A comedonal variant of chronic cutaneous lupus erythematosus: Case report and literature review

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INTRODUCTION

Cutaneous lupus erythematosus (LE) is subdivided into acute, subacute, and chronic forms, each with distinct morphologies. Chronic cutaneous lupus erythematosus (CCLE) most commonly presents in the form of discoid lupus erythematosus (DLE) with pink-to-violaceous coin-shaped plaques with hyperkeratosis and follicular plugging that heal with atrophy, scarring, hyperpigmentation, and telangiectasias. Lesions most commonly occur on the face, ears, scalp, and in sun-exposed areas. On hair-bearing skin, DLE results in cicatricial alopecia.

Although the lesions of DLE are often quite distinct, more than 20 other rare clinical variants of CCLE, such as hypertrophic LE and verrucous LE, have been described that can mimic other dermatologic conditions.[1] In 1972, one such variant consisting of acneiform lesions was introduced by Haroon and Fleming.[2] This variant, although exceedingly rare, is perhaps one of the most critical to recognize, as it can easily be mistaken for one of the most common diseases seen by dermatologists and general practitioners. Here we present a case of comedonal CCLE that was previously misdiagnosed and treated as acne vulgaris.

Abbreviations used:

CCLE:	chronic cutaneous lupus erythematosus
DLE:	discoid lupus erythematosus
LE:	lupus erythematosus

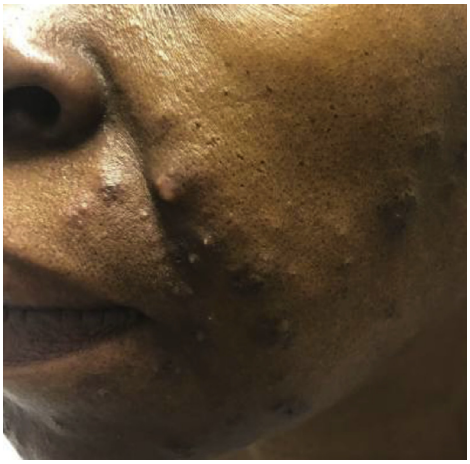

Fig 1. Comedonal CCLE on the cheek, chin, and jawline depicted by cystic pink and tan acneiform papules and pustules.

CASE REPORT

A 57-year-old postmenopausal African-American woman presented to our clinic with recalcitrant acne on the face for 1 year and hair loss for 6 months. The patient's acne did not respond to treatment with topical clindamycin, tretinoin and benzoyl peroxide, and oral doxycycline. Her hair loss was resistant to treatment with clobetasol. She took no other medications. She denied joint pain, oral ulcerations, or photosensitivity. A physical examination found brown papulonodules and cystic acneiform lesions on the bilateral upper neck, cheeks, chin, and forehead (Figs 1 and 2) and purple/brown papules

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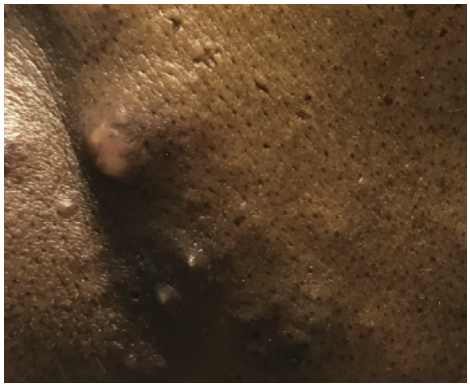

Fig 2. Higher-powered view of acneiform papules and pustules on the left cheek.

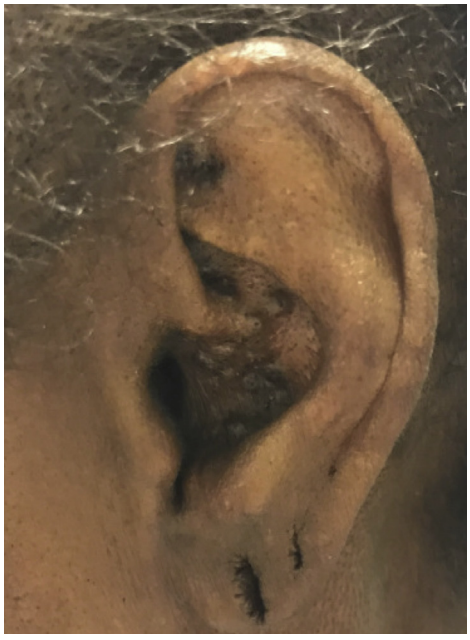

Fig 3. Left conchal bowl with purple/brown comedonal papules with follicular plugging.

and plaques in the bilateral conchal bowls with follicular plugging (Fig 3). The latter morphologically resembled lesions of DLE. Her scalp examination was significant for nonscarring alopecia of the frontotemporal scalp along with discrete circular patches of scarring alopecia of the parietal scalp.

