LUPUS AND DERMATOMYOSITIS IN SKIN OF COLOR

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UT Southwestern Medical Center
DISCLOSURE OF RELATIONSHIPS WITH INDUSTRY

Benjamin Chong, MD, MSCS
S057 – Skin of Color

DISCLOSURES

Biogen Inc.: Investigator – Grants
Daavlin Inc.: Investigator - Grants
OUTLINE

- Cutaneous lupus
  - Skin of color
  - Subtypes
  - Treatments
- Dermatomyositis
  - Skin of color
Subacute Cutaneous Lupus Erythematosus

- Case series of 27 patients
- Associated with anti-Ro antibody

Classification of Cutaneous Lupus Erythematosus

- Acute
- Subacute
- Chronic

1Sontheimer RD et al, Arch Dermatol 1979; 115:1409-15
UT SOUTHWESTERN CUTANEOUS LUPUS REGISTRY INCLUDES PATIENTS OF SKIN OF COLOR

- Established in 2008
- Longitudinal study of cutaneous lupus patients
- Clinical data, blood/skin specimens
- 277 cutaneous lupus patients (701 visits)

<table>
<thead>
<tr>
<th>Race/Ethnicity</th>
<th>N</th>
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<tbody>
<tr>
<td>African American/African</td>
<td>143</td>
<td>51.6%</td>
</tr>
<tr>
<td>Caucasian</td>
<td>92</td>
<td>33.2%</td>
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<tr>
<td>Hispanic</td>
<td>27</td>
<td>9.7%</td>
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<tr>
<td>Asian</td>
<td>11</td>
<td>4.0%</td>
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<tr>
<td>Mixed</td>
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<td>1.4%</td>
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PATIENT CASE #1

- 57 y.o. AA female with history of mildly itchy rash on face, worse with sun exposure
- ROS – joint pains in right knee and hip, fatigue
- Meds - none
- Labs
  - ANA 1:1280 (speckled)
  - ENA – RNP positive
  - Negative dsDNA antibody
  - CBC, BUN/Cr – normal
  - U/A – negative for proteinuria
PATIENT CASE #1: WHAT IS YOUR DIAGNOSIS?

- A) Acute cutaneous lupus erythematosus
- B) Discoid lupus erythematosus
- C) Subacute cutaneous lupus erythematosus
ACUTE CUTANEOUS LUPUS (ACLE) IS ASSOCIATED WITH SLE

- Associated with SLE
- Butterfly rash
- Occurs with sun exposure
- Burning, swelling, itching
- Transient

SUBACUTE CUTANEOUS LUPUS (SCLE) FAVORS THE TRUNK AND UPPER EXTREMITIES

- Sun-exposed areas in trunk and upper extremities
  - Face, scalp less involved

- Two forms:
  - Polycyclic/annular
  - Psoriasiform

- Associated with anti-Ro antibody

- Up to 1/3 of cases may be from a medication

¹Gronhagen CM et al, Br J Dermatol 2012; 167:296-305
CHRONIC CUTANEOUS LUPUS HAS SEVERAL SUBTYPES

- Discoid (DLE) - most common chronic cutaneous lupus

- Other subtypes:
  - Lupus panniculitis
  - Tumid lupus
DISCOID LUPUS ERYTHEMATOSUS HAS SCARRING AND DYSPIGMENTATION

- Most common type of cutaneous lupus
- Head and neck predominantly
- Present in 20% of systemic lupus patients
- May also present in patients without systemic lupus
TUMID LUPUS IS A VERY PHOTOSENSITIVE SUBTYPE OF CUTANEOUS LUPUS

- Most photosensitive variant of cutaneous lupus
- Juicy red edematous papules and plaques in sun-sensitive areas
- Up to 10% have SLE
- Responsive to anti-malarials

LUPUS PANNICULITIS AFFECTS THE LOWER DERMIS AND SUBCUTANEOUS FAT

- Painful -> atrophic nodules
- 2-3% of all cases of cutaneous lupus
- On face, upper arms, buttocks, thighs, trunk (breast)
- 70% have overlying DLE = lupus profundus

MY WORKUP ON CUTANEOUS LUPUS PATIENTS

New CLE diagnosis

Review of systems
Physical examination
Medical chart review

Laboratory Testing:
- CBC with differential
- ANA
- Urinalysis
Consider:
- ENA panel
- Anti-dsDNA antibody
- ESR/CRP
- C3 and C4 complements

ANA positive

Further Laboratory Testing:
- ENA panel
- Anti-dsDNA antibody
- ESR/CRP
- C3 and C4 complements

Meets criteria for SLE

Refer to rheumatology and other specialists as indicated by disease manifestations

CLE without SLE

Start first-line therapy

ANA negative

Start first-line therapy

## TREATMENT LADDER FOR CUTANEOUS LUPUS

### Mild
- Sun protective methods
- Topical Steroids/Immunomodulators
- IL Steroids (5-10 mg/cc)

### Moderate/Refractory Mild
- Low dose prednisone (up to 0.5 mg/kg/day)
- Hydroxychloroquine (<5 mg/kg/day)
- Add quinacrine (100 mg qd)
- Switch hydroxychloroquine to chloroquine (<2.3 mg/kg/day)

### Severe/Refractory Moderate
- High dose prednisone (up to 1 mg/kg/day)
- Mycophenolate mofetil (1000-1500 mg bid)
- Methotrexate (up to 15-25 mg qwk)
- Azathioprine
- Dapsone
- Thalidomide (50-100 mg qd)
If unstable, patients should be followed on a biweekly to monthly basis.

