40-Year-Old Female with Pink Scaly Patches in the Ears and on the Scalp

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Bryce Demoret, Peter Noll, Suchita Sampath, and Shannon C. Trotter

Abstract

Lupus erythematosus is a multisystem disorder that predominantly affects the skin. Discoid lupus erythematosus is the most prevalent subset of chronic cutaneous lupus affecting, most commonly, women in their fourth and fifth decades of life. The lesions of DLE are characteristically well-defined, annular, erythematous patches and plaques and frequently present on photo-distributed areas of the skin. These lesions also tend to have secondary atrophy and scarring. As a result, when present on the scalp, DLE can progress to irreversible scarring alopecia. Although the pathogenesis is not entirely known, there is increasing evidence to suggest that ultraviolet light, environmental, genetic, and immunologic factors may contribute to the development of DLE. The early detection and treatment of DLE are essential to minimize the risk of scarring alopecia. Initial measures for individuals with DLE should involve education regarding exacerbating factors such as UV light exposure and tobacco use. Additionally, patients should be informed of the scarring nature of the disease process and that management aims to improve appearance, control acute lesions and limit further scarring. Treatment with topical and intralesional corticosteroids is considered first-line therapy. Novel treatment methods such as pulsed dye lasers and immune-modulating agents are being investigated for treatment-resistance DLE and have shown promise in initial clinical trials.

Keywords

Discoid lupus: Discoid lupus erythematosus · DLE · SLE · Alopecia · Cicatricial alopecia · Ultraviolet light · Autoimmune

B. Demoret (✉) · P. Noll · S. Sampath
Ohio University Heritage College of Osteopathic Medicine, Athens, OH, USA

S. C. Trotter
Dermatologists of Central States, Canal Winchester, OH, USA
e-mail: strotter@docsdermgroup.com

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A 40-year-old female complained of pink scaly patches on the inside of her ears and scalp. The scalp lesions were associated with hair loss and itched on occasion. She was prescribed hydrocortisone and terbinafine cream by her primary care physician with no improvement. Her health history was significant for migraine headaches.

On physical examination, there were pink scaly plaques noted on the left and right parietal scalp and the concha of the ears bilaterally. The lesions on the scalp lacked hair. A scalp biopsy was performed and showed a dense superficial and deep perivascular and periadnexal lymphocytic infiltrate, basement membrane thickening, follicular plugging, and mucin deposition in the dermis. The eyebrows and fingernails appeared normal.

Based on the clinical case description, what is the most likely diagnosis?

1. Lichen planopilaris
2. Traction alopecia
3. Discoid lupus erythematosus
4. Seborrheic dermatitis

Diagnosis

Discoid lupus erythematosus.

Discussion

Lupus erythematosus is a multisystem autoimmune condition that encompasses a variety of dermatologic manifestations. Discoid lupus erythematosus (DLE) is the most common form of cutaneous lupus erythematosus (CLE) and may or may not be associated with the development of systemic disease [1]. DLE generally follows a more benign disease course compared to other CLE subtypes. If the initial workup for DLE does not reveal any systemic manifestations of systemic lupus erythematosus (SLE), the chance of DLE progressing to SLE is less than 5% [2]. Roughly 60% of patients with DLE have scalp involvement, of which one third will suffer from scarring alopecia [3].

DLE occurs more frequently in women than men, has a higher prevalence in African American women than Caucasian women, and affects individuals in their fourth and fifth decades of life [4]. The lesions are frequently photodistributed and affect the head and neck areas. Due to the scalp being a sun-exposed area, the scalp is a common presentation for DLE [5]. The propensity for the scalp increases the risk for DLE-related scarring alopecia and subsequently can significantly affect patients' quality of life. DLE lesions often appear as well-demarcated, erythematous, scaly macules or papules that gradually develop into an indurated discoid plaque with an adjoined scale. These plaques can extend into the hair follicle, causing alopecia [1] (Fig. 12.1). The lesions are often associated with hyper- and

hypo-pigmented patches (Fig. 12.2). DLE lesions may last for many years and are associated with extensive scarring.