Biopsy of the left conchal bowl showed a follicular infundibular cyst with a lymphocyte-mediated interface dermatitis involving the interfollicular epidermis and the hair follicle. An accompanying moderately dense perivascular lymphocytic infiltrate was also present. A biopsy of the right side of the upper neck found dermal fibrosis consistent with a cicatrix with chronic inflammation and hemosiderin deposition. A third biopsy of the left parietal scalp found mild

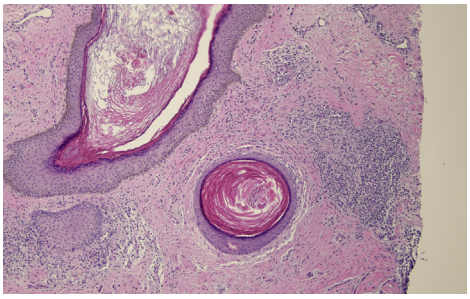

Fig 4. Low-power image shows comedonal dilation of the follicle; however, there is a supervening lymphocyte-mediated interface dermatitis affecting the outer root sheath epithelium.

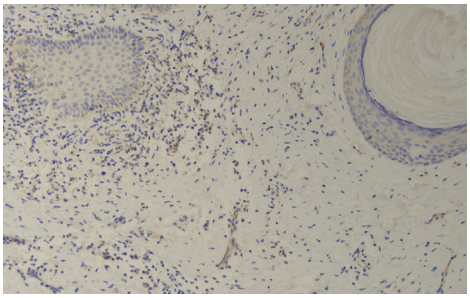

Fig 5. The myxovirus protein stain is a surrogate type I interferon marker that is significantly upregulated in this tissue sample. Normally, the signal for myxovirus protein staining is negative. In this photomicrograph there is a mildly upregulated signal in the follicular epithelium, inflammatory cells, and endothelium.

fibrosing lymphocytic folliculitis with an attendant reduction in terminal hair density. Because of the strong suspicion of discoid LE, a myxovirus protein stain, the surrogate type I interferon marker, was conducted and was strikingly positive in epithelial structures, the endothelium, and amidst inflammatory cells in all specimens. Overall the findings were interpreted as discoid LE manifesting an unusual hyperkeratotic acneiform diathesis compatible with comedomal CCLE (Figs 4 and 5). Despite an antinuclear antibody of 1:80 with speckled pattern, systemic involvement was deemed unlikely given a negative review of systems, negative double-stranded DNA, and normal basic laboratory panels. Although the patient started taking hydroxychloroquine, 200 mg daily, she was lost to follow-up, precluding further laboratory workup, direct immunofluorescence, or monitoring for response to therapy.

DISCUSSION

Comedonal CCLE is an especially rare but documented variant of cutaneous LE. A review of the English-language literature finds only 8 previous

Table I. Summary of reported cases of comedonal lupus in the English-language literature

Case	Age/sex	Time to diagnosis (y)	Sites of acneiform lesions	DIF	Concurrent SLE	Antinuclear antibody	Treatments	Follow-up
Haroon and Fleming[2]	28/F	6	Upper arms, shoulders, back	Positive	No	Negative	—	—
Motel et al[3]	29/F	7	Face, upper trunk	Negative	Yes	Positive (1:2560) homogenous	Tetracycline, erythromycin	No improvement
Motel et al[3]	24/F	—	Face, neck	IgG in nucleoli of focal keratinocytes	Yes	Positive (1:5120) nucleolar	Erythromycin 250 mg QID	No improvement
Chang et al[4]	32/M	3	Right nasolabial fold	IgG + C3 along BMZ	No	Negative	—	—
Stavrakoglou et al[5]	38/M	7	Face, chest, back	Negative	No	Negative	1. Erythromycin, 500 mg BID + adapalene daily → 2. Hydroxychloroquine, 200 mg BID, sunscreen	1. No improvement 2. Marked improvement in 4 mo, clear in 1 y
Hemmati et al[6]	33/F	1.5	Scalp	—	No	—	1. Hydroxychloroquine, 200 mg BID + betamethasone diproprionate 0.05% lotion + ILTAC 5 mg/cc → 2. Tretinoin 0.025% cream + hydroxychloroquine for 2.5 mo → 3. Hydroxychloroquine, 200 mg BID, ILTAC 10 mg/cc, clobetasol, manual extraction, excision	1. No improvement 2. No improvement 3. Lesions flattened and stabilized
Farias et al[7]	35/F	2	Face, ear	—	No	—	1. Tetracycline 500 mg BID × 3 weeks then 250 mg BID × 30 days → 2. Hydroxychloroquine, 400 mg daily	1. Some improvement in pruritus and number of comedones 2. Improvement in 45 d, marked improvement in 6 mo

