Skin Care and Wound Management in Systemic Sclerosis

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Outline

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

Fibroblasts over produce collagen and other extracellular matrix proteins; damage to and subsequent thickening of vessel walls with narrowing (and possible obliteration of the vessel lumen) manifests as vasculopathy and an element of autoimmunity develop.

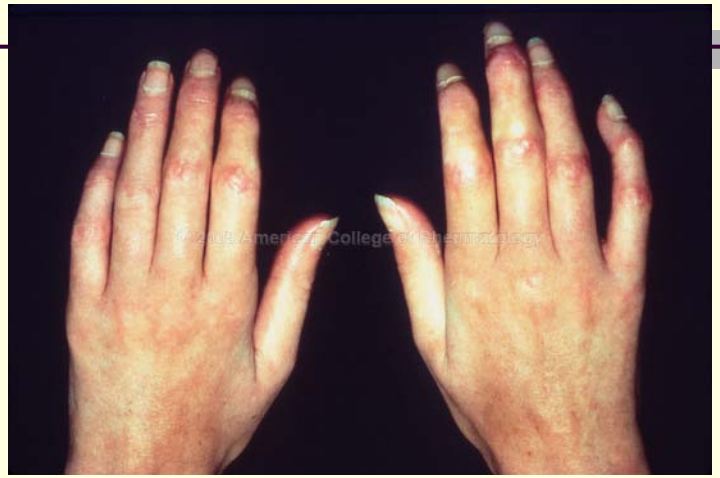
Specifically at the skin level:

Dermal fibrosis loss of subcutaneous fat, epidermal atrophy → loss of sweat glands, hair → thick, tight, dry skin with decreased joint mobility that is more prone to damage/infection. Characteristic “mask-like” face with beak nose and radial perioral furrows

With so many systems affected by this disease (nutrition, circulation, oxygenation, cardiac output etc) and with tight, dry skin the person with SSc can easily be wounded and experience difficulty in healing.

Stages of Skin Changes

- Edematous changes (local inflammation, hydrostatic effect and micro vascular disruption)
  - Painless swelling of hands and fingers
  - Symptoms include morning stiffness, carpal tunnel, 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



- Skin creases become obscure and hair growth decreases
- Skin on neck and face is usually next
- Lips become thin and pursed and radial furrowing around the mouth
- Local skin thickening limits the ability to fully open the mouth and impairs effective dental hygiene
- Prominent localized areas of hyper- and hypo pigmentation may develop

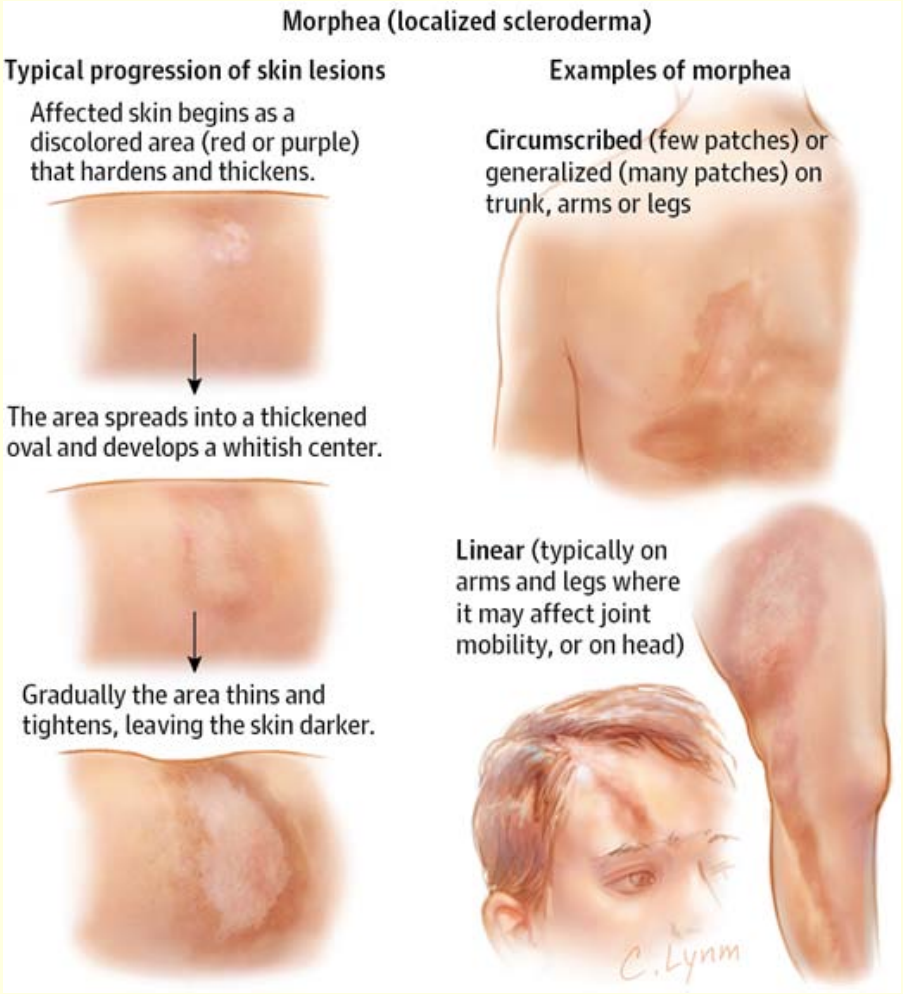
Localized or Morphea Subtypes of Scleroderma

