# Cutaneous manifestations of SLE and other CTD ( dermatomyositis , scleroderma)

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# Objectives

- At the conclusion of these lectures the student will be able to:
  - Differentiate between the various types of Lupus
  - Recognize how Lupus affects the various systems of the body
  - Identify all of the current treatment options available for Lupus

# Objectives

- To learn how to diagnose and investigate dermatomyositis.
- How to manage dermatomyositis.
- To learn the presentation of morphea and systemic sclerosis and ways to manage them.
- This lecture is not meant to be inclusive of all the information about these diseases but to highlight important aspects in their diagnosis and management.

# LUPUS ERYTHEMATOSUS

# LUPUS ERYTHEMATOSUS

- It's a designation of a spectrum of diseases that are linked by distinct clinical findings and distinct patterns of **polycolonal B cell immunity*.**
- It ranges from life threatening manifestations of SLE to the limited and exclusive skin involvement in CCLE.
- More than 85% of patients with LE have skin lesions, which can be classified into **LE-specific & non-specific***.

# Revised ACR's Criteria for Classification of SLE

**Any 4 of the following criteria are required to make the diagnosis:**

- Malar rash
- Discoid rash
- Photosensitivity
- Oral ulcer
- Arthritis
- Serositis
- Renal dis.
- Neurological dis.
- Hematological inv.
- Immunological dis.
- ANA

# Classification of Cutaneous Disease in Lupus Erythematosus

## Lupus Specific eruptions

**I . Acute cutaneous LE (ACLE)**
**Localized, generalized, bullae**

**II. Subacute cutaneous LE ( SCLE)**
- **A. Annular**
- **B. Papulosqamous**
- **C. Syndromes commonly exhibiting similar morphology**
  1. **Neonatal LE (NLE)**
  2. **Complement deficiency syndromes**
  3. **Drug induced**

**III. Chronic Cutaneous LE**
- **A. Discoid LE (DLE)**
  1. **Localized**
  2. **Disseminated**
- **B. Verrucous ( hypertrophic) DLE**
- **C. Lupus erythematosus – lichen planus overlap**
- **D. Chilblain LE**
- **E. Tumid Lupus**
- **F. Lupus panniculitis**

## Lupus- nonspecific eruptions

**Nonscarring alopecia**
**Telangiectasia**
**Livedo Reticularis**
**Palpable Purpura**
**Periungual erythema**

# Acute Cutaneous Lupus Erythematosus

## What is Acute Cutaneous Lupus Erythematosus?

# Acute Cutaneous Lupus Erythematosus

- Acute malar “ butterfly rash” or more generalized photodistributed eruption.
- Nearly ALL patients presenting with ACLE will have systemic lupus erythematosus ( SLE ), often in an acute flare.*
- Patients with ACLE will nearly always have a +ve ANA
- ACLE is transient, improves with improvement of the SLE
- Non scarring

# Acute Cutaneous Lupus Erythematosus

**What is the initial workup of ACLE?**

# Acute Cutaneous Lupus Erythematosus

1. History & Physical examination
2. Skin Biopsy
3. Lupus Band Test
4. Serology
5. CBC, ESR
6. Urine analysis
7. C3,C4: : low levels indicate active disease, often with renal involvement.

# Acute Cutaneous Lupus Erythematosus

**Serology:**

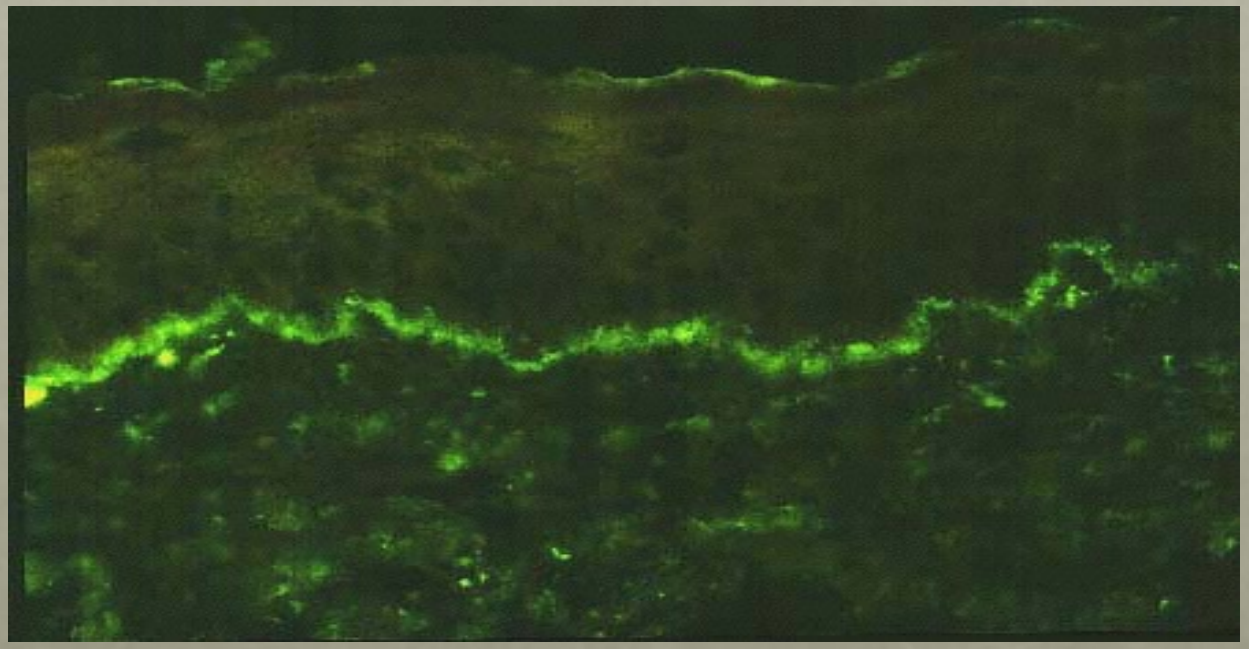
1. ANA :+ve in 95% ,VERY SENSITIVE BUT NOT SPECIFIC
2. Anti-dsDNA (anti- native DNA): Specific but not very sensitive, indicates high risk for renal disease.
3. Anti-smith : most specific +ve in 30%
4. Anti-histone Ab ( drug induced lupus)
5. Rheumatoid factor* : +ve in 30%

# Lupus Band Test

It's preferred to be done on nonlesional nonexposed skin, but can be performed on lesional skin*.

Granular deposits of immunoglobulins and complement are detected in a band-like pattern at the dermal-epidermal junction.

# Lupus Band Test



# Acute Cutaneous Lupus Erythematosus

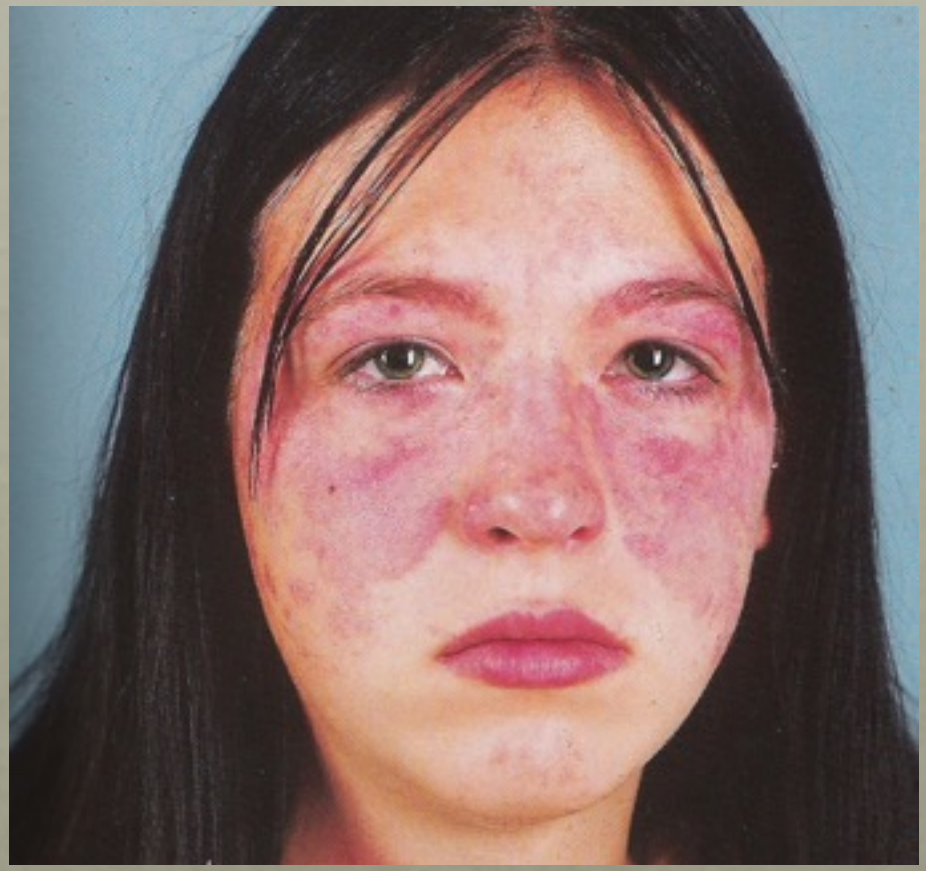
**How is ACLE managed ?**

# Acute Cutaneous Lupus Erythematosus

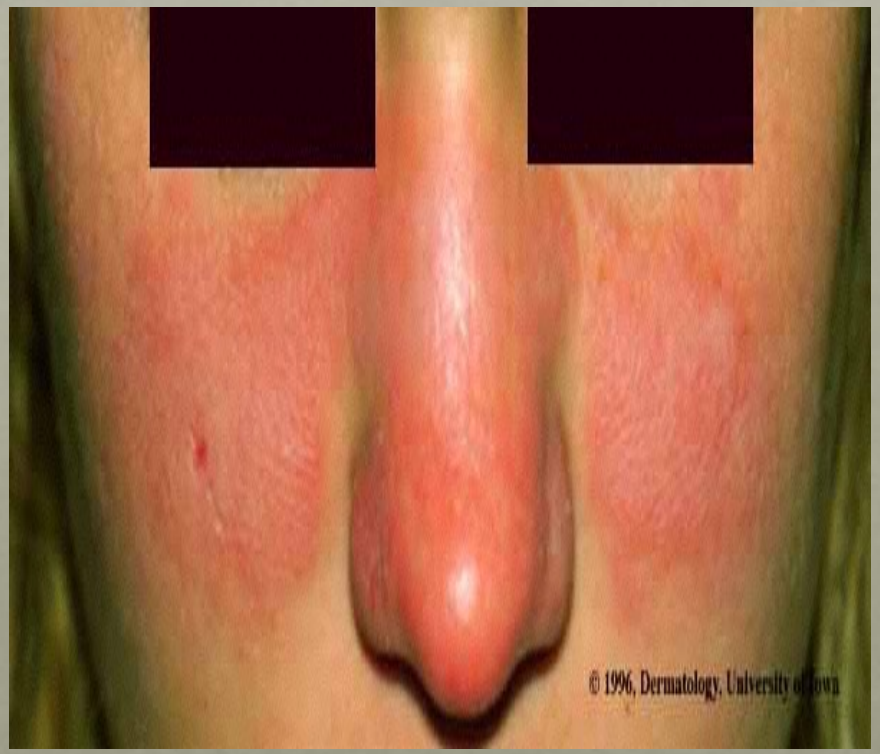
- The aim of treatment for cutaneous LE is to alleviate symptoms and to prevent scarring.

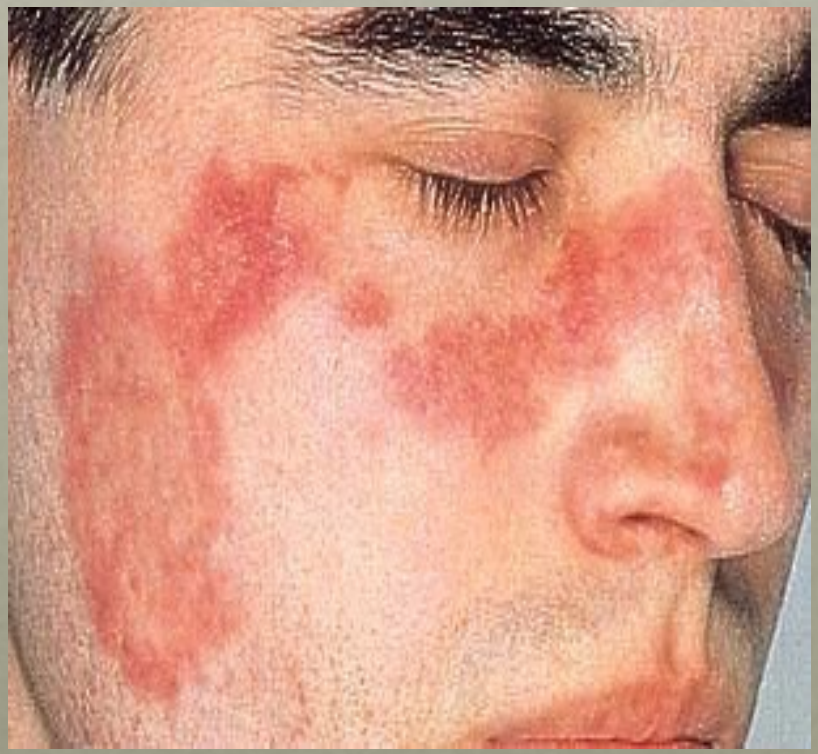
1. Sun protective measures
2. Potent topical steroids
3. Antimalarial drugs.
4. Oral steroids.
5. Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.

