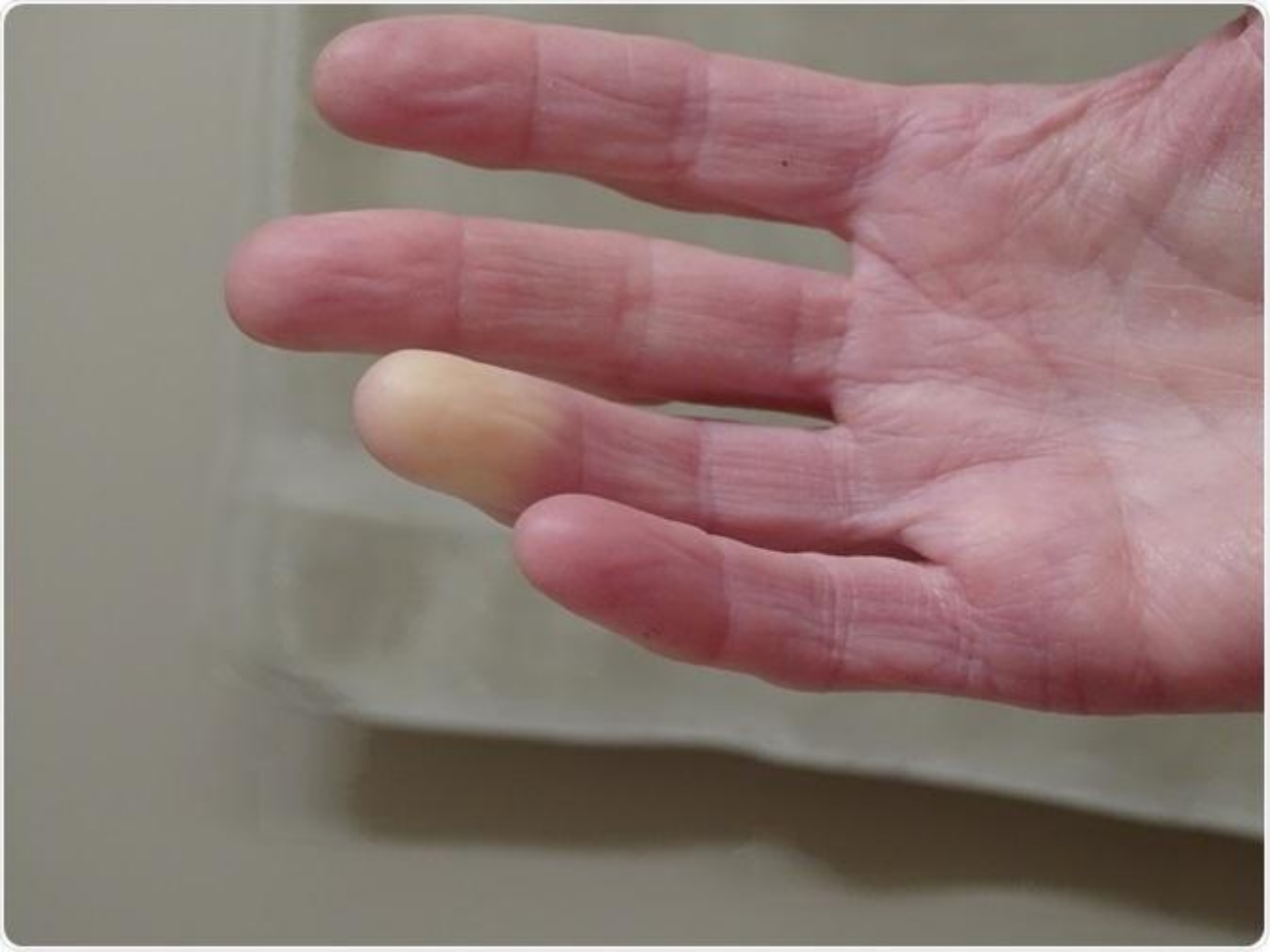
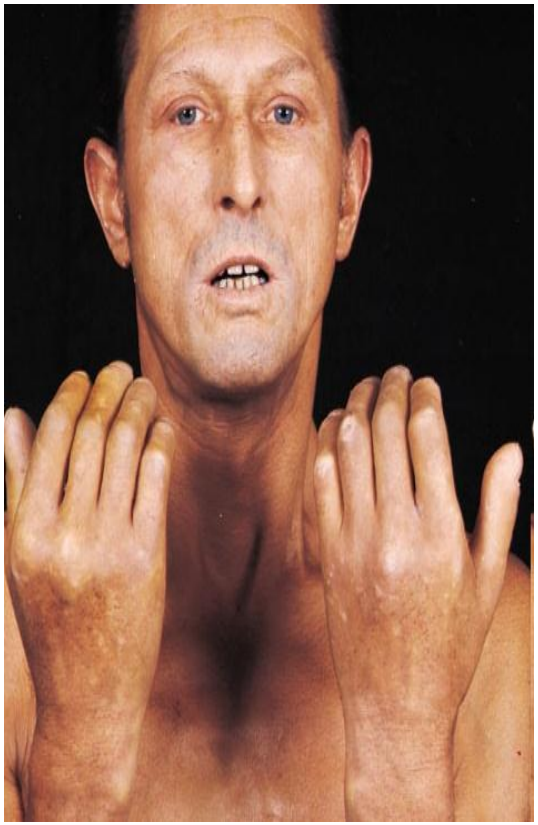


