Skin Care and Wound Management in Systemic Sclerosis

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Major Skin Issues of Systemic Sclerosis

What is going on?
Changes to the skin

What can I do?
Patient management

How can my doctor help?
Prescriptive therapies

Wound Healing in Scleroderma
Localized Sclerosis
Changes to the Skin- What is Happening

Specifically at the skin cell level
Fibroblasts over produce collagen and other proteins →
Dermal fibrosis (skin thickening)
Loss of subcutaneous fat, epidermal atrophy (loss of strength in skin) →
loss of sweat glands, hair →
Thick, tight, dry skin with decreased joint mobility that is more prone to
damage/infection.

AND
Damage to and subsequent thickening of vessel walls with narrowing and
possible obliteration of the vessel lumen

Chapman, 2006; Alivernini, 2009; Zwischenberger, 2011
Phase 1: Edematous Changes

- Painless swelling of hands and fingers
- Symptoms include morning stiffness, carpal tunnel (from nerve compression), pitting edema of fingers and top of hand
- Thickening of the fingers and hands in virtually all cases.
- Skin appears shiny and taut. May be areas of redness.
- Itching is common and intense
- Local inflammation and changes to small blood vessels

- Benadryl over the counter
- Low dose corticosteroid cream

Chapman, 2006; Alivernini, 2009; Zwischenberger, 2011
Phase 2: Thickening

- Thickening will start in hands and fingers in almost all cases
- Progressively skin on the body become tightened and hair growth decreases
- Skin on neck and face is usually next
- Lips become thin and pursed and furrowing around the mouth

Alivernini, 2009; Manetti, 2010; Marvi, 2010; Zwischenberger, 2011
Subtypes of Scleroderma

Linear:
Streaks or bands of hard, waxy skin on arms, legs or face

Circumscribed:
Patches of hard, dry, smooth, usually on the belly, chest and back

Circumscribed
Linear
Systemic Therapeutic Algorithm

ACTIVE
New lesions ≤6 month's duration, disease extension, inflammation (erythema, edema), sclerotic or indurated periphery

SUPERFICIAL

LOCALIZED
+ Topical (bid, occluded)
- Calcipotriene
- Tacrolimus

Phototherapy
- localized or whole body
- NB UVB, BB UVA, UVA-1

Continued progression

Progression with local phototherapy

GENERALIZED

Phototherapy (whole body)
- NB UVB, BB UVA, UVA-1

Continued progression/Deep involvement

DEEP

LOCALIZED
Functional/cosmetic threat

GENERALIZED
Systemic
- MTX
- PCMT

*INACTIVE/DAMAGE
Pigmentary changes, static size, atrophy, central sclerosis

FUNCTIONAL IMPAIRMENT:
- PT/OT
- Orthotics
- Orthopedics
- Podiatry
- Rheumatology
- Oral maxillofacial surgery
- Plastic surgery

COSMETIC IMPAIRMENT
Confirmed long-term disease inactivity

Work-up negative for deep muscle, fascia, bone involvement

Local excision

Inject fillers (face)

Zwischenberger, 2011
Immunomodulators

- Methotrexate orally alone
- Methotrexate with pulsed IV corticosteroids (usually methylprednisone)
- Tacrolimus 0.1% ointment or calcipotriene 0.005% to lesions under occlusive dressings
  - Early inflammatory lesions resolve in approx. 1 month
- Skin softens

Kroft, 2009; Stefanaki, 2008; Zwischenberger, 2011; Kreuter, 2005
Phototherapy

- Used for superficial treatment; not for involvement of subcutaneous tissue, muscle or tendons.

- Ultraviolet light increases enzymes that breakdown collagen in the skin and decrease levels of enzymes the trigger collagen production.

- Considerations of UV therapy are:
  - Side effects: itching, redness, burns, and reactivation of herpes simplex virus.
  - Accelerated skin aging and skin cancer for long term therapy.

Andrea, 2010; Kreuter, 2006; Kroft, 2008
Phototherapy

- 15 minutes, 5 times per week, 8 week course, of UVA1
- In addition to systemic treatment, not stand alone
- Visual improvements, tightness and itchiness significantly decreased, and on US decreased

Kreuter, 2006
A pilot of 17 patients by Sandqvist et al, investigated the effects of hand exercise and the use of warm (50 degrees C) Daily x 30 days
Significant increased mobility and decrease in perceived stiffness of the treatment hand (the opposite hand was used as the control

Pilot study of 26 patients Pfizenmaier et al investigated intermittent pneumatic compression would aid in the healing of ulcers on the upper extremities, as it had with lower extremities.
5 hrs per day demonstrated a 96% (26 out of 27) ulcer healing rates with a mean healing time of 25 weeks

Chapman, 2006
Emollient Creams

- Emollients are non-cosmetic moisturizers which come in the form of creams, ointments, lotions and gels.
- Emollients help skin to feel more comfortable and less itchy.
- They keep the skin moist and flexible, helping to prevent cracks.
- Mineral oil, petroleum and Vaseline
Hydroxyethyl Urea and Diazolidinyl Urea

- Softens skins
- Promotes pH balance to protect acid mantle
- Mild local anesthetic
- Moisturizer for extra dry skin

Miller, 2013
The Acid Mantle

- Shields skin against bacteria and environmental pollutants
- Regulates the skin’s oil and water balance
- Made up of perspiration from the sudoriferous gland and oil from the sebaceous gland
- Only slightly acidic; pH level between 5.5 and 6.5

