Treating dermatomyositis

David Fiorentino, MD, PhD
Stanford University School of Medicine
Department of Dermatology
Department of Medicine (Rheumatology)
September 25, 2010
DM affects many organs
Approaching DM therapy

• Avoid things that make disease worse
Cardinal Rule:
Avoid things that worsen skin disease

• Dry skin
• Smoking
• Sunlight
Sunprotection

MAYBE NEXT TIME YOU'LL TRY A LITTLE SUNSCREEN...
Components of Radiation from the Sun

- Cosmic Rays
- Gamma Rays
- X-Rays
- Ultraviolet
  - Short Wave UV-C Band
  - Medium Wave UV-B Band
  - Long Wave UV-A Band
- Visible Light
- Infrared
- Radio Waves

UV-A-II (320-340)
UV-A-I (340-400)
SPF factor refers to protection from what UV wavelengths?

- UVA
- UVB
- UVA and UVB
- Neither
SPF factor refers to protection from what UV wavelengths?

• UVB
Ultraviolet Light: Physical Protection

- Wear tightly-woven protective *broad-brim* hats and clothing, e.g. long-sleeves and pants
SPF 3-7

SPF 6.5
Ultraviolet Light: Physical Protection

- Wear tightly-woven protective *broad-brim* hats and clothing, e.g. long-sleeves and pants
- Use laundry product that can be added to detergent to increase UV protection in clothing, e.g. Rit Sun Guard (ritdye.com)
- Apply UV blocking films to windows in cars and home
Sunscreen: which one??

- SPF >30
- Look for
  - Titanium dioxide; OR
  - Zinc oxide
Sunscreen: which one??

- SPF >30
- Look for
  - Titanium dioxide; OR
  - Zinc oxide
- Need multiple chemical blockers
Chemical UVA blockers

- Avabenzene (Parsol 1789)
- Oxybenzone
- Mexoryl SX
Use More Than Half a Teaspoon Each on
Head and Neck Area
Right Arm
Left Arm

Use More Than a Teaspoon Each on
Anterior Torso
Posterior Torso
Right Leg
Left Leg
Approaching DM therapy

• Avoid things that make disease worse
• Determine if medical therapy is indicated
Activity ← Damage →
Determining disease activity

- **Muscle**
  - Weakness not necessarily helpful
  - Muscle enzymes in blood
The role of EMG

- Can diagnose a primary myopathy
- Cannot differentiate between myopathies
- Helpful if weak on exam but muscle enzymes normal
The role of muscle biopsy

- Can be diagnostic for dermatomyositis
- Not 100% sensitive
Determining disease activity

• Skin
  – Redness not always helpful
Determining disease activity

- Skin
  - Redness not always helpful
  - ITCH
Approaching DM therapy

• Avoid things that make disease worse
• Determine if medical therapy is indicated
• Select optimal medical therapy
General concepts

• Not every therapy works for everybody
• Therapy is empiric—trial and error
• Don’t give up!
Corticosteroids

- Generally effective for most organ systems
- Act quickly
- Cheap
- Limited by long term side effects
**IVIG**

- Pooled preparation of blood proteins from multiple donors
- Generally effective for skin and muscle
- Acts quickly
- Side effects: allergic response, kidney, rare chance of transmitting infection
- EXPENSIVE
- Tends to lose effectiveness over time
Methotrexate

- Mechanism of action actually not clear
- Often effective for muscle and/or skin
- Slower to act
- Cheap
- Side effects: liver, GI, low blood counts, rare lung inflammation
- Risks of infection, ?cancer (lymphoma)
Azathioprine (Imuran)

- Inhibits ability of lymphocytes to multiply
- Can be effective for muscle or skin (lung)
- Cheap
- Side effects: allergy, low white blood cells, liver irritation
- Risks of infection, ?cancer
Mycophenolate mofetil (Cell Cept)

- Inhibits ability of lymphocytes to multiply
- Can be very effective for muscle/skin
- Lung data look promising
- Side effects: GI upset, low blood counts,
- Risks of infection (?herpes, shingles), cancer?
- EXPENSIVE
Cyclophosphamide (Cytoxan)

