BMJ Best Practice

Discoid lupus erythematosus

Straight to the point of care

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Table of Contents

Summary

Discoid lupus erythematosus is the most common form of chronic cutaneous lupus erythematosus. Lesions are generally well-demarcated erythematous macules or papules with a scaly surface, which frequently evolve into larger coin-shaped plaques; typically neither pruritic nor painful.

Age of onset is usually between 20 and 40 years. Lesions are often precipitated or aggravated by ultraviolet light exposure; smoking is also considered a risk factor.

Over time, lesions slowly expand, producing areas of peripheral inflammation or hyperpigmentation, leaving a central region of scarring with telangiectasia and hypopigmentation. In hair-bearing areas, follicular plugging (scale at hair follicle) is often seen and scarring alopecia may be found.

Diagnosis is usually made on clinical grounds, but if in doubt is confirmed with a skin biopsy.

The mainstay of treatment for localized/limited disease is topical corticosteroids. Disseminated disease, or severe or refractory localized/limited disease, is treated with antimalarials, progressing to immunosuppressants if there is no response.

Definition

Discoid lupus erythematosus (DLE) is a chronic inflammatory condition of the skin. DLE lesions begin as well-demarcated erythematous macules or papules with a scaly surface, and frequently evolve into larger coin-shaped plaques. In most DLE patients the condition remains confined to the skin; anti-nuclear antibodies (ANA) are often negative or present in a low titer.[1] Approximately 5% to 10% of patients may develop a mild form of systemic lupus erythematosus (SLE).[2] DLE-like lesions occur in up to 25% of patients with SLE.[3]

Epidemiology

It is difficult to find reliable epidemiologic data on DLE. Most studies are performed by either rheumatologists or dermatologists; hence, in the absence of systemic lupus erythematosus (SLE), DLE tends to be under-reported by rheumatologists and over-reported by dermatologists. Studies before 1979 did not distinguish between DLE and subacute cutaneous lupus erythematosus (SCLE), and therefore it is difficult to interpret the true prevalence or incidence of classic DLE. The most common age of onset of DLE is between 20 and 40 years.[4] It affects both females and males, with a slight female predominance. The female-to-male ratio has been reported between 3:2 and 2:1 compared with 12:1 in SLE.[5] Unlike SLE, there does not seem to be any racial predisposition to DLE. However, reports in the US suggest that DLE may be slightly more common in black Americans than in white Americans.[6]

Etiology

Several reports have shown that smoking may predispose to the development of DLE, and exposure to ultraviolet radiation may precipitate or aggravate DLE lesions.[7] [8] Nonspecific injury to the skin may also precipitate the development of DLE activity within areas of trauma (known as the Koebner phenomenon). The inflammatory process characteristic of DLE is thought to be driven by autoimmune mechanisms. Associations with some major histocompatibility complex antigens have been reported, but the other genetic factors that increase the risk of DLE are not known.[9]

Pathophysiology

The precise pathophysiology of DLE and of other forms of cutaneous lupus erythematosus (CLE) is not yet fully understood. Interest has been concentrated on the region of the dermal-epidermal junction as the principal site of injury because of the detection of IgG and C3 along the epidermal basement membrane, suggesting an immune-mediated process.[10] The inflammatory cells in DLE are mostly T cells in the dermis. Chronic scarring DLE lesions have a denser inflammatory cell infiltrate, which extends into the deeper reticular dermis and/or subcutis, in contrast with acute CLE and subacute CLE lesions, which contain less dense inflammatory infiltrates confined to the upper dermis. It has been suggested that keratinocyte apoptosis may be the key event in the initiation of CLE lesions.[11] The nucleated layer of the epidermis is generally not thickened and may be atrophic. There is prominent hyperkeratosis and follicular plugging.

Classification

Cutaneous manifestations of lupus erythematosus

No formal classification exists for DLE. However, it is important to understand the various cutaneous manifestations of lupus erythematosus and how DLE fits in with these:

- Acute cutaneous lupus erythematosus (ACLE): characterized by a typical butterfly pattern malar rash involving the central portion of the face and/or a more generalized maculopapular eruption. ACLE is strongly associated with SLE.
- Subacute cutaneous lupus erythematosus (SCLE): characterized by a nonpruritic, nonscarring rash. SCLE may be drug induced or can occur in patients with SLE, Sjogren syndrome, and complement C2 deficiency.

- Chronic cutaneous lupus erythematosus (CCLE): lesions are confined to the skin and oral mucosa; DLE is the most common form.

Subsets of DLE

Localized: only head and neck affected.

Disseminated: other areas affected, regardless of whether head and neck involved.

Case history

Case history #1

A 30-year-old woman presents to her primary care physician with a rash affecting her face, neck, ears, and scalp. The rash started as flat, well-demarcated, red, scaly lesions on her face, sparing the nasolabial folds, and is now confluent with spread to her neck and ears. In addition, the lesions have expanded leaving a depressed central area of scarring. There are no known precipitating factors. She is normally fit and healthy, with no significant past medical history. She smokes 10 cigarettes per day, does not take any regular medication, and has not recently taken any over-the-counter preparations.

Approach

The diagnosis of DLE is primarily clinical. Although it is possible for nonspecialists to make the diagnosis clinically, it is by no means easy. Referral to a specialist dermatologist is usually necessary. It is important to recognize and treat DLE lesions early. If it is left untreated the risk of scarring, permanent alopecia, and hypo- or hyperpigmentation is known to be increased. Chronic refractory lesions are often associated with psychological and social distress.

Clinical presentation

Two subsets of DLE have been recognized: localized/limited DLE, involving only the head and neck; and disseminated DLE, involving other areas regardless of head and neck involvement.[12]

Patients with disseminated DLE are more likely to develop systemic lupus erythematosus (SLE), and are more difficult to treat.

The most common age of onset is between 20 and 40 years.[4] Patients should be questioned about ultraviolet light exposure and smoking history.