Linear:

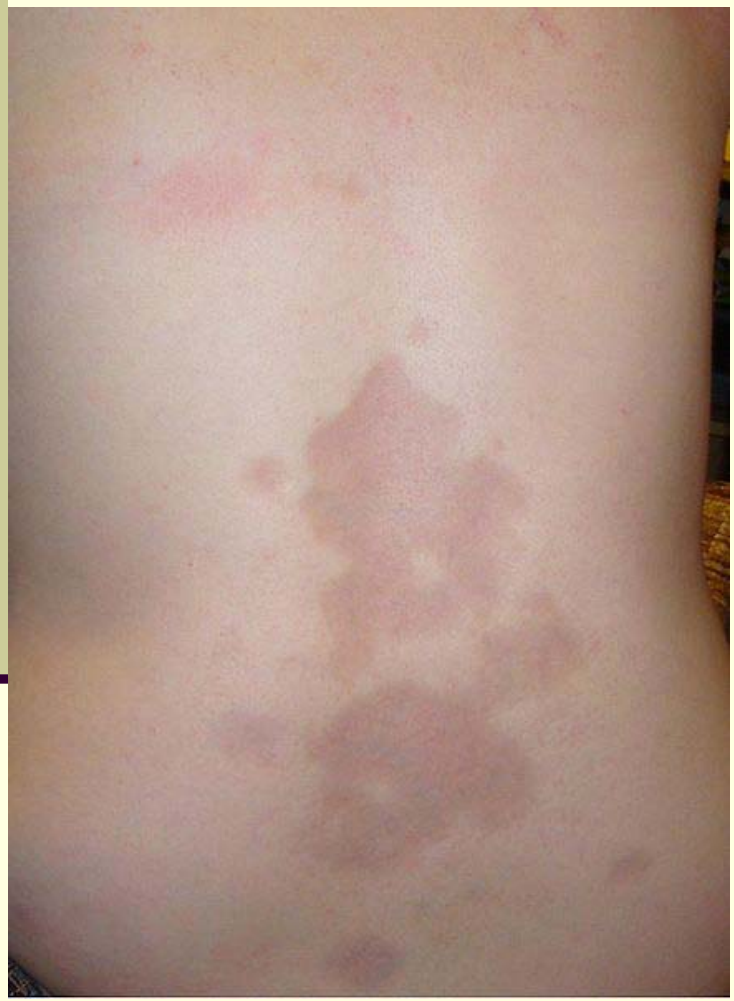
Streaks or bands of hard, waxy skin on arms, legs or face

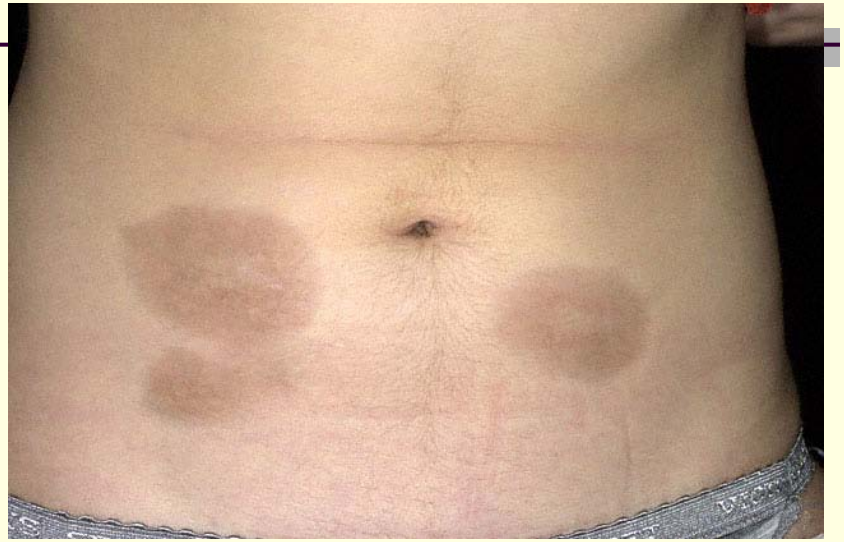
Circumscribed:

