An atypical clinical presentation of alopecia in 2 patients with systemic lupus erythematosus

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INTRODUCTION

Alopecia in varying patterns is a common feature of lupus erythematosus (LE).[1] Several forms of alopecia that are not specific to LE can occur in the setting of lupus, such as alopecia areata, telogen effluvium, and anagen effluvium. LE-specific alopecias are those with a histology consistent with LE and include forms of acute, subacute, and chronic cutaneous lupus. Common patterns of LE-specific alopecia include the nonscarring diffuse hair thinning and fragility of acute LE and the scarring, erythematous scaly plaques with follicular keratotic plugs, peripheral hyperpigmentation, and central hypopigmentation of discoid lupus (DLE).[2] Lupus erythematosus tumidus (LET) may also present on the scalp as well-defined, nonscarring alopecia without overlying scale, atrophy, and dyspigmentation—lesions clinically reminiscent of alopecia areata, albeit with different underlying histopathology.[3]

We report on 2 male patients with large circular nonscarring alopecic plaques on the scalp without overlying scale or erythema but with central hyperpigmentation and scarring. Biopsies found patchy perifollicular and focally lichenoid lymphocytic infiltrate with loss of hair follicles and increased dermal mucin consistent with LE; further workup and serologic testing found systemic lupus. These 2 cases demonstrate an unusual clinical presentation of central scarring alopecia within a larger nonscarring alopecic plaque in the setting of SLE that deviates from typical lupus-related alopecia.

Abbreviations used:

ANA:	antinuclear antibody
CCLE:	chronic cutaneous lupus erythematosus
DLE:	discoid lupus erythematosus
LE:	lupus erythematosus
LET:	lupus erythematosus tumidus
SLE:	systemic lupus erythematosus

CASE REPORT

Patient 1 is a 27-year-old Hispanic man with no medical history who presented with several years of localized but progressive hair loss and scalp discoloration. He reported symptoms of fatigue, unintentional weight loss, night sweats, and joint pain. Examination was significant for a large circular alopecic plaque on the parietal scalp. Central scarring and hyperpigmentation was noted within a larger, smooth, normally pigmented alopecic patch with preserved follicular ostia (Fig 1). No other cutaneous lesions were identified. A punch biopsy from the central area of alopecia found a near—end-stage alopecia with loss of hair follicles, retained fibrous stellae, and perifollicular lymphocytic infiltrate (Fig 2, *A*). Most remaining follicles were in telogen growth phase or miniaturized with no terminal hairs remaining. The infiltrate was focally lichenoid, and there was subtly increased dermal mucin (not shown). Further laboratory testing found antinuclear antibody (ANA) titer of 1:2560, low complement, elevated anti-dsDNA, anti-Ro, anti-Smith, and anti-RNP antibodies, as well as anemia and leukopenia. SLE was diagnosed. Therapy was

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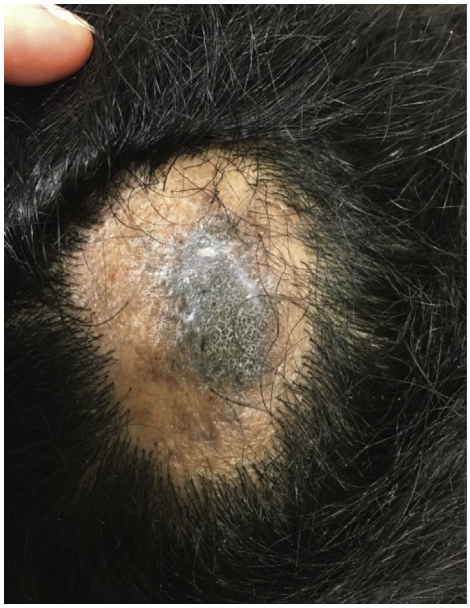

Fig 1. Patient 1 presented with a single large circular nonscarring alopecic plaque without significant scale, erythema, or dyspigmentation but with central hyperpigmentation and scarring.

initiated with hydroxychloroquine, topical betamethasone, and prednisone taper.