The pathogenesis of discoid lupus erythematosus is multifactorial. Research suggests genetic, environmental, ultraviolet light, medications, cigarette smoke, and possibly viral infections can contribute to disease prevalence and severity. Exposure to UV light (UVL) is a well-documented modifiable risk factor that increases the severity of DLE lesions. In particular, UVB and UVA2 wavelengths have been implicated in the formation of DLE skin lesions [2, 6]. After direct UV exposure, the transcription factor IRF5 significantly increases in the skin, potentially leading to the photosensitivity associated with DLE [7]. Several other immunologic factors contribute to the pathogenesis of DLE. These immunologic factors include abnormal antigens and autoantibodies, hypermethylation of MHC class I molecules, plasmacytoid dendritic cell (PDCs), and natural killer cell (NKs) cell proliferation, CD8 T-cell activation, Th1 cell abnormalities, and IgG and IgM deposition in normal skin tissue [2]. This vast immune response plays a prominent role in the primary cicatricial alopecia caused by DLE. Cytotoxic lymphocytes can destroy the stem cells of hair follicles and cause the scarring associated with DLE. The primary histopathologic features associated with DLE are erythema, telangiectasias, follicular plugging, and resolution with scar formation [2].

The lesions are characteristically well-defined, annular erythematous patches or plaques with follicular hyperkeratosis. Plaques that extend into the hair follicle give the lesions a characteristic "carpet tack sign" and frequently result in scarring alopecia and permanent hair loss [8]. Dermoscopy of the scalp in patients with DLE reveals white patches, branching capillaries, keratin plugs, and areas of reduced follicular ostia [9]. Blue-grey dots in a speckled pattern located within white patches of DLE may also be visualized upon dermoscopy [9]. Additionally, plasmacytoid dendritic cells containing the marker CD123 in clusters of 10 cells or more, favors a diagnosis of scarring-DLE alopecia [10].

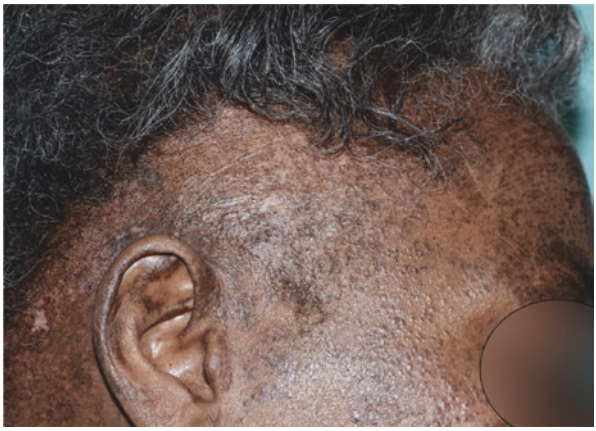

Fig. 12.1 Scaly patches with follicular plugging and scarring alopecia along the temporal and auricular scalp. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

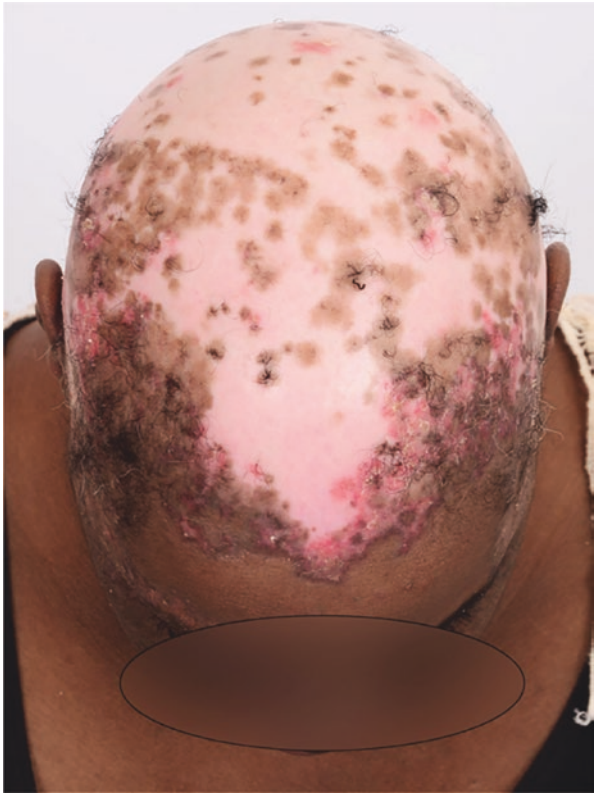

Fig. 12.2 Smooth, hypopigmented patches with scarring alopecia and a scaly, inflammatory anterior border. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