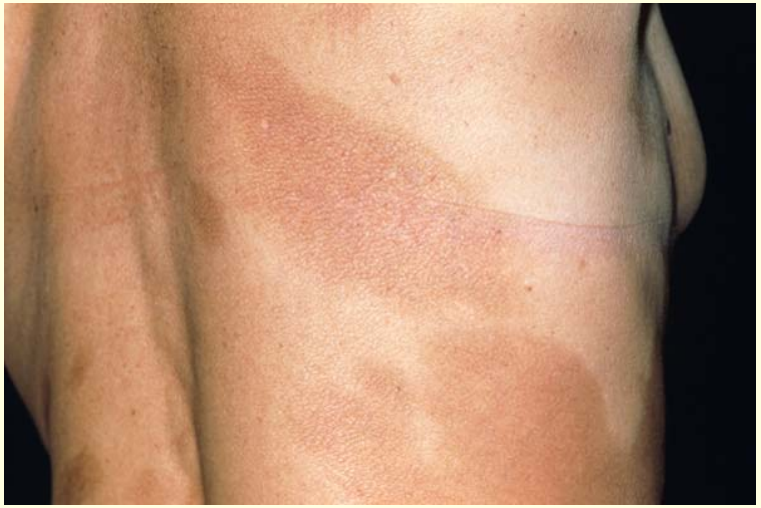
Patches of hard, dry, smooth, usually on the belly, chest and back



