Skin Manifestations of Lupus
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Epidemiology of CLE

- Skin involvement occurs in 72-85% of patients with SLE
- It is the first sign of disease in 23-28%
- Incidence of CLE and SLE are similar
- CLE is more common in men and older adults than SLE

CLE Subtypes

• Chronic cutaneous (CCLE)
  – Discoid (DLE)
  – Lupus profundus
  – Tumid lupus

• Subacute cutaneous lupus (SCLE)

• Acute cutaneous (ACLE)

• Diagnosis relies on the combination of clinical, histologic, and laboratory features
CLASSIFICATION OF LUPUS ERYTHEMATOSUS

Acute cutaneous LE

Subacute cutaneous LE

Chronic cutaneous LE

"Butterfly" rash

Alopecia within lesions

LE tumidus
Lupus panniculitis
Discoid LE
Chilblain lupus

Bologna, J Dermatology
Discoid Lupus

Papules that gradually expand into indurated round plaques. The center develops a depressed scar that is hypopigmented. There is often scale, and follicular plugging may be visible.
Discoid Lupus

- 10-20% of patients with predominantly discoid lupus will have SLE
- The most common areas involved are the head and neck.
- Triggered by sun exposure and also by trauma (Koebner effect).
Scarring Develops
Scarring Alopecia
Scarring is often present in the conchal bowl of the ear.
Subacute Cutaneous Lupus (SCLE)

- Erythematous, scaly, and polycyclic or ring-shaped macules and papules predominantly on the upper chest and back
- Marked photosensitivity
- ~70% have SSA (anti-Ro) antibodies
- May meet criteria for SLE but clinical course is usually benign
Post-inflammatory Hyperpigmentation
Hydroxychloroquine-induced Skin Pigmentation
Subacute Cutaneous Lupus (SCLE)

• Always review the medication list
• >40 different medications have been reported to cause SCLE
• More common causes:
  – hydrochlorothiazide
  – calcium channel blockers (diltiazem)
  – angiotensin-converting enzyme inhibitors
  – terbinafine
Lupus Panniculitis

- Firm or depressed nodules located on the trunk, proximal extremities, face and breasts
- Inflammation is in the deep dermis and underlying fat
- Clinically mimics breast cancer and subcutaneous panniculitis-like T-cell lymphoma
- Dystrophic calcification can occur
Tumid Lupus (LE Tumidus)

• Red urticarial-like plaques on the head and upper body
• Usually in a sun exposed distribution
• Hallmark is pronounced photosensitivity
• Abundant mucin seen in the tissue on skin biopsy
• Not associated with auto-antibodies or systemic lupus
Relationship of Cutaneous Subsets to SLE

Modified from J. Callen,.
Histology of lupus

Histologic findings are similar across LE types

- Interface dermatitis with perivascular and periadnexal inflammation, and mucin

- Does not determine subtype

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Protective Effect of Sunscreen

Tumid lupus

Vehicle

Sunscreen

Untreated

SCLE

UVA

UVB

UVA

UVB

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Patient Recommendations

• Adjust their lifestyle to moderate sun exposure

• Recommend sunscreen, hats and clothing religiously
  – Minimum SPF 50, “broad spectrum” (UVA+UVB)
  – Combination physical blockers (titanium and zinc oxide) with chemical sunscreens (Mexoryl)

• For indoor lighting LEDs may be safer
Vitamin D and Lupus

• In addition to many health benefits, Vitamin D is immunomodulatory and may be disease modifying in lupus
• Low vitamin D levels have been observed in lupus
• Multiple factors likely contribute
  – Sun avoidance
  – Renal disease
  – Darker skin types predominate
• Recommended serum level of at least 30-40 ng/ml 25(OH)D
• Vitamin D supplementation, at a minimum 400iu daily

Remove exacerbating factors

Limited disease
- Topical Steroids
- Intraleisional steroids
- Topical Calcineurin Inhibitors
- Hydroxychloroquine or Chloroquine

Extensive disease
- Add Quinacrine
- Low dose oral steroids
- Methotrexate
- Mycophenolate
- Azathioprine

Refractory disease
- Belimumab
- Thalidomide
- Lenalidomide
- Dapsone
- Retinoids
- Cyclophosphamide
- Rituximab

Smoking cessation
Sun protection