

Lupus

America's Least Known Major Disease

SKIN (CUTANEOUS) LUPUS

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The skin plays a very important role in the early diagnosis of lupus. For example, four of the eleven American Rheumatism Association's revised criteria for systemic lupus erythematosus are skin manifestations: malar rash, discoid rash, photosensitivity and oral ulcerations. Lupus rashes often mimic other more common skin disorders, sometimes making early diagnosis difficult.

Since at least 30% of people who will develop SLE will have skin lesions as their first complaint, it is no surprise that the first question most people newly diagnosed with skin lupus ask is, "What are my chances of developing systemic lupus?". While we know that only about 10% of people with skin lupus, in general, develop systemic disease, there are established "Markers of Transition" from skin-only disease to systemic disease (Table 1). These markers greatly improve your doctor's ability to predict that outcome for you with much greater accuracy. This is especially important if you are in a higher risk category for systemic disease. There are significant advantages to beginning treatment with remittive type treatment (medicine that turns off the disease itself rather than just treating symptoms) that may prevent any further progression of the illness.

Skin lupus is further divided into acute, subacute, and chronic with further subdivisions within each group (Table 2). The importance of recognizing the many forms of skin lupus cannot be overemphasized. Delay in diagnosis may lead to worsening and progression of your illness. For example, many people with the malar (cheeks and nose) "butterfly" rash of lupus are thought to have rosacea, psoriasis, or eczema. If they have hair thinning they may be diagnosed with alopecia areata, a coin shaped area of hair loss. These may initially respond to topical treatments, further confusing and delaying the correct diagnosis. Early diagnosis will allow appropriate treatment that may induce remission of the skin disease as well as, most importantly, preventing progression to more serious systemic disease.

EARLY DIAGNOSIS

If you are being treated for a skin disorder that is not improving despite treatment, or is stubbornly recurrent, a skin biopsy by a dermatologist is the simplest and quickest way to establish the correct diagnosis. Special blood studies may also be helpful in some instances. Using the "Markers of Transition", tell your doctor if you have generalized joint achiness which persists, alert your doctor to any skin rash on the rest of your body, and have your blood and urine checked regularly. Being attentive to these details will greatly empower you over the disease.

MANAGING CUTANEOUS LUPUS

PREVENTION

Most people are anxious to know if anything in their lifestyle or diet caused their lupus. While we don't know all the reasons why certain people get the disease over others, we do know that genetics and environmental triggers play a role. Any blood relative with an autoimmune disease such as thyroid problems, rheumatoid arthritis, etc. may carry the gene that also predisposes you to lupus.

What are the environmental triggers? Photosensitivity (sensitivity to light and other forms of radiation) is a major factor in the induction of most skin lupus. It is extremely important to recognize this and realize that this means an adjustment in lifestyle, not just the use of sunscreen. Photosensitivity may occur 365 days a year, on cloudy as well as sunny days.

Ultraviolet radiation that affects lupus is divided into UVA and UVB. The UVB rays are those that we normally associate with sunburn and tans. The UVA rays penetrate more deeply into the skin and do not cause redness or burn. Therefore, they are more insidious and less likely to be noted by you as the trigger for your lupus. Furthermore, UVA rays are present winter and summer, from dusk until dawn. It is imperative to reduce your exposure to these rays by limiting unnecessary outdoor activity, wearing double layers of clothing, wide brimmed hats, or special sun protective clothing, wearing a broad-spectrum UVA-UVB protective sunscreen every day all year round, preferably SPF 30, and reapply frequently. UVA rays penetrate through window glass, are generated by halogen and fluorescent bulbs, and are emitted from CRT computer screens. Make sure your windows have plastic shields in them (most thermal windows do), use fluorescent and halogen fixtures that are plastic encased, and use a polarizing screen over your computer's CRT. If you are on any medications such as diuretics (water pills), antibiotics, anti-inflammatory pills, hormones (including birth control pills) that may



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increase photosensitivity, alert your doctor and be especially careful.

Smoking has recently been shown to be a factor in skin lupus and smoke cessation should be undertaken with the help of your doctor to ensure that it is successful. Secondary smoke should also be avoided. Eating a healthy, balanced diet is also very important. Alternative medicines and supplements should be discussed with your doctor. Beware of the many unsupported claims on the internet. They can be dangerous if interpreted without the advice of your doctor.

TREATMENT

Localized skin lupus can be successfully treated with a broad array of topical steroid preparations. These come in a number of strengths, and initially the higher potency products are best, followed by a reduction to the lowest potency that remains effective. In addition, steroids can be injected directly into stubborn areas and are especially useful in the scalp for localized areas of hair loss.

Generalized skin lupus, difficult to control local disease, and in those individuals with any "Marker of Transition" regardless of the type of skin lupus, are best treated with remittive medication. The systemic antimalarial drugs such as hydroxychloroquine, quinacrine, or chloroquine are the "standard of care" and offer you the best chance of not only putting your skin lupus under control, but also preventing any chance of progression to more serious forms of systemic disease. Systemic steroids are sometimes used for short periods of time, as well as many other drugs such as azathioprine, retinoids, thalidomide, dapsone, mycophenolate mofetil, cyclosporine, and a number of promising newer agents. Your doctor can provide you with the rationale and indications for their use.

What can be done for lupus scars once the disease has been put into remission? For minor defects, camouflage makeup has many benefits. Initial consultation with a cosmetician experienced in these techniques is preferable, but information can be obtained directly from the companies themselves. Newer laser technology has exciting potential to improve the scarring and pigment disturbances left by skin lupus and is less risky than plastic surgery. Sometimes both techniques are required on one individual. Regardless of the approach, laser surgery or plastic surgery should only be undertaken when your disease is in full remission, and you are maintained on antimalarials to prevent post-operative reactivation. These procedures should be performed by those physicians knowledgeable about lupus scars and in cooperation with the doctor treating your lupus.

TABLE 1

Markers of Transition

- Rash above and below neck
- Associated non-specific skin lesions (skin ulcers, inflammation of blood vessels, calcium deposits, nodules)
- High ANA titers (antinuclear antibody blood test)
- Proteinuria (protein in urine)
- Hematuria (blood in urine)

TABLE 2

Chronic Cutaneous LE

- Localized Discoid
- Generalized Discoid (above and below the neck)
- Hypertrophic Discoid (LP/LE overlap)
- Lupus Profundus (lobular panniculitis)
- Scarring Alopecia
- Acral LE (palmar/plantar)
- Tumid LE

Subacute Cutaneous LE

- Papulosquamous (psoriasiform)
- Annular-polycyclic (+/- vesicles)

Acute Cutaneous LE

- Malar rash
- Bullous LE
- Widespread photodistributed erythema