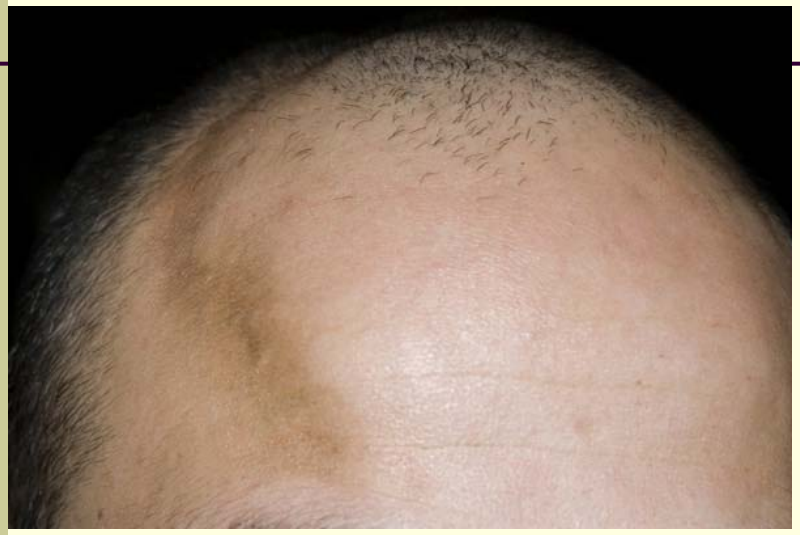
Circumscribed

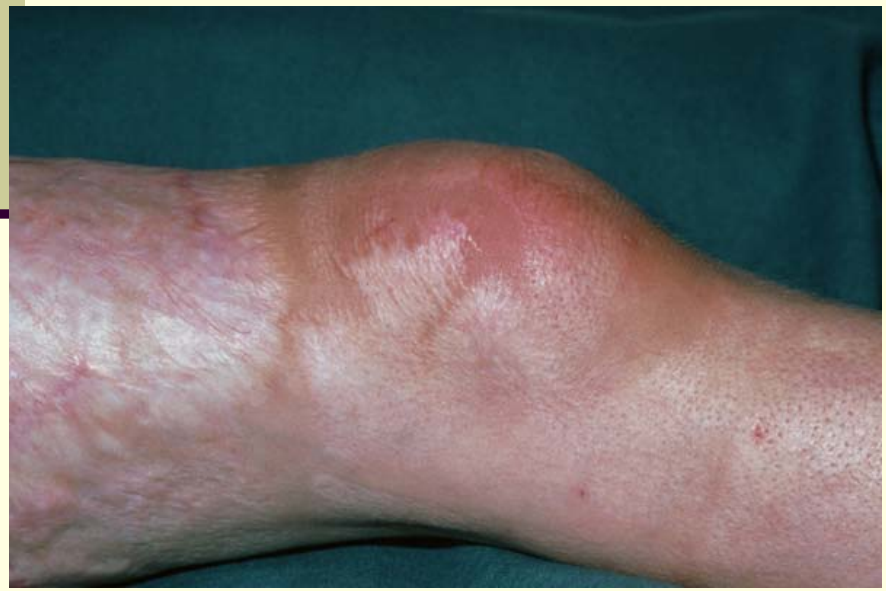




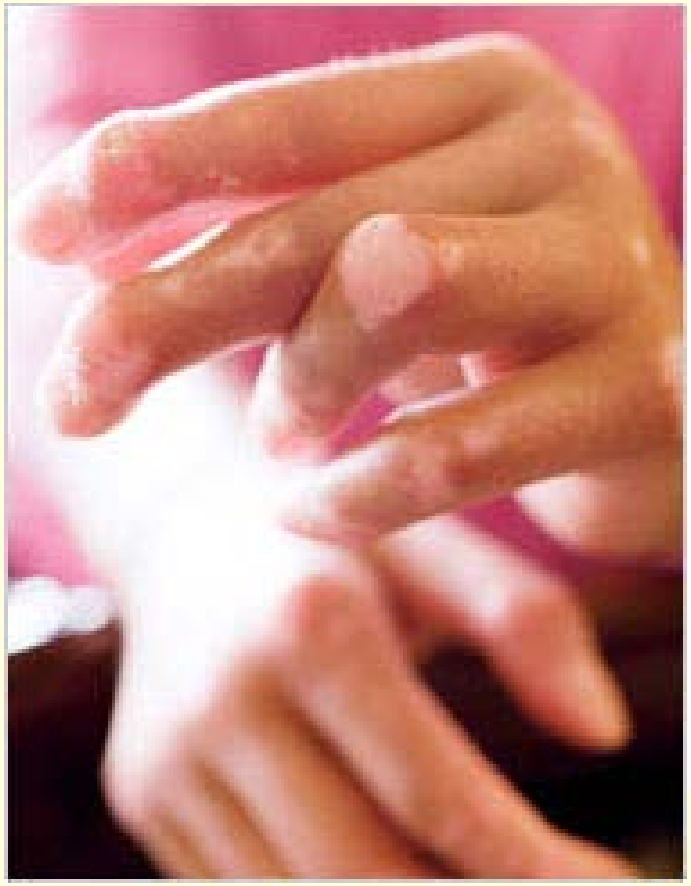


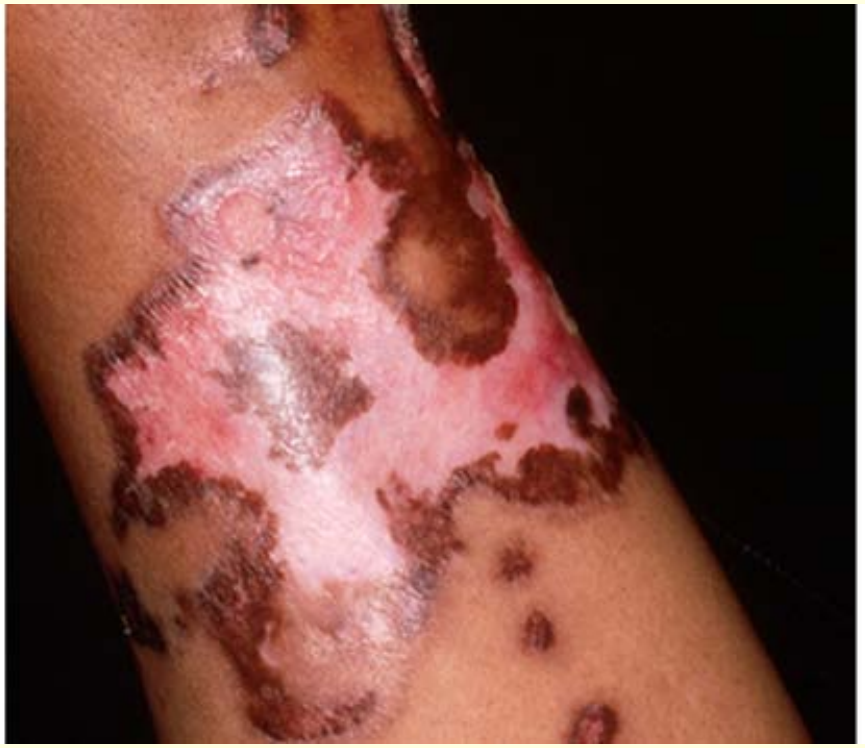
Linear