If stable, check every 3-6 months.

Taper medications during fall/winter.

With SLE, check CBC, U/A, dsDNA and C3 and C4 levels, and ESR at least once a year to check for disease flares.
PATIENT CASE #2

- 46 y.o. AA female with history of persistent itchy rash on face, upper trunk, hands for one year
- Diagnosed with lupus by Rheumatology
- Medications – methotrexate 20 mg qWK, hydroxychloroquine 200 mg BID, prednisone 15 mg QD, IV belimumab
- ROS – muscle weakness
PATIENT CASE #2

- Biopsy – interface dermatitis with mucin
- Laboratory values
  - CK, aldolase high
  - ANA negative
  - ENA negative
  - Mi-2, Jo-1, Scl-70, PM-SCl antibodies negative
PATIENT CASE #2 – WHAT IS THE DIAGNOSIS?

- A) Cutaneous lupus
- B) Dermatomyositis
DERMATOMYOSITIS

- Type of idiopathic inflammatory myopathy
- Inflammatory myopathy with cutaneous eruption
- Clinically amyopathic dermatomyositis presents without muscle involvement

DERMATOMYOSITIS IN PATIENTS WITH SKIN OF COLOR

Heliotrope rash
Courtesy of Dr. Judith Cherit

Poikiloderma
Courtesy of Dr. Lu Le

Shawl sign

Bowman KA, Chong BF. Dermatomyositis. Dermatology Atlas for Skin of Color. 2014
Gottron’s papules

Tendon streaking

Cuticular hypergrowth, periungual telangiectasias

Bowman KA, Chong BF. Dermatomyositis. Dermatology Atlas for Skin of Color. 2014
DERMATOMYOSITIS IN PATIENTS WITH SKIN OF COLOR

Gottron’s sign  | Dermatomyositis panniculitis  | Calcinosis cutis

Courtesy of Dr. Chauncey McHargue

Bowman KA, Chong BF. Dermatomyositis. Dermatology Atlas for Skin of Color. 2014
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<tr>
<th></th>
<th>Cutaneous lupus</th>
<th>Dermatomyositis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Location/Distribution</td>
<td>Between knuckles, V-shaped on back</td>
<td>Favor elbows, knees, knuckles</td>
</tr>
<tr>
<td>Color</td>
<td>Erythematous</td>
<td>Violaceous</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Joint aches more prominent</td>
<td>Scalp pruritus, muscle weakness</td>
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MYOPATHY IN DERMATOMYOSITIS

- Affects proximal muscles
- Symmetrical
- Symptoms
  - Myalgias
  - Fatigue
  - Muscle weakness
- Dysphagia
- Aspiration
Arthralgias/arthritis

Pulmonary involvement

- Interstitial lung disease
  - Often mild and chronic
  - PFTs can normalize with immunosuppressive treatment
- 17-23% of DM patients

Cardiac involvement (rare)

WORKUP FOR DERMATOMYOSITIS PATIENTS

- **Skin biopsy**
  - Gottron’s papules – low yield
- **Laboratory tests**
  - ANA
  - Muscle enzymes (CK, aldolase, LDH, ALT)
  - Antibodies **(only if diagnosis in question)**
    - Mayo Clinic myositis panel – Mi-2, PL-7, PL-12, EJ, OJ, SRP, Ku, U2-snRNP
    - Jo-1 (Part of ENA panel)
    - MDA5 autoantibodies
    - TIF-1γ autoantibodies
WORKUP IN DERMATOMYOSITIS PATIENTS

- MRI
- EMG
- Muscle biopsy
  - Perifascicular atrophy, inflammation around blood vessels, – dermatomyositis
  - Inflammation around muscle fibers - polymyositis

Mammens AL, Ann NY Acad Sci 2010; 1184:134-53
TREATMENT LADDER FOR DERMATOMYOSITIS

**Mild (skin-limited)**
- Sun protective methods
- Topical Steroids/ Immunomodulators
- Hydroxychloroquine (<5 mg/kg/day)
- Add quinacrine (100 mg QD)
- Switch hydroxychloroquine to chloroquine (<2.3 mg/kg/day)

**Moderate/Severe (with systemic symptoms)**
- Prednisone (0.5-1 mg/kg/day)
- Mycophenolate mofetil (1000-1500 mg BID)
- Methotrexate (15-25 mg QWK)
- IV Immunoglobulin
- Rituximab
- Tofacinitib
Cutaneous lupus and dermatomyositis can be distinguished by color, scalp involvement, and lesion distribution.

- Pigmentary changes are pronounced in DLE and dermatomyositis in patients with skin of color.

Carefully screen patients for systemic involvement with thorough history and physical exam and selected lab testing.

Treatments are similar, but immunosuppression can be more aggressively used in patients with dermatomyositis.
REFERENCES

- Bendewald MJ et al, Arch Dermatol 2010; 146:26-30
- Bowman KA, Chong BF. Dermatomyositis. Dermatology Atlas for Skin of Color. 2014
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- Sontheimer RD et al, Arch Dermatol 1979; 115:1409-15
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ACKNOWLEDGEMENTS
THE DERMATOLOGY FOUNDATION HAS SUPPORTED & ADVANCED MY CAREER.