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Dermatology Online Journal, 29(6)

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Publication Date

2023

DOI

10.5070/D329663001

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Volume 29 Number 6|November/December 2023|
29(6):13

Cutaneous erythematous lupus with acneiform presentation

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Abstract

We present a 57-year-old woman with cutaneous lupus erythematosus (CLE), initially treated as acne. She noted blemishes, including nodules and facial swelling for nine months associated with discrete itching of the ears. Examination showed multiple malar nodules, comedones, pustules, atrophic scars, and hyperpigmentation. A biopsy was performed and revealed atrophic epidermis, discrete hyperkeratosis, vacuolar degeneration of basal layer, basal membrane zone with upper dermal lymphohistiocytic inflammatory infiltrate and deep perivascular and peri-adenexal lymphocytes, vascular ectasia, and mucin deposits. The acneiform presentation of CLE is commonly underdiagnosed due to the similarity with inflammatory acne. Histopathologic diagnostic in acneiform lupus is of extreme importance. This case emphasizes the relevance of knowing the notable variety of presentations of CLE and considering this diagnosis.

Keywords: acne, lupus, skin

Introduction

Cutaneous lupus erythematosus (CLE) is an autoimmune disease of various presentations that can mimic other pathologies. The acneiform presentation of CLE is uncommon [1], with only few cases described in the literature, but may be underdiagnosed (**Table 1**). We report a case of CLE initially treated as acne.

Case Synopsis

A 57-year-old woman, with systemic arterial hypertension, osteoporosis, and smoking history, complained of blemishes on the face with swelling. This was associated with facial nodules and itching of ears. Medications included losartan, hydrochlorothiazide, calcium carbonate, and vitamin D. Upon physical examination, multiple malar nodules, comedones, pustules, and atrophic scars were noted with hyperpigmentation (**Figure 1**). In the concha of the ears, hyperpigmentation and follicular plugging were observed. No lesions in other body areas, joint pain, or other systemic symptoms were noted. She had been previously treated with manual extraction of comedones and oral antibiotics based on a diagnostic of acne vulgaris, without improvement.

A cutaneous biopsy was performed on the right cheek region, which revealed an atrophic epidermis

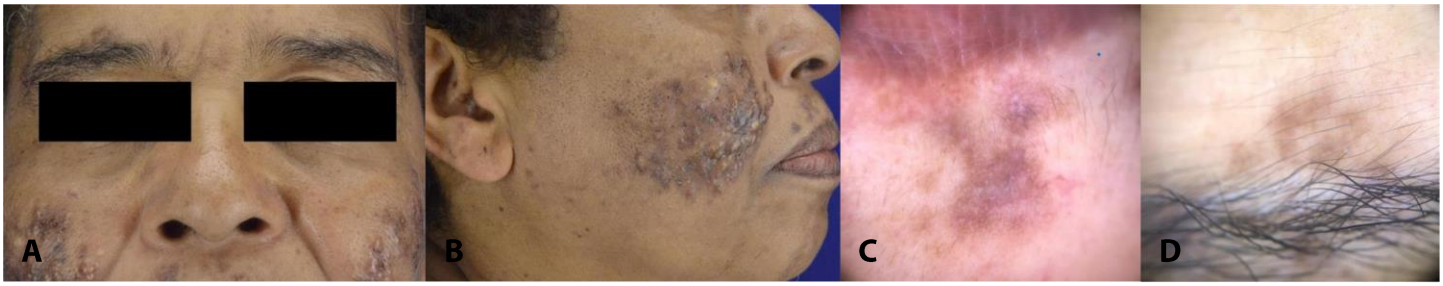

Figure 1. ***A, B)*** *Brown-violaceous nodules, pustules, open comedones (pinhead), atrophic scars.* ***C, D)*** *After treatment with hydroxychloroquine.*

with discrete hyperkeratosis and extensive vacuolar degeneration in the basal layer (**Figure 2A-D**). The basal membrane zone was thickened on Schiff-periodic acid staining, with an upper dermal lymphohistiocytic inflammatory infiltrate and deep perivascular and periadenexal lymphocytes (**Figure 2E**); mucin deposits were confirmed by colloidal iron stain.

Laboratory testing examinations showed antinuclear antibodies 1/640 in a thick nuclear dotted line pattern and positive anti-RNP antibodies. The clinical, laboratory, and histopathology were consistent with CLE. Treatment with hydroxychloroquine 400 mg/day for 8 months and topical photo-protection resulted in remarkable regression of the lesions.

Case Discussion

The acneiform presentation of chronic CLE is uncommon [1-3] and commonly underdiagnosed due to the similarity with inflammatory acne in some cases.

Common acne usually manifests as comedones, pustules, and localized inflammatory nodules, generally in the face and upper trunk [3]. Our patient exhibited a clinical case remarkably similar to the inflammatory disease acne and was treated as such several times. Cystic acne and hidradenitis suppurative were the main conditions in the differential diagnostics.

