

# Acneiform Presentation of Chronic Cutaneous Discoid Lupus Erythematosus, Treated with Isotretinoin: A Case Report

Lencinas VN<sup>1</sup>, Guidi AE<sup>2</sup>, Consigli CA<sup>1</sup>

<sup>1</sup>Department of Dermatology, National Hospital Clinics, National University of Córdoba, Argentina

<sup>2</sup>Department of Pathology, Fabiola's Queen University Clinic, Catholic University of Córdoba, Argentina

\*Correspondence: Lencinas VN, Department of Dermatology, National Hospital Clinics, National University of Córdoba, Argentina

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## Abstract

Chronic cutaneous discoid lupus erythematosus (CCDLE) is the most common clinical variant of chronic cutaneous lupus erythematosus (CCLE). There are rare forms of CCDLE, one of them is acneiform presentation, which may include comedones, scars, and depressions or dimples. It is often underdiagnosed because of its similarity with inflammatory acne vulgaris. We present a case of Acneiform presentation of CCDLE, located on the left cheek of a young woman, who responded well to treatment with Isotretinoin per os.

**Key words:** Chronic cutaneous lupus erythematosus; Chronic cutaneous discoid lupus erythematosus; Sistemic erythematosus lupus; Acneiform presentation; Comedones; Isotretinoin.

## INTRUDUCTION

Chronic cutaneous discoid lupus erythematosus (CCDLE) is an autoimmune disease, whose pathogenesis involves environmental, hormonal, and genetic factors.<sup>1</sup> It is the most common clinical variant of chronic cutaneous lupus erythematosus (CCLE) [1]. It has a chronic and insidious evolution, characterized by the absence of regression of lesions and resistance to treatment [2,3]. It is more frequent between the '20s and the '40s, although it can be seen in infants and the elderly. It affects all races, but there are studies that suggest a higher prevalence in black people. It is more common in women [1,2]. CCDLE presents circular, erythematous, and well-defined plates, covered with adherent scales; removing them causes pain and follicular plugging; in its evolution, erythema and hyperpigmentation appear in the periphery, and an atrophic distinctive central scar, telangiectasia, and hypopigmentation. It affects visible areas of skin exposed to sunlight, such as the face, scalp, arms, and hands [4,5]. CCDLE lesions may evolve with scars, disfiguring ones in some cases, causing psychosocial disorders [1,4,5]. In older lesions, atrophic scars associated with warty hyperkeratosis can be seen, and on several occasions of chronic evolution, scars can develop malign tumors [3,6].

Exacerbating factors include sun exposure, cold, trauma, stress, progesterone, estrogen, and drugs (griseofulvin, dapson, D-penicillamine, procainamide, isoniazid) [3].

Astoptical treatment of CCDLE, glucocorticoids, calcineurin inhibitors, and intralesional triamcinolone acetonide are used. As a systemic treatment, hydroxychloroquine sulfate, chloroquine, and quinacrine, azathioprine, dapson, thalidomide, and oral retinoids are prescribed, among other medications [1,7]. As a treatment for scar sequelae, the erbium-YAG, and CO<sub>2</sub> lasers, have obtained a remarkable aesthetic improvement [8].

We present a case of Acneiform presentation of Chronic Cutaneous Discoid Lupus Erythematosus, located on the left cheek of a young woman, who responded well to treatment with Isotretinoin per os.

## CASE REPORT

A 24-year-old female, with a history of CCDLE, 4 years of evolution, no personal or family acne history, treated with corticosteroids and hydroxychloroquine 200mg/day during the last three months with little clinical improvement. At the time of consultation, she had an indurated, well-defined, erythematous plate, with a 6 x 7 cm diameter and whitish adherent scales.

Regarding nasolabial folds, it also presented areas with hyperkeratosis, scars, atrophy, and comedones grouped by sectors. On the nose bridge, we observed 3 discoid plates (2 cm in diameter) with irregular edges like the previously described lesion (Figure 1).

Complementary examinations were requested: Hemogram, blood glucose, uremia, creatinemia, complete urine, CPK, liver function tests, Complementemia, cholesterol, and triglycerides, resulting in normal parameters. Dosage of B subunit of human chorionic gonadotropin (hCG) was also performed to rule out pregnancy and immunological profile for lupus. Both studies were negative. The liver function tests, lipid profile, and the dosage of B subunit of hCG were repeated on the second and fourth months of treatment.

A biopsy of the lesion was performed, which revealed thinned epidermis with hyperparakeratosis, comedogenic expansion in one of the margins, and chronic periadnexal and perivascular inflammatory infiltrate (Figure 2).