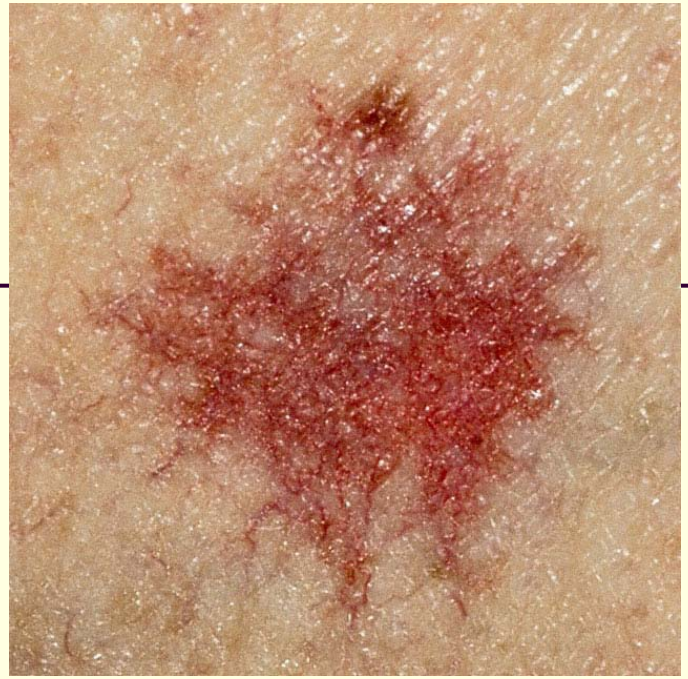
Hyper- and Hypopigmentation

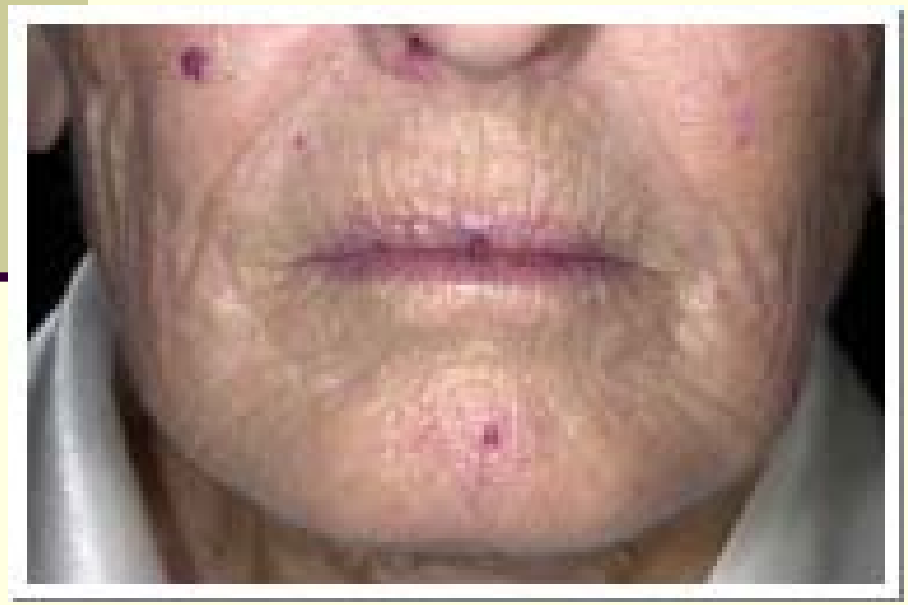


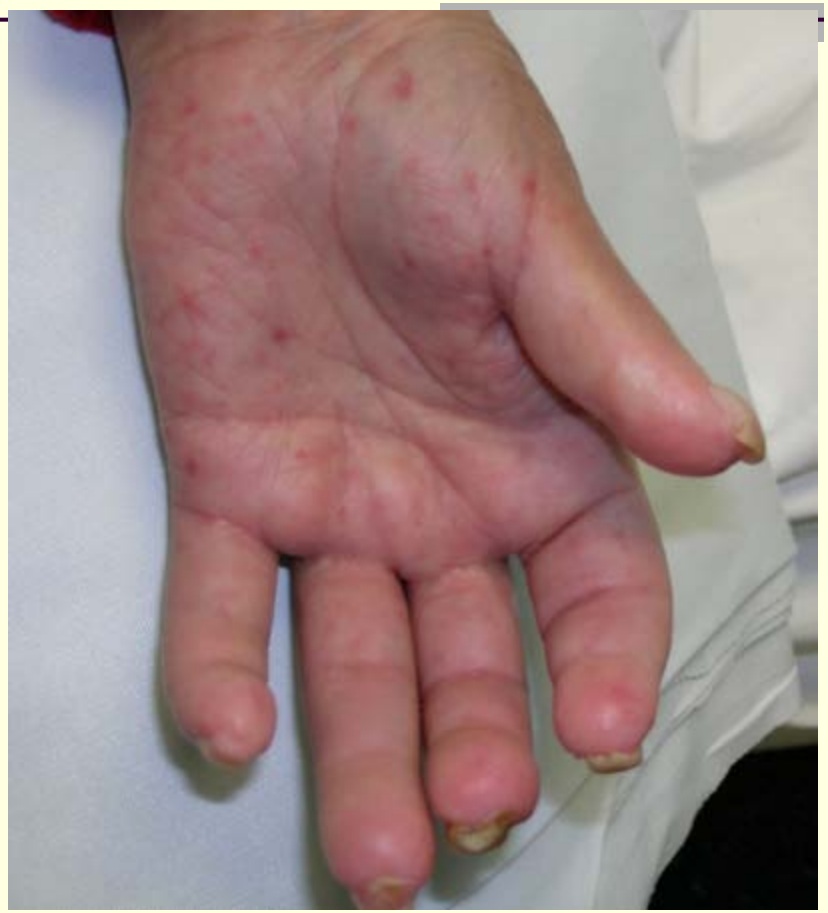


Telangiectasia (Spider Veins)