# Acute Cutaneous Lupus Erythematosus



# Acute Cutaneous Lupus Erythematosus



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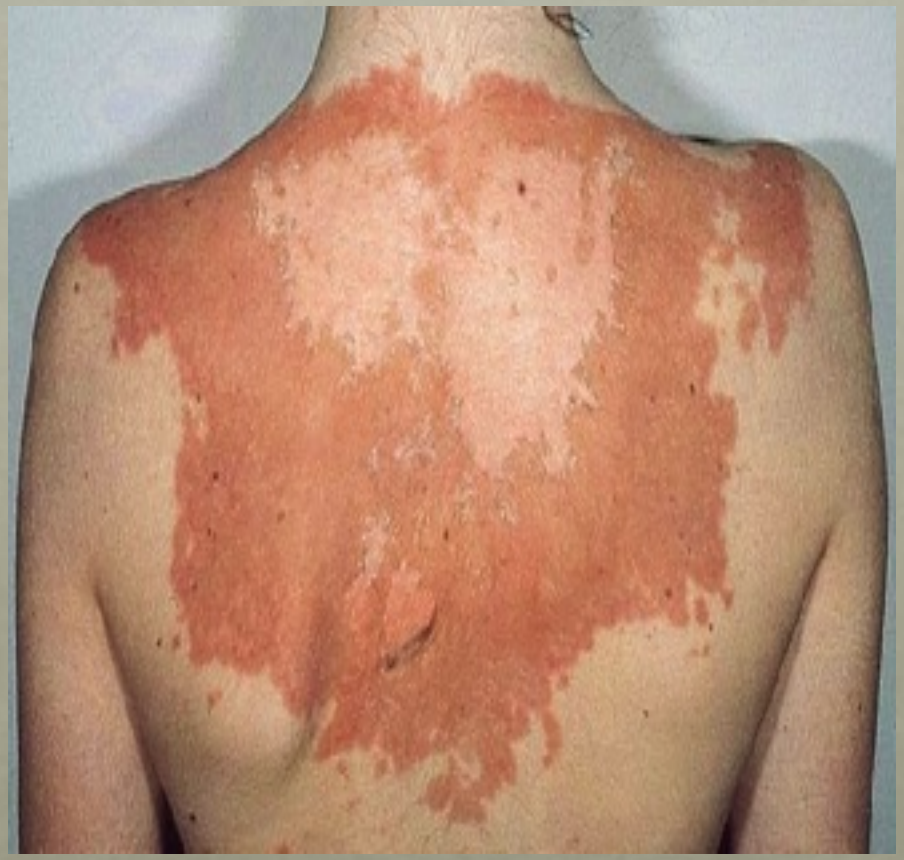
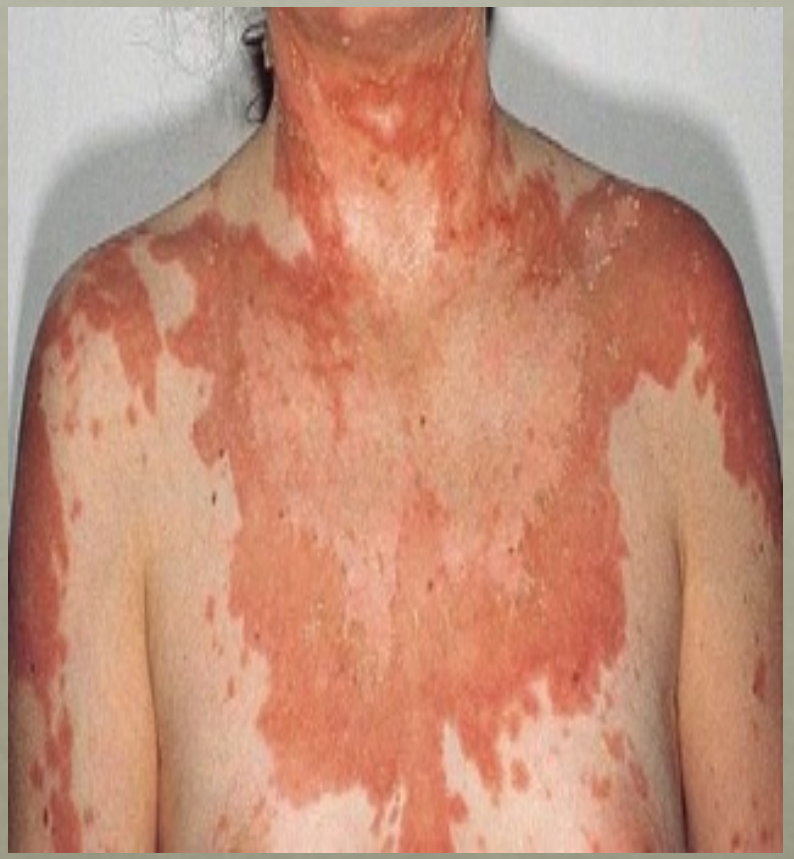




# Subacute Cutaneous Lupus Erythematosus

**What is subacute cutaneous lupus erythematosus?**

# Subacute Cutaneous Lupus Erythematosus

- More persistent than those of ACLE ( weeks- months)
- Scaly, superficial, inflammatory macules, patches, papules and plaques that are photodistributed, particularly on the upper chest & back, lateral neck, and dorsal arms & forearms.
- Morphologic subtypes:
  - Annular/polycyclic lesions " ring-shaped"
  - Papulosquamous lesions:
    - psoriasiform
    - pityriasiform

# Subacute Cutaneous Lupus Erythematosus

**Do patients with SCLE have SLE?**

# Subacute Cutaneous Lupus Erythematosus

- About 1/2 patients with SCLE will have 4 or more criteria for the classification of SLE
- Skin disease, photosensitivity, musculoskeletal complaints*
- SCLE could occur in patients with Sjögren syndrome, deficiency of the second component of complement (C2d), or it may be drug induced

# Subacute Cutaneous Lupus Erythematosus

**How do you make the diagnosis of SCLE?**

# Subacute Cutaneous Lupus Erythematosus

- SCLE  is a clinical diagnosis based on presence of:
  - typical photodistributed eruption
  - skin biopsy
- Direct Immunofluorescence*
- A strong association exists with anti-Ro/SS- A autoantibodies*, and a lesser extent will have anti-La/SS-B

# Subacute Cutaneous Lupus Erythematosus

**What is the initial workup of SCLE?**

# Subacute Cutaneous Lupus Erythematosus

1. History & Physical examination
2. Laboratory testing
3. Medication History

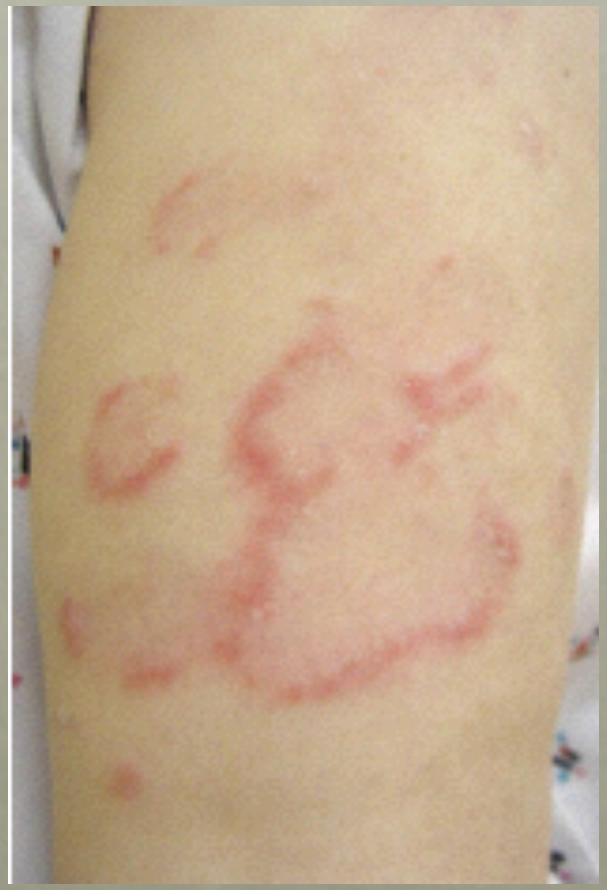
# Subacute Cutaneous Lupus Erythematosus

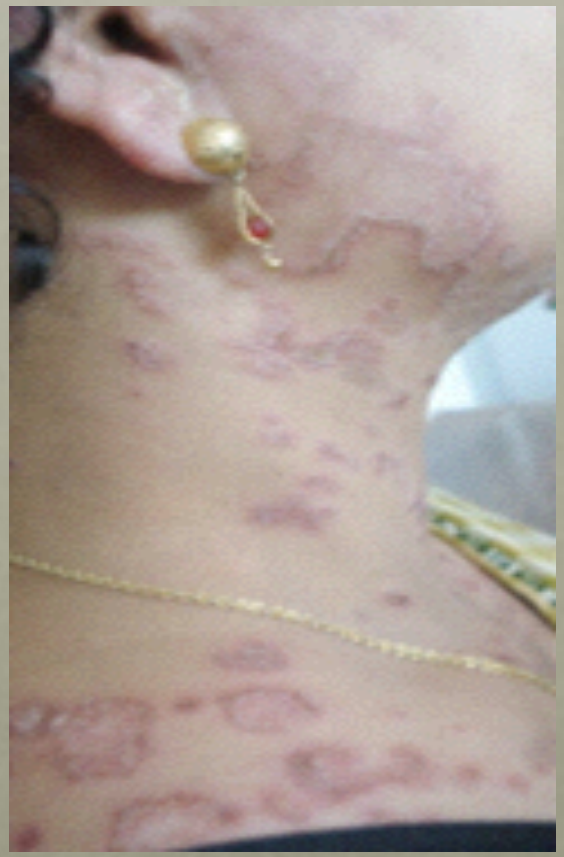
**How is SCLE managed ?**

# Subacute Cutaneous Lupus Erythematosus

- Broad-spectrum sunscreens
- Sun-protective measures
- Topical steroids
- Antimalarial drugs

# Subacute Cutaneous Lupus Erythematosus





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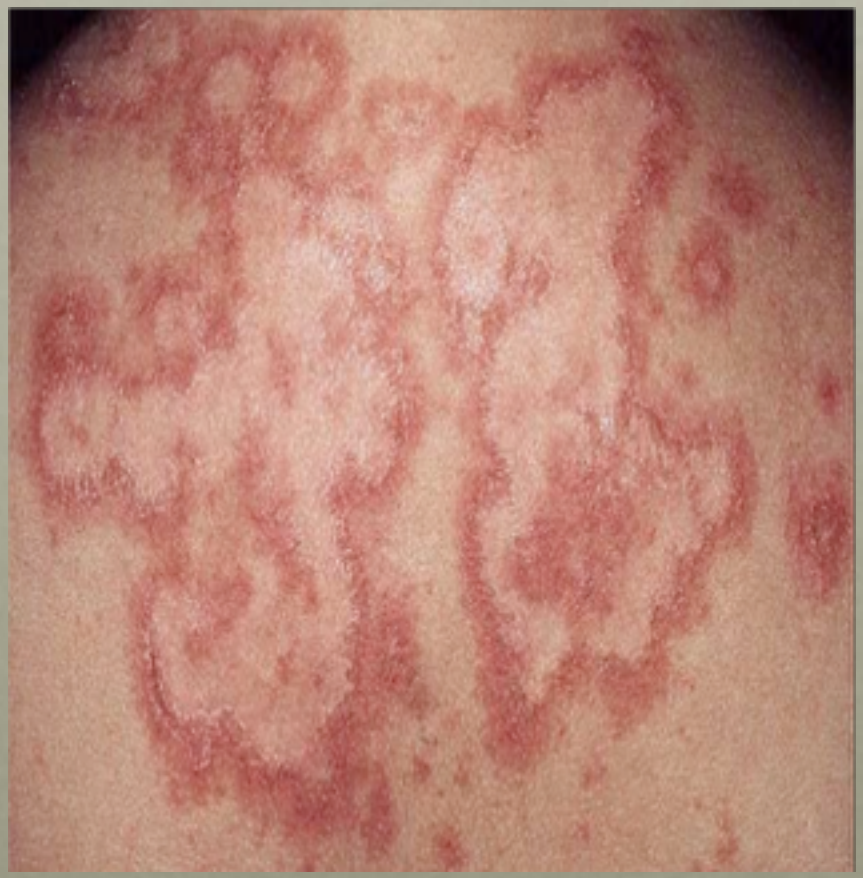
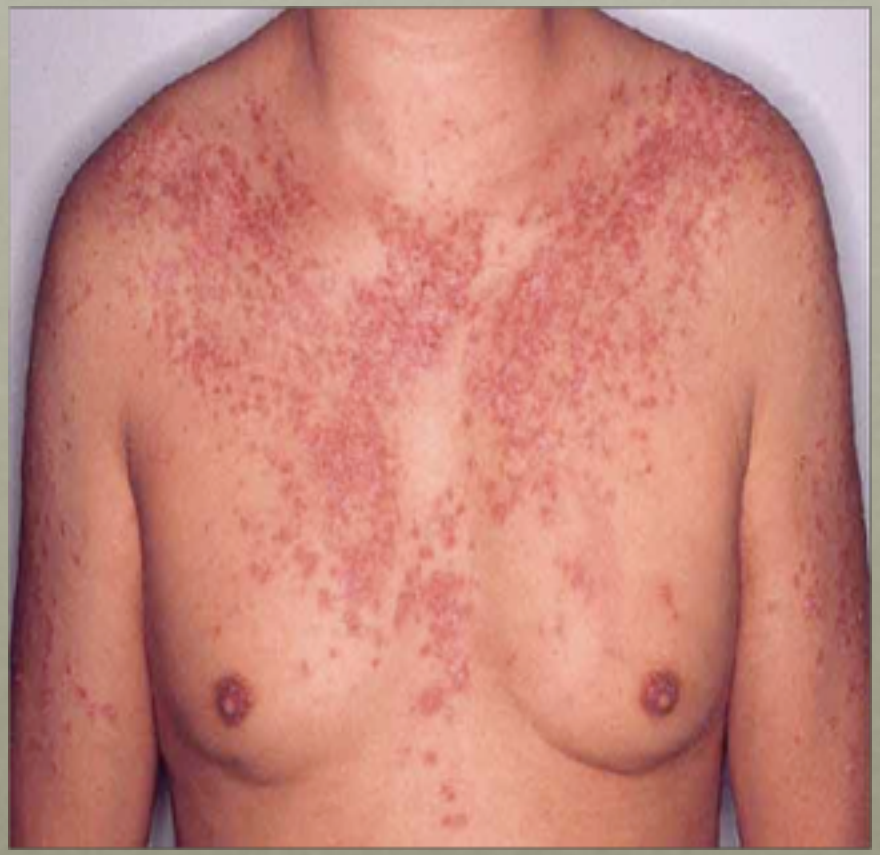
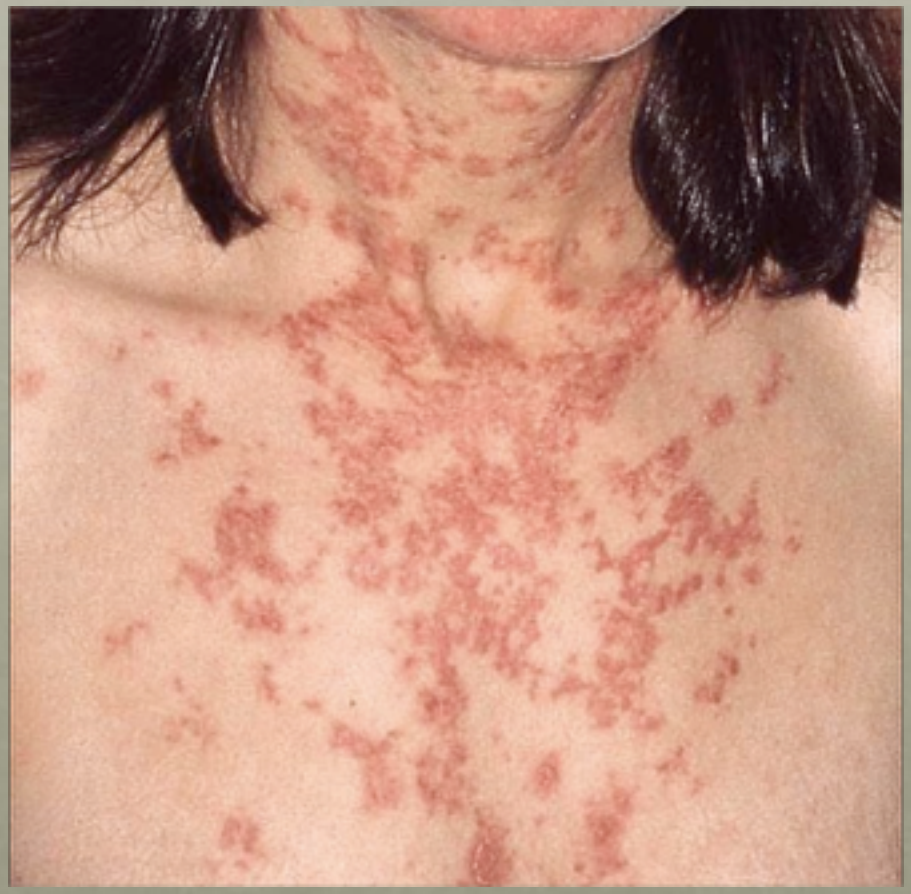