Raynaud's syndrome

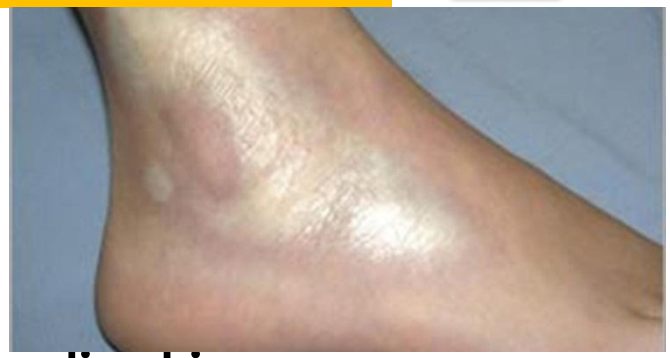
- **Common in young women5-10%**
- **No ulceration or ischemia.**
- **If History and examination is reasuring.**
- **No investigations is necessary.**
- **Avoid cold**
- **rare need ca channel blockes.**

Scleroderma

- group of disorders chacterized by thickened skin .
  - Scleroderma literally means hard skin.
- Unknown etiology
- Autoimmune reaction leads to stimulation of fibroblasts (cells that make collagen). And vascular changes result is accumulation of collagen and other connective tissue components in parts of the body such as skin, lungs ECT…



Types of Scleroderma



- Localized :Affecting cutaneous and subcutaneous tissues only
  - Morphea-Localized patches of sclerotic skin