Patient 2 is a 31-year-old African-American man with recent history of myocarditis of unclear etiology who presented with several years of localized but progressive scalp hair loss. He did not respond to antifungal treatment for presumed tinea capitis. On examination were several large circular normally pigmented nonscarring alopecic patches with preserved follicular ostia but with distinct central areas of scarring and hyperpigmentation (Fig 3, *A*). A punch biopsy from the hyperpigmented area of alopecia found dropout of hair follicles associated with perifollicular lymphocytic infiltrate. Most remaining hairs were in the anagen growth phase (Fig 2, *B*). Of note, ANA was positive (1:80) during autoimmune workup for myocarditis. The patient reported excessive fatigue and family history of lupus in his brother. Further laboratory testing found elevated anti-dsDNA, anti-Smith, and anti-RNP antibodies. These findings, along with chronic cutaneous lupus erythematosus (CCLE) and alopecia, met 5 Systemic Lupus International Collaborating Clinics criteria and qualified for SLE diagnosis. Therapy was initiated with hydroxychloroquine and topical halobetasol. At 6-week follow-up, he exhibited vellus hair regrowth surrounding the central scarred area (Fig 3, *B*). Unfortunately, he presented months later with several new hyperpigmented, erythematous, scaly plaques on bilateral upper extremities. Biopsy found epidermal atrophy, periadnexal lymphocytic infiltrate, and increased dermal mucin consistent with CCLE. A prednisone taper was added to his treatment regimen.

DISCUSSION

These 2 cases of scalp alopecia display features of central scarring with surrounding nonscarring alopecia that deviate from classical clinical findings of LE-related alopecia. Specifically, these patients presented with large circular nonscarring alopecia with preserved follicular ostia and absence of overlying scale, erythema, and dyspigmentation—findings reminiscent of LET or alopecia areata. However, both cases also exhibited centrally hyperpigmented, scarring plaques as well as serologic and constitutional signs of systemic lupus. Notably, LET is very rarely associated with SLE.[4] Scalp lesions were also distinct from DLE in that no significant erythema, scale, keratotic plugs, central hypopigmentation, or peripheral hyperpigmentation were noted. Other than scarring alopecia, these lesions do not resemble clinical findings or demographics of lichen planopilaris, central centrifugal cicatricial alopecia, or fibrosing alopecia in a pattern distribution.[5]

Histologically, both patients displayed perifollicular lymphocytic infiltrate, follicular dropout, and increased dermal mucin. Patient 1 had more advanced disease than patient 2, demonstrated by more significant fibrosis and mucin deposition; we speculate that these 2 cases may be on a continuum of severity. Furthermore, both patients subsequently had SLE, and antimalarial therapy with hydroxychloroquine was initiated. Patient 2 had initial clinical response and hair regrowth after 6 weeks of treatment in concordance with the milder histopathology. This response to treatment is more reminiscent of LET, which is remarkably responsive to antimalarial therapy in as little as 4 to 6 weeks,[4] than of DLE, where clinical response may not be evident for several months.[6] Unfortunately, both patients were lost to follow-up, and their long-term response to therapy with hydroxychloroquine remains unknown.

Interestingly, Stead et al[7] previously reported on 2 patients in whom LET and DLE, as well as LET and SLE, coexisted—although neither patient exhibited alopecia.[7] Lehrhoff et al[8] also described a case of multiple distinct alopecic plaques consistent with LET adjacent to those consistent with DLE[8]; features of nonscarring and scarring alopecia were not present within the same lesion as described here. Indeed, hydroxychloroquine resulted in complete hair regrowth in LET-like but not DLE-like lesions at an unspecified time of follow-up.[8] We describe 2