Classic DLE presents with disk-shaped lesions, primarily affecting the face and scalp; the neck, ears, and extensor surfaces of the arms are also commonly involved at presentation. At onset, the lesions are scaly, erythematous macules or papules. The scale is follicular in all cases, and this is an early and diagnostic sign, preceding more general scaling of the lesion. In hair-bearing areas, follicular plugging (scale at hair follicle) is often seen. Characteristically, the lesions are neither painful nor pruritic, but some patients may experience occasional pain within the lesion and/or mild pruritus. Over time, the lesions slowly expand, producing areas of peripheral inflammation or hyperpigmentation, leaving a central region of scarring with telangiectasia and hypopigmentation. If left untreated, DLE scalp lesions inevitably result in alopecia, which becomes permanent once scarring occurs.

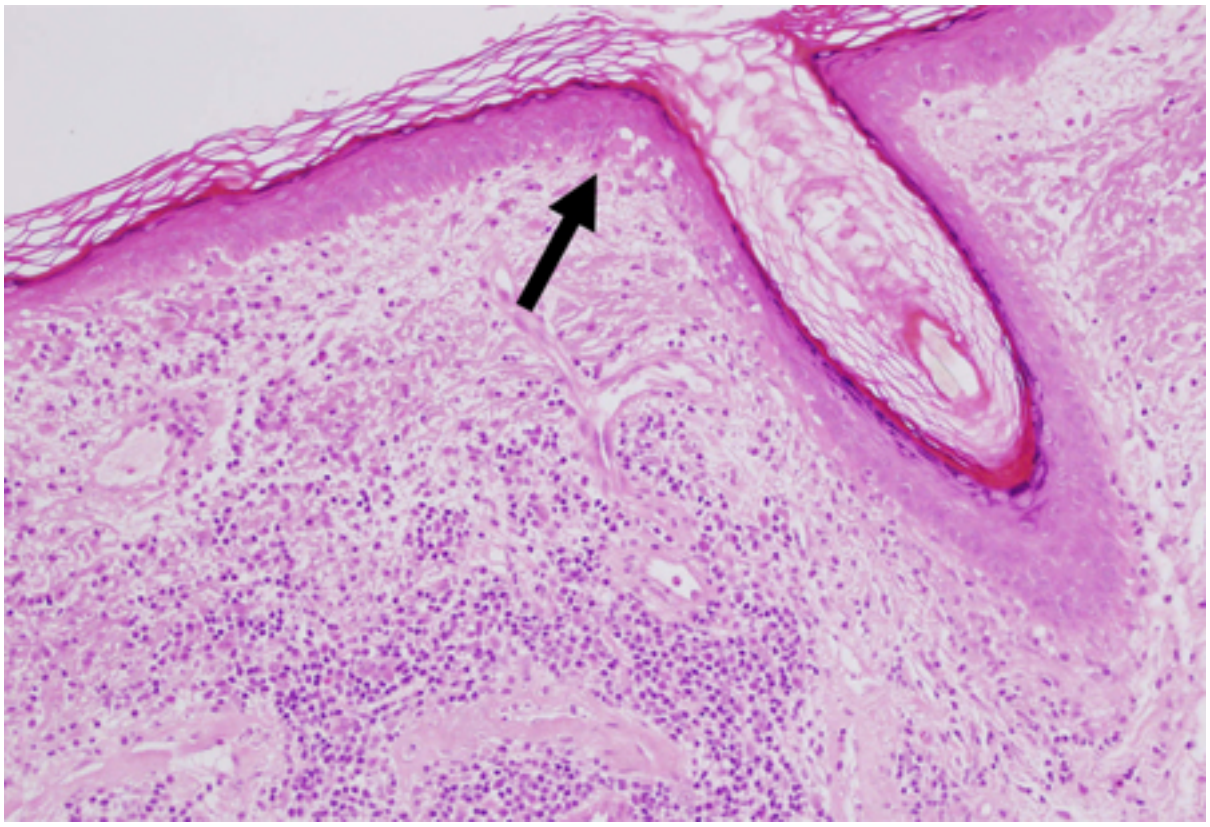
Approximately 5% to 10% of patients develop a mild form of SLE.[2] Therefore, a complete physical exam should be performed to look for signs suggestive of underlying systemic disease such as arthritis, serositis (pleuritis, pericarditis), central nervous system involvement (seizures, psychosis), or renal involvement. DLE is considered to have progressed to SLE if systemic features are present and the patient meets the criteria for diagnosis.[13] However, if the patient does not meet diagnostic criteria for SLE, a diagnosis of undifferentiated connective tissue disease is made.

Investigations

Serologic tests should be performed in all patients presenting with DLE, to detect progression to SLE as early as possible. Antinuclear antibody (ANA) titer is low in 30% to 40% of patients. High-titer ANA, anti-double-stranded DNA antibodies, and anti-ribonucleoprotein antibodies may suggest SLE or overlapping connective tissue disease. Anti-Ro (SS-A) antibodies can occasionally be found in patients with DLE, while anti-La (SS-B) antibodies are distinctly unusual in patients with isolated DLE.

Complete blood count (CBC) and erythrocyte sedimentation rate (ESR) should also form part of the initial workup. Although CBC is usually normal, a few patients may have a mild leukopenia. ESR may also be mildly elevated. In addition, BUN, electrolytes, and urinalysis are indicated to exclude any systemic involvement, particularly renal impairment, associated with SLE.

If the diagnosis is in doubt, a skin biopsy is indicated. Characteristic changes include hyperkeratosis, follicular plugging, basal layer vacuolar changes, and mononuclear cell infiltration at the dermal-epidermal junction.[12] [14]



Skin biopsy showing hyperkeratosis, thinning of the epidermis, degeneration of the basal layer of the epidermis (arrow), and a predominantly lymphocytic inflammatory cell infiltrate in the dermis

From the collection of Dr K. Blessing, used with permission

History and exam

Key diagnostic factors

disk-shaped erythematous maculopapular scaly lesions (common)

- DLE lesions typically present on the face, neck, scalp, ears, and extensor surfaces of arms.
- The lesions usually begin as flat or slightly elevated, well-demarcated, erythematous macules or papules with a scaly surface.
- These lesions commonly become larger and coalesce, leading to erythematous plaques with prominent, adherent scale extending into dilated hair follicles.

age 20 to 40 years (common)

- The most common age of onset.[4]

history of ultraviolet light exposure (common)

- Can precipitate or aggravate DLE lesions, with UV-A and UV-B wavelengths being implicated.

smoking history (common)

- Has been linked both to the development and the severity of DLE.[7] [8]