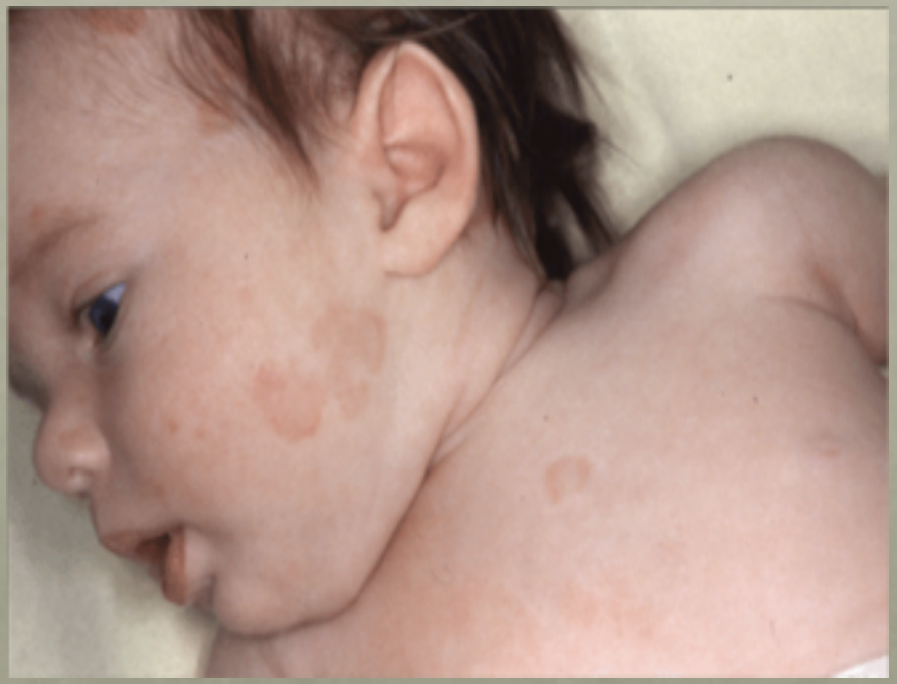
# Subacute Cutaneous Lupus Erythematosus

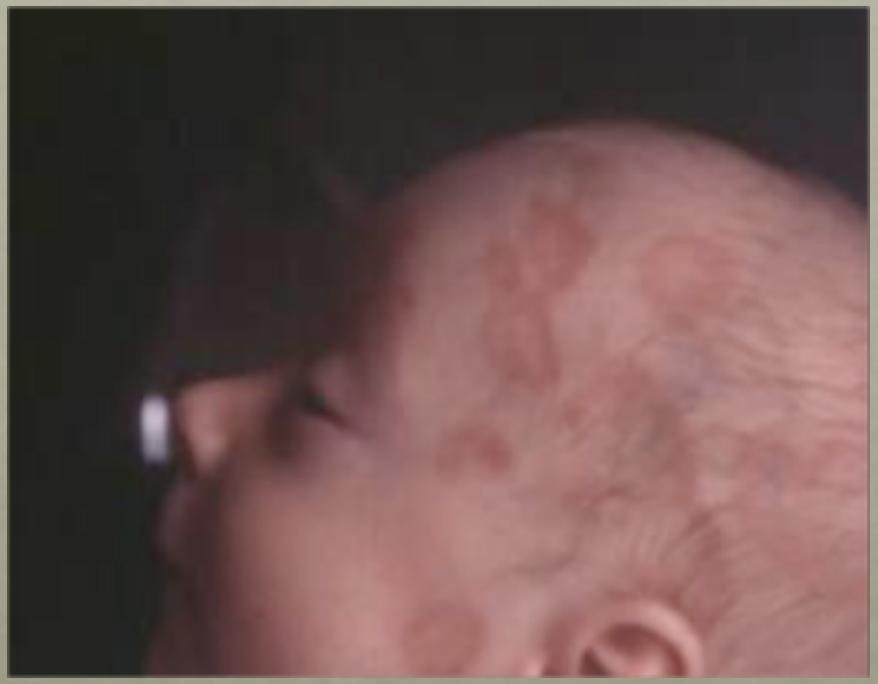


# Neonatal Lupus Erythematosus

- The skin lesions occur on the face and head, morphologically resemble SCLE lesions, they are transient, resolving within a few months*.

# Neonatal Lupus Erythematosus





# Neonatal Lupus Erythematosus

- In NLE , infants develop skin disease (50%), heart disease (50%), or both (10%).
- The heart disease usually manifests as isolated complete heart block.
- It's usually permanent and may require a pacemaker.
- 10% of infants with NLE and heart disease die from cardiac complications.
- Thrombocytopenia/ liver disease*.
- Nearly all have anti-Ro/SS-A and sometimes anti-La/SS-B antibodies, as will their mothers*

# Chronic Cutaneous Lupus Erythematosus

**What is chronic cutaneous lupus erythematous ?**

# Chronic Cutaneous Lupus Erythematosus

- Several types of cutaneous LE that are **very persistent** are termed Chronic cutaneous lupus erythematosus.
- The **most common** of these chronic forms is Discoid lupus erythematosus " DLE"
- Serologic abnormalities are uncommon

# Chronic Cutaneous Lupus Erythematosus

**Describe the skin changes that occur with discoid lupus erythematosus?**

# Chronic Cutaneous Lupus Erythematosus

- **<u>Discoid lupus erythematosus:</u>**
  - Localized or generalized
  - Chronic, fixed, indurated, erythematous papules and plaques often distributed over the head& neck.
  - Scarring*
  - Pigmentary changes ( hypo/hyperpigmentation)
  - Epidermal changes: scales, keratotic plugging of hair follicles, crusting
  - External ears*

# Chronic Cutaneous Lupus Erythematosus

**Do patients with DLE develop systemic lupus erythematosus?**

# Chronic Cutaneous Lupus Erythematosus

- Risk of developing SLE is 5% ( slightly higher risk if DLE is generalized ).

- 25% of SLE patients will develop lesions of DLE at some time during the course of their disease.

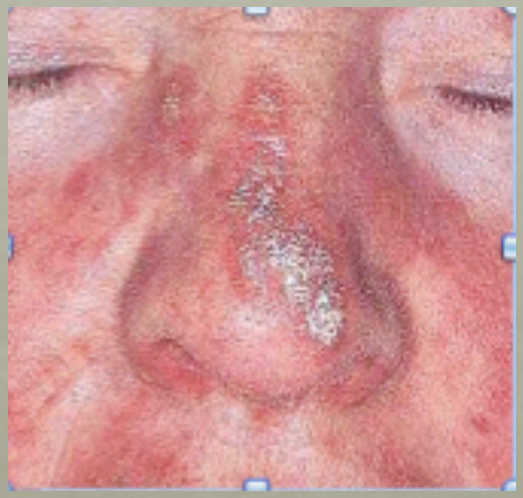
# Chronic Cutaneous Lupus Erythematosus

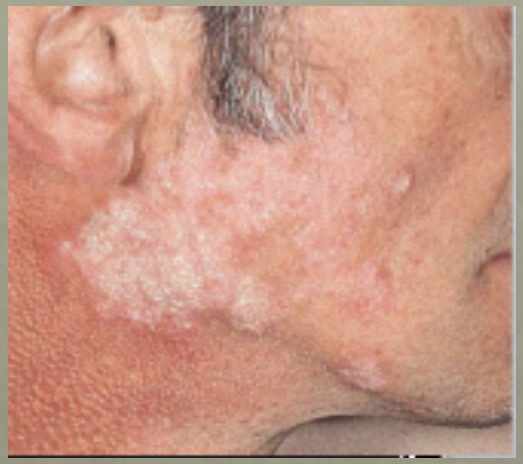
**How is Discoid Lupus treated?**

# Chronic Cutaneous Lupus Erythematosus

- Sunscreens
- Sun-protective measures
- Potent topical steroids/ intralesional steroids
- Antimalarial drugs

# Chronic Cutaneous Lupus Erythematosus





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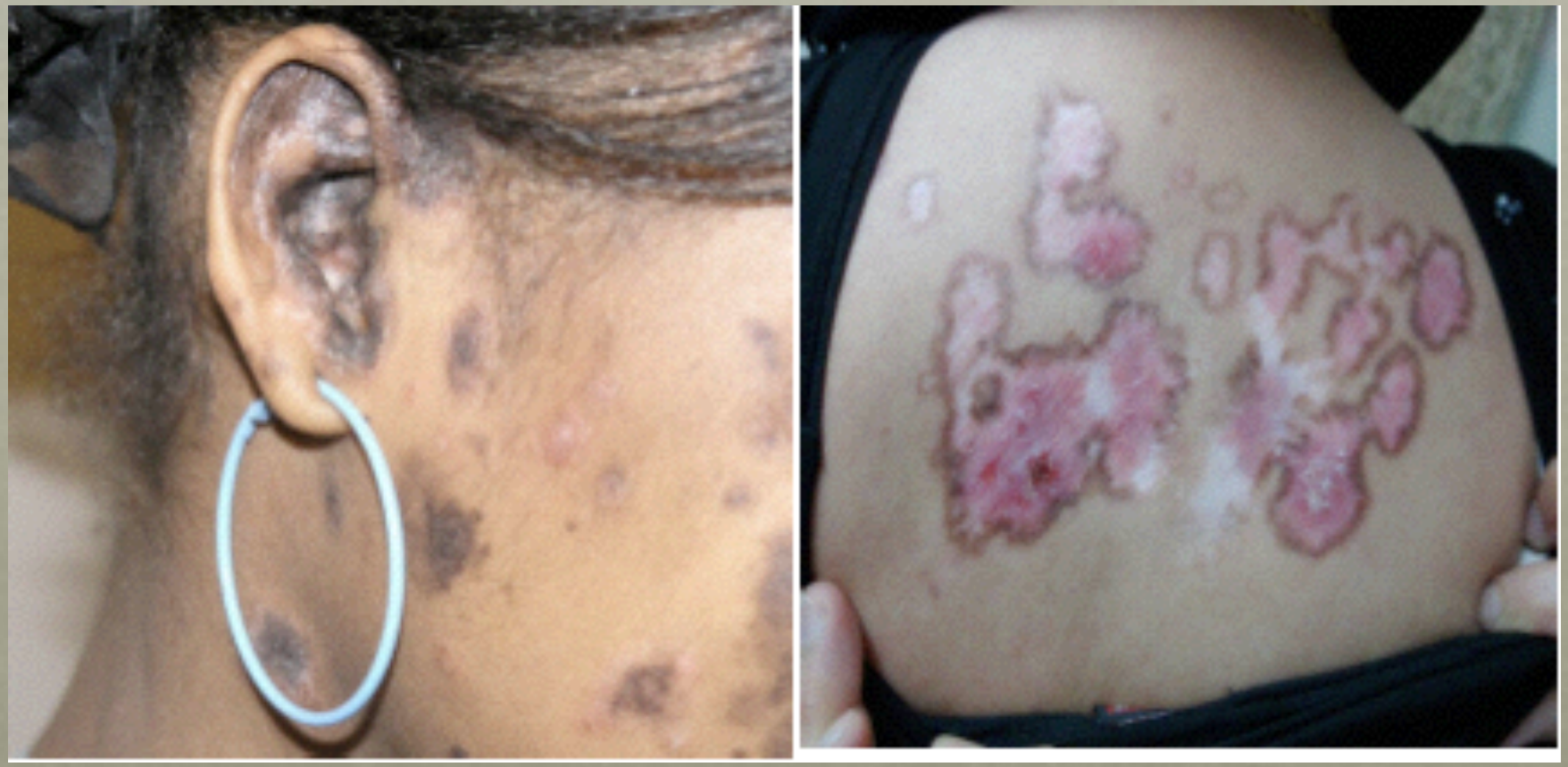