With the histopathological findings, laboratory and clinical characteristics, the diagnosis of CCDLE with



Figure 3: The histopathological findings, laboratory and clinical characteristics.

the acneiform presentation was confirmed. Treatment with Isotretinoin 0.75 mg/kg/day during 5 months, contraception, and sunscreen were indicated. In the clinical control, the patient had a noticeable improvement, which was demonstrated by the reduction of erythema, infiltration, number of comedones, and the disappearance of the scales (Figure 3).

## DISCUSSION

There are other rare forms of CCDLE, as discoid lesions on eyelids, injuries with an acneiform appearance, nails' lesions with hyperkeratosis of the nail bed, and periungual inflammation [4]. The acneiform lesions may include comedones, scars, and depressions or dimples [9-11]. The CCDLE with acneiform presentation or CCDLE comedogenic is an uncommon clinical manifestation, and it is often underdiagnosed because of its similarity with inflammatory acne vulgaris [9,12-14]. The mean time to diagnosis among reported cases is 3.7 years [15]. 31% of individuals with Comedonal CLE have concomitant systemic erythematosus lupus, with significant potential morbidity associated with diagnostic and therapeutic delay [12]. Itching and photo sensibility in acneiform lesions are important features to guide diagnostic suspicion of CLE [13].

The cause of this type of disease is not clear yet and its prognosis is uncertain [9]. Yu-Hao et al. described a case of CCDLE with acneiform pitting scars in a 32-year-old man. They considered that the lesions were possibly caused by a destructive process of the pilosebaceous units, attributed to a periadnexal mononuclear inflammatory cell infiltration [10]. Among the differential diagnoses of CCDLE comedogenic, it can be included: acne vulgaris, Favre-Racouchot disease, milium, milia en plaque, syringoma, trichoepithelioma, clustered dilated pores, and nevus comedogenic [9,16].



Figure 1: The patient experienced photosensitivity.

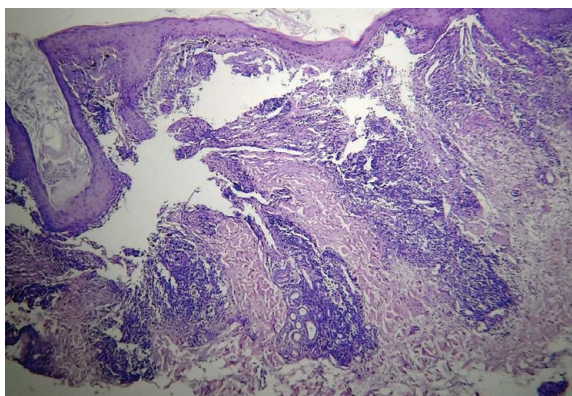


Figure 2: A skin lesion biopsy.

The first use of isotretinoin in CCLE or lupus erythematosus subacute was reported by Newton et al. in 1986 [17]. In the study 8 patients were treated with isotretinoin 80 mg/day (40 mg twice daily), all showing improvement at 16 weeks. The first change observed in all patients was a simultaneous decrease in erythema and peeling, at 4 weeks of treatment [18]. Shornick et al. treated 6 patients with cutaneous lupus erythematosus with isotretinoin at a dose of 1 mg/kg/day, resulting in effective and well-tolerated [17]. All patients in both studies showed resistance to conventional therapies.

Shornick et al. reported that is often possible to reduce the dose to 0.5 mg/kg/day, after initial improvement, but their experiences indicate that there is little benefit to be achieved below this level [17]. However, a report of Vena, et al. described 24 patients with CCLE and subacute cutaneous lupus erythematosus that had been treated with isotretinoin 0.15 to 0.5 mg/kg/day for 16 weeks, in which 87% of the patients showed clearing or improvement of clinical lesions and histopathologic changes [19].

The action of retinoids on suppression of sebaceous activity, its effects on the microflora, its anti-inflammatory and immunomodulatory effects on cell growth and differentiation, reduction follicular keratosis and cosmogenesis are well known (apparently by decreasing formation and/or growth on the degree of separation of the corneocytes within the ductus pilosebaceous) [20]. All these actions are important in the treatment of a wide range of cutaneous diseases, mainly keratinization disorders and proliferative diseases [19].

The indicated dose is usually 1mg/kg/day for 11 to 16 weeks [17,19,21,22]. In a case report of a young woman with CCDLE, isotretinoin was used as a maintenance dose (20 mg daily or 40 mg every other day), after finishing treatment [23].

In conclusion, we report a very rare case of CCDLE with the acneiform presentation that is barely mentioned in the classical books for the study of this area of expertise.

Due to certain aspects of our clinical case presentation, such as the lack of response to conventional therapy and the presence of comedones, added to the bibliographic history of use of oral retinoids in CCDLE, it was decided the use of oral isotretinoin. This drug has shown to be effective in the treatment of CCDLE resistance to other treatments and intermittent treatment of acute flares in selected patients and in CCDLE of atypical presentation [18,24].

## CONFLICT OF INTEREST

None.

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