Other diagnostic factors

absence of pruritus and/or pain (common)

- Characteristically, the lesions are neither painful nor pruritic, but some patients may experience occasional pain within the lesion and/or mild pruritus.

telangiectasia, hyperpigmentation, and/or hypopigmentation (common)

- Over time, the lesions slowly expand, producing areas of peripheral inflammation or hyperpigmentation, leaving a central region of scarring with telangiectasia and hypopigmentation.
- Hyperpigmentation is a late sign and usually postinflammatory in white skin but more pronounced in darker skin types. Hypopigmentation may be postinflammatory or a sign of scarring.

permanent scarring alopecia (common)

- If left untreated, DLE scalp lesions inevitably result in alopecia, which becomes permanent once scarring occurs.

systemic features (arthritis, pleuritis, pericarditis, seizures, psychosis) (uncommon)

- Approximately 5% to 10% of patients develop a mild form of systemic lupus erythematosus (SLE).[2] Therefore, a complete physical exam should be performed to look for signs suggestive of underlying systemic disease, such as arthritis, serositis (pleuritis, pericarditis), central nervous system involvement (seizures, psychosis), or renal involvement.
- DLE is considered to have progressed to SLE if systemic features are present and the patient meets the criteria for diagnosis.[13] However, if the patient does not meet diagnostic criteria for SLE, a diagnosis of undifferentiated connective tissue disease is made.

Risk factors

Strong

age 20 to 40 years

- DLE more frequently develops between the ages of 20 and 40 years; the average peak age of incidence is 38 years.[4]

ultraviolet#ight exposure

- DLE lesions may be precipitated or aggravated by ultraviolet (UV) light exposure, with UV-A and UV-B wavelengths being implicated.

smoking

- Several reports have suggested that smoking may predispose to the development of DLE. One study noted a higher smoking prevalence (84.2%) in the DLE group than the controls (33.5%). Smokers also tend to have more extensive involvement than nonsmokers.[8]

Weak

female sex

- DLE affects both men and women. There is a slightly higher female predominance, with reports of a female-to-male ratio between 3:2 and 2:1 (female-to-male ratio reported as high as 12:1 in systemic lupus erythematosus).[5]

nonspecific skin injury

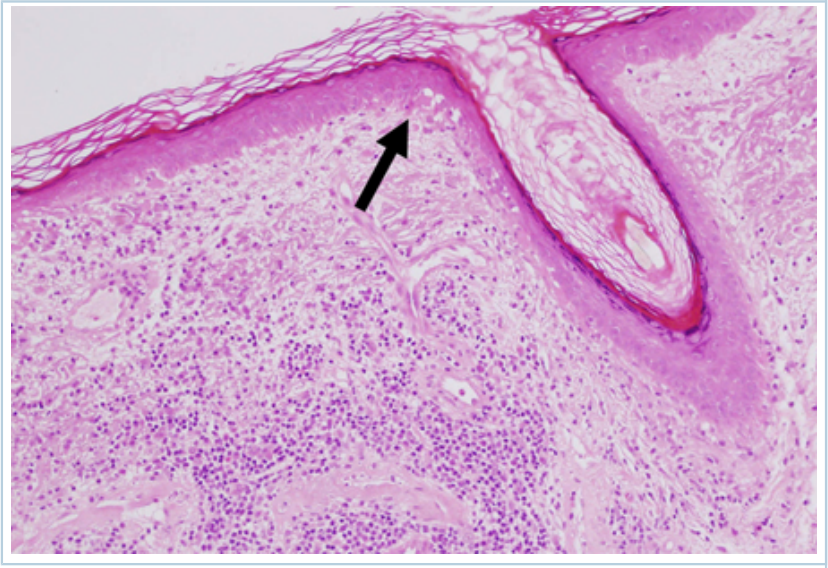
- DLE may be precipitated by nonspecific injury to the skin (known as the Koebner phenomenon).

Tests

1st test to order

Test	Result
antinuclear antibodies, double-stranded (ds) DNA • Should be performed in all patients presenting with DLE lesions to detect progression to systemic lupus erythematosus (SLE) as early as possible. • Antinuclear antibody (ANA) titer is low in 30% to 40% of patients. • High-titer ANA, anti-dsDNA antibodies, and anti-ribonucleoprotein antibodies may suggest SLE or overlapping connective tissue disease. Anti-Ro (SS-A) antibodies can occasionally be found in patients with DLE, while anti-La (SS-B) antibodies are distinctly unusual in patients with isolated DLE.	**positive or negative**
CBC • Should be performed in all patients presenting with DLE lesions. • Although typically normal, a few patients may have mild leukopenia. Modest to severe leukopenia may suggest underlying systemic lupus erythematosus.	**normal or mild leukopenia**
ESR • Should be performed in all patients presenting with DLE lesions. • Moderate or marked elevation is more likely to be associated with active systemic lupus erythematosus or underlying infection.	**normal or slightly elevated**
BUN and electrolytes • Should be performed in all patients presenting with DLE lesions to exclude any systemic involvement, particularly renal impairment, associated with systemic lupus erythematosus.	**normal**
urinalysis • Should be performed in all patients presenting with DLE lesions to exclude any systemic involvement, particularly renal impairment, associated with systemic lupus erythematosus.	**normal**

Other tests to consider

Test	Result
skin biopsy • Confirms diagnosis. Usually indicated if the diagnosis is in doubt.  *Skin biopsy showing hyperkeratosis, thinning of the epidermis, degeneration of the basal layer of the epidermis (arrow), and a predominantly lymphocytic inflammatory cell infiltrate in the dermis* *From the collection of Dr K. Blessing, used with permission*	**prominent hyperkeratosis, follicular plugging, vacuolar degeneration, thickened epidermal basement membrane, periappendageal inflammation**