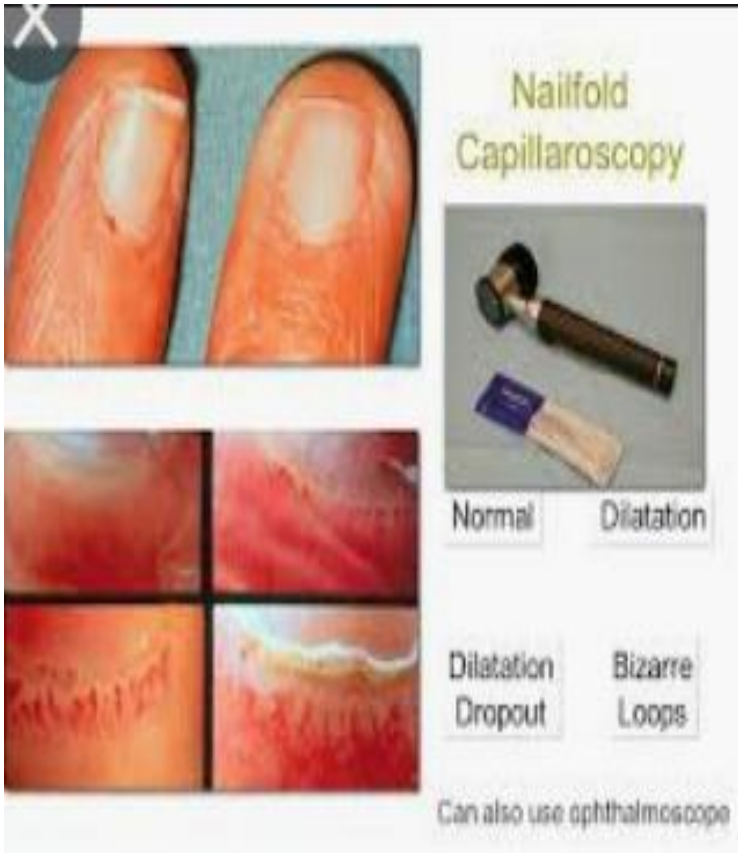
- Systemic
  - Affecting Skin and internal organ involvement
    1. Limited (CREST)
    2. Diffuse(Systemic Sclerosis).

Systemic Sclerosis

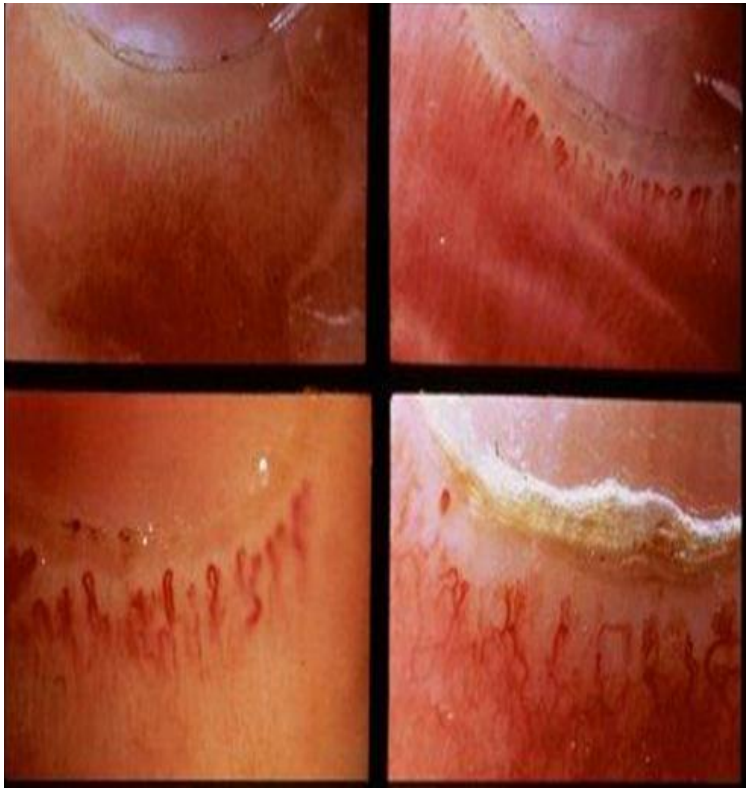
- Raynaud's is the first manifestation .
- Skin tightness extends proximal to MCPs, involves skin of chest and back.
- Multi -organ involvement.
- Anti-topoisomerase-I (Scl-70) antibodies (30% of patients)

Signs and Symptoms

- Skin
  - early(Puffy)edematous then Tightening and Thickening.
  - Telangiectasis.
  - Calcinosis
  - Periungual erythema. dilated nail fold capillary loops.
  - Ulcers on fingertips and gangrene.

