

Comedonic lupus: a rare presentation of discoid lupus erythematosus *

Lupus comedônico: rara apresentação do lúpus eritematoso discóide

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Abstract: Chronic cutaneous lupus erythematosus is a polymorphous autoimmune disease which may mimic some other clinical conditions, causing diagnostic difficulties. Acneiform lesions, including comedones and pitting scars are occasionally atypical presentations of cutaneous discoid lupus erythematosus. **Keywords:** Lúpus erythematosus, cutaneous; Lúpus erythematosus, discoid; Lupus vulgaris

Resumo: Lúpus eritematoso cutâneo crônico é doença autoimune, com apresentações polimorfas que podem, eventualmente, mimetizar outras condições clínicas, causando dificuldade diagnóstica. Quadro acneiforme, com comedões e lesões atróficas pontuadas (pitting scars) pode constituir apresentação atípica do lúpus eritematoso cutâneo discóide.

Palavras-chave: Lupus eritematoso cutâneo; Lupus eritematoso discóide; Lúpus vulgar

INTRODUCTION

Chronic cutaneous discoid lupus erythematosus (CCDLE) or discoid lupus erythematosus (DLE) is a rare chronic auto-immune dermatosis and the most common clinical variant of the lupus erythematosus (LE). It is caused or triggered by exposure to ultraviolet radiation, cold and drugs and develops gradually. The diagnosis of DLE is confirmed by histopathological examination. Uncommon clinical manifestations, like comedonic lupus can mimic other diseases like acne vulgaris, Favre-Racouchot disease, milium, milia *en plaque*, syringoma, tricoepithelioma, clustered dilated pores and nevus comedogenic.

CASE REPORT

35 years old female, reported a pruriginous acneiform eruption on the face for two years, which did not respond to conventional treatments for acne. The dermatologic examination showed lightly-colored violaceous, infiltrated plaques of various sizes and atrophic scars on the right ear, dorsal aspect of the

nose and chin. Comedones and pitting scars were noted on a partially infiltrated plaque on the chin (Figure 1). CCDLE was considered. The histopathological examination showed acanthosis, vacuolar degeneration of the basal cell layer, pigmentary incontinence, periadnexial mononuclear infiltrate, follicular plugging and comedones. PAS staining evidenced thickening of the basal membrane (BM) of the epidermis, more intense around the hair follicles (Figure 2). Treatment started with sunscreen applied to the skin and systemic use of 500mg of tetracycline twice a day for three weeks and then 250mg twice a day for another thirty days. There was improvement of the pruritus and the number of comedones (Figure 3). With discontinuation of the tetracycline there was an acute recurrence of the lesions. The patient was then started on hydroxychloroquine 400mg daily with clinical improvement in 45 days (Figure 4). After six months of use of hydroxychloroquine there was marked improvement (Figure 5). During twelve months of fol-

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FIGURE 1: Infiltrated erythematous-violaceous plaque with comedones and pitting scars



FIGURE 3: Partial improvement after use of tetracycline

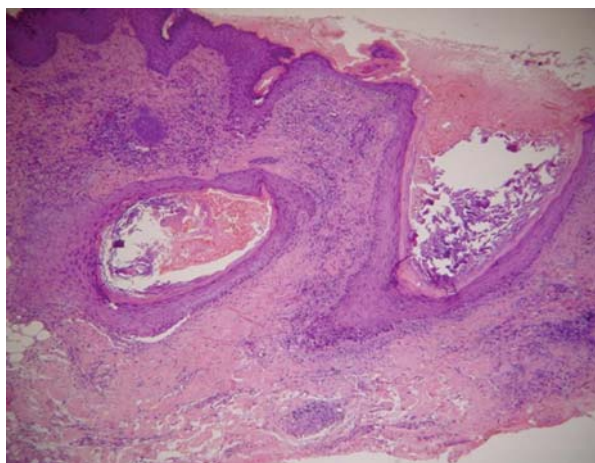


FIGURE 2: Vacuolar degeneration of the basal cell layer, periadnexial inflammatory infiltrate, follicular plugging and comedones, (PAS, 10X)



FIGURE 4: Partial improvement on the 45th day treatment with hydroxychloroquine

low-up no signs or symptoms of systemic disease were observed.

DISCUSSION

The clinical manifestations of CCLE are multiple and varied. The acneiform presentation of discoid CCLE is rare and only six cases have been reported so far.¹⁻⁷ this presentation is usually under diagnosed due to its similarity to inflammatory acne vulgaris.¹ The cases reported showed a predominance of women aged 25 to 35 and the symptoms that most commonly lead to the diagnosis were pruritus and photosensitivity. The cause of the comedogenic form of the CLE is not yet clear and the prognosis is uncertain. It is supposed that at least half of the patients with this condition have the tendency to develop systemic LE.⁵

The differential diagnosis of comedogenic dis-

coid CLE are: acne vulgaris, Favre-Racouchot disease, milium, milia em plaque, syringoma, tricoepithelioma, clustered dilated pores and nevus comedogenic.^{5,8} Acne vulgaris usually manifests as comedones and inflammatory papules and nodules, most commonly on the face and trunk. Favre-Racouchot disease, also known as 'cutaneous nodular elastoidosis with cysts and comedones' is characterized by volumous open, black comedones, located on the sun damaged skin of the elderly.⁸ The nevus comedogenic, uncommon variant of the adnexial hamartoma, appears as linear groups of open comedones and, in 50% of the cases, they are present at birth.⁵ Colloid milium is a rare degenerative disease, with development of small translucent, yellow to brown papules, nodules or plaques usually on sun damaged areas.⁹ Milia *en plaque* is a rare clinical variant of the milium, characterized by multiple milium-like lesions over ery-



FIGURE 5: Marked improvement after 6 months' treatment with hydroxychloroquine

thematous, edematous plaque, usually in middle-aged women.¹⁰ Syringomas are adnexial tumors, more common in women, typically seen as soft, translucent or lightly yellow papules around the eyes.¹⁰ Tricoepithelioma is a rare benign lesion, originated from the hair follicles, developing mostly on the face and scalp.¹¹

The diagnosis of comedonic DLE was confirmed mainly by the histopathological changes. The histological findings are comparable to those on the literature review, like: hydropic degeneration of the

basal layer, thickening of the BM, follicular dilation, hyperkeratosis, lymphocyte inflammatory infiltrate, and melanophages on the dermis.¹² From all those, the most relevant findings to the establishment of the diagnosis of LE are the hydropic degeneration of the basal layer and the thickening of the BM, that occurs due to the deposition of reactive immune complexes and, in many cases, is seen only with the progression of the disease.¹³ On the present case the identification of the comedones on the anatomopathological examination associated to the related changes permitted the confirmation of the diagnosis of comedonic LE.

Tetracycline was suggested at the beginning of the treatment and an antiinflammatory and immunoregulator agent that has been successful on the treatment of acneiform conditions.¹⁴ Other therapeutic options include oral hydroxychloroquine (400mg daily), topical tretinoin (0.025% cream), oral isotretinoin (1mg/kg/day), triamcinolone acetate (injected, 10mg/cc every 6 weeks), topical clobetasol (0.05% lotion twice a day) and mechanical extraction of the comedones.^{1,3}

The present case reinforces the importance of taking into consideration the various forms of presentation and the atypical behavior of discoid CLE, like the presence of acneiform lesions, accompanied by suggestive signs of CLE, that do not respond to conventional treatment to acne vulgaris. □

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