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Systemic lupus erythematosus (SLE)	• Skin manifestations are a common presentation of SLE. The photosensitive malar or butterfly rash is characteristic. This erythematous rash extends from the cheeks over the bridge of the nose, sparing the nasolabial folds. It can be painful and pruritic, usually lasts a few days, heals without scarring, but often recurs after sun exposure. • The diagnosis is made using 2019 criteria recommended by the American College of Rheumatology and the European League Against Rheumatology.[13]	• High-titer ANA, anti-double-stranded DNA antibodies, and anti-ribonucleoprotein antibodies are strongly suggestive.
Subacute cutaneous lupus erythematosus (SCLE)	• SCLE lesions begin as small, erythematous, slightly scaly papules that evolve into either psoriasiform (papulosquamous) or annular forms. The lesions typically have erythematous crusted margins, usually sparing the face and mostly affecting the neck, arms, and upper torso. • Although telangiectasia may be seen, permanent pigment changes and scarring are absent.	• Findings on skin biopsy are similar to those of DLE, except that basement membrane thickening is generally absent or minimal.
Psoriasis	• Lesions are red, inflamed, silvery-white scaly and circumscribed papules and plaques; often affecting the elbows, knees, extensor limbs, and scalp. Psoriatic nails have a pitted surface, onycholysis, and/or hypertrophic (subungual) changes.	• Skin biopsy findings include intraepidermal spongiform pustules and Munro neutrophilic microabscess within the stratum corneum.
Polymorphous light eruption (PMLE)	• Considered an exclusively photo-triggered dermatosis that can express several clinical forms.	• Skin biopsy shows edema in the upper part of the dermis.

Condition	Differentiating signs / symptoms	Differentiating tests
	• PMLE lesions are usually itchy papules, eczematous plaques, or vesicles. They typically develop within 24 hours of sun exposure and there is often associated urticaria.	
Rosacea	• Common chronic disorder of the skin characterized by redness, flushing, and other cutaneous findings that often include telangiectasia, papules and/or pustules, rhinophyma, and general inflammation that can resemble acne. • Typically affects the convexities of the central face, including the nose, cheeks, eyelids, and forehead.	• Skin biopsy shows nonspecific granulomatous or lymphohistiocytic infiltrate. Associated edema, telangiectasia, and sebaceous hyperplasia may also be present.
Lichen planopilaris	• Refers to lichen planus of the hair follicles. Presents with alopecia associated with hyperkeratotic papules and perifollicular erythema with scale. • Typical lichen planus lesions affecting skin, nails, and buccal mucosa may also be present.	• Skin biopsy shows lichenoid lymphocytic infiltrate under the epidermis.

Approach

The primary goals of treatment are to improve physical appearance, control existing lesions and limit scarring, and prevent the development of new lesions.

Lifestyle changes

Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B) high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

Cosmetic camouflage may be used to improve the appearance of lesions.

Nonsevere localized/limited disease

Localized/limited disease involves only the head and neck.

Topical corticosteroids are used as first-line treatment for patients with localized disease.[12] [15] [16]

Calcineurin inhibitors (e.g. pimecrolimus, tacrolimus) are recommended as an alternative first-line option if topical corticosteroids are contraindicated in patients with concomitant acne/rosacea.[12] [15] [16]

Initial treatment with a potent/very potent topical corticosteroid for severe disease is recommended for 4 weeks (stepped down once symptoms are controlled), or 12 weeks with a calcineurin inhibitor.[15] Topical corticosteroids of different potencies may then be used in combination depending on the patient's symptoms. Potent corticosteroids (e.g., betamethasone valerate 0.1%) and very potent corticosteroids (e.g., clobetasol propionate 0.05%) are often used to treat the trunk and limbs including the hands, as well as the scalp. Potent or very potent formulations can be considered for severe disease on the head and neck for short periods. Moderate-potency corticosteroids (e.g., triamcinolone acetonide 0.1%) are used in areas more prone to atrophy, such as the face and neck. Mild-potency corticosteroids (e.g., hydrocortisone 1%) are typically reserved for the eyelids. Scalp involvement may be treated with foam or lotion formulations.

The effectiveness of calcineurin inhibitors as an alternative treatment of cutaneous lupus erythematosus (CLE), including DLE, has been assessed in randomized trials with variable results. One vehicle-controlled trial of topical tacrolimus in patients with CLE (14 with DLE, 4 with subacute CLE) found that tacrolimus significantly improved skin lesions at 4 and 6 weeks, but not at 12 weeks, compared with vehicle.[18] However, when pimecrolimus was compared to betamethasone in a small randomized controlled trial, no significant difference was found in efficacy between the treatments for patients with DLE at 8 weeks. An 86% decrease in clinical severity score was seen for pimecrolimus, compared with a 73% decrease with betamethasone (P=0.043).[19] Small uncontrolled studies have shown topical tacrolimus or pimecrolimus to be effective alternatives in the treatment of cutaneous lupus, including DLE.[20] [21] [22]

Intralesional injection of a corticosteroid may be considered for individual lesions and is recommended for sites at higher risk of atrophy in patients with localized DLE, or as an adjunct treatment for persistent lesions.[12] [15]

Disseminated disease or severe or refractory localized/limited disease

First-line therapy

For patients with disseminated disease or severe or refractory localized/limited DLE, an antimalarial drug (e.g., hydroxychloroquine or chloroquine), either as monotherapy or with adjunctive topical corticosteroids, is recommended as first-line treatment.[12] [15] [16] Adjunctive topical corticosteroid treatment may be stopped, or used as required, once the antimalarial is fully effective.

Chloroquine should only be considered as an option when hydroxychloroquine is ineffective or not tolerated.[12]

Patients with DLE are at a greater risk of scarring than patients with other subtypes of CLE; therefore, the following options should be considered for initial treatment:[12] [15]

- A higher dose of an antimalarial drug
- Short-term concomitant use, or tapering courses, of systemic corticosteroids.

Patients who receive long-term oral corticosteroids (>3 weeks' duration), or those who require frequent courses (3-4 per year), should be monitored regularly to prevent corticosteroid-induced osteoporosis and adrenal insufficiency.[12]

Treatment with antimalarials is recommended for a period of 1-2 years to fully suppress cutaneous lupus activity.[23]

Second-line therapy

If antimalarials are ineffective, immunosuppressant therapy (e.g., methotrexate, mycophenolate) should be considered.[12] [15] [16] [24] [25] [26]

Combination treatment with methotrexate or mycophenolate plus an antimalarial may be considered in patients with a partial response to an antimalarial plus topical therapy.[12]

A retinoid (e.g., acitretin) or dapsone may be considered as monotherapy (or in addition to an antimalarial for patients with refractory DLE).[12] [15]

One randomized double-blind study comparing acitretin with hydroxychloroquine for the treatment of facial lesions in 58 patients with CLE reported similar rates of complete clearing or marked improvement in both groups at 8 weeks.[27] The rate of adverse effects was higher in the acitretin group, leading to discontinuation of treatment in 4 patients.