Treatment

The early detection and treatment of DLE are essential to minimize scarring and ultimately prevent permanent alopecia. General measures for individuals with DLE should involve non-pharmacologic methods such as eliminating exacerbating factors such as UVL [11]. Physical blockage of UVL with brimmed hats and the use of broad-spectrum sunscreens with an SPF of ≥30 are recommended [12]. Additional preventative measures include tobacco cessation, the avoidance of photosensitizing medications, and medications causing drug-induced lupus. When non-pharmacological management of DLE cannot be achieved, medications should be considered in the treatment plan. DLE management aims to improve the patient's appearance, control acute lesions, and limit scarring alopecia [13].

First-line therapy of acute DLE consists of topical corticosteroids, topical calcineurin inhibitors, and intralesional corticosteroid injections. Recent studies suggest that patients with DLE saw greater clinical improvement when treated with high-potency topical corticosteroids, such as fluticasone 0.05% cream, compared to medium- or low-potency topical corticosteroids (hydrocortisone 1.0% cream) after 6 weeks [14]. Although high-potency topical corticosteroid creams are preferred, patients must be educated about the possible side effects of prolonged usage

(cutaneous atrophy, telangiectasia, striae), especially when treating lesions on the face. If not resolved with topical corticosteroids, topical calcineurin inhibitors and intralesional corticosteroids should be considered.

Topical calcineurin inhibitors have specific advantages when treating facial lesions as they are not associated with cutaneous atrophy. Side effects of these medications include transient pruritus and burning, which typically resolve after one to two weeks [15]. The approved topical calcineurin inhibitors for DLE include tacrolimus 0.03% and 0.1% ointment and pimecrolimus 1.0% cream. Upon initiation, the treatment response with calcineurin inhibitors is usually seen between 4–8 weeks. Unlike corticosteroids, long-term therapy with calcineurin inhibitors is well tolerated and helpful in sustaining remission [15]. Unsuccessful treatment with calcineurin inhibitors or topical corticosteroids may warrant intralesional steroid injections with triamcinolone (3–5 mg/mL). If DLE does not respond to topical corticosteroids or calcineurin inhibitors, intralesional corticosteroid injections such as triamcinolone (3–5 mg/mL, multiple 0.1 mL, spaced 1 cm) are warranted.

Systemic therapy is considered second-line due to a greater risk of adverse events and is reserved for patients with DLE refractory to topical treatment regimens. The antimalarial hydroxychloroquine (200–400 mg/day) has historically been used in CLE management and is considered the first-line systemic agent for DLE [16]. A retrospective study of 200 patients with DLE demonstrated an adequate clinical response in 60% of patients within the first six months after long-term treatment with hydroxychloroquine [17]. Retinopathy is the most common adverse event associated with hydroxychloroquine and recommended guidelines for ophthalmologic examinations should be followed. For patients who do not respond to hydroxychloroquine therapy alone, quinacrine (100 mg/day) may be added in combination for additional benefit [18].

Other treatment modalities such as pulsed dye lasers, azathioprine, systemic retinoids, methotrexate, and thalidomide are documented in patients with refractory DLE [16]. However, these treatment options are generally considered third-line therapy. More recent clinical trials involving biological and immunological agents such as ruxolitinib, etanercept, and secukinumab are under investigation. One potential treatment option for treatment-resistant DLE is ASF-1096, which is primarily composed of R-salbutamol in an oil-in-water emulsion [19]. R-salbutamol has anti-inflammatory properties and hence could help treat glucocorticoid-resistant lesions. One study found that new and non-hypertrophic lesions responded well to treatment, but old hypertrophic lesions were difficult to treat [19]. The most common side effects associated with R-salbutamol included mild irritation to the applied area [19]. While this is a novel treatment option, it has shown promise in treating DLE-resistant lesions and warrants more extensive clinical trials.

Key Points

- Discoid lupus erythematosus (DLE) most commonly presents in isolation from systemic lupus and is more prevalent in middle-aged African American females.