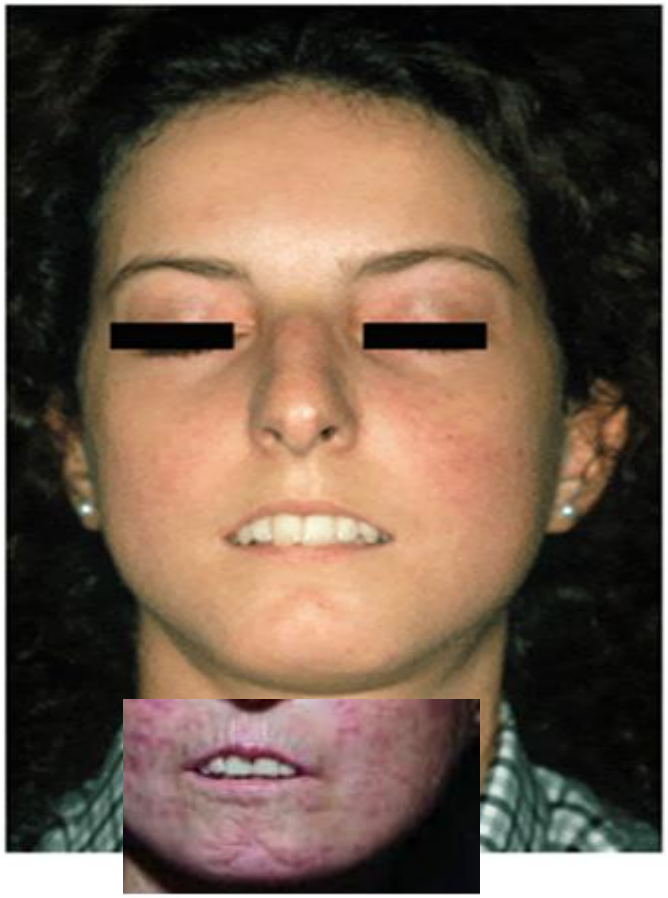


Nailfold Capillaroscopy
Used for dx

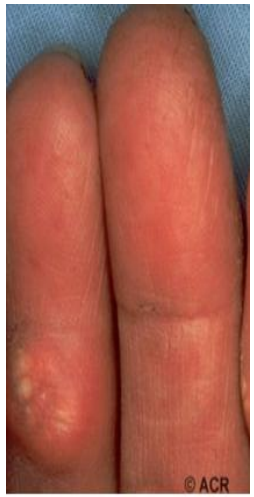


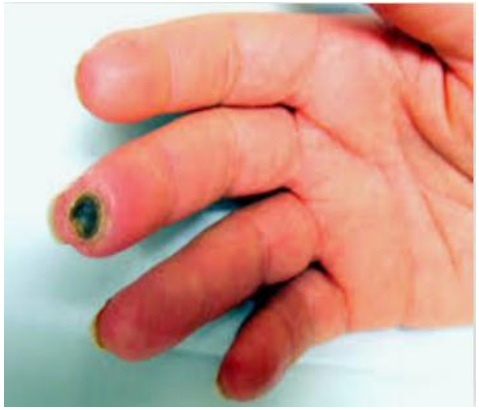
Hands and face in SS

Arthritis/ sclerodactyle/ulcers

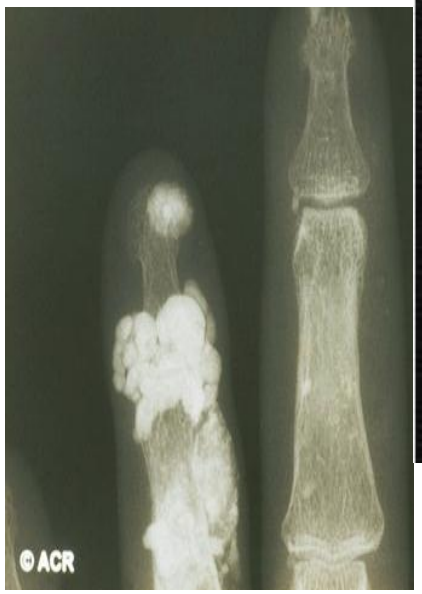


Decreased mouth opening