Evidence is limited for the treatment of DLE with dapsone. One small study reported that out of 11 patients with DLE, 9 patients showed improvement in skin disease at 16 weeks.[28] One retrospective review of 33 patients with DLE who received dapsone for 1-27 months found that treatment gave excellent results in 8 patients (24%), and some effect in 8 patients (24%), while no response was seen in 17 patients (52%).[29]

Consultation with a specialist should be sought before initiating immunosuppressant therapy.

Third-line therapy

Thalidomide should be considered as treatment for cutaneous lupus, including DLE, that has not responded to other treatments.[12] [15] [16] [30] [31]

Due to the high incidence of neurotoxicity associated with thalidomide, which does not appear to be dose dependent, it has been suggested that thalidomide should be used as a remission-inducing drug only for patients with severely refractory CLE or who are at high risk for severe scarring.[32] [33] However, in clinical practice, low doses of thalidomide can be effective without significant risk of neurotoxicity.

Laser or surgical treatment

Pulsed dye laser treatment is generally not recommended but may be considered as an adjunct treatment for telangiectasia.[15] [16]

A small study involving patients with active chronic DLE lesions concluded that pulsed dye laser treatment is an effective and safe therapy for patients with refractory chronic DLE.[34]

Surgical excision followed by skin graft may be considered for patients with refractory DLE with localized lesions in cosmetically unacceptable areas when topical and systemic treatments have failed or are not tolerated.[15] Burned-out scarred lesions may be excised surgically, although reactivation of inactive lesions following surgical excision has been reported.[35] Therefore, surgical intervention should be combined with medical treatment with antimalarials and/or systemic corticosteroids.[15]

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Ongoing (summary)		
nonsevere localized/limited disease		
	1st	**topical treatment**
	plus	**lifestyle changes**
	adjunct	**intralesional corticosteroid injection**
	adjunct	**pulsed dye laser**
severe or refractory localized/limited disease; disseminated disease		
	1st	**antimalarial**
	plus	**lifestyle changes**
	adjunct	**topical corticosteroid**
	adjunct	**systemic corticosteroid**
	2nd	**immunosuppressant therapy**
	plus	**lifestyle changes**
	adjunct	**antimalarial**
	3rd	**thalidomide**
	plus	**lifestyle changes**
	4th	**surgical excision**
	plus	**lifestyle changes**

Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Ongoing

nonsevere localized/limited disease

1st **topical treatment**

Primary options

» hydrocortisone topical: (1%) apply to the affected area(s) once or twice daily
May be used on eyelids.

OR

» triamcinolone topical: (0.1%) apply to the affected area(s) once or twice daily
May be used on face and neck.

OR

» betamethasone valerate topical: (0.1%) apply to the affected area(s) once or twice daily
May be used on body/limbs and scalp. May be used on the face if other treatments are ineffective. May be used for severe disease on the head and neck for short periods.

OR

» clobetasol topical: (0.05%) apply to the affected area(s) twice daily
May be used on body/limbs or scalp. May be used for severe disease on the head and neck for short periods.

OR

» tacrolimus topical: (0.1%) apply to the affected area(s) twice daily

OR

» pimecrolimus topical: (1%) apply to the affected area(s) twice daily

» Localized/limited disease involves only the head and neck.

Ongoing

» Topical corticosteroids are used as first-line treatment for patients with localized disease.[12] [15] [16]

» Initial treatment with a potent/very potent topical corticosteroid is recommended for 4 weeks (stepped down once symptoms are controlled), or 12 weeks with a calcineurin inhibitor.[15] Topical corticosteroids of different potencies may then be used in combination depending on the patient's symptoms. Potent corticosteroids (e.g., betamethasone valerate 0.1%) and very potent corticosteroids (e.g., clobetasol propionate 0.05%) are often used to treat the trunk and limbs including the hands, as well as the scalp. Potent or very potent formulations can be considered for severe disease on the head and neck for short periods. Moderate-potency corticosteroids (e.g., triamcinolone acetonide 0.1%) are used in areas more prone to atrophy, such as the face and neck. Mild-potency corticosteroids (e.g., hydrocortisone 1%) are typically reserved for the eyelids. Scalp involvement may be treated with foam or lotion formulations.

» Calcineurin inhibitors (e.g., pimecrolimus, tacrolimus) are recommended as an alternative first-line option if topical corticosteroids are contraindicated in patients with concomitant acne/rosacea.[12] [15] [16]

» The effectiveness of calcineurin inhibitors as an alternative treatment of cutaneous lupus erythematosus (CLE), including DLE, has been assessed in randomized trials with variable results. One vehicle-controlled trial of topical tacrolimus in patients with CLE (14 with DLE, 4 with subacute CLE) found that tacrolimus significantly improved skin lesions at 4 and 6 weeks, but not at 12 weeks, compared with vehicle.[18] However, when pimecrolimus was compared to betamethasone in a small randomized controlled trial, no significant difference was found in efficacy between the treatments for patients with DLE at 8 weeks. An 86% decrease in clinical severity score was seen for pimecrolimus, compared with a 73% decrease with betamethasone (P=0.043).[19] Small uncontrolled studies have shown topical tacrolimus or pimecrolimus to be effective alternatives in the treatment of cutaneous lupus, including DLE.[20] [21] [22]

plus **lifestyle changes**

Treatment recommended for ALL patients in selected patient group

MANAGEMENT

Ongoing

» Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

» Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

» Cosmetic camouflage may be used to improve the appearance of lesions.

adjunct **intralesional corticosteroid injection**

Treatment recommended for SOME patients in selected patient group

Primary options

» triamcinolone acetonide: consult specialist for guidance on intralesional dose

» Intralesional injection of a corticosteroid may be considered for individual lesions and is recommended for sites at higher risk of atrophy in patients with localized DLE, or as an adjunct treatment for persistent lesions.[12] [15]

adjunct **pulsed dye laser**

Treatment recommended for SOME patients in selected patient group

» Pulsed dye laser treatment is generally not recommended but may be considered as an adjunct treatment for telangiectasia.[15] [16]