- Early identification and management of discoid lupus erythematosus (DLE) can help prevent permanent scarring and related alopecia.
- Patient education should focus on avoiding exacerbating factors such as excessive sun exposure, tobacco smoking, and associated provoking medications.
- Medical management begins with topical and intralesional immunosuppressant therapy and more extensive or refractory cases may warrant systemic anti-inflammatory agents.
- Ongoing clinical trials with novel treatment regimens and promising biologics are currently under investigation to manage DLE scarring alopecia.

References

1. Okon LG, Werth VP. Cutaneous lupus erythematosus: diagnosis and treatment. Best Pract Res Clin Rheumatol. 2013;27(3):391–404.
2. Li Q, Wu H, Zhou S, Zhao M, Lu Q. An update on the pathogenesis of skin damage in lupus. Curr Rheumatol Rep. 2020;22(5):16.
3. Udompanich S, Chanprapaph K, Suchonwanit P. Hair and scalp changes in cutaneous and systemic lupus erythematosus. Am J Clin Dermatol. 2018;19(5):679–94.
4. Izmirly P, Buyon J, Belmont HM, Sahl S, Wan I, Salmon J, et al. Population-based prevalence and incidence estimates of primary discoid lupus erythematosus from the Manhattan lupus surveillance program. Lupus Sci Med. 2019;6(1):e000344.
5. Hordinsky M. Cicatricial alopecia: discoid lupus erythematosus. Dermatol Ther. 2008;21(4):245–8.
6. McGrath H. Ultraviolet-A1 irradiation therapy for systemic lupus erythematosus. Lupus. 2017;26(12):1239–51.
7. Achtman JC, Werth VP. Pathophysiology of cutaneous lupus erythematosus. Arthritis Res Ther. 2015;17:182.
8. Inamadar AC. Perforation of paper with pen: simple technique to explain the carpet tack sign in discoid lupus erythematosus. J Am Acad Dermatol. 2019;81(6):e159–60.
9. Duque-Estrada B, Estrada BD, Tamler C, Sodré CT, Barcaui CB, Pereira FB. Dermoscopy patterns of cicatricial alopecia resulting from discoid lupus erythematosus and lichen planopilaris. An Bras Dermatol. 2010;85(2):179–83.
10. Rakhshan A, Toossi P, Amani M, Dadkhahfar S, Hamidi AB. Different distribution patterns of plasmacytoid dendritic cells in discoid lupus erythematosus and lichen planopilaris demonstrated by CD123 immunostaining. An Bras Dermatol. 2020;95(3):307–13.
11. Kuhn A, Sonntag M, Richter-Hintz D, Oslislo C, Megahed M, Ruzicka T, et al. Phototesting in lupus erythematosus: a 15-year experience. J Am Acad Dermatol. 2001;45(1):86–95.
12. Herzinger T, Plewig G, Röcken M. Use of sunscreens to protect against ultraviolet-induced lupus erythematosus. Arthritis Rheum. 2004;50(9):3045–6.
13. Oboite ME, Love PB. Discoid lupus erythematosus. In: Love PB, Kundu RV, editors. Clinical cases in skin of color: medical, oncological and hair disorders, and cosmetic dermatology. Cham: Springer International Publishing; 2016. p. 3–13.
14. Wolverton SE. Controversies in dermatologic drug therapy. Dermatol Ther. 2009;22(5):397.
15. Sárdy M, Ruzicka T, Kuhn A. Topical calcineurin inhibitors in cutaneous lupus erythematosus. Arch Dermatol Res. 2009;301(1):93–8.
16. Company-Quiroga J, Alique-García S, Romero-Maté A. Current insights into the management of discoid lupus erythematosus. Clin Cosmet Investig Dermatol. 2019;12:721–32.

17. Wahie S, Daly AK, Cordell HJ, Goodfield MJ, Lovell CR, et al. Clinical and pharmacogenetic influences on response to hydroxychloroquine in discoid lupus erythematosus: a retrospective cohort study. J Invest Dermatol. 2011;131(10):1981–6.
18. Cavazzana I, Sala R, Bazzani C, Ceribelli A, Zane C, Cattaneo R, et al. Treatment of lupus skin involvement with quinacrine and hydroxychloroquine. Lupus. 2009;18(8):735–9.
19. Wulf HC, Ullman S. Discoid and subacute lupus erythematosus treated with 0.5% R-salbutamol cream. Arch Dermatol. 2007;143(12):1589–90.