

Dark brown scaly plaques on face and ears

A 27-year-old woman, otherwise healthy, presented for evaluation of a mildly pruritic eruption of plaques on her face and ears. The eruption started as a few small, scaly red papules on her cheeks and nose about 3 months earlier. The papules slowly expanded to form well-demarcated, rounded, dark brown plaques covered with superficial scale. Similar lesions appeared on the concha of both ears. The older lesions started to regress after 2 months. Complete regression was followed by residual scarring and hyperpigmentation.

Physical examination revealed multiple well-defined, erythematous, hyperpigmented, round plaques covered with adherent scale affecting her nose, cheeks, and the concha of both ears (**FIGURE 1**). The mucosae and the rest of her skin and adnexa were unaffected. She had no personal or family history of similar skin findings or autoimmune disorders. She also had no history of any drug intake prior to the eruption. Her routine blood tests and urinalysis results were unremarkable, and the serological analysis for antinuclear antibodies had negative results.

A punch biopsy was taken from one of the lesions for histopathology. The epidermal histopathological findings were remarkable for hyperkeratosis, follicular plugging, pigment incontinence, and vacuolization of the basal cell layer. There was a predominantly lymphocytic

FIGURE 1

Facial plaques



Well-demarcated dark brown-colored scaly erythematous plaques. Hyperpigmented scars resulting from the spontaneous healing of older lesions are also visible.

infiltrate of the subepidermal and perivascular dermal areas.

- **What is your diagnosis?**
- **How would you treat this patient?**

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■ **Diagnosis: Discoid lupus erythematosus**

Discoid lupus erythematosus (DLE) is an autoimmune inflammatory disorder of the skin that often leads to scarring and alopecia. It may be localized to sun-exposed areas such as the face, ears, and scalp but occasionally is much more extensive, involving the trunk and extremities. Most patients are otherwise healthy, and DLE may be the only clinical finding.

Although its prevalence is not known, DLE is not uncommon. It affects females twice as often as males, and patients fall between the ages of 25 to 45 years, with no racial predilection. While about 15% to 20% of patients with systemic lupus erythematosus (SLE) manifest DLE lesions, only about 5% to 10% of patients with DLE go on to develop SLE.¹

Like SLE, DLE is believed to be an autoimmune disorder. Unlike SLE, however, DLE patients do not have similar serologic abnormalities. Skin trauma and ultraviolet light exposure have been reported to induce or exacerbate the lesions of DLE. Sex hormones may also play a role: exacerbation may occur during pregnancy, during menstrual or premenstrual periods, or while taking oral contraceptives. Drugs such as procainamide, hydralazine, isoniazid, diphenylhydantoin, methyl dopa, penicillamine, guanides, and lithium may also precipitate DLE lesions.²

DLE usually begins as dull red macules with adherent scales on sun-exposed areas. If the overlying scales are removed, an undersurface of horny plugs is revealed. These plugs fill the follicles and resemble carpet tacks or a cat's tongue (*langue au chat*). The macules slowly expand to form large plaques.

Usually only a mild pruritus and tenderness is seen with DLE lesions. As the lesions progress, the scale may thicken and pigmentary changes become evident. The lesions heal with atrophy, scarring, telangiectasias, and changes in pigmentation. Morphological appearance of older lesions may vary from erythematous plaques to hyperkeratotic, dark gray plaques with

centrally depressed scars.³ Scalp involvement in DLE results in more sclerotic and depressed scars with subsequent scarring alopecia.⁴

Differential diagnosis is wide. The differential diagnosis of DLE is extensive and includes actinic keratosis, dermatomyositis, granuloma annulare, granuloma faciale, keratoacanthoma, lichen planus, subacute cutaneous lupus erythematosus, psoriasis, rosacea, sarcoidosis, squamous cell carcinoma, syphilis, and nongenital warts.

■ **Histopathology**

Histopathological findings include hyperkeratosis, parakeratosis, follicular plugging, telangiectasias, and atrophy of the epidermis. Liquefaction or hydropic degeneration of the basal layer leads to pigmentary incontinence. A perivascular and perifollicular mononuclear inflammatory cell infiltrate is present in the superficial and deep dermis.^{5,6}

Direct immunofluorescence demonstrates immunoglobulins and complement deposits at the dermoepidermal junction.⁵ This test was not recommended for this patient, as the diagnosis of DLE had already been confirmed on clinical and histopathologic grounds.

