

Medical and Surgical Treatment for Discoid Lupus Erythematosus

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SUMMARY A case is presented of a 27-year-old Caucasian woman that presented with an oval purplish patch with defined borders on her right cheek. It was swollen and tense, with a mottled surface due to scarring. Histologic examination indicated discoid lupus erythematosus and the patient was initially treated with medical therapy. The lesion relapse and cosmetic results convinced us to propose surgical option with aggressive medical treatment and follow up. In our patient, this combination therapy proved effective.

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INTRODUCTION

Discoid lupus erythematosus (DLE) is a chronic benign skin disease frequently involving the face and characterized by red papular lesion flatly delimited, with adherent scales, scarring and pigmentary changes (1). In many cases, cosmetic disfigurement makes this clinical condition more severe and it is very difficult for the dermatologist to manage such cases only by medical therapy.

We report on a female patient affected by severe DLE on the face and treated with combined medical and surgical therapy. Surgery could be an important option in the treatment of DLE forms

resistant to medical therapy and characterized by persistent scarification and cosmetic disfigurement.

CASE REPORT

A 27-year-old woman developed papular-pustular lesions on her right cheek, misdiagnosed in the past as psoriasis and annular granuloma. After three months, these lesions turned to a single confluent tumor-like infiltrated lesion. The patient presented with a 6x5 cm oval infiltrated patch with defined borders. This lesion was characterized

by a central erythematous and squamous area, surrounded by papules and diffused scarification and scales. The borders were erythematous and surrounded the entire patch, resembling lupus tumidus. It was not painful and the patient did not report any other symptoms. Physical examination of the skin was negative for other similar lesions. This clinical aspect suggested biopsy in search for DLE (Fig. 1).

Histology is crucial to make the diagnosis. Atrophic epidermis with characteristic keratotic plugging is one of the salient features; moreover, liquefaction degeneration of the basal layer is present in the context of hyalinization and edema of the connective tissue (2). Histopathology confirmed an interface dermatosis with hypergranulosis and follicular hyperkeratosis, dystrophic epidermal pattern, and an important perivascular and periadnexal involvement, consistent with the clinical diagnosis of DLE. An additional characteristic of DLE might be intense periadnexal and perivascular infiltrate extending into the interstitium and subcutaneous tissue, and consisting of lymphocytes, histiocytes, eosinophilic granulocytes and plasma cells (3,4).

Moreover, granular deposits of IgG, IgM, IgA, C1q, C3, C4, C5 and C9 at the dermoepidermal junction were found by direct immunofluorescence. The patient underwent additional diagnostic work-up for possible systemic involvement.



Figure 1. A 6x5 cm oval red purplish infiltrated patch on the patient's right cheek.

Laboratory testing did not suggest any systemic damage or involvement. Erythrocyte sedimentation rate, blood count and serum immunoglobulins were normal; ANA, ENA n-DNA were negative, and complement (C3-C4) was normal. Creatinine level was within the normal range and there was no microscopic hematuria. Chest x-ray and abdominal ultrasonography were unremarkable. The patient was treated with hydroxychloroquine 200 mg daily and thalidomide 100 mg daily for six months, and the lesion disappeared completely in four months. However, the patient developed relapse of the lesion that showed no substantial improvement upon the introduction of chloroquine 250 mg daily for six months. The failure of first-line systemic therapy required patient admission to the hospital to initiate administration of mycophenolate mofetil 1 g daily and methylprednisolone 8 mg daily. After five months of therapy, we observed mild improvements, without enlargement or new lesions, with infiltration reduction and appearance of normal skin in the center of the lesion. Nevertheless, the slow course of consistent improvement suggested conversion to combined medical and surgical treatment. This option included a series of surgical interventions in general anesthesia. In the first act, the lesion was removed and repaired by an expanded skin flap, after removing the skin expander that had been previously inserted in her right cheek. The surgical procedure was successful and the patient continued her medical therapy for one year to avoid relapses. Then she decided to stop immunosuppressant therapy, however, relapse developed in several months. Another similar infiltrated patch grew on her right cheek and she underwent surgical removal of the relapsing lesion. The greatest part of the lesion was repaired by an expanded skin flap and a small nasogenian area was repaired by a skin graft obtained from the redundant expanded skin derived from the skin expander previously localized in the preauricular zone.

Up to now, after these surgical interventions, she has continued therapy with mycophenolate mofetil and methylprednisolone, in consultation with plastic surgeon and dermatologist. The clinical and cosmetic results are excellent and no sign of relapse was found during almost two-year follow up (Fig. 2).

DISCUSSION

This clinical condition is characterized by frequent relapses and management is often difficult.



Figure 2. The patient's right cheek after surgical interventions.

The first-line medical therapy includes potent topical steroids, with chloroquine or hydroxychloroquine as the mainstay of treatment. Some cases of DLE can be refractory to standard therapy; in these cases retinoids, thalidomide, and topical tacrolimus offer alternatives, as do immunosuppressants like azathioprine, cyclosporine, mycophenolate mofetil, and methotrexate (5). In the end, there is surgical option but it is strictly limited to few cases. DLE typically involves the face and its effects can be devastating and become a handicap for the patient.

The young age of our patient, the continuing relapses of the disease on her cheek, and inadequate response to the first-line medical therapy suggested a combination of medical, non standard alternative and surgical approach. Upon excision of the lesion, genetic predisposition remains the

same and precipitating factors can still influence the disease (6). So, in our case, it was very important to combine these two therapeutic methods in an attempt to achieve an immediate cosmetic result and relapse-free condition at long term. By appropriate clinical management and tight follow up the common collateral damages associated with immunosuppressant therapy were successfully avoided.

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