» A small study involving patients with active chronic DLE lesions concluded that pulsed dye laser treatment is an effective and safe therapy for patients with refractory chronic DLE.[34]

severe or refractory localized/limited disease; disseminated disease

1st **antimalarial**

Primary options

» hydroxychloroquine sulfate: 200-400 mg/day orally given in 1-2 divided doses, maximum 5 mg/kg/day

Secondary options

Ongoing

» chloroquine phosphate: consult specialist for guidance on dose

» For patients with disseminated disease or severe or refractory localized/limited DLE, an antimalarial drug (e.g., hydroxychloroquine, chloroquine), either as monotherapy or with adjunctive topical corticosteroids, is recommended as first-line treatment.[12] [15] [16]

» Chloroquine should only be considered as an option when hydroxychloroquine is ineffective or not tolerated.[12]

» Patients with DLE are at a greater risk of scarring than patients with other subtypes of CLE; therefore, a higher dose of an antimalarial should be considered for initial treatment.[12] [15]

» Treatment with antimalarials is recommended for a period of 1-2 years to fully suppress cutaneous lupus activity.[23]

» Hydroxychloroquine and chloroquine are associated with retinal toxicity. Regular screening for retinal changes is required according to local guidelines. Maximum doses of <5 mg/kg/day (hydroxychloroquine) or <2.3 mg/kg/day (chloroquine) are recommended (based on real body weight) to reduce the risk of retinal toxicity.[16]

plus **lifestyle changes**

Treatment recommended for ALL patients in selected patient group

» Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

» Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

» Cosmetic camouflage may be used to improve the appearance of lesions.

adjunct **topical corticosteroid**

Ongoing

Treatment recommended for SOME patients in selected patient group

Primary options

» hydrocortisone topical: (1%) apply to the affected area(s) once or twice daily
May be used on eyelids.

OR

» triamcinolone topical: (0.1%) apply to the affected area(s) once or twice daily
May be used on face and neck.

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» betamethasone valerate topical: (0.1%) apply to the affected area(s) once or twice daily
May be used on body/limbs and scalp. May be used on the face if other treatments are ineffective. May be used for severe disease on the head and neck for short periods.

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» clobetasol topical: (0.05%) apply to the affected area(s) twice daily
May be used on body/limbs or scalp. May be used for severe disease on the head and neck for short periods.

» May be used with initial antimalarial treatment. Adjunctive topical corticosteroid treatment may be stopped, or used as required, once the antimalarial is fully effective.

» Topical corticosteroids of different potencies may be used in combination depending on the patient's symptoms. Potent corticosteroids (e.g., betamethasone valerate 0.1%) and very potent corticosteroids (e.g., clobetasol propionate 0.05%) are often used to treat the trunk and limbs including the hands, as well as the scalp. Potent or very potent formulations can be considered for severe disease on the head and neck for short periods. Moderate-potency corticosteroids (e.g., triamcinolone acetonide 0.1%) are used in areas more prone to atrophy, such as the face and neck. Mild-potency corticosteroids (e.g., hydrocortisone 1%) are typically reserved for the eyelids. Scalp

Ongoing

involvement may be treated with foam or lotion formulations.

adjunct **systemic corticosteroid**

Treatment recommended for SOME patients in selected patient group

Primary options

» methylprednisolone sodium succinate: 1 g intravenously once daily for 3-5 days

OR

» prednisone: 0.5 to 1.5 mg/kg/day orally for 2-4 weeks, then taper gradually

» Patients with DLE are at a greater risk of scarring than patients with other subtypes of CLE; therefore, a short-term concomitant use, or tapering courses, of a systemic corticosteroid (e.g., intravenous methylprednisolone or oral prednisone) should be considered for initial treatment.[12] [15]

» Patients who receive long-term oral corticosteroids (>3 weeks' duration), or those who require frequent courses (3-4 per year), should be monitored regularly to prevent corticosteroid-induced osteoporosis and adrenal insufficiency.[12]

2nd **immunosuppressant therapy**

Primary options

» methotrexate: 7.5 mg orally once weekly on the same day each week initially, increase by 2.5 mg/week increments at monthly intervals according to response, maximum 20 mg/week

OR

» mycophenolate mofetil: 0.25 to 1 g orally twice daily

OR

» acitretin: consult specialist for guidance on dose

OR

» dapsone: consult specialist for guidance on dose

Ongoing

» If antimalarials are ineffective or not tolerated, immunosuppressant monotherapy (e.g., methotrexate, mycophenolate) should be considered.[12] [15] [16] [24] [25] [26]

» A retinoid (e.g., acitretin) or dapsone may be considered as monotherapy (or in addition to antimalarials for patients with refractory DLE).[12] [15]

» One randomized double-blind study comparing acitretin with hydroxychloroquine for the treatment of facial lesions in 58 patients with CLE reported similar rates of complete clearing or marked improvement in both groups at 8 weeks.[27] The rate of adverse effects was higher in the acitretin group, leading to discontinuation of treatment in 4 patients.

» Evidence is limited for the treatment of DLE with dapsone. One small study reported that out of 11 patients with DLE, 9 patients showed improvement in skin disease at 16 weeks.[28] One retrospective review of 33 patients with DLE who received dapsone for 1-27 months found that treatment gave excellent results in 8 patients (24%), and some effect in 8 patients (24%), while no response was seen in 17 patients (52%).[29]

» Consultation with a specialist should be sought before initiating immunosuppressant therapy.

plus **lifestyle changes**

Treatment recommended for ALL patients in selected patient group

» Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

» Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

» Cosmetic camouflage may be used to improve the appearance of lesions.

adjunct **antimalarial**

Treatment recommended for SOME patients in selected patient group

Ongoing

Primary options

» hydroxychloroquine sulfate: 200-400 mg/day orally given in 1-2 divided doses, maximum 5 mg/kg/day

Secondary options

» chloroquine phosphate: consult specialist for guidance on dose

» Combination treatment with an antimalarial plus methotrexate or mycophenolate may be considered in patients with a partial response to antimalarials and topical therapy.[12]
An antimalarial may also be considered in combination with a retinoid (e.g., acitretin) or dapsone for patients with refractory DLE.[12] [15]