- 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

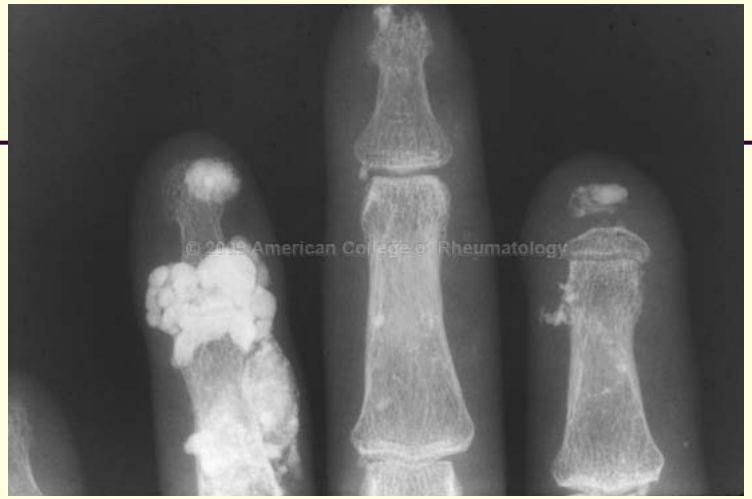






Calcinosis

- Besides digital calcinosis patients may develop calcium deposits along the forearm, elbow, buttocks, thigh, knee or shin.
- Cannot to prevent or dissolve deposits with probenecid and warfarin.
- 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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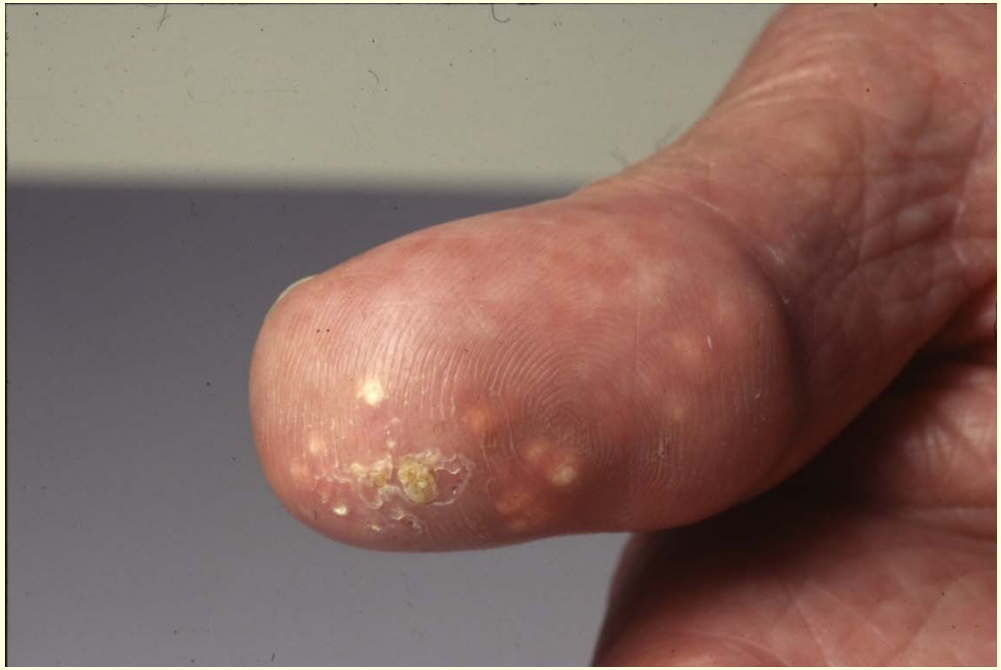


What you can do

- Over the counter antihistamines (Benadryl) may work but cause drowsiness.
- Low-dose oral glucocorticoids sometimes work for severe itching (need a prescription)
- Completely avoid smoking
- Tight control of diabetes and hypertension

What You Can Do

- Patient should avoid cold and wear hats and mittens to prevent distal extremities from entering into vasospasms
- Emollients may be beneficial to maintain a certain moisture level in the skin in an effort to reduce the likelihood of skin drying and breakdown.

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

What Your Doctor Can Do

- Goal of therapy is not to completely return to new skin but to improve redness, softening lesions and local hair regrowth.
- As disease progresses the goal becomes preventing new lesions and limiting disease spreading
- In patients with generalized scleroderma topical therapies are often ineffective. Phototherapy and systemic treatment
- For those with deep tissue involvement topical therapies are not effective.
- Quickly progressing disease should be addressed with systemic treatment

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

Phototherapy

Topical Tacrolimus 0.1%

- For active inflammatory morphea
- It is immunosuppressive

Topical and intra lesion corticosteroids injections

- Topical steroids are usually applied twice or once daily.
- If there is no response in 10-12 weeks then treatment should be discontinued.
- Injections of steroid in conjunction with phototherapy or systemic treatment has had varied results.
- Injection of Triamcinolone acetonide 5 to 10 mg/mL.
- Patients who respond should receive repeat injections every 4 to 6 weeks until sufficient improvement occurs

Topical Vitamin D (Calcipotriol)

- Inhibits fibroblast proliferation, collagen synthesis and T lymphocytes activation.
- In patients where topical steroids alone have failed, twice daily applications of topical calcipotriene 0.005% ointment under a dressing clinically improved some patients in the study
- In some cases a combination of calipotrol and betamethasone dipropionate impro
lesions

