A Mashup of Instructive Cases Presenting With Subacute Cutaneous LE (SCLE) Skin Lesions

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Potential Conflicts of Interest
March, 2014

• **Consultant**
  - Centocor (Remicade-infliximab)
  - Genentech (Raptiva-efalizumab)
  - Alexion (eculizumab)
  - MediQuest Therapeutics
  - P&G (ChelaDerm)
  - Celgene
  - Sanofi

• **Research collaboration**
  - 3Gen, LLT

• **Paid speaker**
  - Winthrop (Sanofi)
    - Plaquenil (hydroxychloroquine)
  - Amgen (etanercept-Enbrel)
  - Connetics/Stiefel

• **Royalties**
  - Lippincott, Williams & Wilkins

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*Cutaneous Manifestations of Rheumatic Diseases*
Edition: 2nd
Editors: Richard D. Sontheimer, Thomas T. Provost

“Subacute cutaneous lupus erythematosus”

Original Contributions

Subacute Cutaneous Lupus Erythematosus

A Cutaneous Marker for a Distinct Lupus Erythematosus Subset

Richard D. Sontheimer, MD; Jesse R. Thomas; James N. Gilliam, MD
38 yrs of SCLE
The First Two Decades: 1979-1996
Dallas

• Prognostic significance of SCLE lesions' morphology and anatomic distribution
• SCLE and “pseudolupus”
• SjS complicating SCLE
SCLE

Papulosquamous
(Psoriasiform Array)

Annular
(Polycyclic array)
Risk of Systemically-Significant SLE in SCLE
Relative Sparing of Central Face
Risk Factors SCLE → Clinically-Significant SLE (?)

- Papulosquamous/psoriasiform morphology
- Central facial involvement (ACLE)
- Resistance to antimalarials
- Leukopenia
- High titer ANA
- Circulating dsDNA antibodies

Subacute cutaneous lupus erythematosus: a decade's perspective.
Sontheimer RD.
PMID: 2671533 [PubMed - indexed for MEDLINE]
SCLE and “Pseudolupus”
Dallas – 1990s
“…anything happening to a patient with SLE which is not immediately otherwise explicable will automatically be blamed on the lupus, regardless of pathophysiologic validity.”
Greenwald’s “Law of Lupus”
LE

DM
Greenwald’s “Law of Lupus”
Norwegian Scabies
Greenwald’s “Law of Lupus”
Pediculosis Capitis
Lesson Learned

Individuals with cutaneous lupus lesions are at risk for also contracting one or more of the other 1,999 skin disorders that are listed in our textbooks.
Sjögren’s Syndrome complicating patients presenting with SCLE skin lesions
SCLE SjS Overlap (10-40%)
SCLE Patient 10 Years Later

Cutaneous small vessel LCC vasculitis
~30% of primary SjS
SCLE/SjS Overlap Patient 3 Years Later

- Brought to clinic by pt’s family on a stretcher. Too weak to standup.
SCLE/SjS Overlap Patient 3 Years Later

- Admission W/U
  - Serum potassium – 2.6 mmol/L
- Muscle weakness resolved quickly upon IV potassium repletion
Hypokalemic paralysis in Sjögren’s syndrome secondary to renal tubular acidosis.

Christensen KS.

Abstract
A 62-year-old woman with Sjögren's syndrome, distal renal tubular acidosis and hypokalemic muscle paralysis is described. The sicca syndrome was nearly subclinical and went unrecognized for several years. The main and first manifestation to be expressed was that of hypokalemic muscle paralysis secondary to renal tubular acidosis. In the last decade several reports have appeared indicating that renal tubular acidosis is associated with Sjögren's syndrome. The data in this report support the view that adult onset distal renal tubular acidosis is often a disorder of an autoimmune disease, frequently that of Sjögren's syndrome. The complications to renal tubular acidosis such as hypokalemic muscle paralysis or chronic muscle weakness, nephrolithiasis, and osteomalacia can be avoided if the diagnosis of renal tubular acidosis is made and corrective alkali therapy is maintained.

PMID: 4001878 [PubMed - indexed for MEDLINE]
The 3rd Decade: 2000-2010
Iowa City & Oklahoma City

Drug-induced SCLE
Drug-induced SCLE

- 1985 - Reed et al at the Univ. of Colorado (Ann Intern Med 103:49–51)
  - 5 patients with typical SCLE skin lesions developed while on hydrochlorothiazide (HCTZ)
  - All 5 patients, the SCLE lesions resolved following HCTZ discontinuation
Drug-induced subacute cutaneous lupus erythematosus: a paradigm for bedside-to-bench patient-oriented translational clinical investigation

Richard D. Sontheimer · Clifford L. Henderson · Renee H. Grau
Systematic Review of Drug-induced SCLE

• 71 reported cases (as of 2007)
  – 47% - Cardiovascular disease (esp. anti-HBP)
• Epidemiology
  – Demographics - Caucasian females, mean age - 59.5 yrs
  – “Incubation period” - 2 weeks - 3.2 years
  – Mean duration to clearance of lesions when drug was stopped - 5.76 weeks (1 - 24 weeks)
Fig. 1. Rates of DI-SCLE by drug class

- Calcium channel blockers
- Antifungals
- Diuretics
- Antiepileptics
- Betablockers
- Antihistamines
- Chemotherapeutics
- Immune modulators
Pantoprazole (Protonix)
24 patients—lansoprazole (12), omeprazole (six), esomeprazole (four) and pantoprazole (two).
Pseudo SCLE still lives!

- Most SCLE patients that are referred to me now appear to be drug induced
  - Recurrent tinea corporis in a man with terbinafine-induced SCLE was confused with reactivation of SCLE
Lesson Learned

Antimalarial-resistant SCLE?
“Recurring” SCLE
Think drug-induced SCLE
Need of a Biomarker for Drug-Induced LE

- Drug-induced T cell stimulation assay
- Experimental model
  - Chronic eczematous drug eruption of the aged
The Next 35 Years?
The End
Thank You
The End
SCLE Follow-Up Study - 1995

Changes in Serum Creatinine

Years of Follow-Up

Serum Creatinine

Upper limits of normal

Skin Dis. Activity at F/U

Inactive
Active
Acute cutaneous lupus erythematosus: a decade's perspective.

Attehimer RD.


ID: 2671533 [PubMed - indexed for MEDLINE]
## Skin Disease in LE Patients

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• Risk for clinically-significant SLE in patients presenting with papulosquamous SCLE skin lesions with central facial involvement
• Greenwald’s Law of Lupus
  – Norwegian scabies; head lice
• Beware SjS complications in SCLE patients
Skin Findings in SjS

- Dryness/Pruritus
- Raynaud's phenomenon
- Angular cheilitis
- Vasculitis
- Annular erythema of SjS
Learning Objective

Present a series of SCLE cases that illustrate salient clinical points of understanding that I have personally gained over the past 4 decades of caring for patients with this form of cutaneous LE
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**Secondary to Treatment of LE**
- Steroid acne
- Drug eruptions
- Opportunistic Infections

**True-True and Unrelated**
- Psoriasis
- Eczema
- BCC

Greenwald’s Law of Lupus