Taken from [http://www.besthealthfoodstore.net/healthy_living_blog/2015/3/pHace-The-Facts-of-pH-Balance-for-Healthy-Skin-Care](http://www.besthealthfoodstore.net/healthy_living_blog/2015/3/pHace-The-Facts-of-pH-Balance-for-Healthy-Skin-Care) on August 10, 2015
Normal pH

- Sebum membrane
- Natural moisturising factor
- Lipid

Allergenic & non-allergenic stimulus
Bacteria & Virus

Moisture

Nerve (itchiness)

Sebaceous gland

Disrupted pH

- Allergenic & non-allergenic stimulus
- Bacteria & Virus

Moisture

Nerve (itchiness)

Sebaceous gland

What to Avoid in Creams

- Retinol, Retinoid, Retin A
- Vitamin A
- Vitamin C
- Alpha hydroxyl acid

They encourage production of collagen and stop collagen break down - great for antiaging, worsens scleroderma skin changes

Fraser, 1996; Telang, 2013; Kornhauser, 2010
Sodium Lauryl and Lauretha Sulphate

INGREDIENTS: Aqua, Sodium Lauryl Sulfate, Sodium Laureth Sulfate, Cocamidopropyl Betaine, Sodium Chloride, Sodium Citrate, Citric Acid, Sodium Xylenesulfonate, Parfum, Cocamide MEA, Sodium Benzoate, Tetrasodium EDTA, PEG-60 Almond Glycerides, Butylphenyl Methylpropional, Panthenyl Ethyl Ether, Panthenol, Guar Hydroxypropyltrimonium Chloride, Benzyl Alcohol, Linalool, Hexyl Cinnamal, Limonene, Benzyl Salicylate, Magnesium Nitrate, Hydroxyisohexyl 3-Cyclohexene Carboxaldehyde, Methylchloroisothiazolinone, Magnesium Chloride, Methylisothiazolinone, Fumaric Acid

For more information on the ingredients, please visit www.mywomenstuff.com.

Irritant, Sensitivity and Allergens

- SLS is a known skin irritant—as per the MDS sheets
- Research in medical journal The Lancet demonstrates SLS damages the skin barrier function, altering skin cells and enhancing any allergic response to other toxins and allergens.
- Hair follicles are larger than regular pores on the face
- This ingredient alone can be responsible for dry hair and itchy scalp.
- SLS in toothpaste can be linked with canker sores

Skin may actually soften

Telangiectasia (spider veins) common on hands and face, especially in limited cutaneous SSc

Calcinosi? may form on pads of finger tips and over joints in the fingers

Painful digital ulcers on fingertips resulting from local ischemia and vascular insufficiency

Hypo- or hyperpigmentation in areas of skin, loss of hair follicles and loss of sebaceous glands with dryness as a result

Pruritus associated with dry skin can be so intense it can impact quality of life

Alivernini, 2009; Manetti, 2010; Marvi, 2010; Zwischenberger, 2011; Muratore, 2013;
Telangiectasia (Spider Veins)

- Damage of the small blood vessels
- No pain associated
- Cosmetic problem, particularly on the face.
- Can be covered with subtle green foundation makeup
- Laser or other light therapy may be useful in larger lesions
Calcinosis

- Besides digital calcinosis patients may develop calcium deposits along the forearm, elbow, buttocks, thigh, knee or shin.
- Mechanism is not completely understood
- 1/2 of patients with calcinosis have no symptoms.
- Can be painful and inflamed may respond to Colchicine (anti-inflammatory used to treat gout) and NSAIDS
- Calcinosis responsible for ulceration lesions require antibiotics for infection and occasionally surgical debulking.
- Surgical removal of calcinosis is indicated when wound healing is not occurring
- Low dose warfarin (1mg per day) in small and new onsets has been effective within 2 months
- Chronic and large calcinosis no effect

Sparsa, 2005; Cukierman, 2004
Hyper- and Hypopigmentation
Raynaud's Phenomenon

- Complex etiology of development of vasculopathy, activation of cellular and humoral immune responses and progressive fibrosis.
- Episodic event of vasoconstriction of the digital arterial structures
- Usually occurs in one or several digits after exposure to the cold or stressful situation

Alivernini, 2009; Chapman, 2006; Manetti, 2009; Muratore, 2013; Marvi, 2010
Evaluation Algorithm

Marvi, 2010
Approach to Digital Ulcers

- Keep warm
- Smoking cessation
- Decrease activity level
- Tight control of blood sugars in diabetes
- Tight control of hypertension
- Basic moist antimicrobial dressing to prevent infection and promote skin integrity
  - PolyMem (Ag)
  - Medihoney (Manuka Honey)
  - Polysporin
- If suspect osteomyelitis treat with IV antibiotics
- Pain management
- Aggressive vasodilation
  - Start with oral calcium channel blockers
  - IV prostacyclin (not only vasodilator but also inhibits platelet aggregation)
- Surgical intervention

Chapman, 2006; Kreuter, 2005; Manetti, 2010; Shanmugan, 2010; Vilel, 2010; Zwischenberger, 2011
Approach to Wound Healing

Treat the Cause
Systemic treatment of sclerosis
Anticoagulation
Vascular supply

Local Wound Care

Debridement
Surgical, Enzymatic, Autolytic
Biological, Mechanical

Infection
Local vs systemic
Treat with local antimicrobial dressing vs antibiotic support

Patient Centered Concerns
Pain
Cosmetic concerns
Limitation to activity
Lifestyle modifications

Moisture Balance
Dressing choice
If it is wet dry it, if it is dry wet it, if it is moist protect it

Stalled healing
Steroid injections
Negative pressure therapy
Hyperbaric therapy
Electric stimulation
Dressings
Patient centered Multidisciplinary approach is BEST!!


References