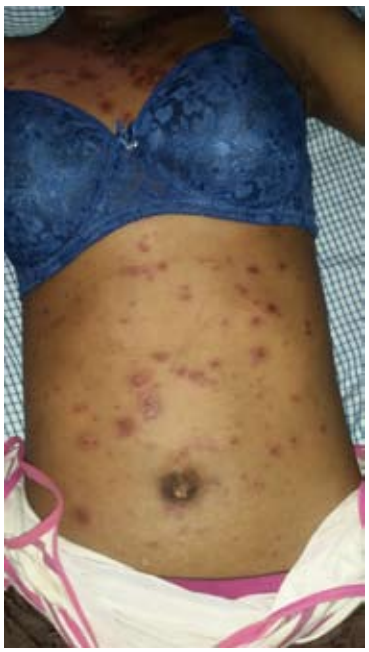
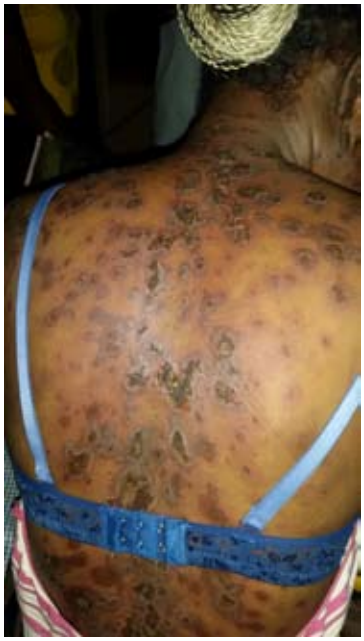
Rowell Syndrome in Nigeria: Systemic Lupus Erythematosus Presenting as Recurrent Erythema Multiforme in a Young Woman

Dear Editor,

Systemic lupus erythematosus (SLE) is a heterogeneous autoimmune disease characterized by diverse patterns of auto-antibody production with multi-organ affectation. Cutaneous involvement, either alone or in association with other systemic illnesses, is one of its most common manifestations (1). Dermatologic disorders like malar and discoid rashes are quite suggestive of SLE. However, the occurrence of non-specific skin lesions like erythema multiforme (EM) in patients with SLE (Rowell syndrome) can rarely occur (1). In such patients, a diagnosis of SLE may be missed or delayed in the absence of other overt clinical features of lupus. Herein we report a case of recurring EM-like eruptions as the cardinal cutaneous manifestation of previously undiagnosed, active SLE in a young Nigerian woman.

A 26-year-old Nigerian woman presented with a three-day history of non-pruritic, generalized, and target-like, erythematous annular patches and plaques which mostly affected the trunk. A few lesions had presented with crusting and erosions at the time of examination (Figure 1). Associated symptoms included oral painful ulcers, low grade fever, and malaise. The patient had no other systemic symptoms and her prior drug history was not remarkable. Her erythrocyte sedimentation rate (ESR) was 66 mm/hour using the Westergren method. Screening for HIV and hepatitis B and C was negative. Herpes simplex, cytomegalovirus, and Epstein Barr viruses could not be screened for. Other baseline investigations (complete blood count, electrolytes, urea and creatinine as well as urinalysis) were within normal limits. The patient was managed as a case of EM of an unidentified inciting agent and her symptoms resolved with supportive care and antibiotics. However, she developed a recurrence about 5 weeks later, with more extensive and coalescent skin lesions (Figure 2). Additionally, there was a new onset of alopecia and pain in the small joints of the hands as well as both knees and ankles. At this time, the patient's ESR had gone





up to 112 mm/h and she had developed significant proteinuria, with a protein creatinine ratio of 1.3 g/g (reference <0.5 g/g). Her antinuclear antibody (ANA) titer was high (1:320) with a speckled pattern. Anti-Smith antibody was also positive. A renal biopsy was declined. A tentative diagnosis of Rowell syndrome was made. The patient was started on high-dose steroids and hydroxychloroquine 200 mg twice daily. Subsequent care included the use of mycophenolate mofetil 1 g twice daily for 6 months. This was then changed to azathioprine at 50 mg twice daily. Follow-up after 6 months showed sustained clearance of skin lesions, resolution of fever and joint pains, as well as improvement in the renal profile, with a urine protein-creatinine ratio of 0.77 g/g.

The presence of systemic lupus erythematosus, EM-like lesions, and a speckled pattern of antinuclear antibody in our patient fulfils the revised diagnostic criteria for RS put forward by Zeitouni *et al*. at the turn of the twenty-first century (2).

Considering the rarity of EM-like lesions in SLE and the possibility of constitutional symptoms in EM, a diagnosis of RS may be readily overlooked in patients like the one described, whose major cutaneous manifestation of severe active SLE was EM-like lesions. In contrast to classic EM, where skin lesions are concentrated in the extremities, a predominant truncal distribution of EM-like lesions as found in our patient may favor a clinical consideration of RS (3).

However, some authors have challenged the existence of Rowell syndrome as a distinct clinical laboratory entity. Arguments put forward in this regard include the fact that none of the immunological markers that have been described in RS are specific to any disorder. Additionally, the annular polycyclic dermatosis seen in sub-acute cutaneous lupus erythematosus (SCLE) can be difficult to clinically and histologically differentiate from EM (4,5). Patients with SLE also have a higher likelihood of developing adverse drug reactions (6).

The inherent complexity of SLE may make for delayed and oftentimes difficult diagnosis, especially in a country where immunologic tests are expensive and rheumatologists are scarce. When patients do occasionally present with recurrences of skin lesions in the spectrum of EM, Steven-Johnson syndrome, and toxic epidermal necrolysis in the absence of a definite inciting agent, undiagnosed lupus may indeed be present in some of these individuals and should be considered in the differential diagnosis.

In conclusion, while it is very rare, SLE may present first with recurrent episodes of EM-like rash. Despite the various possibilities which underlie their association, prompt identification and treatment of SLE in patients presenting with EM is important to prevent death or irreversible organ damage.

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Richard Oluyinka Akintayo[1], Gboyega Musbau Olarinoye[2], Foluke Comfort Akintayo[3], Omotoyosi Nike Ilesanmi[2]

[1]Division of Rheumatology, Department of Medicine, University of Ilorin Teaching Hospital Ilorin, Ilorin, Nigeria

[2]Division of Dermatology, Department of Medicine, University of Ilorin Teaching Hospital Ilorin, Ilorin, Nigeria

[3]Department of Family Medicine, Ladoke Akintola University of Technology Teaching Hospital, Ogbomoso, Nigeria

Corresponding author:

Richard Akintayo, MD

Department of Medicine

University of Ilorin Teaching Hospital

Ilorin

Nigeria

richocounlimited@gmail.com

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