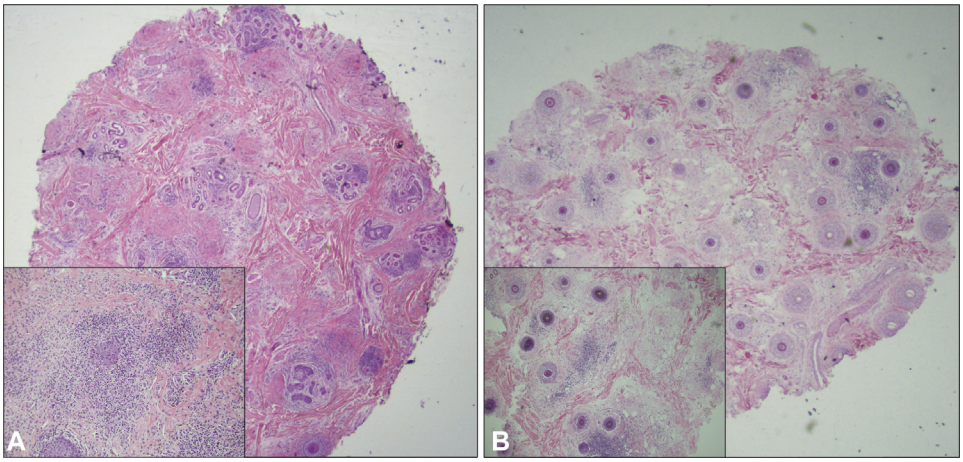

Fig 2. **A**, Patient 1: scanning image of horizontally oriented scalp skin at the mid to lower dermal level shows marked dropout of hair follicles with patchy lymphocytic infiltrate around remaining hair follicles and eccrine glands (original magnification ×20). Higher-power inset shows dense lymphocytic infiltrate around a degenerated hair follicle (original magnification ×100). **B**, Patient 2: scanning image of horizontally oriented scalp skin at the mid to lower dermal level shows scattered dropout of hair follicles with remaining follicles in anagen growth phase. Patchy lymphocytic infiltrate is present near remaining hair follicles (original magnification ×20). Higher-power inset shows fibrous stellae, retained anagen hairs, and lymphocytic infiltrate juxtaposed to remaining hair follicles (original magnification ×40).

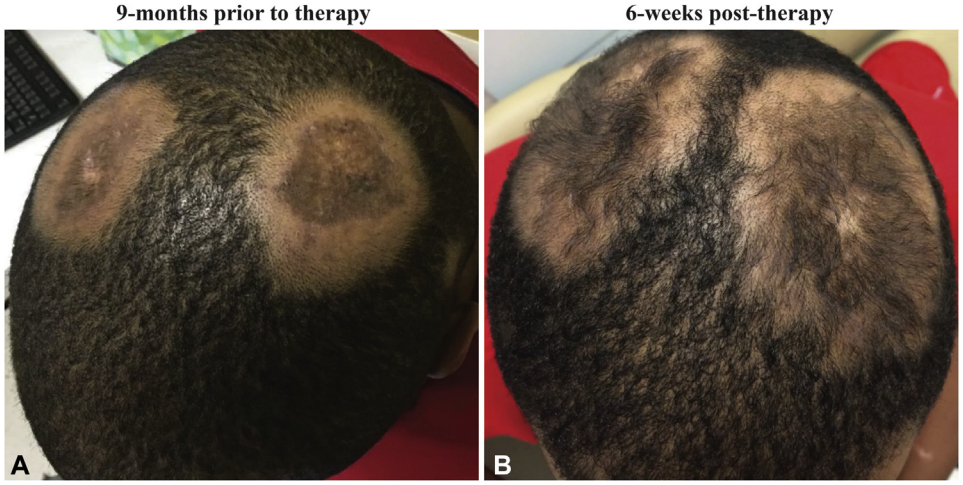

Fig 3. Patient 2 presented with several large circular nonscarring alopecic patches without significant scale, erythema, or dyspigmentation but with central hyperpigmentation and scarring. A new, smaller lesion with no surface change is noted on the left scalp above the ear. **A**, Nine months before initiating antimalarial therapy. **B**, Six weeks after therapy shows significant vellus hair regrowth.

interesting cases that demonstrate a novel clinical pattern of nonscarring alopecia with central scarring in systemic lupus that has not been described to our knowledge. Furthermore, these cases underscore the morphologic heterogeneity of CCLE and need for thorough evaluation of systemic disease in patients presenting with both scarring and nonscarring alopecia.

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