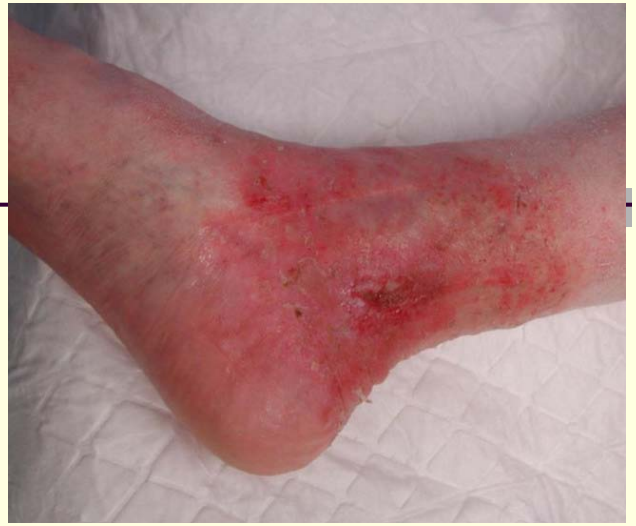
Imiquimod

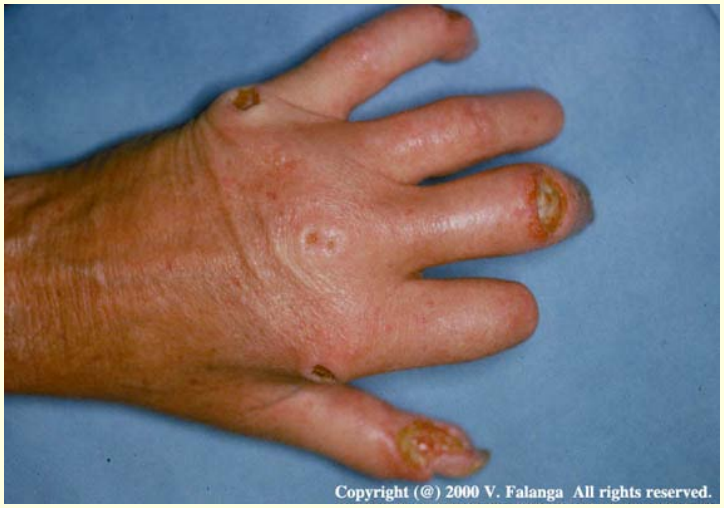
- Topical immunomodulator that induces interferon gamma (a cytokine that can inhibit TGF-beta and the production of extracellular matrix proteins) i.e. it slows the process of skin thickening
- Application 3 days per week was associated with improvements of dyspigmentation, induration and erythema

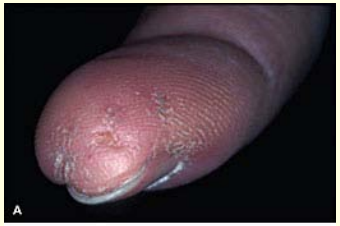
Systemic Therapy (Medications)

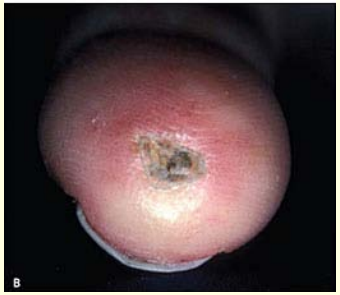
- Methotrexate and glucocorticoids (mostly prednisone or methylprednisone) in combination
- Methotrexate orally (initial dose 15 mg/week) and pulse IV methylprednisone (1000mg for three days once monthly) for at least six months was associated with softening of sclerotic skin and reduced inflammation
- Or Methotrexate orally or subcutaneously (initial dose 15 mg/week) and prednisolone (1 mg/kg per day for 2 to 4 weeks then tapered as tolerated) significantly improves deep tissue scleroderma on MRI
- Once medications stopped there is a high relapse rate therefore therapy is suppressive rather than curative