# Chronic Cutaneous Lupus Erythematosus

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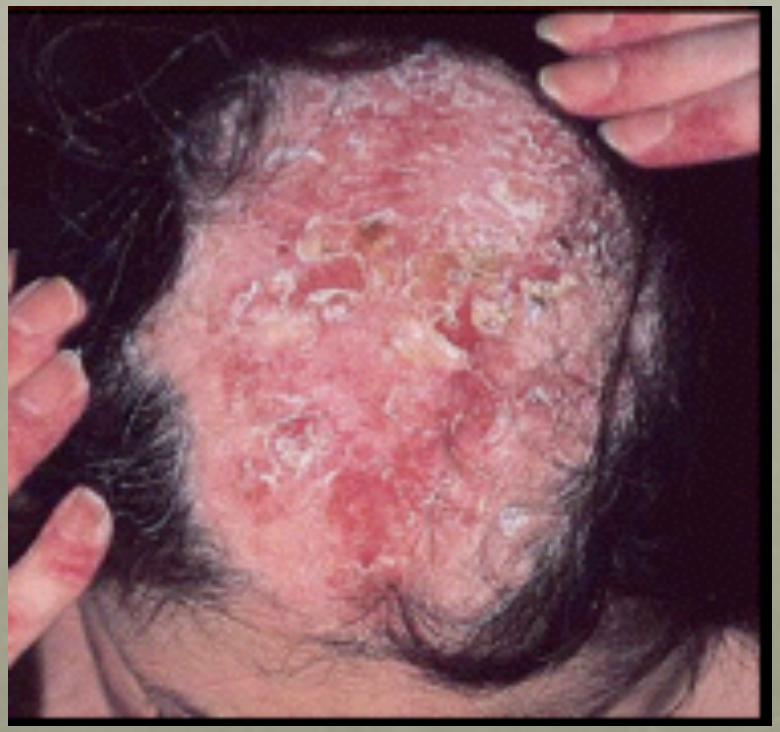

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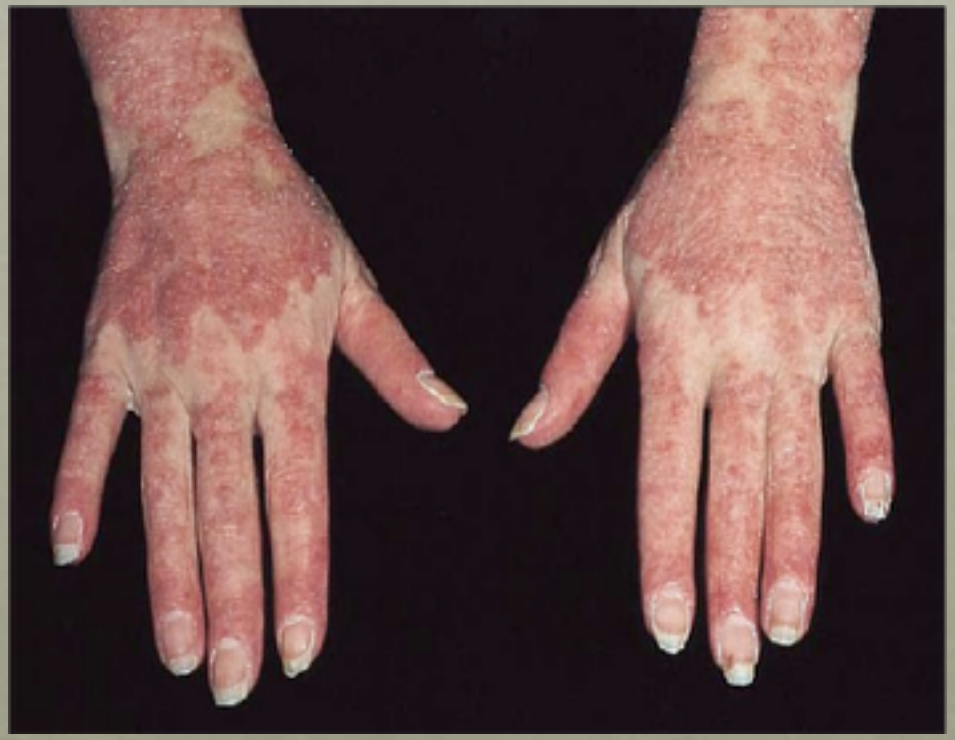
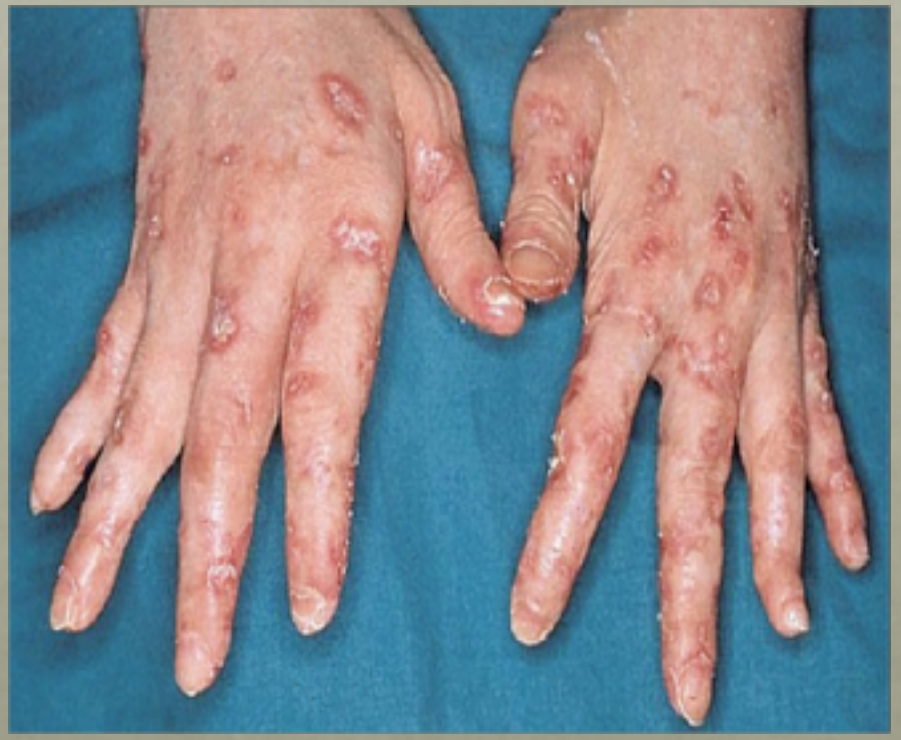




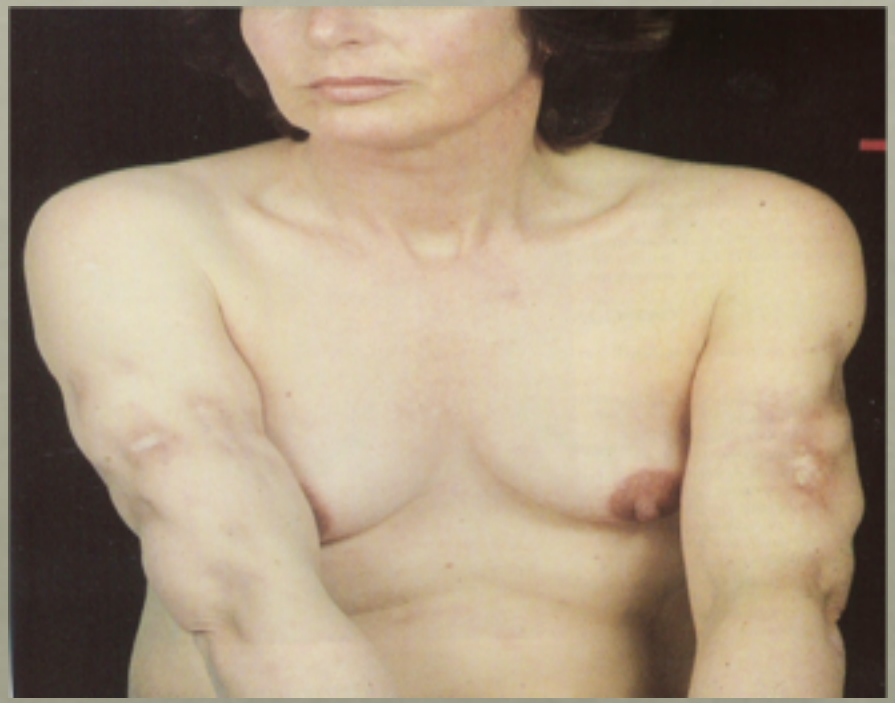
# Lupus Panniculitis

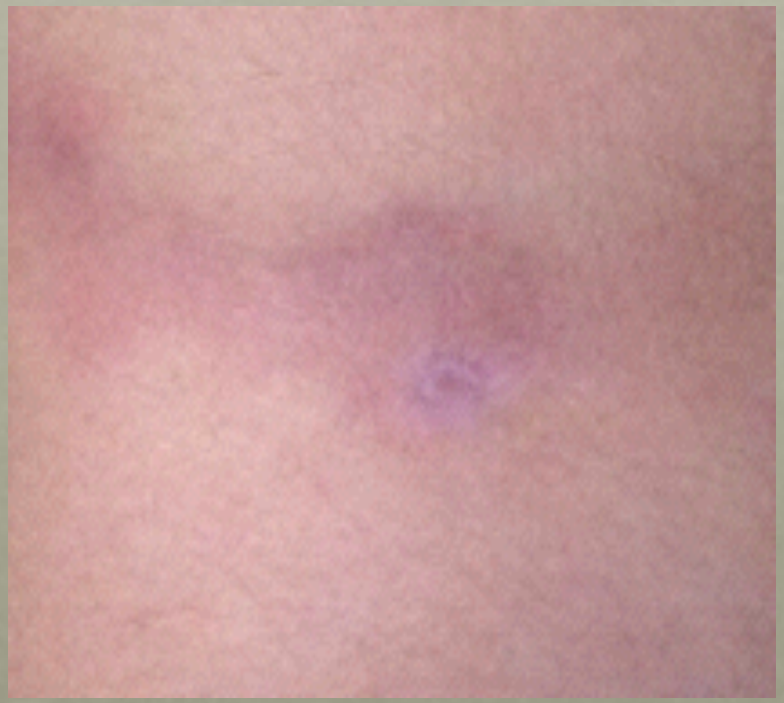
**What is Lupus Panniculitis?**

# Lupus Panniculitis

- Inflammation involving the subcutaneous tissue, resulting inn inflamed nodules that often resolve with depressed scars.
- They could have overlying DLE lesions " Lupus Profundus"
- About 1/2 of patients will have four or more criteria for urge classification of SLE.
- Diagnosis confirmed by excisional biopsy
- Treatment of choice: Antimalarial drugs.

# Lupus Panniculitis





# Drug-Induced Lupus Erythematosus

Drug-induced lupus differs from SLE by the following features:

- Sex ratios are nearly equal.
- Nephritis and central nervous system features are not commonly present.
- No antibodies to native DNA or hypocomplementemia are present.
- When the drug is discontinued, the patient has resolution of clinical manifestations and reverting of abnormal laboratory values to normal.

# Drug-Induced Lupus Erythematosus

- Drugs associated with lupus erythematosus:

| | |
|---|---|
| Chlorpromazine | Isonizide |
| Hydralazine | procinamide |
| Methyldopa | Quinidine |
| Anti- TNF | Minocyclin |
| Penicillamin | IFN-alpha, IFN-beta |

# DERMATOMYOSITIS

# Dermatomyositis

- An idiopathic chronic inflammatory disease involving the skin and skeletal muscles.
- Muscle involvement usually presents with proximal muscle weakness.
- **Amyopathic dermatomyositis-** in some instances, muscle involvement may not be detectable.