Continued

Table I. Cont'd

Case	Age/sex	Time to diagnosis (y)	Sites of acneiform lesions	DIF	Concurrent SLE	Antinuclear antibody	Treatments	Follow-up
Vieira et al[8]	32/F	2	Face	IgM along BMZ and upper dermal vessels	No	Negative	Prednisone 40 mg × 1 mo then tapered over 4 mo + chloroquine, 250 mg daily	Significant improvement at 3 mo, no new lesions at 4 ys
Current case	57/F	1	Face, ears, neck	—	No	Positive (1:80) speckled	Topical clindamycin, tretinoin, benzoyl peroxide, oral doxycycline	No improvement

BID, Twice daily; *BMZ*, basement membrane zone; *DIF*, direct immunofluorescence; *ILTAC*, intralesional triamcinolone; *SLE*, systemic lupus erythematosus; *QID*, 4 times daily.

cases (Table I).[2-8] An analysis of the reported cases in Table I is helpful in characterizing the distinctive features that define this apparently rare variant of CCLE. First, it predominates in young women (78% of reported cases occurred in women, 86% of whom were in their third or fourth decade of life). Akin to DLE, this subtype of CCLE also tends to favor the face, ears, scalp, and sun-exposed areas. The mean time to diagnosis among reported cases is 3.7 years, reflecting the diagnostic challenge posed by its resemblance to acne vulgaris. Pruritus appears to be a significant symptom in many previously reported cases.[8] Two of 9 cases reported concomitant systemic involvement. Regarding treatment, antimalarials such as hydroxychloroquine and chloroquine have the best-reported efficacy, perhaps in combination with intralesional steroids and manual extraction for resistant cases.[6] Notably, as with our patient, typical acne treatments do not appear to be effective in the treatment of comedonal CCLE. Like many subtypes of CCLE, the pathogenesis behind this acneiform variant is not well elucidated. Because DLE can demonstrate a Koebner response, perhaps an isomorphic DLE response in a lesion that is otherwise characteristic for acne vulgaris is possible.[3] Others have suggested that acneiform lesions result from the destructive process of mononuclear cell infiltration of the pilosebaceous units.[4] Intrinsic to the pathology of both DLE and acne vulgaris is follicular hyperkeratosis. In this regard, it would not be surprising that on occasion an exaggeration of the follicular hyperkeratosis intrinsic to DLE could translate clinically and histologically into a comedonal process. Finally, there is already substantial precedent regarding other T-cell lymphocyte–rich reactions that can result in a follicular cystic pattern of hyperkeratosis resembling a comedone such as the striking comedonal lesions seen in follicular mycosis fungoides.[9] It has been shown that a critical event that presages ductal hypercornification in the setting of acne vulgaris is one of follicular epithelial hyperplasia stimulated by T cells primarily of the CD4 subset.[10] The role of T cells in the pathogenesis of acne is also reflected by the association of acne with T-cell–rich autoinflammatory and autoimmune T-cell disorders such as pyoderma gangrenosum and inflammatory bowel disease.

This case lends credence to a slowly growing body of literature supporting the existence of this subtype of CCLE, and it highlights the importance of considering comedonal CCLE in patients with apparently ordinary acne vulgaris not responding to conventional treatment. The scarring nature of CCLE, its preference for cosmetically sensitive areas, and the

resultant significant potential morbidity associated with a delay in its diagnosis underscore the importance of diagnostic recognition of CCLE clinically and pathologically.

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