- Inhibits ability of lymphocytes to multiply
- Generally reserved for severe disease
- Can be useful for ulcerations/gangrene of skin
- Side effects: low blood counts, ovarian failure, bladder irritation/cancer
Skin-specific therapies
ITCH
Itch: Basic Treatments

• Dry skin care
  – Moisturizing soaps
  – Apply moisturizing cream within 3 minutes after bathing and again during day
  – Avoid hot showers; keep less than 10 min
  – Consider use of humidifiers
  – Minimize use of central heating
Itch: Topical Therapy

- Immediately soothing
  - Menthol-based (i.e. Sarna lotion)
  - Pramoxine-based (i.e. Prax lotion, pramasone)
  - Avoid topical anti-histamines
    - May cause allergies over time
  - Cool compresses or ice
Itch: Prescription Therapy

- **Systemic**
  - Anti-histamines (hydroxyzine, doxepin)
  - Naltrexone
  - Mirtazapine
  - Gabapentin
  - Thalidomide
  - Butorphanol
Best therapy for itch......

Treat underlying inflammation!
Topical therapies for DM

• Corticosteroids
Topical Corticosteroids

- **Group I steroids** (e.g., Temovate) - use cyclically in others
- **Group II steroids** (e.g., Lidex)
- **Group III-IV steroids** (e.g., triamcinolone acetonide 0.025-0.1%) for general widespread use on trunk and extremities
- **Group VI-VII steroids** (e.g., hydrocorticone cream 0.1%) for face and flexural areas/body folds
Topical Corticosteroids

• **Vehicles**
  - sprays
  - solutions
  - foams
  - gels
  - creams
  - ointments

Patient acceptance & compliance

Moisturization
Topical therapies for DM

• Corticosteroids
• Immunomodulators
  – Tacrolimus (Protopic)
  – Pimecrolimus (Elidel)
Hydroxychloroquine (Plaquenil)

- Mechanism of action unknown—generally NOT immunosuppressive
- Can be effective for skin, not muscle
- Side effects: GI, rare liver/muscle inflammation, rare eye toxicity
- SKIN RASH (up to 30% DM patients)
- Consider switch to chloroquine
Dapsone

- Sulfa type drug
- Blocks lymphocyte function
- Can be effective for mild skin disease
- Side effects: allergy, anemia, liver irritation, nerve damage
Leflunomide (Arava)

• Blocks ability of lymphocytes to multiply
• Approved for Rheumatoid Arthritis
• 2 Case Reports of effectiveness for skin and muscle disease
• Side effects: GI, low blood counts, liver
Approaching DM therapy

• Avoid things that make disease worse
• Determine if medical therapy is indicated
• Select optimal medical therapy
• Damage reversal
Muscle damage

• Non inflammatory weakness

• Determine cause
  – Atrophy from previous DM inflammation
  – Corticosteroids
  – Lack of use

• Physical therapy

• Low weight bearing exercise is helpful
Skin damage

- Enlarged blood vessels
Skin damage

• Enlarged blood vessels
  – Laser (intense pulsed light) therapy
Skin damage

• Enlarged blood vessels
  – Laser (intense pulsed light) therapy
• Calcium deposits
Managing calcinosis cutis

• Aggressive prevention is the key
• Treatments
  – Treat superinfection and pain
  – Aluminum hydroxide, warfarin, Ca^{++} channel blockers, bisphosphonates
  – Local surgical removal
New therapies?

• Rituximab
  – Kills B lymphocytes
  – Preliminary data suggest it can help muscle disease; we have not found it effective for skin disease
  – Large, controlled trial (RIM) completed and await data
New therapies?

• Blocking interferon signaling
  – Evidence that interferon is overactive in skin, muscle and blood of DM patients
  – MEDI 545 (Medimmune)—early phase I trial completed; no data released.

www.medimmune.com
What will improve our therapy?

• Need “biomarkers” to determine how much of symptoms will be reversible with medication
• Need better classification of disease so we can individualize therapy
• Better understanding of what actually causes the disease