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## LUPUS AND THE SKIN

***Taken from Transcript of a talk by Dr. Christina Saywell (Skin & Cancer Foundation) and Dr. Laura Saywell (Sydney) as printed in the Lupus Association of NSW Newsletter August 1995.***

Lupus Erythematosus (LE) can produce a wide variety of skin (cutaneous) lesions. However there are four basic categories of cutaneous lupus:

1. **Discoid LE**
2. **Sub Actue Cutaneous LE**
3. **Neonatal LE**
4. **Systemic LE**

Each group is characterised by relatively specific skin lesions. Furthermore Lupus tends to run quite a characteristic course within a certain group, quite separate and different from the other categories. This means that a patient who is initially diagnosed with Discoid LE (category 1) is generally unlikely to progress to Systemic LE (category 4). There are a few notable exceptions however which will be illustrated later.

The aims of this talk are to firstly discuss the main features of each category of cutaneous lupus, highlighting the main differentiating factors. The latter half will review recent treatment updates of cutaneous lupus.

### **DISCOID LE:**

Discoid LE is a relatively benign (non serious) form of Lupus which predominantly affects the skin and spares the internal organs. The characteristic lesion is, not surprisingly the "discoid" lesion which is usually a red scaly patch that generally heals eventually but leaves scars or permanent discolouration of the skin. Lesions are usually confined to areas that receives a high degree of sun exposure eg, the cheeks, nose, back of hand but occasionally are quite widespread. (The importance of sun exposure and photosensitivity i.e. sun sensitivity in all forms of cutaneous lupus will be emphasised in more detail later).

Some DLE patients also develop permanent bald patches due to discoid lesions in the scalp (scarring alopecia), blotchy discolouration of the legs (reticulate telangiectasia), chilblains and Raynaud's Phenomenon, where the hands and feet turn sequentially white then blue then red due to excessive shut down of the arteries on exposure to cold temperature. Others may suffer from joint pains and up to 55% have at least one blood test abnormality eg, low haemoglobin (anaemia) or a positive ANA (anti nuclear antibody) but these factors do NOT necessarily imply that such patients have a bad outlook or that they will go on to develop systemic (internal) disease.

Debate still exists as to whether Discoid LE (DLE) is a milder variant of Systemic LE (SLE) or is in fact a separate entity.

The evidence supporting the notion that DLE is a milder variant of SLE includes:

- “Discoid” lesions occur in both diseases, 5% of patients who initially present with discoid lesion eventually progress to SLE. Up to 20% of patients with SLE have a discoid lesion at some stage during the course of their disease.
- Similar blood test abnormalities and microscopic findings on skin biopsy.
- Involvement of the fatty tissues below the skin, called Lupus Profundus may occur in both DLE and SLE.

The evidence that DLE and SLE are separate entities includes:

- The very small risk that DLE patients will progress to SLE (5%).
- Different immunofluorescence findings (a specialised pathology test) on skin biopsy.
- Genetic differences between DLE and SLE patients ie. people are genetically programmed to develop one form but not the other. This genetic programming can't be changed. It is imprinted for life. Overall it seems that DLE and SLE are probably closely related diseases but due to differences in genetic make up and other possible unknown factors the risk of progression from DLE and SLE seems very low.

Factors that indicate that the development of SLE is more likely in a patient with initially discoid lesions include:

- Widespread lesions
- No response to treatment
- Internal organ disease
- Persistently abnormal blood count and certain specific abnormalities eg. positive SS-DNA and DS-DNA tests. The latter, (DS-DNA) is a very specific antibody test for detecting SLE.

#### **SUB ACUTE CUTANEOUS LE:**

This is a comparatively uncommon form of lupus, comprising only 10% of case. Two thirds of these patients present with a red scaly eruption usually involving the chest, neck and arms. The remainder have annular (or ring shaped) lesions. Unlike DLE, the lesions do not scar but as they resolve they may leave telangiectases (visible dilated blood vessels) and altered skin pigmentation. Very marked photosensitivity is characteristic.