# Dermatomyositis

Criteria for diagnosing dermatomyositis:

- Progressive proximal symmetrical weakness
- Elevated muscle enzyme levels
- Abnormal findings on electromyograms
- Abnormal findings from muscle biopsy.
- **Compatible cutaneous disease.**

Diagnosis :

Proximal muscle weakness with two of the three laboratory criteria

# Dermatomyositis

**Are there skin changes that are diagnostic of dermatomyositis?**

# Dermatomyositis

Two cutaneous findings have been describes as "pathognomonic" of dermatomyositis:

1. Gottron's papules

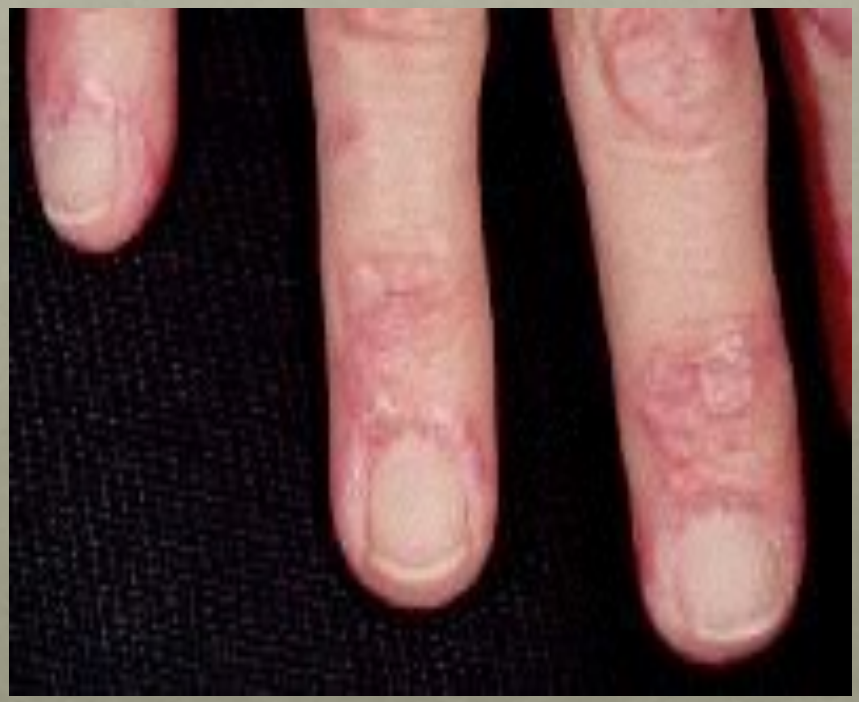
2. Gottron's sign

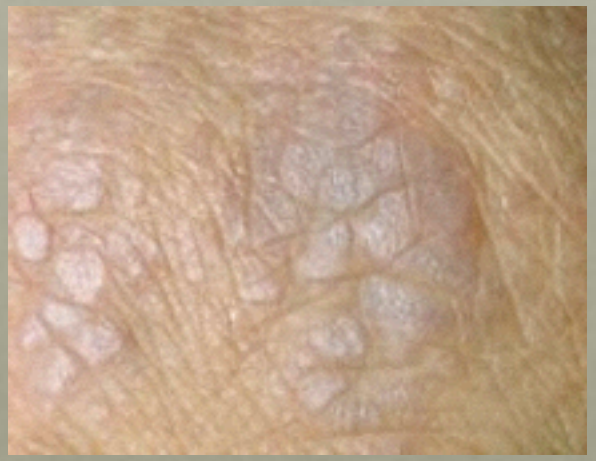
# Dermatomyositis

**What is the difference between Gottron's papules and Gottron's sign?**

# Dermatomyositis

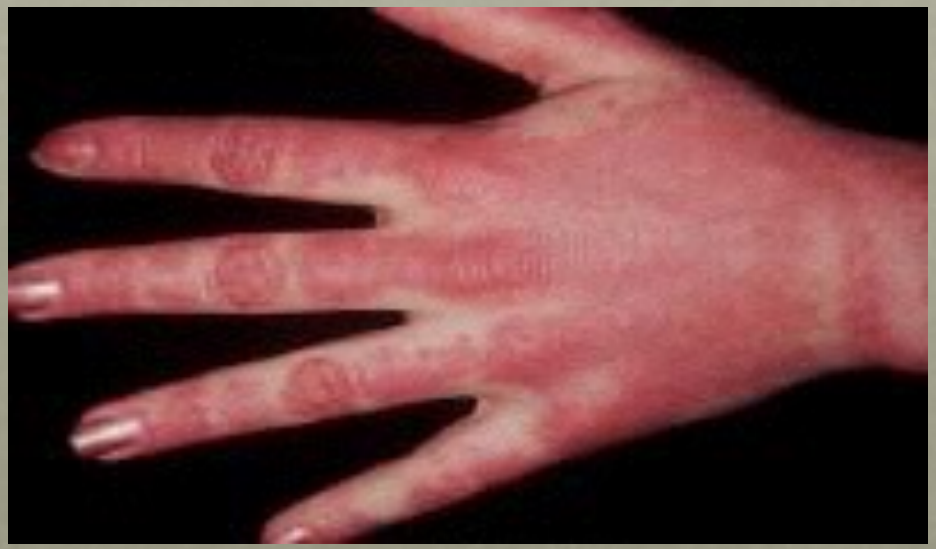
**Gottron's papules**- are erythematous to purplish flat papules on the extensor surfaces of the interphalangeal joints





# Dermatomyositis

- **Gottron's sign**- consist of symmetric violaceous erythema, sometimes with edema, over the dorsal knuckles of the hands, elbows, knees, and medial ankles.

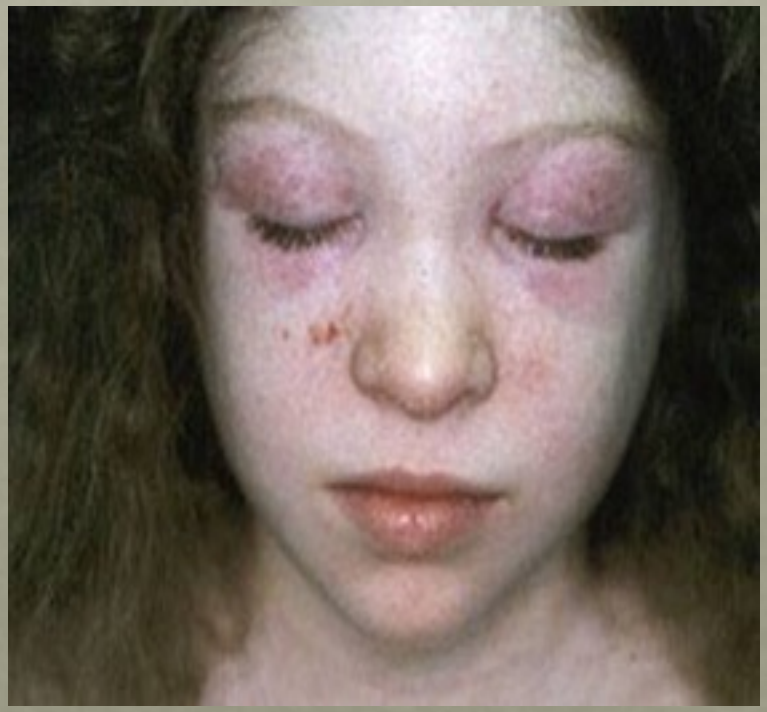


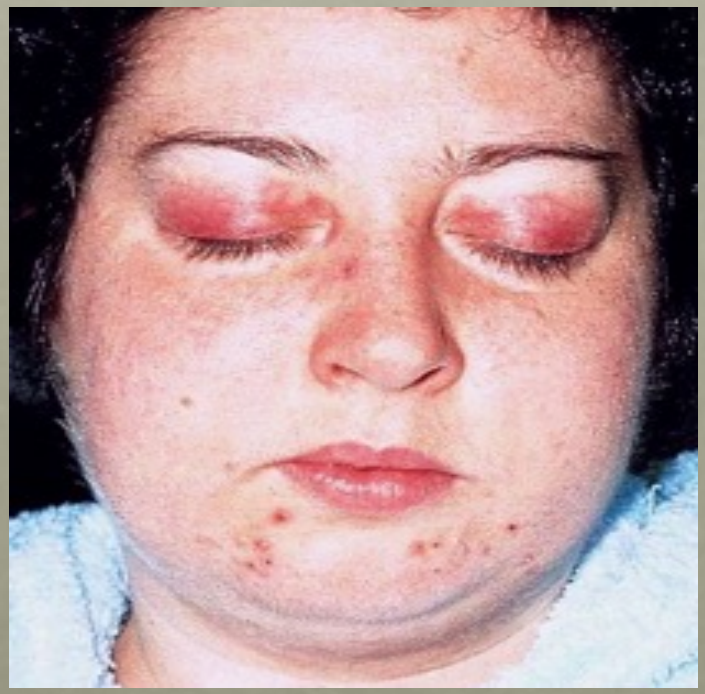
# Dermatomyositis

**Are there other skin findings that are characteristic of dermatomyositis?**

# Dermatomyositis

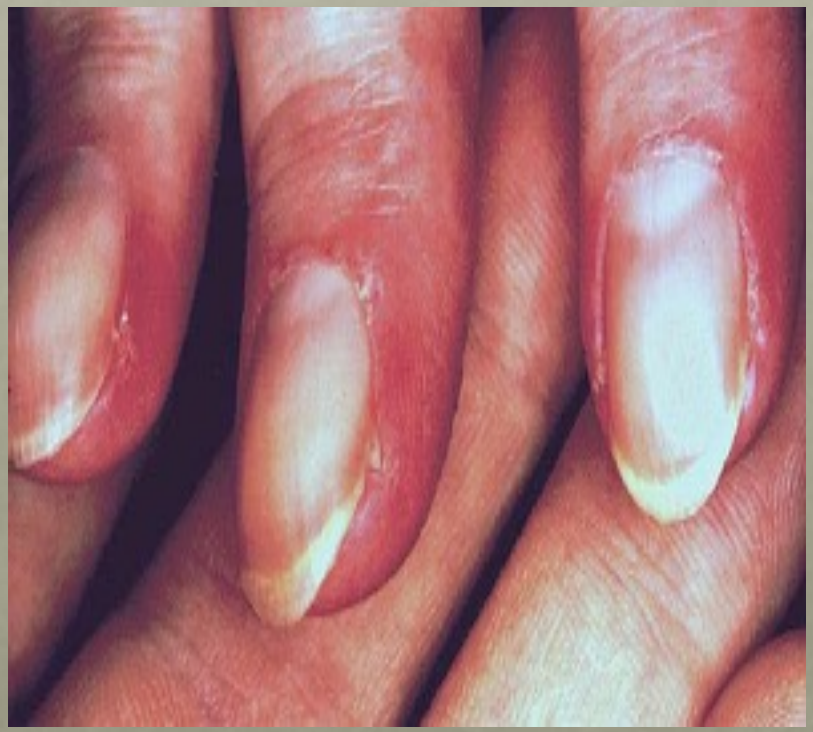
- **Heliotrope rash**- symmetrical periorbital edema with a violaceous (lilac) dusky erythema

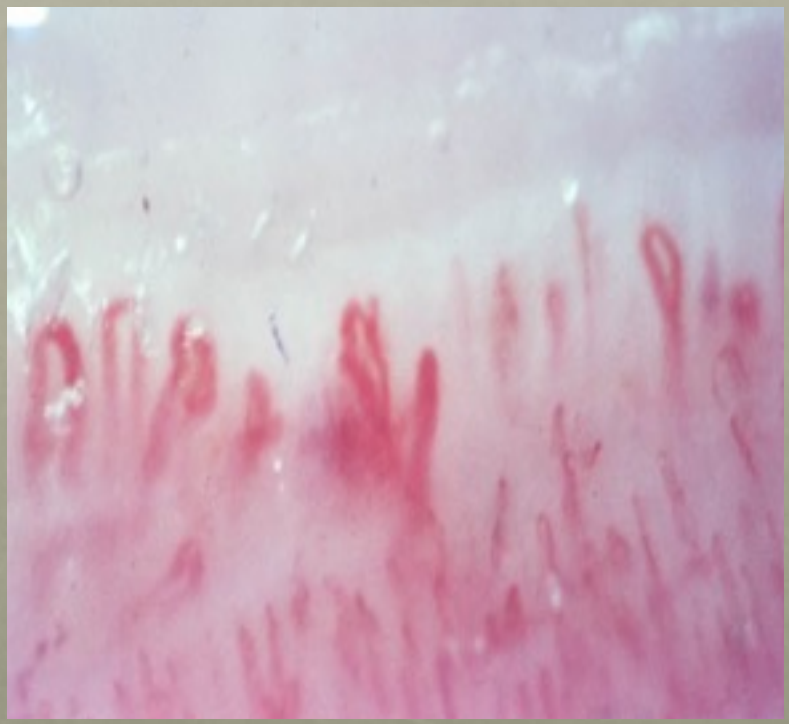




# Dermatomyositis

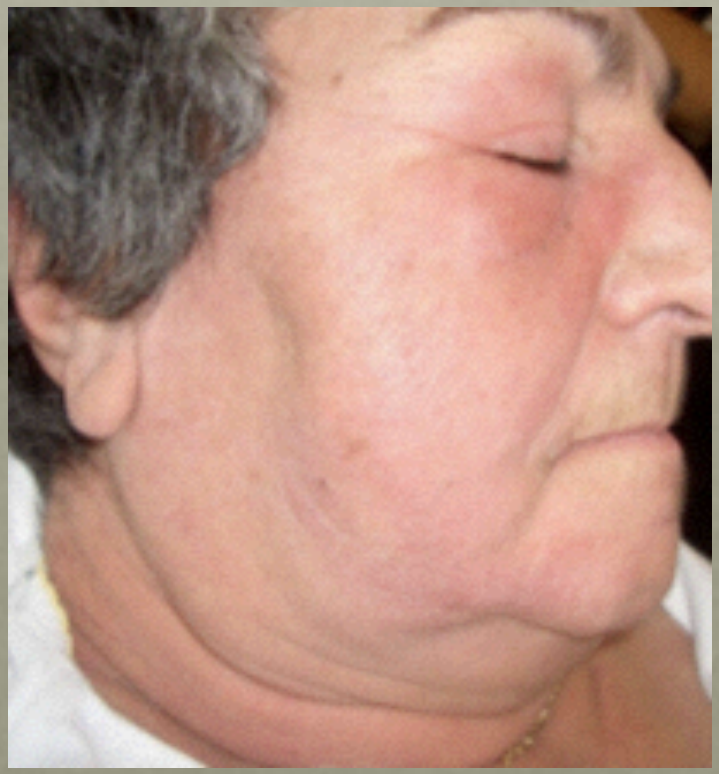
- Periungual talangiectasia with cuticle atrophy

