition of dapsone, 25 mg/d, resulted in complete



**Fig 3.** Back with concentric patches of postinflammatory hypopigmentation 2 weeks after starting dapsone.

resolution of the eruption within 2 weeks with subsequent postinflammatory hypopigmentation in concentric patches (Fig 3). Oral prednisone was tapered and discontinued. His treatment course was complicated by hemolytic anemia necessitating the discontinuation of dapsone. Mycophenolate mofetil, 1500 mg twice daily, was initiated, and he has remained clear to date.

## DISCUSSION

Bullous lupus erythematosus is an SLE variant characterized by rapid onset of tense vesiculobullous lesions typically in a photodistributed pattern on the face, trunk, and upper extremities. Less commonly, lesions can appear on mucous membranes and non-photo-exposed skin.<sup>1,3</sup> Affected patients usually experience constitutional symptoms of SLE including fever, fatigue, weight loss, and arthralgia. Histopathology findings show subepidermal vesiculation, neutrophilic microabscesses in the papillary dermis, mucin deposition in the dermis, dermal edema, and absence of eosinophils. Direct immunofluorescence shows IgG, IgA, IgM, and complement components at the dermal-epidermal junction in a granular or linear pattern. Depositions can be focal or continuous and may extend into the dermis and perivascular region.<sup>4</sup> Most patients with bullous lupus erythematosus have a positive antinuclear antibody and frequently will have positive anti-double-stranded DNA antibody and anti-collagen type VII antibody.<sup>5</sup> First-line treatment of bullous lupus erythematosus is with dapsone, although it is contraindicated in patients with glucose-6-phosphate dehydrogenase deficiency. Dapsone results in cessation of new blister formation within days for most patients.<sup>6</sup> Prednisone is used as an alternative or adjunct, especially for patients that require treatment for concomitant systemic manifestations of SLE. Other treatment options include rituximab, methotrexate,

azathioprine, and mycophenolate mofetil, although data on their use are limited.<sup>1</sup> Lesions heal without scarring but often with evident postinflammatory hypopigmentation or hyperpigmentation.<sup>7</sup>

The presented patient met 2012 Systemic Lupus International Collaborating Clinics criteria for diagnosis of SLE and had immunohistopathologic and clinical evidence of bullous lupus, with blisters developing in a wood grain, concentric morphology reminiscent of EGR. EGR is a clinical entity with nonspecific histopathologic findings. Most patients are found to have an associated malignancy; therefore, affected patients must undergo malignancy workup.<sup>2</sup> Other diseases including lepromatous leprosy, limited systemic sclerosis, and psoriasis are reported to present in an EGR pattern.<sup>2,7</sup> Finally, there is an entity known as lupus erythematosus gyratus repens, a rare variant of subacute cutaneous lupus erythematosus, which was a diagnostic consideration in this case. In this entity, lesions typically present as widespread, concentric, erythematous plaques, often with double-contoured edges. The plaques are often moderately to deeply infiltrated with overlying crusted erosions and scaling. Histopathologic characteristics include hydropic degeneration of the basal layer, keratinocyte necrosis, and atrophy of the epidermis.<sup>8</sup>

To our knowledge, blisters of bullous lupus erythematosus presenting in a centrifugally expanding, concentric pattern are not reported. There is 1 report of infiltrated erythematous plaques arranged in an EGR-like pattern in a patient with SLE.<sup>9</sup> Another patient with SLE had leukocytoclastic vasculitis in an EGR-like pattern. Histopathologic findings included

a neutrophil predominant vasculitis.<sup>10</sup> These findings appear to be similar clinical entities. We suspect that the EGR-like pattern in patients with SLE, and forms of SLE such as bullous lupus erythematosus, may be an uncommon morphology that is associated with a neutrophilic predominance on histopathology.

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