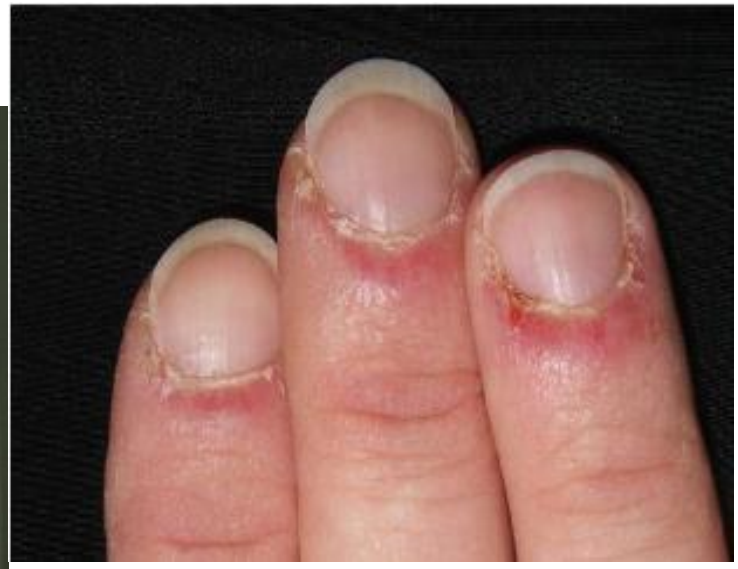




Digital ischemic ulcers



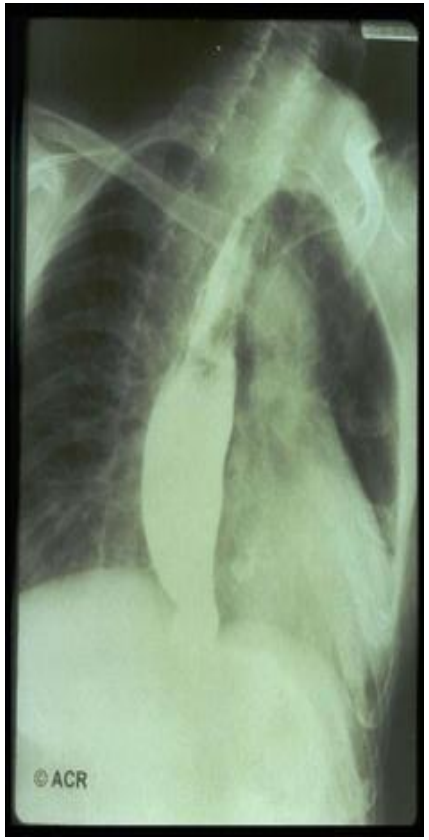
calcinosis



Periungual Erythema

Gastrointestinal involvment

1. Decreased esophageal mobility & Dysphagia AND Heart burn(GERD).
2. SOME patients Constipation .
3. Others Mal-absorption & Watery diarrhea