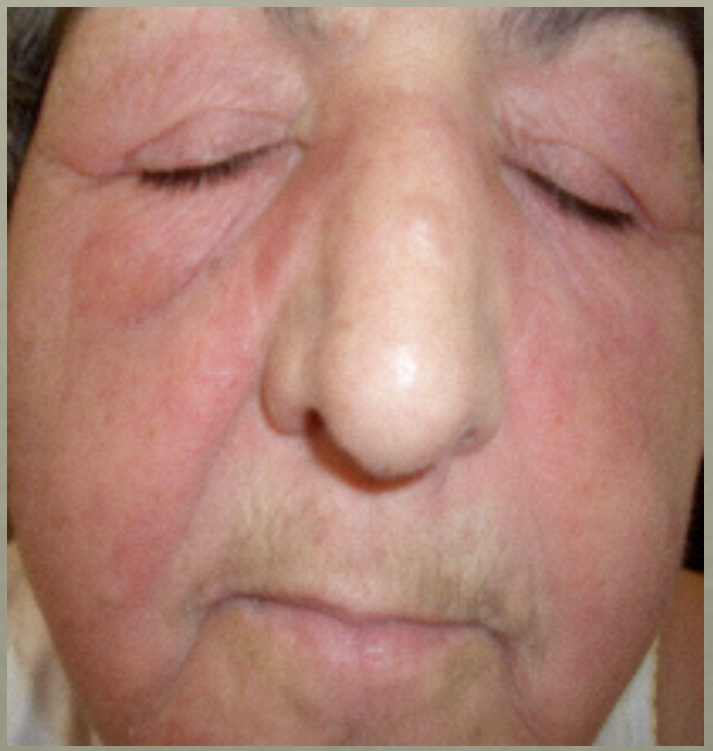




# Dermatomyositis

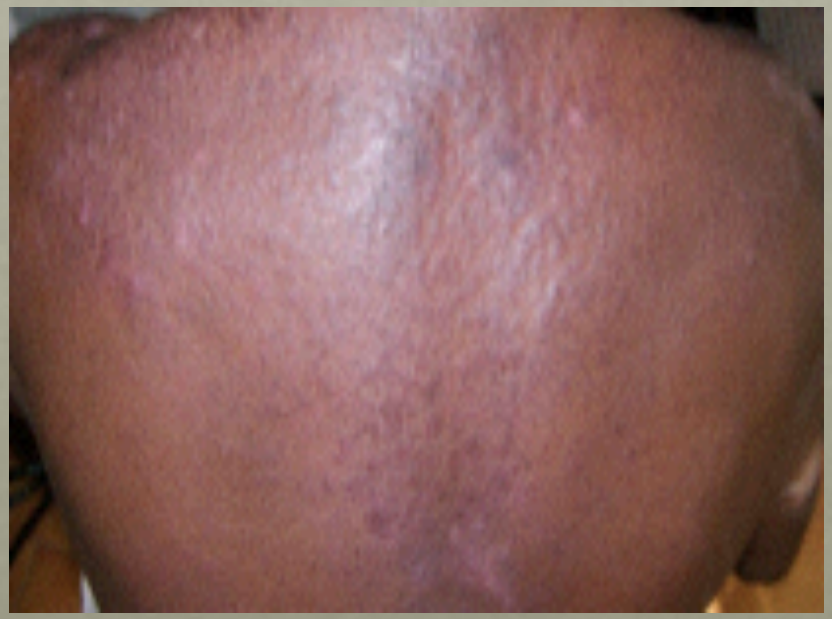
- Photodistrbuted violaceous erythema of the face, sun-exposed areas of the neck, upper chest, shoulders, dorsal arms, forearms, and hands.

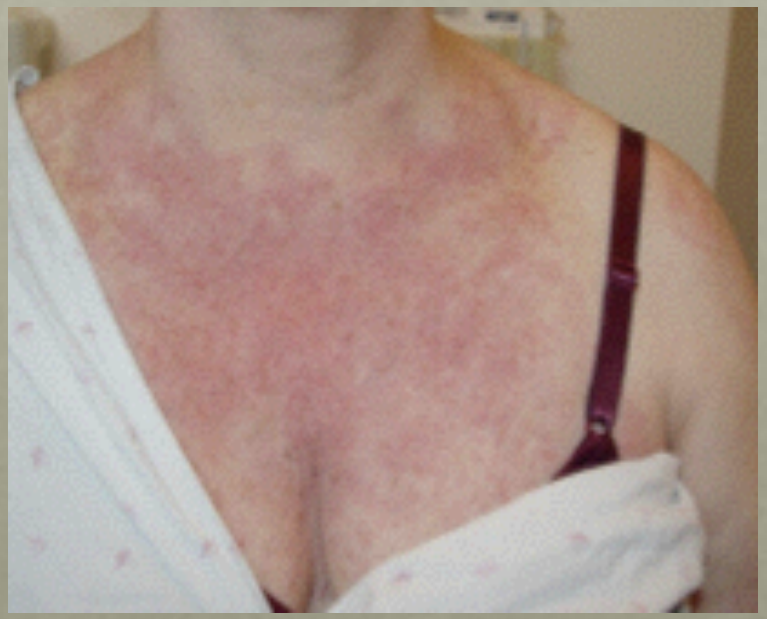




# Dermatomyositis

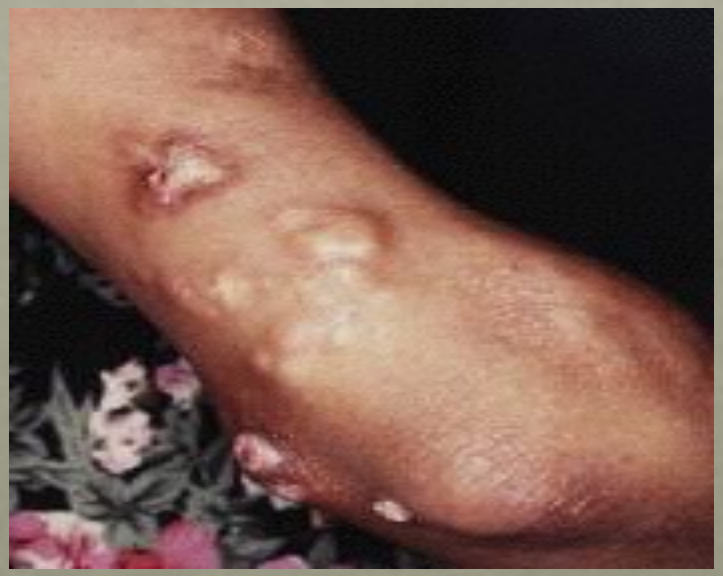
- **Shawl sign-** highly associated with interstitial lung disease





# Dermatomyositis

- **Calcinosis-** of the skin or the muscle.
  - manifests as firm, yellow or flesh-colored nodules, often over bony prominences.



# Dermatomyositis

**Are there any diseases associated with dermatomyositis?**

# Dermatomyositis

- It can be associated with other connective tissue diseases such as lupus, rheumatoid arthritis, scleroderma and Sjogren's syndrome.
- Adults with dermatomyositis have been reported to have a variety of malignancies (cervix, lungs, pancreas, breasts, ovaries and gastrointestinal tract) that sometimes follow a clinical course of exacerbation and remission in concert with the dermatomyositis.
- Female patients should be carefully screened for ovarian cancer.

# Dermatomyositis

**How do you diagnose dermatomyositis?**

# Dermatomyositis

1. History & Physical examination
2. Serum levels of muscle enzymes- creatine phosphokinase (CPK) level is most reliable indicator of disease activity
3. Serology-
   - ANA in < 60%
   - Anti-Jo-1 (anti-histidyl-t-RNA synthase) in 30%
   - Anti-Mi-2 (highly specific), but it lacks sensitivity because its present in only 25% of patients, indicates good prognosis.
4. Magnetic resonance imaging (MRI)
5. Electromyogram
6. Muscle biopsy- Inflammatory cell infiltrations & necrosis of muscle cells.
7. Skin biopsy- suggestive but not diagnostic, shows interface dermatitis.

# Dermatomyositis

**How is dermatomyositis treated?**

# Treatment of Dermatomyositis

- Oral steroids are the mainstay treatment.
- Steroid sparing agents- Methotrexate, azathioprin, mycophenolate mofetil, cyclosporine, cyclophosphamide, IVIG, and Rituximab.
- Topical steroids and antimalarial medications are used to improve the cutaneous rashes.
- Physiotherapy to improve strength and flexibility of the muscles.
- Surgical excision or Co2 laser could be utilized to remove tender calcium deposits .

# Scleroderma

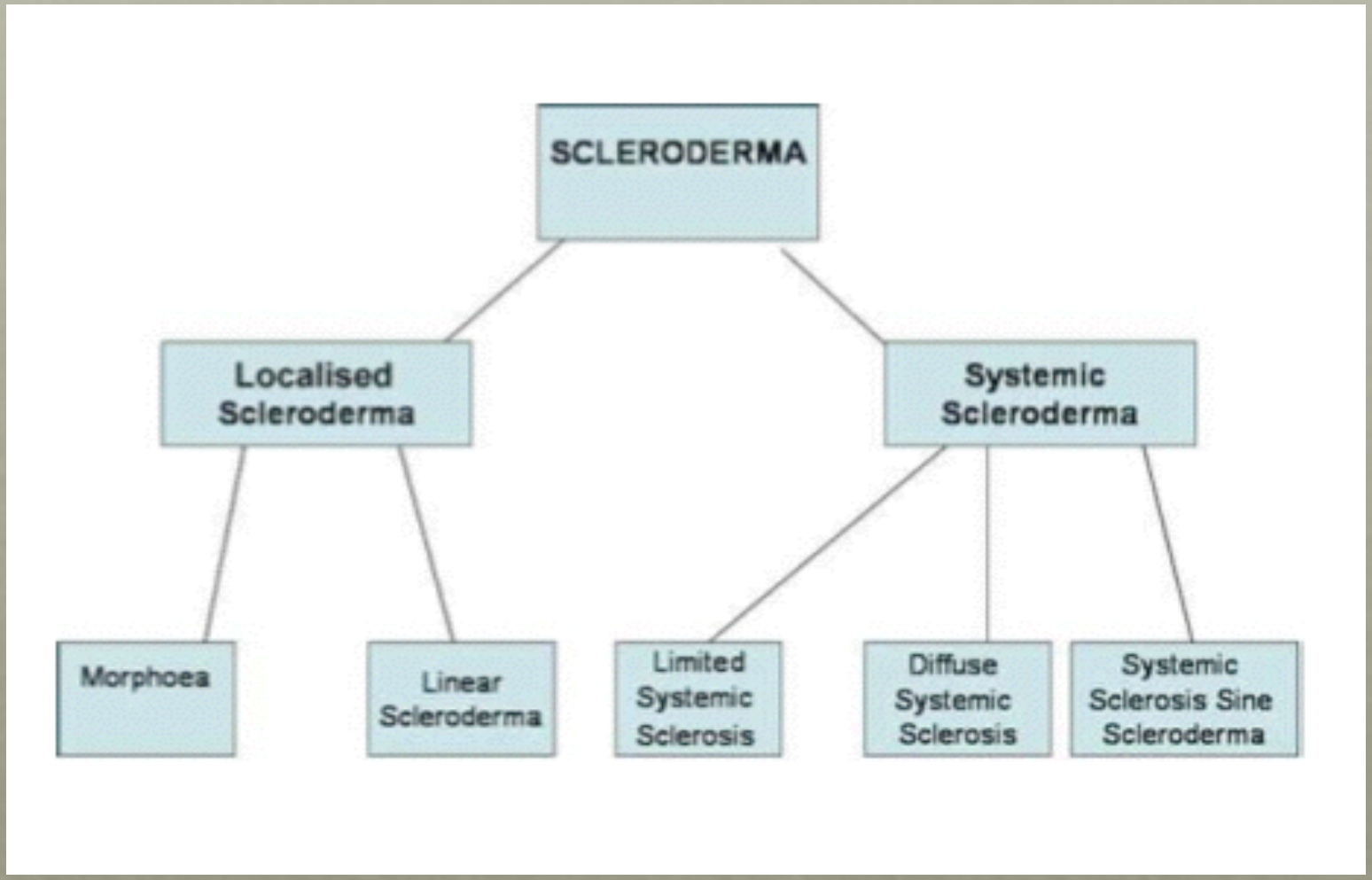
# Scleroderma

**What is scleroderma?**

# Scleroderma

- It's a chronic disease that involves the microvasculature and connective tissue and results in fibrosis.
- There is an increase in dermal collagen & decrease in the elastic tissues which leads to typical thickening & immobility
- It may be localized, as in **morphea**, or more generalized and involving visceral organs, as in **progressive systemic sclerosis.**