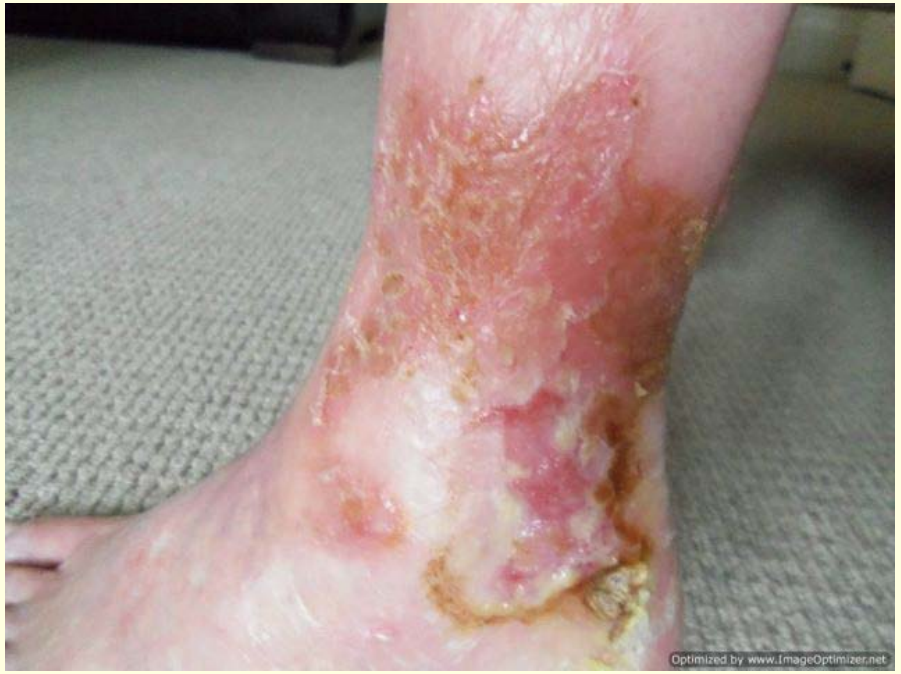
Wounds/Ulcers









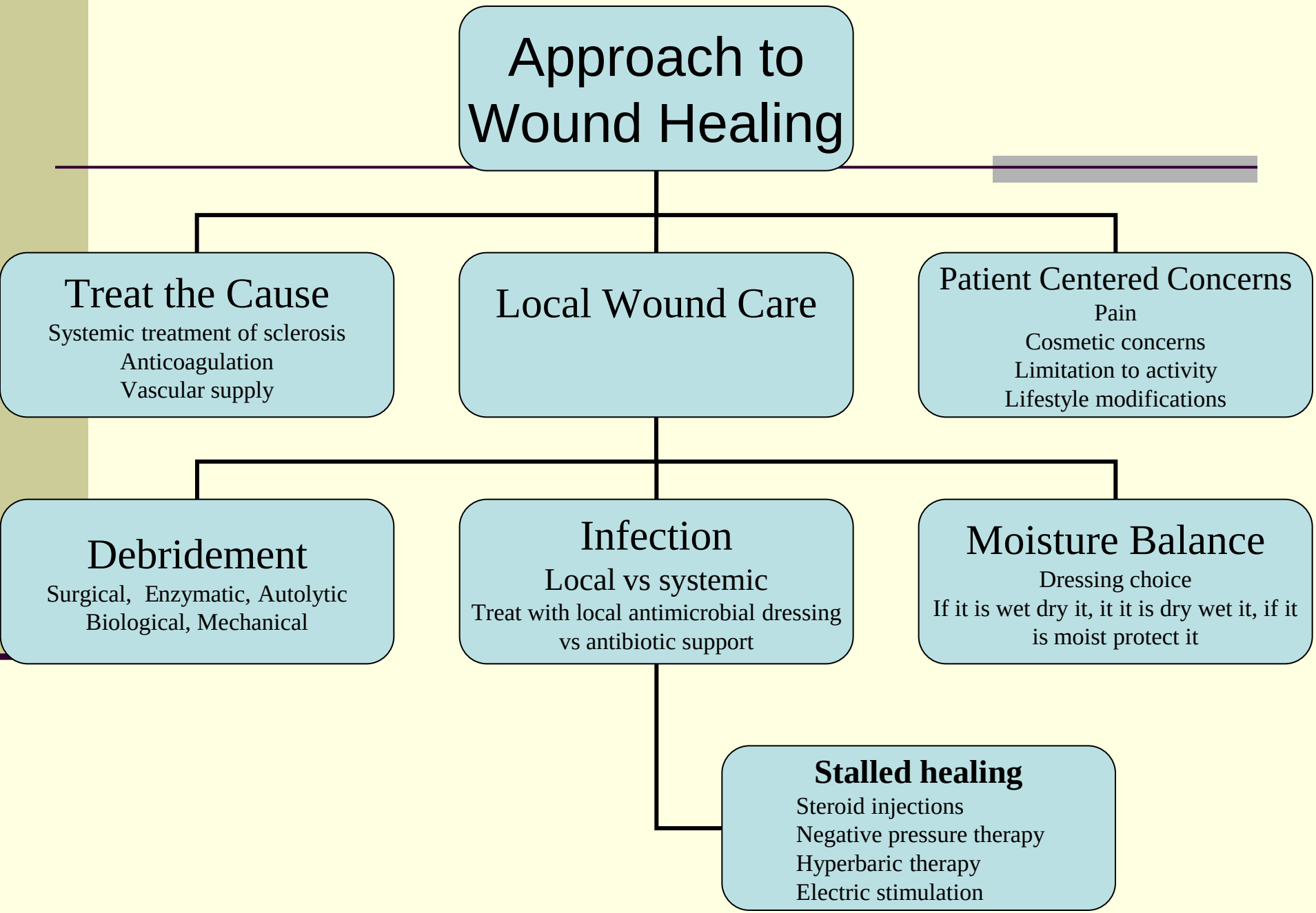


Leg/Foot and Finger Ulcers

- Cause is from damage to the structure and functioning of the blood vessels due to systemic scleroderma and possible preexisting conditions such as hypertension, peripheral vascular disease or diabetes
- Even once circulation is optimized fibroblasts over production of collagen and inflammation response makes it difficult to heal a wound.

Approach

- Venous and arterial testing (blood flow tests)
  - Doppler of affected limb
- Blood work
  - Anti phospholipids
- Optimizing vascular by referring to vascular surgery service
- Evaluate for infection in wounds
- Aggressive local wound therapy
  - Wound preparation paradigm
- Systemic therapy
  - Low-dose, low-molecular-weight heparin (Enoxaparin 40 mg sub cut daily)
- Pain and quality of life


Approach to Wound Healing
Treat the Cause
Systemic treatment of sclerosis
Anticoagulation
Vascular supply
Local Wound Care
Patient Centered Concerns
Pain
Cosmetic concerns
Limitation to activity
Lifestyle modifications
Debridement
Surgical, Enzymatic, Autolytic
Biological, Mechanical
Infection
Local vs systemic
Treat with local antimicrobial dressing vs antibiotic support
Moisture Balance
Dressing choice
If it is wet dry it, it it is dry wet it, if it is moist protect it
Stalled healing
Steroid injections
Negative pressure therapy
Hyperbaric therapy
Electric stimulation

Patient centered, multi disciplinary approach will best serve the patient

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