Respiratory involvment

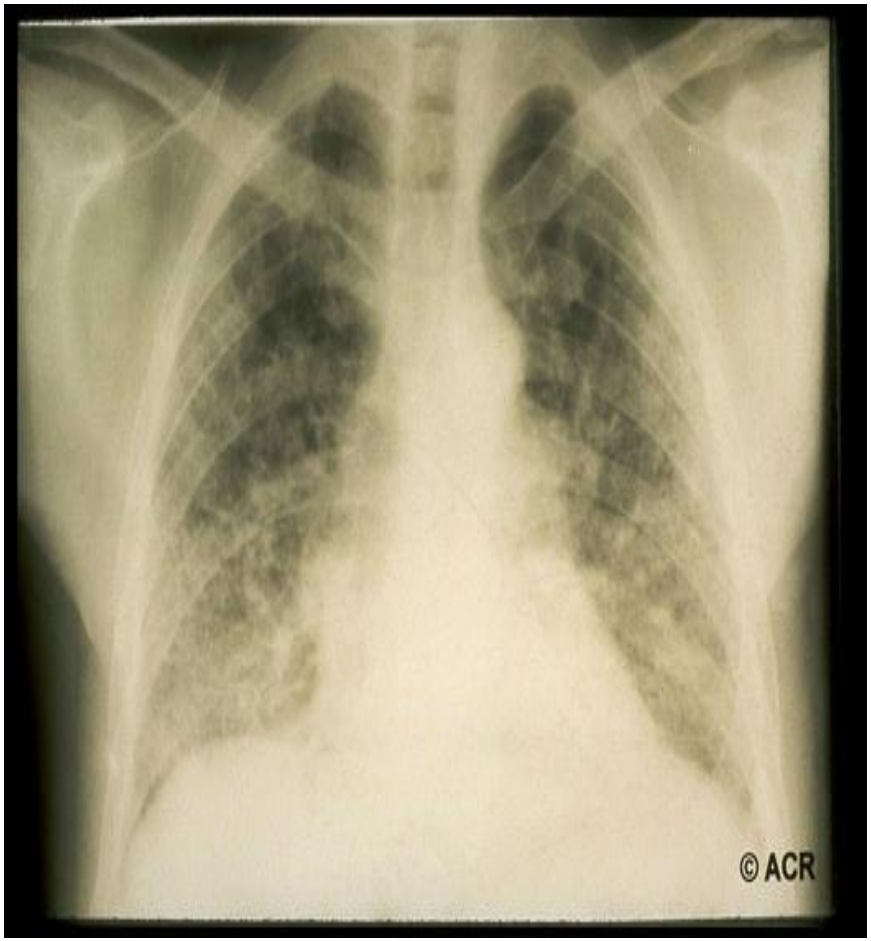
interstitial pneumonitis

↓

Pulmonary fibrosis

↓

Pulmonary hypertension



Pulmonary Fibrosis

Renal

- Proteinuria, hypertension
- Scleroderma renal crisis – 10-15% **********
- Hypertensive encephalopathy
- Headache and Blurred vision.
- +/-generalized seizures.
  - Acute onset hypertension.
  - Renal impairment (increase creatinine)
  - Pulmonary oedema.
  - Microangiopathic haemolytic anaemia

Sclerordema renal crisis Risk Factors

- High dose corticosteroid predisposes to crisis
  - so use prednisolone only if needed temporarily, & keep dose <15 mg/day.

Treatment of renal crisis

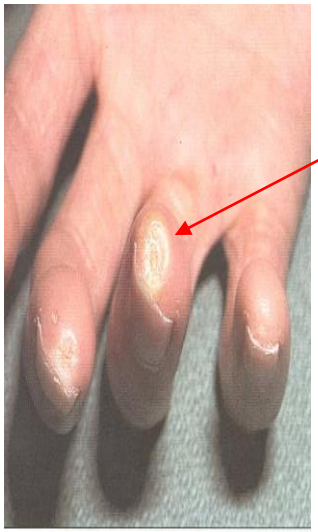
ACE inhibitor is the agent of choice- leading to an improvement in blood pressure ****