# Scleroderma



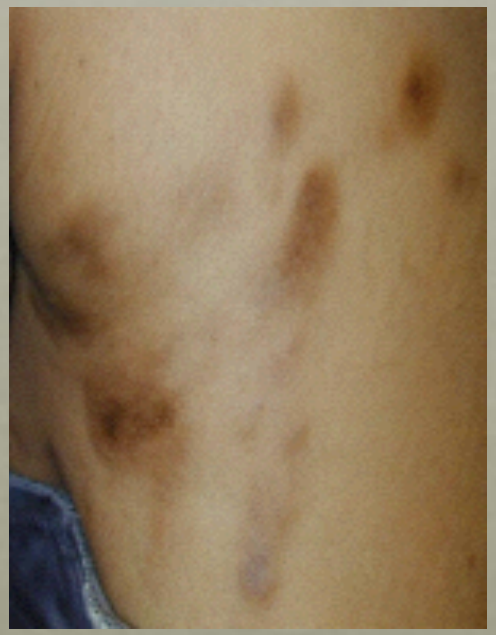
# Morphea

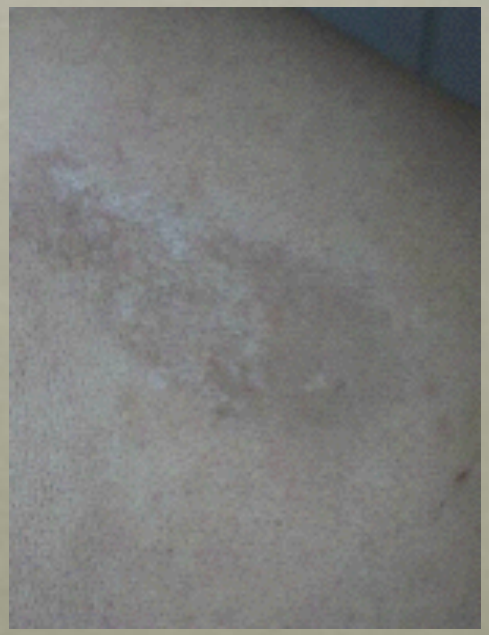
**Describe the skin changes that occur with morphea?**

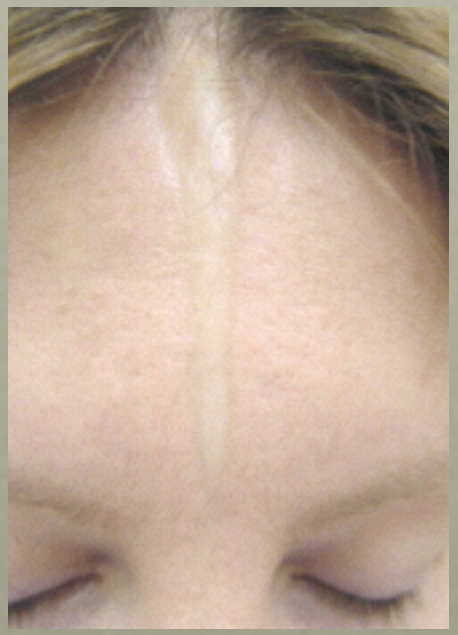
# Morphea

- Sclerotic, indurated plaques that may be solitary, multiple, linear, or generalized.
- The surface is usually smooth, with the center of the lesion a whitish or ivory color, whereas the border of active lesions is usually violaceous.
- It usually involves the skin and subcutaneous tissues but involve deeper structures, even bone.

# Morphea







# Morphea

**Do patients with morphea develop systemic sclerosis?**

# Morphea

**How is morphea treated ?**

# Morphea

- Morphea has no known cure.
- Treatment of morphea focuses on controlling signs and symptoms and slowing spread.
- Topical and intralesional steroids , phototherapy, systemic steroids, azathioprine, methotrexate, and cyclosporine might be used in severe cases.
- Physical therapy could be of help if the involvement is close to joints and cause contracture and difficulty in movement.

# CREST Syndrome

**What is CREST syndrome?**

# CREST Syndrome

- It's considered a type of limited systemic scleroderma

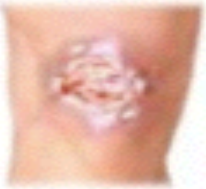
C = **C**alcinosis cutis
R = **R**aynaud's phenomenon
E = **E**sophageal dysfunction
S = **S**clerodactyly
T = **T**alangiectasia

- Most patients with CREST syndrome have circulating antibodies to centromeres, called " anti-centromere antibodies"

# CREST Syndrome

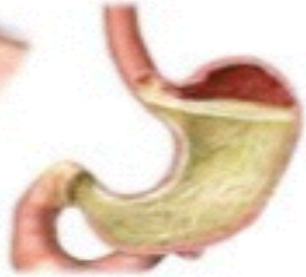
The limited symptoms of scleroderma are referred to as CREST

**C**alcinosis- calcium deposits in the skin



**R**aynaud's phenomenon- spasm of blood vessels in response to cold or stress

**E**sophageal dysfunction- acid reflux and decrease in motility of esophagus



**S**clerodactyly- thickening and tightening of the skin on the fingers and hands

**T**elangiectasias- dilation of capillaries causing red marks on surface of skin

A.D.A.M.

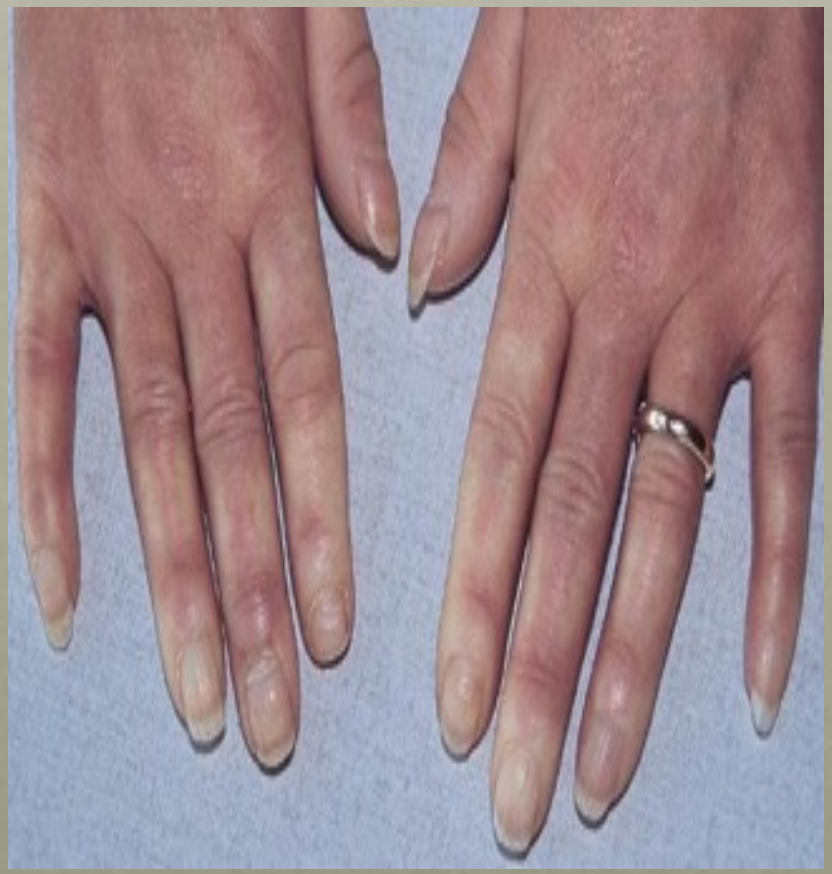
# Progressive Systemic Sclerosis

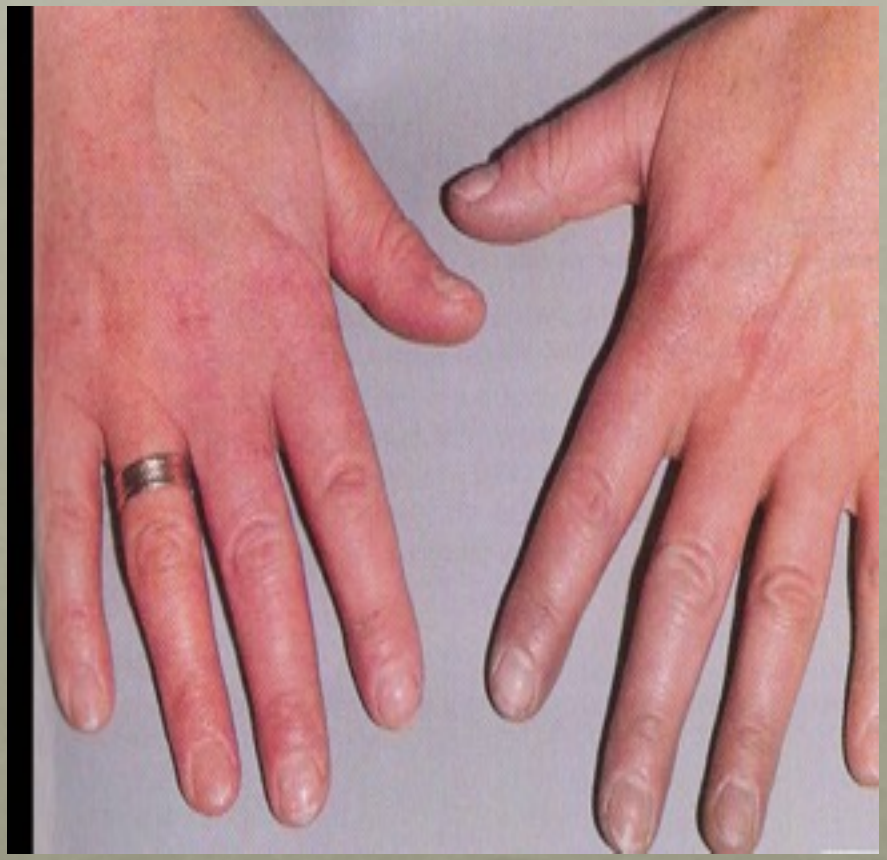
**What are the cutaneous findings in progressive/ diffuse systemic sclerosis?**

# Progressive Systemic Sclerosis

1. Swelling of the hands and feet and/or Raynaud's phenomenon
2. Telangiectasia
3. Proximal nail fold changes ( avascular areas)
4. Thickening and sclerotic changes involving the face& extrimities- progressive
   - Sclerodactyly- tapering of fingers "madonna fingers", with waxy , shiny hardened skin,which is tightly bound down & doesn't permit folding or wrinkling
   - Loss of normal facial lines ( mask like ) patient looks younger than they are
   - Thinning of lips, microstomia, radial perioral furrowing , small sharp nose
5. Digital ulcers +/- loss of digits

# Progressive Systemic Sclerosis





# Raynaud's Phenomenon

- It is digital ischemia that occurs on exposure to cold and/ or as a result of emotional stress.

Causes:

1- Rheumatic disorders( SS 85%,SLE 35%, DM 30%, RA, PAN)

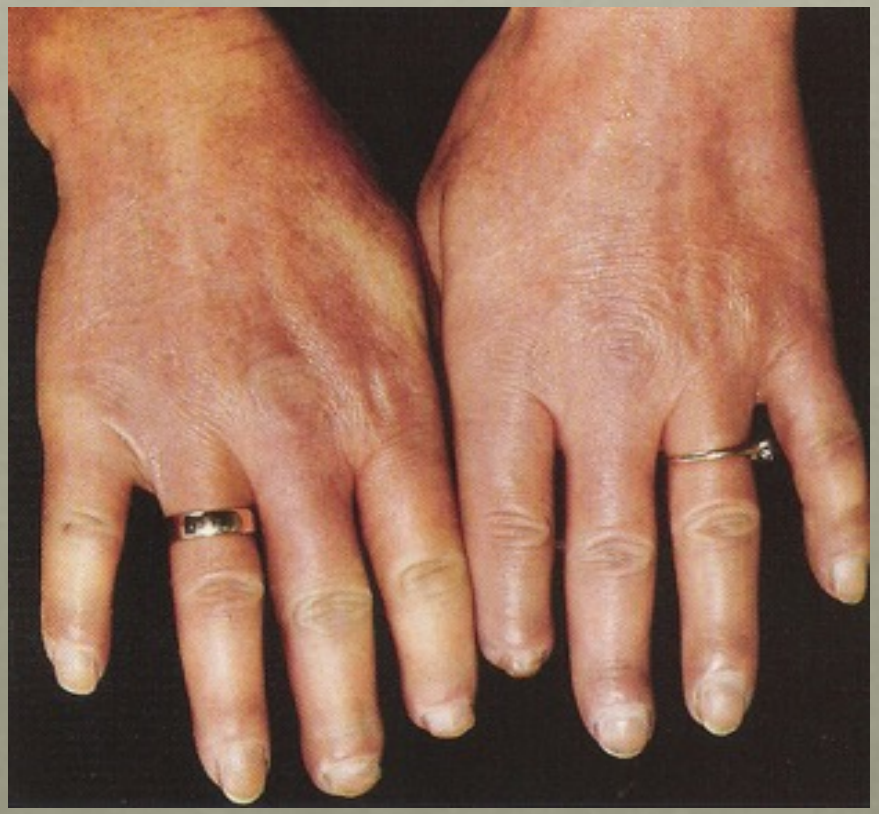
2- Diseases with abnormal blood proteins ( cryoprotein, macroglobulins)

3- Drugs ( b-adrenergic blockers, nicotine,cyclosporine)

