Notes & Comments

Comedonal variant of chronic cutaneous lupus erythematosus causing mutilation of the earlobe



To the Editor: We read with interest the article by Droesch and Magro¹ reporting on the comedonal variant of chronic cutaneous lupus erythematosus (CCLE).

This case report highlighted the importance of recognizing this extremely rare variant of cutaneous lupus, which is easily mistaken for acne vulgaris, but can lead to important scarring if misrecognized and mistreated and can also represent the warning sign of concomitant systemic involvement, requiring adequate management. We report a similar case to further highlight this entity and the significant consequences it can lead to, if neglected.

A 45-year-old woman with a history of alcohol abuse presented to our clinic with pruritic, brown-red, infiltrated plaques of both ears for 1 year, with open comedones and atrophic scarring. The right earlobe was disfigured by a destructive scarring process (Figs 1 and 2).

She also had typical discoid lesions with dyspigmentation and scarring on her face, especially on the nose, that she said had been present for 25 years. Informed consent was obtained from the patient.

Histopathology confirmed CCLE, showing epidermal atrophy, extensive vacuolar degeneration of the epidermal basal cell layer, superficial and deep dermal perifollicular and perivascular lymphocytic infiltrate, and marked follicular hyperkeratosis and plugging. Direct immunofluoresshowed granular deposits of IgG, immunoglobulin M, and C3 at the dermoepidermal junction and around hair follicles. Test results for antinuclear antibodies and antiextractable nuclear antigens were negative. Laboratory investigations revealed high levels of liver enzymes and megaloblastic anemia. Because of hepatic impairment, we did not administer hydroxychloroquine and the patient was advised to apply only sunscreens meticulously.

During follow-up, most of the cutaneous lesions remained almost stable and the patient never developed signs of systemic erythematous lupus, antinuclear antibodies, or antiextractable nuclear antigens. The destructive scar on her right earlobe deteriorated further. She died of cirrhosis at aged 54 years.



Fig 1. Comedonal chronic cutaneous lupus erythematosus on the ears, depicted by brownred, infiltrated plaques, with open comedones in the right ear's concha and atrophic scarring. The right earlobe is disfigured by a mutilating scarring process.

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Fig 2. Close-up of the open comedones (arrow) of the right ear's concha.

Comedonal CCLE is an exceedingly rare but documented variant of CCLE: only 13 cases have been reported in the literature,70% of which occurred in women with a mean age of 32 years. 1-5 Concurrent systemic erythematous lupus was reported in 4 cases. 1,4,5 Comedonal CCLE can be mistaken for benign dermatologic conditions such as nevus comedonicus, Favre-Racouchot disease, agminate lichen follicularis with cysts and comedones, and especially acne vulgaris.^{3,6} However, unlike these benign entities, CCLE bears a highly destructive potential, with atrophic scarring and even mutilations occurring in untreated disease, as in our case. Also, 31% of individuals with comedonal CCLE have concomitant systemic erythematous lupus, with a significant potential morbidity associated with diagnostic and therapeutic delay. 1,4,5

We add to the existing literature on this less well-known subtype of CCLE. We hope our article adds further value to the case report by Droesch and Magro, promoting caution regarding acnelike lesions that are not responsive to acne therapy, especially in young women, because a comedonal CCLE may be the underlying cause of the lesions and may become irreversibly mutilating and even life threatening in concomitantly systemic cases if misdiagnosed and untreated.

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