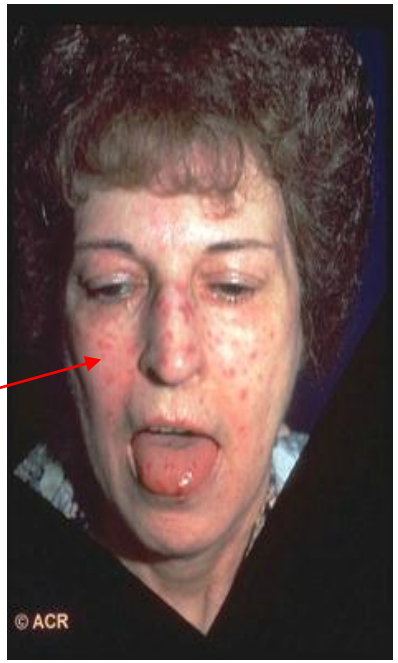
Scleroderma : Treatment

- Assess patient determine extent and Organs involved rx is organ based
  - GERD: PPI,
  - Alveolitis : steroids,and cyclophosphamide
  - Pulm HTN: similar to idiopathic
  - Renal: ACE inhibitors
- Raynaulds ⟹ Keep warm ca channel blockers .
- IV prostaglandins in Cases of digital ulcers or pulmonary hypertension .
- Bosentan endothelin receptor antagonist vasodilator in pulmonary hypertension

Limited cutenous systemic sclerosis(CREST)

- **CREST**- was called crest because of pattren of involvment.
  - **C-Calcinosis=ca deposits.**
  - **R-Raynaud's**
  - **E-Esophageal dysfunction**
  - **S-Sclerodactyly.**
  - **T-Telangiectasia.**





Two important features

1. A shows anticentromere antibody, and is highly specific
2. Development of pulmonary hypertension.

Sjögren's syndrome (SS)

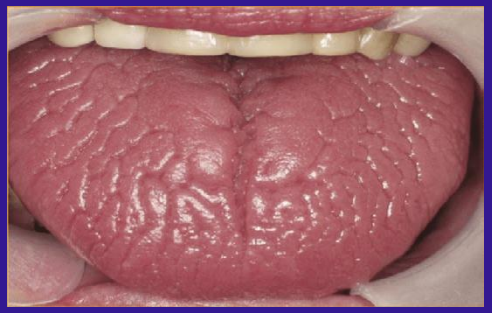
A chronic inflammatory disorder characterized by lymphocytic infiltration of the lacrimal and salivary glands.

types

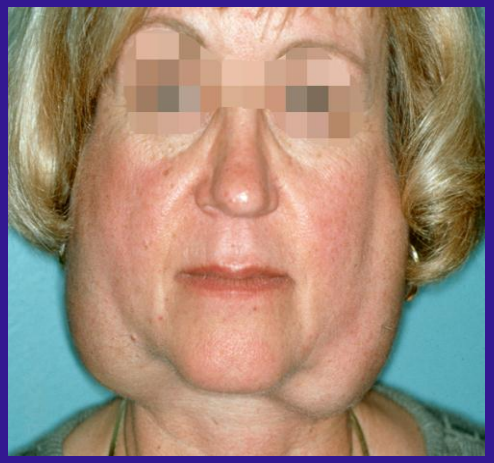
1. **a primary sj .**
2. **secondary sj in association with another ed autoimmune dis such as rheumatoid arthritis, systemic lupus erythematosus (SLE) .**

clinical features

- Keratoconjunctivitis sicca (dry eyes)
- Xerostomia (dry mouth)
- Salivary gland enlargement
- arthritis .
- Fatigue .



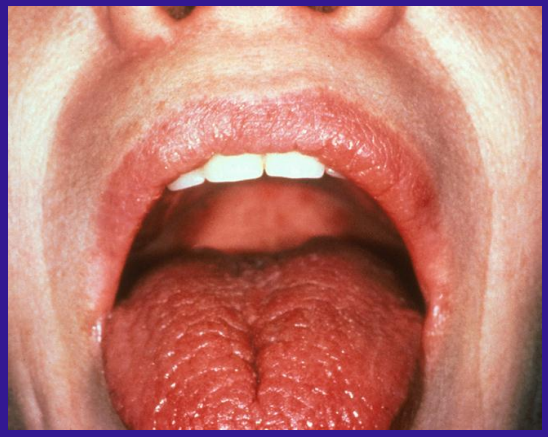
Dry mouth

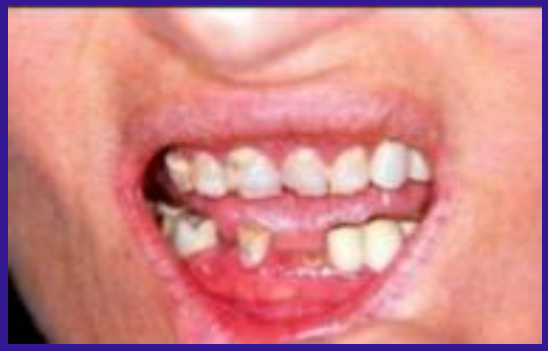


Bilateral enlarged parotid gland

Xerostomia

- Dental caries(loss of teeth).
- oral candidiasis .
- **changes in taste.**
- **Dysphagia.**