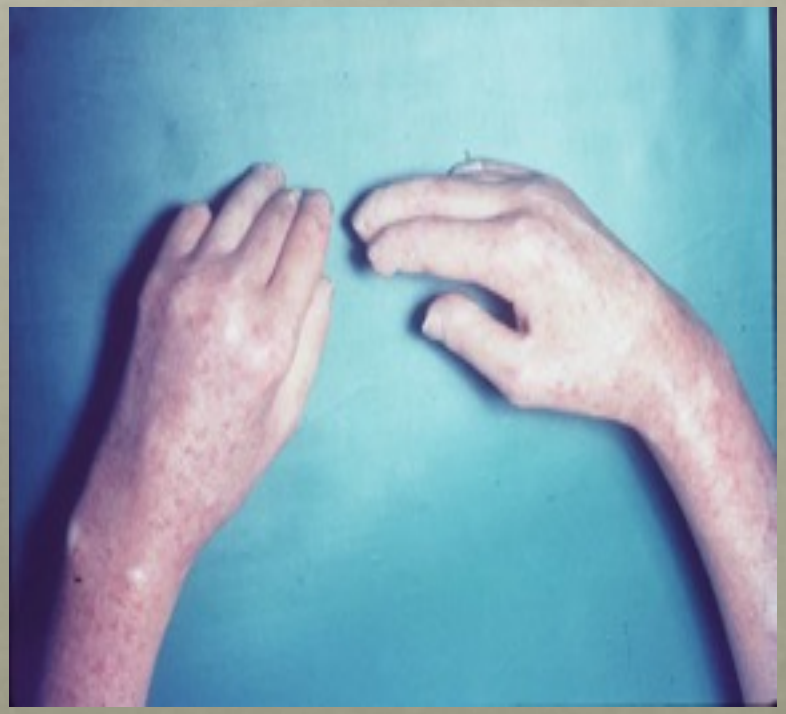
4- Arterial disease (atheriosclerosis obliterans)

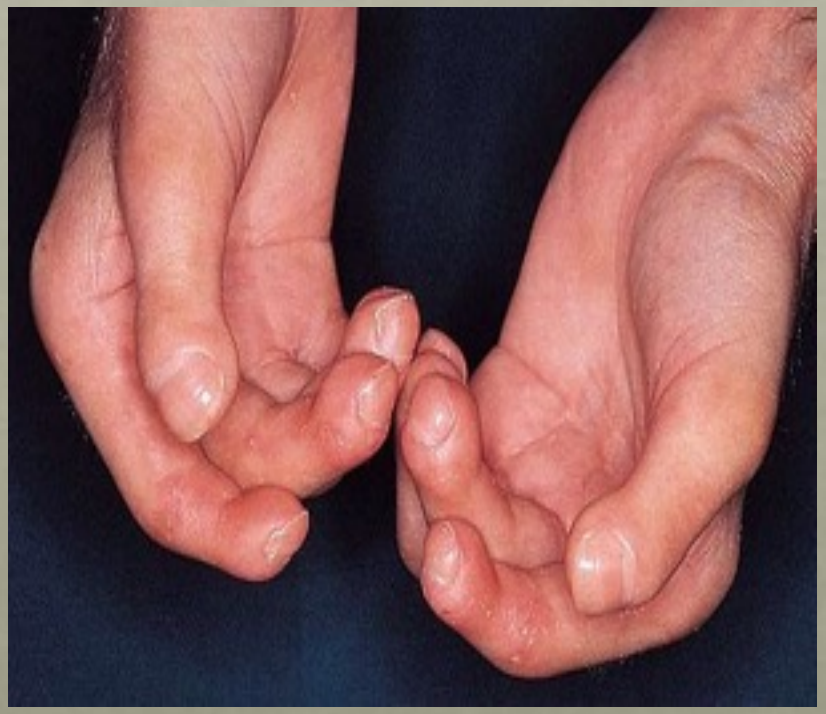
# Progressive Systemic Sclerosis

- Nonpitting edema of the hands & feet.



# Progressive Systemic Sclerosis





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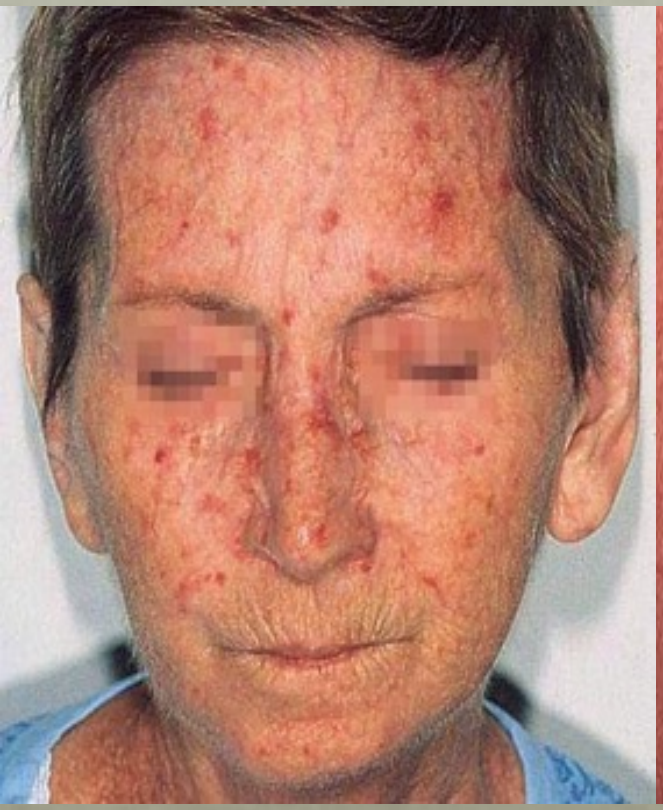

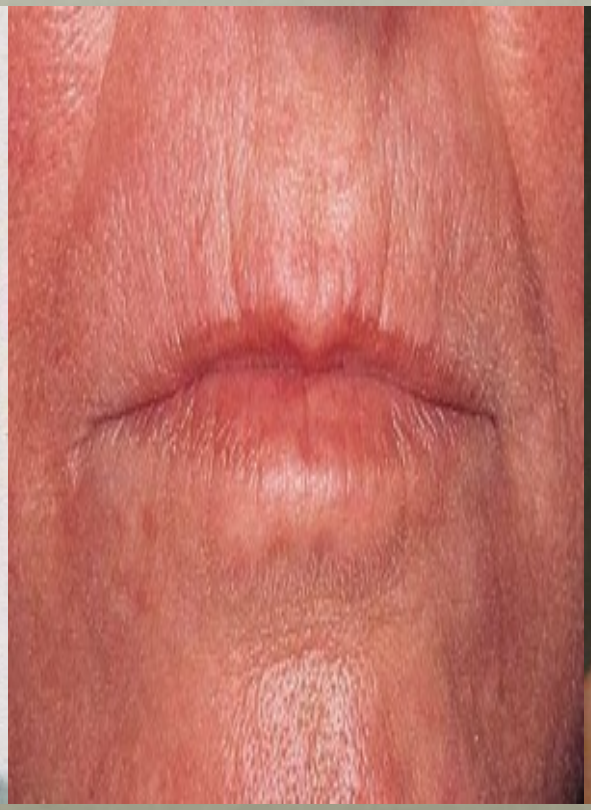

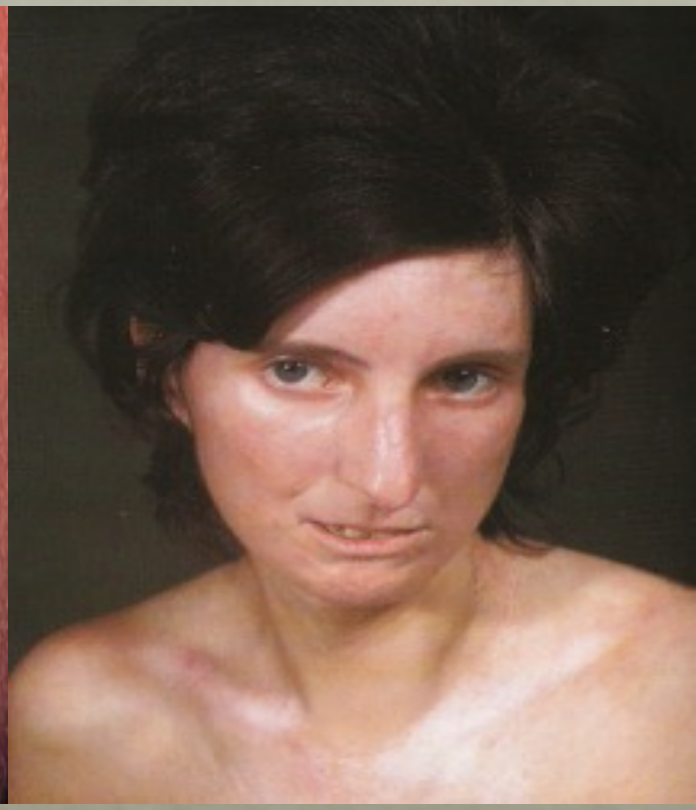

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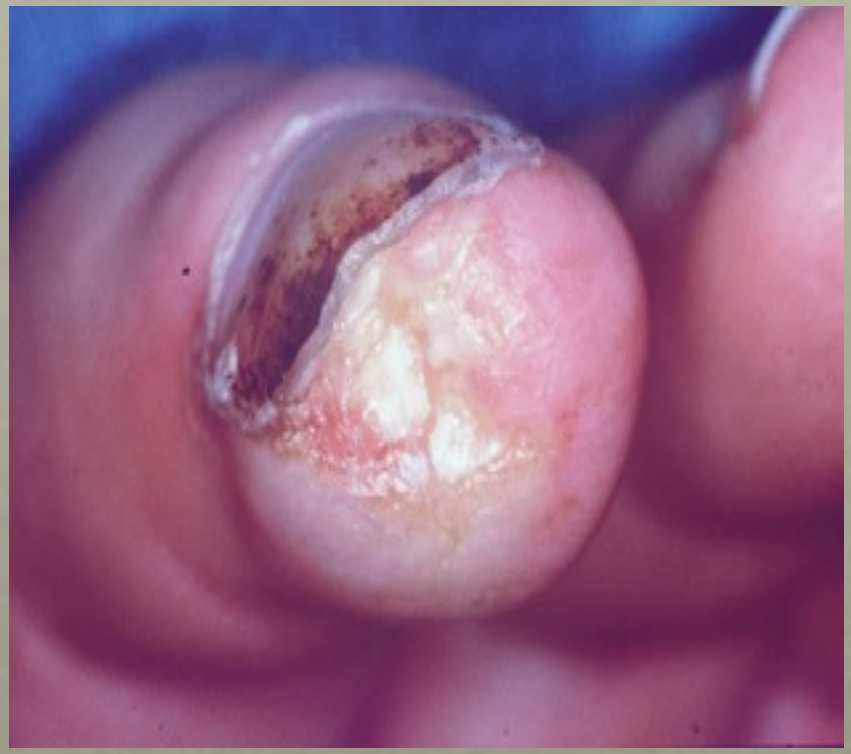

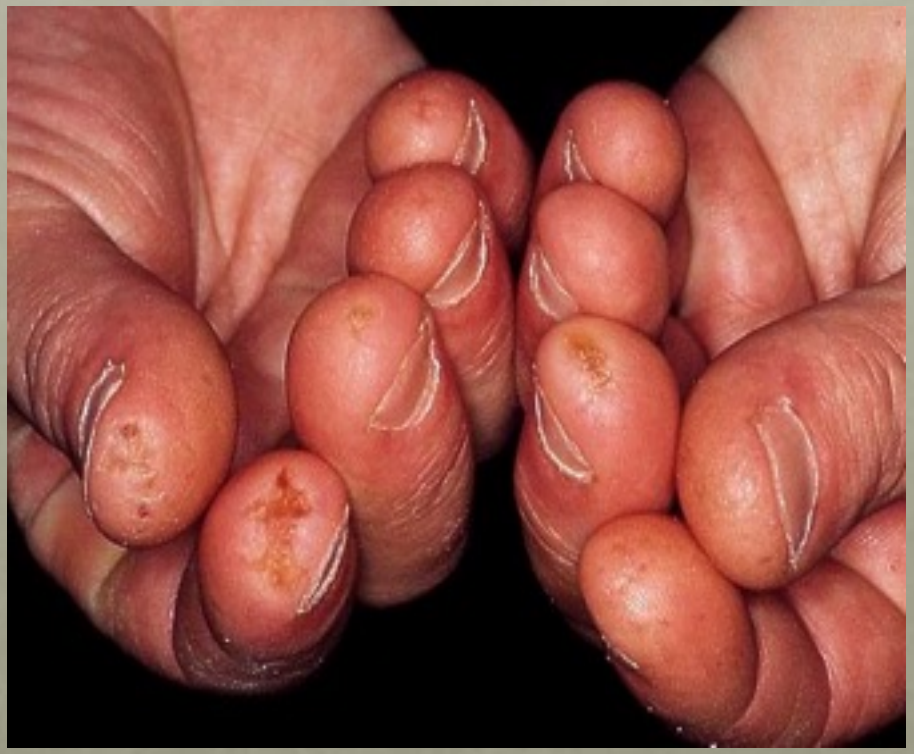

# Scleroderma

**How do you diagnose scleroderma?**

# Scleroderma

1. History & physical examination- characteristic skin changes
2. Serology
   - ANA ( often +ve)
   - Anti-centromere antibodies — 71% LSSc " CREST"; 21% of DSSc " progressive"
   - Anti-Scl-70 " anti-topoisomerase I" — 33% of dSSc; 18% of CREST
3. Skin biopsy- skin atrophy with preservation of skin appendages.

# Scleroderma

**How do you manage a patient with scleroderma?**

# Scleroderma

- Treatment is symptomatic.
- Raynaud's phenomena:
    - Stop smoking
    - keep hands warm and decrease trauma
    - calcium channel blockers* ( nifedipine)
    - aspirin and
    - vasodilating drugs ( iloprost )
- Calcinosis cutis: nifedipine, surgical or laser excision.
- Skin sclerosis: physiotherapy, phototherapy.
- GI: proton pump inhibitor, surgery for strictures.
- Kidney: ACE inhibitors.
- In severe cases: immunosuppressant , D-Penicillamine might be used ( blocks aldehyde groups involved in intermolecular cross-links in collagen)