Lupus and Scarring Alopecia

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Introduction

- Autoimmune disorder
- Affects skin, kidney, joints, heart, lungs
- Signs and symptoms
  - Butterfly rash
  - Joint pain, stiffness, swelling
  - Fatigue, shortness of breath, chest pain
  - Raynauds, photosensitivity
Epidemiology

- 6:1 Ratio Women to men
- 4x higher in African American women than in caucacian women
- African americans higher incidence
  - Nephritis, pneumononitis, discoid lesions
  - Develop disease at an earlier age
  - Higher mortality rate
Pathogenesis

- Cause is not known
- Some agent (?infection) triggers the immune system to react by attacking "self."
- Genetics play a role
- Environment may also play a role
Classification of Skin Lesions

- Acute
- Subacute
- Chronic
CHARACTERISTIC SITES OF INVOLVEMENT FOR THE THREE MAJOR SUBTYPES OF CUTANEOUS LUPUS ERYTHEMATOSUS

Acute cutaneous LE
- "Butterfly" rash

Subacute cutaneous LE

Chronic cutaneous LE
- Alopecia within lesions

- LE tumidus
- Lupus panniculitis
- Discoid LE
- Chilblain lupus
Classification of Skin Lesions

• **Acute Cutaneous Lupus**
  – Butterfly malar rash
  – Follows sun-exposure
  – Associated with systemic lupus
Acute Cutaneous LE
Acute Cutaneous LE

- Associated with systemic LE
- Malar ‘butterfly’ rash
  - Erythema
  - Scale
  - Edema
  - Papules
  - Follows sun exposure
- Other areas can also be involved
Systemic LE

- Women of childbearing age
- Skin involvement in ~80%
  - 4 of ACR’s criteria for SLE are mucocutaneous
- Prevalence 4x greater in AA
  - ↑ nephritis, pneumononitis, discoid lesions
  - ↓ photosensitivity
  - earlier age of onset
  - higher mortality
ACR’s Criteria for SLE

- Malar rash
- Discoid rash
- Photosensitivity
- Oral ulcers

- Positive ANA
- Arthritis
- Proteinuria – >0.5 g/day or casts
- Neurologic disorder – seizures, psychosis
- Pleuritis/pericarditis
- Blood abnormalities – hemolytic anemia, leukopenia, thrombocytopenia
- Immunologic disorder – anti-dsDNA, anti-Sm, anticardiolipin antibodies
Cutaneous findings in SLE

- **Vascular lesions**
  - Erythema, edema of fingers, toes
  - Periungual tlg’s
  - Red or spotted lunulae
  - EM like lesions (Rowell syndrome)
  - Livedo pattern (r/o Sneddon syn)

- **Diffuse, nonscarring alopecia**
  - Lupus hairs

- **Mucous membrane ulcerations**
- **Multiple, eruptive dematofibromas**
- **Leg ulcers**
- **Calcinosis cutis**
Classification of Skin Lesions

• **Subacute**
  – Confined to sun-exposed areas
  – Photosensitive
  – Lesions DO NOT SCAR
  – Can be caused my medications (HCTZ, diltiazem, terbinafine)
  – Anti-Ro Antibodies
Classification of Skin Lupus

• Chronic Cutaneous Lupus
  – Discoid Lupus
    • Discoid lesions on face, scalp & ears
    • Can occur on mouth and lips
    • Disfiguring scarring & hair loss
    • Goes deep into skin to affect hair follicles
    • Discoloration-white centers and dark edges
    • Can develop skin cancer within lesions
    • 5-10% can develop systemic lupus
Discoid LE

- Young adults, women:men = 2:1
- Indurated scaly plaques
  - Above neck: face, scalp, ears
- Atrophy, scarring, pigment changes
- 95% only skin involvement
Generalized DLE

- Generalized variant
  - Less common
  - Head, Neck, Thorax, Extremities
  - ANA, ESR, ssDNA
  - Leukopenia
- Adnexal involvement
  - Follicular plugging
  - Scarring alopecia
- Mucosal involvement
- Early perifollicular erythema
- Easily extracted anagen hairs

- Smooth, shiny scarring
- Loss of follicles
Mucosal erosions with rim of erythema
Progression to SLE uncommon
- ~ 5-10%
- Higher risk with extensive disease
- 1/11 criteria for SLE

Pts with SLE freq have DLE
- Fever, arthralgias
- ANA, dsDNA

Development of SCC in chronic lesions
Chronic Cutaneous Lupus

• **Lupus Panniculitis**
  – Affects fat within the skin
  – Leads to depressed areas
  – Occurs on face, upper arms, **scalp**, buttocks, thighs, breast, trunk
Lupus panniculitis
Lupus panniculitis/profundus

- Women 20-45
- Firm, nontender SQ nodules
- Proximal extremities
- Overlying skin normal, +/- DLE
- Heal with depressions
- Path: Lymphoid follicles in septae, fat necrosis, “Frost on a window pane” – MC in lipodermatoscl but may be present
Work-up for Lupus
Labs

• Biopsy the affected areas
• CBC: Hemolytic anemia, thrombocytopenia
• Lymphopenia
• ESR $\rightarrow$ correlates with activity
• IgG elevated, serum globulin increased ($\gamma$)
• Urine $\rightarrow$ albumin, RBCs, casts
Immunologic findings in SLE

- **ANA very sensitive (95%)**
- **Anti-dsDNA, most specific (~60%)**
  - Correlates with disease activity
  - Renal disease
- **Anti-Smith**
  - Low sensitivity, very specific
- **Anti-rRNP (~7%)**
  - MCTD, neuropsychiatric manifestations
- **Lupus band (DIF)**
  - Continuous granular deposits of Ig and complement along DEJ ('full house')
Immunologic findings in SLE

- Anti-La → SCLE, Sjogren syndrome

- Anti-Ro → 25% SLE, 40% Sjogrens, SCLE (70%), neonatal LE (95%), C2-C4 deficient LE (50-75%), late onset LE (75%)
Treatment of Cutaneous LE

- Sun avoidance and sunscreen
- Topical steroids
- ILK
- Antimalarials – first line therapy
  - Plaquenil. Dose?
    - 6.5 mg/kg/day IBW
- Corticosteroids
  - Calcium + Vit D, bisphosphonates
- Steroid sparing agents
Other Treatment Options

• Azathioprine  
  – 1.5-2.5 mg/kg/day

• Mycophenolate mofetil  
  – 2.5-3.5 g/day

• Methotrexate  
  – 7.5-25 mg po or SQ/week
Lupus and Hair Loss
Hair Loss

- Caused primarily by Discoid lupus
- Also seen in Lupus profundus
- Affects deeper layers of skin where hair follicles and fat are located
Discoid Lupus
Lupus Panniculitis
Treatment

- Topical steroids
- Intraleisional steroids
- Antimalarials (hydroxychloroquine)
- Systemic steroids
- Steroid sparring agents: azathioprine, mycophenolate mofetil
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Not For Use By Women

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Thank You
Questions

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