» Hydroxychloroquine and chloroquine are associated with retinal toxicity. Regular screening for retinal changes is required according to local guidelines. Maximum doses of <5 mg/kg/day (hydroxychloroquine) or <2.3 mg/kg/day (chloroquine) are recommended (based on real body weight) to reduce the risk of retinal toxicity.[16]

3rd **thalidomide**

Primary options

» thalidomide: consult specialist for guidance on dose

» Thalidomide should be considered as treatment for cutaneous lupus, including DLE, that has not responded to other treatments.[12] [15] [16] [30] [31]

» Due to the high incidence of neurotoxicity associated with thalidomide, which does not seem to be dose dependent, it has been suggested that thalidomide should be used as a remission-inducing drug only for patients with severely refractory CLE or who are at high risk for severe scarring.[32] [33] However, in clinical practice, low doses of thalidomide can be effective without significant risk of neurotoxicity.

plus **lifestyle changes**

Treatment recommended for ALL patients in selected patient group

» Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum

Ongoing

(both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

» Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

» Cosmetic camouflage may be used to improve the appearance of lesions.

4th **surgical excision**

» Surgical excision followed by skin graft may be considered for patients with refractory DLE with localized lesions in cosmetically unacceptable areas when topical and systemic treatments have failed or are not tolerated.[15] Burned-out scarred lesions may be excised surgically, although reactivation of inactive lesions following surgical excision has been reported.[35] Therefore, surgical intervention should be combined with medical treatment with antimalarials and/or systemic corticosteroids.[15]

plus **lifestyle changes**

Treatment recommended for ALL patients in selected patient group

» Lifestyle changes should be discussed with patients.[12] [15] Patients are advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16] All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

» Smoking cessation is highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

» Cosmetic camouflage may be used to improve the appearance of lesions.

Emerging

Alternative immunotherapy or biologic treatments

In the UK, clofazimine, intravenous immune globulin, lenalidomide, belimumab, or rituximab may be considered as third-line treatments, although these recommendations are not supported by non-UK guidelines.[12][15][16]Belimumab or rituximab are only recommended for patients with CLE with systemic involvement.[15][16] See also the management section of Systemic lupus erythematosus .

Anifrolumab

Anifrolumab is a type 1 interferon receptor antagonist. Two cases of severe and refractory DLE showed improvement when treated with anifrolumab.[36] However, further evidence is needed to determine the safety and efficacy of this drug in treating DLE.

Litifilimab

Litifilimab is a humanized monoclonal antibody that binds to blood dendritic cell antigen 2 (BDCA2). Treatment with litifilimab was superior to placebo in a phase 2 trial involving patients with CLE.[37] However, further evidence is needed to determine the safety and efficacy of this drug in treating DLE.

Patient discussions

Lifestyle changes should be discussed with patients.[12] [15] Patients should be advised to limit their exposure to the sun, cover up exposed areas of skin, and apply a broad-spectrum (both ultraviolet A and ultraviolet B), high-SPF sunscreen every 2 hours.[12] [15] [16]

All patients who use long-term sun protection should consider vitamin D supplementation.[12] [15] [16]

Smoking cessation should be highly recommended, as smoking is associated with more severe disease and decreases the efficacy of antimalarials.[7] [12] [15] [17]

Patients may want to consider cosmetic camouflage to improve the appearance of lesions.

Monitoring

Monitoring

Patients with DLE should be monitored every 6-12 months if lesions are inactive, and more frequently if lesions are active and difficult to treat.[15] Follow-up with complete blood count and urinalysis is sufficient for patients with inactive lesions. However, for patients with active and more resistant lesions, erythrocyte sedimentation rate, serologic tests, blood urea nitrogen, and a serum electrolyte panel should also be performed at every follow-up visit to detect progression to systemic lupus erythematosus as early as possible.[15]

Patients taking immunosuppressant agents should be evaluated for complications of treatment.[15]

Regular ophthalmologic visits are necessary for those taking an antimalarial agent, to monitor for signs of ocular toxicity. Annual screening for retinopathy is recommended in patients who have received hydroxychloroquine or chloroquine for 5 years, and may be considered for patients who have additional risk factors for retinal toxicity, such as concomitant tamoxifen therapy, or impaired renal function (estimated glomerular filtration rate <60 mL/ minute/1.73 m^2), or if the daily dose of hydoxychloroquine is 5 mg/kg or more.[12]

Complications

Complications	Timeframe	Likelihood
squamous cell carcinoma of the skin	**long term**	**low**
Squamous cell carcinoma (SCC) can occasionally develop in chronic DLE lesions.[39] Multiple methods exist for treating SCC and vary depending on tumor type, size, and location; patient history; and practitioner. They include surgical excision, Mohs micrographic surgery, and radiation therapy.		
systemic lupus erythematosus	**variable**	**medium**
Patients with DLE have approximately a 5% to 10% risk of eventually developing systemic lupus erythematosus (SLE).[2] The risk may depend on the extent and distribution of DLE lesions; patients with widespread DLE have a higher rate of immunologic abnormalities and a greater risk of progressing to SLE.		

Prognosis

DLE may occasionally resolve spontaneously, but, if it is left untreated, most patients tend to have progressive lesions that spread to produce larger areas of scarring. With scalp involvement permanent scarring alopecia can result, which may be emotionally and psychologically disabling. Acute exacerbations may occur, particularly in spring and summer, most likely as a consequence of ultraviolet light exposure.