Histopathologic studies in cases of acneiform lupus are of diagnostic importance. The most relevant findings for the establishment of CLE diagnostic are vacuolar degeneration of the basal layer of the epidermis and the thickening of basal membrane zone due to immunoreactive complex deposits [3]. The histopathologic findings of our patient were compatible with the literature [4,5]. This case emphasizes the relevance of knowing the different presentations of CLE and considering this condition in the differential diagnosis of inflammatory acne resistant to conventional acne treatment. Occasionally severe acne may be misdiagnosed as CLE [5]. This disease has an insidious evolution, and can be aggravated by ultraviolet radiation and some drugs [3].

Conclusion

Chronic cutaneous LE discoid-like lesions are in most cases distinct and well identifiable. However, they can sometimes mimic other dermatological conditions such as acne vulgaris leading to delayed diagnosis or inadequate treatment.

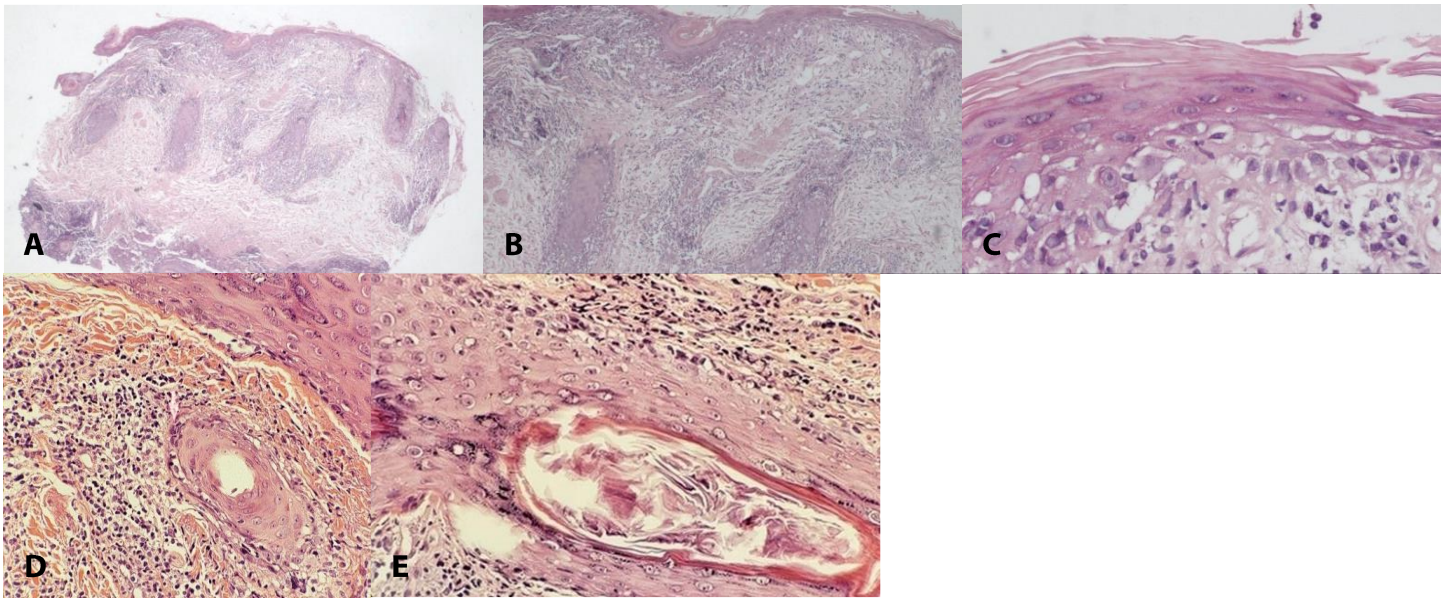

Figure 2. ***A)*** *Full-skin section showing upper and deep dermal cell infiltrate. H&E, 40×.* ***B)*** *Detail of the inflammatory cell infiltrate in vacuolar pattern bellow epidermis and around hair follicles. H&E, 100×.* ***C, D)*** *Detail of the vacuolar degeneration of the basal epidermal layer. H&E, 200×.* ***E)*** *lympho-histiocytic inflammatory infiltrate involving a hair follicle. H&E, 400×.*

Potential conflicts of interest

The authors declare no conflicts of interest.

