



## Discoid Lupus

### What is discoid lupus erythematosus?

Lupus erythematosus (LE) is a group of diverse persistent autoimmune inflammatory diseases and often affects the skin. Discoid lupus erythematosus (DLE) is the most common chronic form of cutaneous lupus. It is characterised by persistent scaly, disk-like plaques on scalp, face and ears that may cause pigmentary changes, scarring and hair loss.

### What causes discoid lupus erythematosus?

Factors leading to DLE include:

- Genetic predisposition
- Exposure to sunlight (often several weeks prior to presentation)
- Toxins such as cigarette smoke
- Hormones
- The manifestations of DLE are due to loss of regulation of the immune system in the skin.

### Who gets discoid lupus erythematosus?

DLE can affect males and females of any age. DLE is 5 times more common in females than males, and onset is most often between the ages of 20 and 40 years. DLE is more common than systemic lupus erythematosus (SLE). The estimated prevalence is around 20–40 people in every 100,000.

DLE may be more common in patients with darker coloured skin than in fair Caucasians.

DLE is more common and more severe in smokers compared to non-smokers. Smoking also reduces the effectiveness of antimalarials and other therapies.

### Clinical features of discoid lupus erythematosus

Most patients with DLE just have skin involvement (cutaneous LE). Between 5% and 25% of patients with DLE develop SLE, in which there may be other forms of cutaneous lupus and other organs may develop disease. Typically, systemic symptoms are mild in these patients.

DLE may be localised (above neck in 80%) or generalised (above and below neck in 20%).

### **Signs of localised DLE include:**

- Initial lesions are dry red patches
- These evolve to indurated red or hyperpigmented plaques with adherent scale
- Follicular keratosis, ie plugs of keratin within hair follicles, is noted when surface scale is removed, eg with tape (carpet-tack sign)
- Older lesions are hyperpigmented, especially on the edge of the plaques
- Scarring results in central loss of pigment (white patches) and skin atrophy (tissue loss)
- DLE is typically located on nose, cheeks, ear lobe and concha
- It may involve lips, oral mucosa, nose or eyelids
- Scalp lesions cause temporary or permanent patches of hair loss
- Hypertrophic (warty) lupus erythematosus describes red, very thickened plaques

### **Signs of generalised DLE include:**

- Plaques on anterior chest, upper back, backs of hands
- Sometimes, plaques on upper and lower limbs
- Can affect palms and soles
- Can affect anogenital mucosa

### **How is discoid lupus erythematosus diagnosed?**

DLE is often diagnosed from its distribution in sun-exposed sites and the clinical appearance of the plaques. After a careful history, the patient with DLE should undergo a thorough general examination, to find out if other forms of lupus may be present.

The diagnosis is usually confirmed by skin biopsy, in which typical features of lupus are noted: interface and periadnexal dermatitis, follicular plugging, atrophy and scarring. Direct immunofluorescence is often positive in lesional skin in DLE (positive lupus band test).

### **Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI)**

The Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) was developed in an attempt to classify the severity of cutaneous LE. [2] A score of activity and damage due to the disease is calculated in each of 12 anatomical locations (refer to original published paper for details).

Total activity score is made up of:

Degree of redness (0–3) and scale (0–2)

Mucous membrane involvement (0–1)

Recent hair loss (0–1), nonscarring alopecia (0–3)

Total damage score is made up of:

The degree of dyspigmentation (0–2), and scarring (0–2)  
Persistence of dyspigmentation more than 12 months doubles the dyspigmentation score  
Scalp scarring (0, 3, 4, 5, 6)

### **Blood tests**

Patients with DLE will usually have blood tests at the time of diagnosis and from time to time afterwards.

Full blood count

Renal function test

Inflammatory markers such as C-reactive protein (CRP)

Antinuclear antibody (ANA, ANF; if present, they are usually in low titre)

Extractable nuclear antibody (ENA)

Anti-annexin 1 antibodies—these may be a diagnostic marker for discoid CLE

Circulating autoantibodies are found in about 50% of patients with DLE.

### **What is the treatment for discoid lupus erythematosus?**

#### **Preventative measures**

The following measures are important to reduce the chance of flares of DLE.

Careful year-round protection from sun exposure using clothing, accessories and thickly applied SPF 50+ broad-spectrum sunscreens. Sunscreens alone are not adequate.

Indoors, some patients may also need to stay away from glass windows or these can be treated with UV-blocking films.

Vitamin D supplements should be recommended for those who strictly avoid the sun.

Smoking cessation.

#### **Topical therapy**

Intermittent courses of potent topical corticosteroids are the main treatment for DLE. They should be applied accurately to the skin lesions for several weeks. Potency should be selected to suit body site and thickness of the plaque. Very potent topical steroids may cause thinning of the surrounding skin and increase blood vessel formation (telangiectasia). Intralesional injections of corticosteroids are sometimes used, especially for hypertrophic DLE.

The calcineurin inhibitors tacrolimus ointment and pimecrolimus cream can also be used.

Camouflage makeup is useful to improve appearance.

#### **Systemic therapy**

Typically, any of the following drugs may be used to treat DLE alone or in combination.

Hydroxychloroquine and other antimalarials—response rates are about 80% in CLE. It is thought to be less effective in smokers.

Systemic corticosteroids such as prednisone or prednisolone. These are rarely required for DLE.

Methotrexate—best response in subacute CLE and discoid CLE

Retinoids isotretinoin and acitretin

Mycophenolate

Azathioprine

Dapsone

### **What is the outlook for discoid lupus erythematosus?**

DLE tends to persist for years or decades. In some patients, all signs of active disease resolve in time.

Squamous cell carcinoma can rarely arise within a longstanding DLE plaque in the skin or mucous membrane. It presents as an enlarging warty growth or ulcer. It is usually treated surgically.

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