Some patients will also have features of DLE eg scarring alopecia of the scalp and even more significantly 50% will have enough general symptoms to fulfil the ARA criteria for SLE. (In this classification system patients must have 4 or more of 11 criteria eg mouth ulcers, joint pains, photosensitivity, positive ANA blood test to be diagnosed as having SLE). It would therefore seem reasonable to presume that patients with Subacute LE would often progress to full blown SLE. However this is not the case. The prognosis (outlook) in Subacute LE is generally better than SLE with minimal risk of kidney involvement.

Blood test results help to define this category of lupus. Most patients have a positive anti-Ro antibody but negative results to the antibodies that often occur in SLE patients eg DS-DNA, Sm and RNP. Therefore whilst there may be some overlap in skin and systemic signs between Subacute LE and other forms of LE, this is essentially a distinct group.

#### **NEONATAL LE:**

This is a very rare entity and the vast majority of women with LE should not be concerned about transmitting it to their new born babies. A small percentage of women ( $\leq 1\%$ ) who possess the anti-Ro antibody are at risk of doing so. The mother's anti-Ro antibody crosses the placenta and causes the disease in the growing foetus. The babies are born with, or develop within a few months of birth, skin lesions like Subacute LE mainly around the eyes. The lesions usually resolve without problem by 12 months of age as the mother's antibodies are gradually lost from the baby's circulation. The babies do not really have lupus themselves as all the effects are due to their mother's antibody not their own. Importantly however some of these babies do develop permanent heart block which may require a pacemaker indefinitely.

Interestingly, up to 50% of mothers are completely well during pregnancy with no signs of lupus or any abnormalities other than the anti-Ro blood test. This antibody is not routinely tested for in pregnancy so it is not usually discovered until testing is performed because of the baby's rash. Many of these women later develop connective tissue diseases like lupus, often after many years.

#### **SYSTEMIC LE:**

Skin lesions are but one of many symptoms that may affect patients with SLE. They may seem relatively less significant in severity in comparison to the internal manifestations of SLE but as our skin is an essential contact with the outside world facial rashes and hair loss can be very disabling. Other skin conditions eg, ulcers and weals are important signs of blood vessel damage. 80% of SLE patients develop skin lesions at some stage in the course of their disease and from 25% the skin shows the first sign of lupus. The butterfly rash (over the cheeks and nose) is the most characteristic skin sign of SLE but actually occurs in only one third of patients. More common is a patchy red scaly rash on sun exposed sites. Again photosensitivity is important and although probably less of a problem than in Subacute LE and DLE it should never be ignored in any SLE patient. Sun exposure may exacerbate both internal disease and the skin lesions. As previously mentioned a significant number of SLE patients develop discoid lesions.

Hair loss in SLE can be very obvious but fortunately unlike DLE it is reversible. Most common is diffuse loss due to telogen effluvium which is a process that occurs a few months after the body suffers a major stress. This occurs in otherwise healthy people eg after childbirth, high fevers and can occur with a flare of lupus. Once the stressful event is removed, the hair regrows. More rarely "lupus hair" develops when the disease is very active. The hair is weak, breaks easily and appears coarse, dry and unruly. The nails may show changes such as divots, ridges and lifting up of the free edge (onycholysis). Mouth ulcers, usually painless occur in 25% of patients usually on the roof of the mouth. Many different lesions result from blood vessel involvement including Raynaud's Phenomenon, chilblains, leg ulcers, livedo reticularis (mottling of the legs) and urticaria (seen in 10% of lupus patients as the first clinical feature) (weals). There are a wide variety of skin signs that may rarely be associated with SLE including blistering, altered pigmentation, facial swelling, nodules etc.

**Reviewed by:** Assoc/Prof. Glenn Reeves – Senior Specialist Immunology (20 March 2003)

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