Diagnostic guidelines

International

British Association of Dermatologists guidelines for the management of people with cutaneous lupus erythematosus (https://www.bad.org.uk/guidelines-and-standards/clinical-guidelines) [12]

Published by: British Association of Dermatologists **Last published:** 2021

Guideline for the diagnosis, treatment and long-term management of cutaneous lupus erythematosus (https://pubmed.ncbi.nlm.nih.gov/34364171) [15]

Published by: Asian Dermatological Association, Asian Academy of Dermatology and Venereology, and the Lupus Erythematosus Research Centre of the Chinese Society of Dermatology **Last published:** 2021

Treatment guidelines

International

Guideline for vaccinations in patients with rheumatic and musculoskeletal diseases (https://rheumatology.org/clinical-practice-guidelines) [38]

Published by: American College of Rheumatology **Last published:** 2023

British Association of Dermatologists guidelines for the management of people with cutaneous lupus erythematosus (https://www.bad.org.uk/guidelines-and-standards/clinical-guidelines) [12]

Published by: British Association of Dermatologists **Last published:** 2021

Guideline for the diagnosis, treatment and long-term management of cutaneous lupus erythematosus (https://pubmed.ncbi.nlm.nih.gov/34364171) [15]

Published by: Asian Dermatological Association, Asian Academy of Dermatology and Venereology, and the Lupus Erythematosus Research Centre of the Chinese Society of Dermatology **Last published:** 2021

S2k guideline for treatment of cutaneous lupus erythematosus - guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV) (https://eadv.org/publications/clinical-guidelines) [16]

Published by: European Academy of Dermatology and Venereology **Last published:** 2016

Key articles

- O'Kane D, McCourt C, Meggitt S, et al; British Association of Dermatologists' Clinical Standards Unit. British Association of Dermatologists guidelines for the management of people with cutaneous lupus erythematosus 2021. Br J Dermatol. 2021 Dec;185(6):1112-23. Full text (https://onlinelibrary.wiley.com/doi/10.1111/bjd.20597) Abstract (http://www.ncbi.nlm.nih.gov/pubmed/34170012?tool=bestpractice.bmj.com)
- Lu Q, Long H, Chow S, et al. Guideline for the diagnosis, treatment and long-term management of cutaneous lupus erythematosus. J Autoimmun. 2021 Sep;123:102707. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/34364171?tool=bestpractice.bmj.com)
- Kuhn A, Aberer E, Bata-Csörgő Z, et al. S2k guideline for treatment of cutaneous lupus erythematosus - guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV). J Eur Acad Dermatol Venereol. 2017 Mar;31(3):389-404. Full text (https://onlinelibrary.wiley.com/doi/10.1111/jdv.14053) Abstract (http://www.ncbi.nlm.nih.gov/pubmed/27859683?tool=bestpractice.bmj.com)

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Images

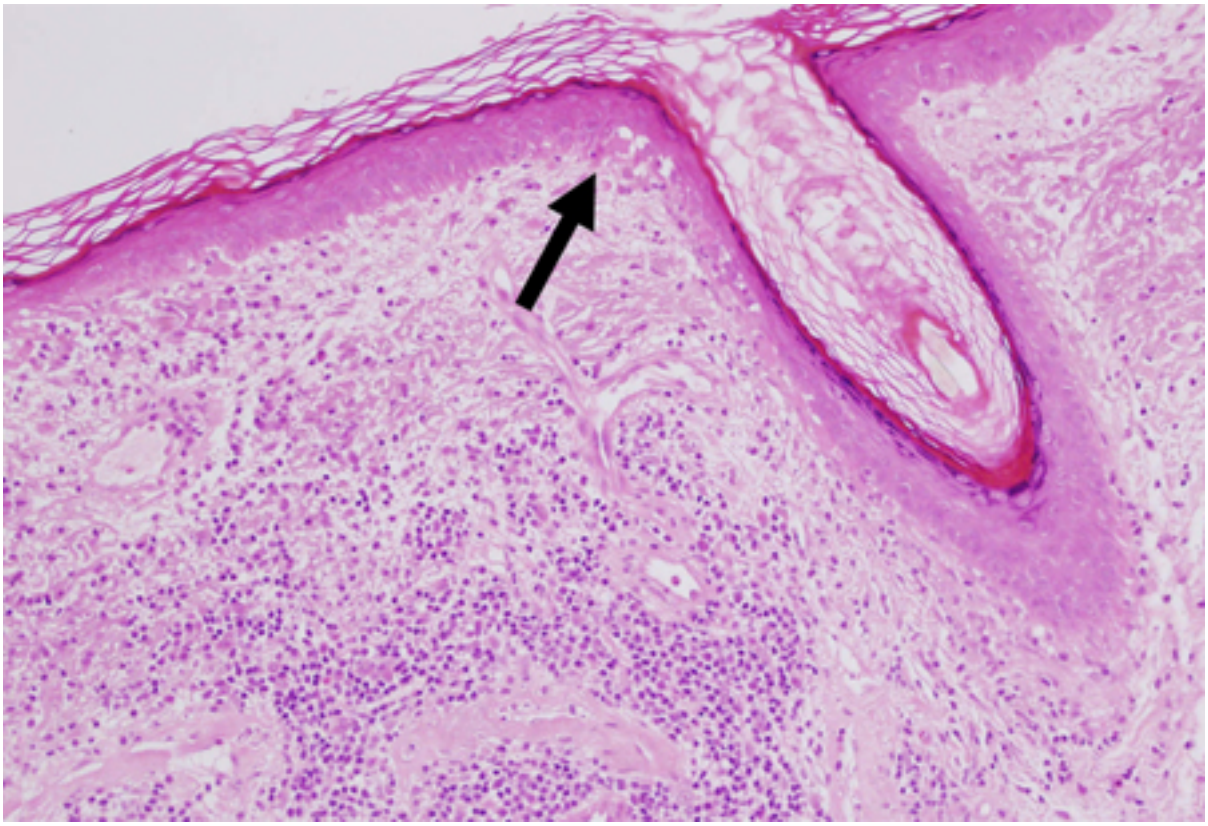

Figure 1: Skin biopsy showing hyperkeratosis, thinning of the epidermis, degeneration of the basal layer of the epidermis (arrow), and a predominantly lymphocytic inflammatory cell infiltrate in the dermis

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Where your version of BMJ Best Practice does not integrate with a local drug formulary, you should consult a local pharmaceutical database for comprehensive drug information including contraindications, drug interactions, and alternative dosing before prescribing.

Interpretation of numbers

Regardless of the language in which the content is displayed, numerals are displayed according to the original English-language numerical separator standard. For example 4 digit numbers shall not include a comma nor a decimal point; numbers of 5 or more digits shall include commas; and numbers stated to be less than 1 shall be depicted using decimal points. See Figure 1 below for an explanatory table.

BMJ accepts no responsibility for misinterpretation of numbers which comply with this stated numerical separator standard.

This approach is in line with the guidance of the International Bureau of Weights and Measures Service.

Figure 1 – BMJ Best Practice Numeral Style

5-digit numerals: 10,000

4-digit numerals: 1000

numerals < 1: 0.25

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