■ **Workup: Exam, biopsy**

In addition to a routine history and physical examination, the workup for DLE should include a complete blood count, antinuclear antibody levels, anti-Ro, anti-La, hepatic and renal function tests, and urinalysis. Consider a diagnosis of SLE by using the American College of Rheumatology criteria. A biopsy for histopathology of a fresh lesion or a biopsy for immunofluorescence of an old lesion can confirm the diagnosis.

■ **Therapeutic options are varied**

Effective early therapies for DLE are available, but patients who do not

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The primary therapeutic approach: educate patients regarding exposure to sunlight

respond appropriately may end up with deep scars, alopecia, and pigmentary changes that are considerably disfiguring, especially for dark-skinned people. Therefore, the goal of treatment is not only to improve the appearance of the skin by minimizing the scarring and preventing further lesions, but also to prevent future complications.

Avoiding sunlight. The primary therapeutic approach is to educate patients regarding exposure to sunlight. Sun-protective measures include the use of high-SPF sunscreen lotions and protective clothing, such as baseball caps without vent holes and wide-brimmed hats.⁷

Medication options. Current treatment options for DLE include antimalarial agents such as chloroquine, topical and intralesional glucocorticoids, and thalidomide. The use of potent topical steroids may prevent significant scarring and deformity, especially of the face. Common side effects include steroid withdrawal syndrome, perioral dermatitis, steroid acne, and rosacea. All of these side effects can be treated and result in less long-term deformity than untreated DLE (FIGURE 2).

Systemic agents. Widespread disease may require you use systemic agents, such as antimalarials. Chloroquine has long been considered the gold standard in the treatment of DLE.⁸ Due to the frequency of ocular side effects with chloroquine, hydroxychloroquine is by far the most widely used agent. Hydroxychloroquine at a dose of 6.5 mg/kg/d for 3 months may lead to resolution of lesions for many patients.⁷ In resistant cases, higher dosages (eg, 400 mg/d) or combinations (eg, hydroxychloroquine 200 mg/d plus quinacrine 50 to 100 mg/d) may be required for months or even years. An ophthalmological evaluation is advisable before starting antimalarial treatment, and you should repeat it at 4- to 6-month intervals during treatment.⁹ Systemic corticosteroids may be needed to obtain timely initial control, especially for widespread and disfiguring lesions.

When this therapy is inadequate,

FIGURE 2

After treatment



Perceptible regression of the lesions after one month of therapy with topical steroid cream and oral hydroxychloroquine.

other treatments for DLE include azathioprine, retinoids, and dapsone. Recent reports confirm the efficacy of thalidomide for cutaneous lupus erythematosus in dosages ranging from 50 to 200 mg/d.¹⁰ Twice-daily application of tacrolimus 0.01% has also been shown to be effective in a few clinical trials.¹¹

Surgery. Surgical intervention is useful to remove scarred lesions, and laser therapy has also been effective, especially for lesions with prominent telangiectasias.¹²

Patient education. Patient education plays an important role in the treatment of DLE. Advising patients on how to avoid the sun and instructing them in the proper use of sunscreens is extremely important. Smoking cessation has also been shown to be beneficial.¹³

Patients with DLE generally have a favorable prognosis with regards to morbidity and mortality. Because DLE is usually self-limited, the course is most often benign; therefore, early recognition and adequate therapy may prevent clinical complications. Many patients with DLE

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Widespread disease may require systemic agents (eg, anti-malarials); surgery is useful to remove scarred lesions

go on to develop destructive or deforming scarring or pigmentary disturbances,¹⁴ especially in the spring and summer months when the sun is the strongest.

■ This patient's outcome

Our patient was treated with topical fluocinolone acetonide 0.025% ointment and oral hydroxychloroquine 200 mg/d for 1 month. The patient responded well to the treatment and the skin lesion regressed perceptibly (**FIGURE 2**). The treatment was nevertheless continued for another 5 months, and resulted in complete regression of the lesions, with minimal residual scarring.

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