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Table 1. *A summary of chronic cutaneous erythematosus mimicking acne reported in the literature indexed on PubMed until October 19, 2022.*

Ref	Age	Sex	Race	Lesions Location	Morphology	Path	Treatment	Result	Labs
Present case	57	Female	Black	Malar area, concha bowls	Multiple nodules, hyperpigmented, comedones, pustules, atrophic scars, brownish plate and follicular plugging	Right cheek region: atrophic epidermal layer, discrete hyperkeratosis, extensive vacuolar degeneration in the basal layer; the basal membrane zone was thickened on Schiff-periodic acid staining, with upper dermal lympho-histiocytic inflammatory infiltrate and deep perivascular and perianexial inflammatory cells, constituted overall by lymphocytes and vascular ectasia, and evidence of mucin deposits revealed by colloidal iron stain	Hydroxychloro-quine 400mg/day for 8 months, trimethoprim-sulfamethoxazole (800:400mg) b.i.d. during the first month and topical photo-protection	Remarkable regression of the lesions	ANA 1:640 and anti-RNP reagent
Ref 1	20	Female	Indian	Face, neck, back, and abdomen	Erythematous-infiltrated papules. Mild hair loss		Methotrexate 15mg once a week, folic acid 5mg once daily for 2 days in a week, and hydroxychloro-quine 200mg once daily. Deflazacort 15mg daily for 6 weeks followed by 12mg daily for another 6 weeks	She improved dramatically and the macula-papulopustular lesions became less prominent, with only residual skin pigmentation	Hemoglobin 13.2g/dL, leucocyte 9700/mm3, platelet 107,000/mm3, Antinuclear antibodies and anti double-stranded DNA highly positive. Negative anti-SSA, anticardiolipin, IgM, and IgG antibodies to B2 glycoprotein I complex and normal lupus anticoagulant Serum complement levels (C3, C4). Direct Coombs test positive. Normal serum vitamin B12 and folic acid were normal. Thyroid-stimulating hormone was found to be 24.2IU/mL. Serum vitamin D was at 15.1ng/ mL

Ref 2	57	Female	African-American	Bilateral upper neck, cheeks, chin, and forehead. Bilateral conchal bowls. Scalp	Brown papulonodules and cystic acneiform lesions and purple/brown papules and plaques with follicular plugging. resembling lesions of DLE. Nonscarring alopecia of the frontotemporal scalp and discrete circular patches of scarring alopecia	Follicular infundibular cyst with a lymphocyte-mediated interface dermatitis involving the interfollicular epidermis and the hair follicle. Moderately dense perivascular lymphocytic infiltrate. Upper neck: dermal fibrosis consistent with a cicatrix with chronic inflammation and hemosiderin deposition. Parietal scalp: mild fibrosing lymphocytic folliculitis with an attendant reduction in terminal hair density. 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	Hydroxychloro-quine, 200mg daily	She was lost to follow-up, precluding monitoring for response to therapy	Antinuclear antibody of 1:80 with speckled pattern, negative review of systems, negative double-stranded DNA, and normal basic laboratory panels
Ref 3	37	Female		Right ear, dorsal aspect of the nose and chin	Pruriginous acneiform eruption. Lightly-colored violaceous, infiltrated plaques of various sizes and atrophic scars. Comedones and pitting scars on a partially infiltrated plaque	Acanthosis, vacuolar degeneration of the basal cell layer, pigmentary incontinence, periadnexial mononuclear infiltrate, follicular plugging and comedones. Periodic acid-Schiff staining evidenced thickening of the basal membrane of the epidermis, more intense around the hair follicles	Topical sunscreen and systemic use of 500mg of tetracycline twice a day for three weeks and then 250mg twice a day for another thirty days. Then started on hydroxycloro-quine 400mg daily	Improvement of pruritus and the comedones. Discontinuation of the tetracycline resulted in acute recurrence of the lesions. After six months of use of hydroxycloro-quine there was improvement. During 12 months of follow-up no signs or symptoms of systemic disease were observed	

