Lupus and the Skin

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Objectives

- Define the term “photosensitivity” and the role of sunlight in lupus

- Familiarize with the most common skin manifestations in lupus

- Understanding the principles of therapy
Photosensitivity

- Photosensitivity refers to the development of a rash after exposure to UV-B radiation found in sunlight or fluorescent lights.
- It occurs in up to 60 percent of patients with systemic lupus erythematosus (SLE).
- Some patients are also sensitive to UV-A (as from a photocopier), and may even be sensitive to the visible light spectrum.
Photosensitivity

- Glass protects individuals sensitive to UV-B, but only partially protects those sensitive to UV-A.
- Blonde, blue eyed, fair skinned individuals are much more photosensitive than brunettes or individuals with pigmented skin.
- It is important to recognize that not all photosensitive patients have lupus.
Sunlight – The Damage!

- The mechanism whereby UV radiation causes skin lesions is not clear:
- Damage to DNA (the backbone of the genes in the nucleus) and/or proteins in the skin leads to antibody formation to these altered molecules.
- Subsequent the antibodies attack new cells causing a local inflammatory reaction.
- In addition to the local effects in skin, UV radiation may also increase the degree of autoimmunity and trigger a flare of the disease!
The Spectrum of Lupus

- Systemic Lupus Erythematosus
  SLE is a prototypic autoimmune disease characterized by the production of antibodies to components of the cell nucleus. Patients with SLE are subject to myriad symptoms, complaints, and inflammatory involvement that can affect virtually every organ.

- Cutaneous Lupus Erythematosus
The Spectrum of Cutaneous Lupus

- Acute Cutaneous (skin) Lupus/ the Butterfly Rash
- Subacute Cutaneous Lupus
- Chronic Cutaneous Lupus/ the Discoid Rash
Butterfly Rash

- The classic acute butterfly rash, characterized by redness (erythema) over the cheeks and bridge of the nose (show picture 1), appears usually after sun exposure.
- The involved skin feels warm and appears slightly puffy (edematous).
- Application of alcohol (found in many sunscreens) can enhance the redness, due to increased circulation to the skin.
Some other skin lesions may be quite similar to the classic acute butterfly rash of SLE. Rosacea, for example, may also present as “red chicks”. A skin biopsy may be needed to distinguish between the two lesions. Other causes of facial erythema include seborrheic dermatitis, corticosteroid-induced dermal atrophy, and flushing.
Discoid Lupus

- Chronic discoid lesions may occur in the absence of any other clinical feature of SLE.
- Patients with only cutaneous discoid lupus generally have a negative antinuclear antibody (ANA) titer
- Patients with cutaneous discoid lupus have approximately a 5 to 10 percent risk of eventually developing SLE, which tends to be mild
- 25 percent of patients with SLE can have discoid lesions
Discoid Lupus

- Discoid lesions are red (erythematous) plaques covered by an adherent scale.
- Discoid lesions are most often seen on the face, neck, and scalp, but also occur on the ears, and infrequently on the upper torso (show picture).
- They tend to heal by leaving central scars, and discolored skin hyperpigmentation = darker than normal
- Depigmentation = lighter than normal skin
Subacute Cutaneous Lupus

- **Subacute cutaneous lupus — (SCLE)**
  Approximately 50 percent of affected patients have systemic disease (internal organ involvement), and about 10 percent of patients with SLE have this type of skin lesion.

- The most frequently affected areas in SCLE are the shoulders, forearms, neck, and upper torso. The face is usually spared.
Subacute Cutaneous Lupus

- SCLE lesions begin as small, red (erythematous), slightly scaly bumps (papules) that evolve into polycyclic or figurative patterns.
- Different from the discoid lesions, permanent color changes, and scarring do not occur.
- There is a strong association with antibodies to Ro/SSA.
Subacute cutaneous lupus  Annular lesions of subacute cutaneous lupus on the back of a 37 year-old woman who was ANA negative and anti-Ro positive. Courtesy of Samuel Moschella, MD.
**Subacute cutaneous lupus erythematosus**
Patient with a photosensitive eruption on the forearms and back of hands. The lesions are hyperkeratotic, making one suspect psoriasis. The history of photosensitivity suggests SLE, which confirmed by biopsy. Courtesy of Peter H Schur, MD.
Involvement of the Mouth and Nose

- Mucous membrane involvement occurs in 25 to 45 percent of patients with SLE.
- Mouth (oral) lesions may be the first sign of lupus. The oral ulcers are usually painless. There is no apparent association between the presence of such ulcers and systemic activity.
- Nasal ulcers have been noted in some patients.
Alopecia in Lupus

- Hair loss occurs in a majority of patients with SLE at some time during their illness. In some cases, it can precede other manifestations of lupus. Lupus alopecia may involve the scalp, eyebrows, eyelashes, beard, and/or body hair.
- Hair loss in active SLE usually is diffuse and responds well to treatment of the lupus.
- In comparison, in the discoid lesions the hair loss is usually permanent due to the scarring.
Treatment - Prevention

- **Prevention** — Patients who are photosensitive should avoid high sun exposure (beaches, snow, lakes), especially between 10 AM and 3 PM.

- Photosensitive patients should also use sunscreens daily.

- The sunscreen should be applied 30 to 60 minutes prior to exposure and reapplied every four to six hours. Sunscreens of at least SPF 30 should be used; higher SPFs are available for more sensitive patients.
Treatment - Topical

- **Local therapy** — A lupus rash should initially be treated with topical corticosteroids.
- For early superficial involvement hydrocortisone may suffice, but more potent steroids (particularly the fluorinated preparations) should be used for thicker lesions (but should be used for no more than two weeks).
- Chronic use of topical steroids, especially fluorinated steroids may lead to skin thinning, color changes
Treatment - Topical

- **Local therapy** - Other topical agents are being investigated. Few reports suggest the immunomodulatory agent, tacrolimus, may have some benefit for facial skin lesions due to lupus.

- **Cosmetic** — Erythematous lesions can be disguised with the use of cosmetics such as Cover Mark™, Dermablend™, or a green foundation.
Mucous membrane lesions respond well to topical corticosteroids, 0.1% tacrolimus ointment, intralesional corticosteroids and systemic antimalarial drugs.

The response to topical steroids (usually Orabase mixed with either 0.1 % triamcinolone or 0.05 % clobetasol) takes a few days to weeks and the response hydroxychloroquine takes weeks to months.
Treatment - Systemic

- **Antimalarial drugs** — Patients with persistent rashes should be treated with an antimalarial.
- Currently, the most popular antimalarial in SLE is hydroxychloroquine (less than 6.5 mg/kg per day to a maximum of 400 mg/day).
- Chloroquine (250 to 500 mg/day) is somewhat more potent but has a higher risk of eye damage.
Treatment - Education

- Internet resources with advice on sun-protection for adults include the Safe Sun site: http://tray.dermatology.uiowa.edu/SafeSun/SafeSun-1.html

- Guides to clothing with SPF of 30 or greater are available at some internet sites, including: sunprecautions.com, sunproof.com, sunprotectiveclothing.com, and sungrubbies.com.
Thank You for Your Attention!

Questions & Answers