Cutaneous lupus erythematosus

What is cutaneous lupus erythematosus?

Cutaneous lupus erythematosus (CLE) is a form of lupus that predominantly affects the skin. The most common forms are subacute cutaneous lupus erythematosus (SCLE) and discoid lupus erythematosus (DLE).

There is a very small chance that these cutaneous forms of lupus may develop into systemic lupus erythematosus (SLE) with internal organ involvement.

What causes cutaneous lupus erythematosus?

CLE is thought to be an "autoimmune" condition where specific antibodies are produced and cause damage to the skin resulting in the appearance of a rash. The exact cause is unknown. However, genetic and environmental factors are thought to play a role. Sunlight, certain medications and hormones have been shown to trigger or exacerbate the condition.

CLE tends to be more common in females and amongst certain racial groups. It is not hereditary or contagious.

What does cutaneous lupus erythematosus look like?

The appearance of CLE varies greatly depending on the subtype.

Subacute cutaneous lupus erythematosus (SCLE)

- The rash usually appears as red, non-itchy patches on sun-exposed areas such as the upper chest and back and arms.
- There are two main forms of SCLE:
    - Papulosquamous SCLE occurs in approximately two thirds of cases and consists of red scaly patches.
    - Annular (or polycylic) SCLE occurs in approximately one third of cases and causes a red ring or "arcuate shaped" rash.
- SCLE heals without scarring, although the rash may fade with some colour changes.
- Rarely there may be mild symptoms such as tiredness, weakness or joint aches.

Discoid lupus erythematosus (DLE)

- DLE occurs most commonly on sun-exposed sites, in particular the face and scalp.
- It appears as red scaly patches and bumps which can sometimes be thickened or warty.
- If DLE affects the hair then red scaly areas develop around the base of the hairs and result in patches of hair loss (alopecia) which may be permanent.
- DLE may occasionally be uncomfortable or itchy.

- DLE often heals with scarring and/or skin colour changes. The scarring can sometimes be disfiguring and difficult to treat.

In some cases, there may be overlap between the different forms of CLE present, including the presence of chilblains (perniosis).

What other problems can occur in cutaneous lupus erythematosus?

A minority of cases (less than 5%) of CLE may become systemic lupus erythematosus (SLE). Very rarely, some lesions in DLE may turn into skin cancer (such as squamous cell carcinoma or basal cell carcinoma).

How is cutaneous lupus erythematosus diagnosed?

To confirm the diagnosis of CLE, a skin biopsy is usually required for routine histopathology (viewing under microscope) and immunofluorescence (specific stains). In addition, blood tests are usually taken to look for markers of inflammation and autoantibodies, the latter often being negative in CLE.

How is cutaneous lupus erythematosus treated?

There is no cure for any form of CLE. However, all forms of the condition may respond to treatment and the passing of time. The main goals of treatment are to reduce symptoms and the appearance of the rash as well as prevent long-term complications such as scarring and hair loss (alopecia).

General measures are important in the treatment of all forms of CLE. These include strict sun protection which should always include avoiding direct sunlight (e.g. by seeking out shade, wearing tightly woven longer sleeve clothing, broad brimmed hats and sunglasses) and wearing a broad spectrum SPF 50+ sunscreen. Ask your dermatologist for further advice. Avoiding soaps which can further dry and irritate the skin and using a good moisturiser regularly is important. Camouflaging the lesions with makeup may be beneficial in DLE. Stopping smoking also helps improves most cases of CLE.

Topical treatments for CLE include topical steroids and non-steroid based anti-inflammatory ointments (e.g. pimecrolimus or tacrolimus).

Oral medications may be needed to treat CLE. Hydroxychloroquine, an antimalarial tablet, has been used for many years to treat most forms of CLE. Oral medications that suppress the immune system (such as azathioprine, methotrexate, cyclosporin or mycophenolate mofetil) may also be used in CLE.

Specific treatments for DLE include oral retinoid medications (such as acitretin or isotretinoin) or physical treatments (such as steroid injections).

What is the likely outcome of cutaneous lupus erythematosus?

SCLE uncomplicated by systemic disease often has a chronic course with intermittent flare-ups. It often flares up during sunnier weather. Sometimes there is spontaneous remission. The skin usually heals without scarring or permanent marks. It can be managed with treatment.

In DLE there may be permanent loss of pigmentation, scarring of the skin and/or alopecia (hair loss) leading to disfiguration and psychological distress. Early treatment may reduce the risk of scarring.