Ref 4	53	Female		Scalp, face, arms, and chest	Bright pink to violaceous, atrophic plaques. Extension of erythematous crusted papules onto the upper extremities and chest	Atrophic vacuolar interface dermatitis suggestive of connective tissue disease. Patulous follicles intimately associated with a vacuolar interface reaction and a permeative perijunctional lymphocytic infiltrate. Enlarged pale cells with abundant, vacuolated cytoplasm were situated in loose perifollicular collections in an elastotic dermis. S100 and adipophilin were negative in the cells of interest. CD68 labeled all of the pale vacuolated cells, confirming the presence of widely dispersed histiocytes. Of note, CD68 also co-labeled many of the lymphocytes. CD123 highlighted small clusters of perijunctional plasmacytoid dendritic cells, but was negative in the histiocytic infiltrate. Colloidal iron showed interstitial dermal mucin and peppered the intracytoplasmic material, substantiating histiocytes with engulfed ground substance	Macitentan was subsequently discontinued. Topical steroids were initiated along with recommend ations for photoprotection. Intralesional corticosteroid injection	The eruption gradually improved over several months. The lesion resolved after Intralesional corticosteroid	Positive antinuclear antibodies (1:640) and positive ribonucleoprotein. The following labs were negative or within normal limits: anti-Smith antibodies, aldolase, creatinine kinase, anti-Scl70 antibodies, and anti-ds-DNA antibodies. A serum lipid panel was within normal limits
Ref 5	12	Male		Face, forehead, glabella and both of cheeks. After on nose, chin, and both of temples	Multiple erythematous plaques, scars, opened and closed comedones, papules, and pustules. The lesion on the malar sparing of his nasolabial fold areas, seen in a slight sight, was similar to that of a malar rash in systemic lupus erythematosus	Mild spongiosis and neutrophil infiltrate in the epidermal layer, mixed infiltrate of lymphocytes, histiocytes, and neutrophils, a couple of foreign body granulomas in the dermal layer, and fibrotic tissue at the border of incision. Immunoglobulin (IgA, IgM, and IgG), C3 and C1q, and fibrinogen deposit were negative according to DIF staining examination	Topical alpha-hydroxy acid 10% lotion BID, topical benzoyl peroxide and clindamycin gel, topical retinoic acid 0.05% cream and glycolic acid face soap. Intralesional injection of 0.5mL triamcinolone acetonide 10mg/mL was also administered once a week for the management of the hypertrophic scar	Some improvements were observed: fewer comedones, papules, and pustules; a more diffuse erythematous base; and minimal shrinkage of the hypertrophic scars	Routine hematological testing, complete urinalysis, complement 3 level, C4 level, anti-double-stranded deoxyribonucleic acid level, and antinuclear antibody level. Antinuclear antibody with a titer of 1:100 and a speckled pattern

Ref 6	52	Female	South American	On the surface located on the pinnae, both nasolabial folds, and chin. Exposed areas of arms and shoulders	Multiple tumoral plaques with areas of cystic and pustular appearance. Small erythematous plaques with shiny-white appearance surrounded by brownish red areas	Interface dermatitis with vacuolar degeneration of the basal layer, superficial and deep periadnexal and perivascular lymphocytic infiltrates. The epidermis showed marked hyperkeratosis, vacuolar degeneration, and interface dermatitis. Areas of thickening of basement membrane. Multiple cysts and comedonal dilation of the infundibula of the hair follicles with marked plugging and periadnexal and perivascular superficial and deep mononuclear cell infiltrates, with clusters of CD123+ cells	Hydroxychloro-quine. A course of oral corticosteroid was also prescribed	Little improvement. Corticosteroid was also prescribed with significant improvement	Complete blood test, with autoantibodies and autoimmune serology, showed normal or negative results, except for a low titre of ANA (1/160)
Ref 7	32	Female		Right chin area, ear concha and abdomen, and scalp	Pruritic acneiform and comedonal eruption. Erythematous-infiltrated plaque, papules, open comedones, pitting scars and hypopigmented atrophic scars. Erythematous plaques, some with atrophy	Scalp: dense superficial and deep perivascular and periadnexal lymphocytic infiltration, and signs of lichenoid infiltrate. Chin: basal layer vacuolar degeneration, superficial and deep perivascular, and periadnexal mononuclear infiltration at the dermis and exuberant follicular hyperkeratosis and plugging. Melanophages. Direct immunofluorescence showed presence of homogeneous continuous deposition of immunoglobulin M in the basement membrane zone and positivity stain of upper dermal vessels	40mg of prednisone for one month with a progressive dose reduction for four months, and 250mg of chloroquine diphosphate	Significant improvement with less inflammation and pruritus. After four years the lesion is stable, with some scarring, but without pruritus, inflammation, or new lesions	Blood inflammatory markers, antibody profile, serology for HIV, Hepatitis B, C, venereal disease research laboratory, and radiological investigation all negative
Ref 8	32	Male		Right nasolabial fold	Conspicuous acneiform pitting scars, that were preceded by erythematous plaques. Perilesional erythematous infiltration and telangiectasia	Irregular acanthosis, follicular plugging, vacuolar degeneration of the basal cell layer with marked melanin incontinence, and heavy periadnexal mononuclear cell infiltration. Direct immunofluorescence studies displayed continuous granular deposition of immunoglobulin G (IgG) and C3 along the dermo-epidermal junction			The hemogram, antinuclear antibody test, complement, and urinalysis were within normal limits

Ref 9	30	Female		Face	Inflammatory lesions and comedones after pregnancy. Raynaud phenomenon and diffuse alopecia	Characteristic of chronic lupus erythematosus. Immunofluorescence microscopy of lesional skin showed a lupus band deposit	Chloroquine	Successfully treated. Three years later, the patient presented with photodistributed eruption. Antinuclear antibodies were still positive and in addition anticardiolipin antibodies were found	Antinuclear antibodies were highly positive

ANA, anti nuclear antibody; F, female; M, male;.