Less common features

- Interstitial lung disease
- Renal tubular acidosis
- Lymphoma.

If **massive lymphadenopathy** or develops during the disease course, biopsy should be performed to detect malignancy (lymphoma)***

Investigations

- CBC -Anemia of ch dis.

And high ESR.

one or more of these autoantibodies

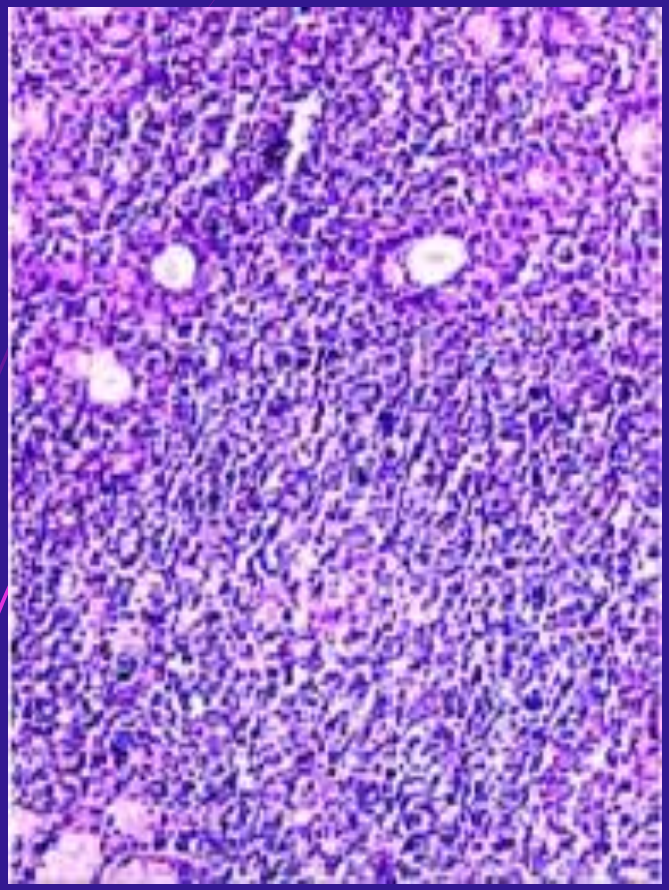
1. SS-A (anti-Ro)*
2. SS-B (anti-La) *
3. Rheumatoid factor(+).
4. ANA (+) .

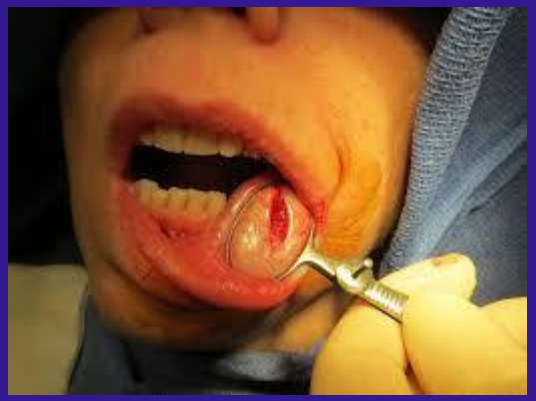
Schirmer tear test
To dx dry eyes :

- **measures flow of tears over 5 minutes using absorbent paper strips placed in the lower lachrymal sac;**

a normal result is greater than 6 mm of wetting.

Pathological diagnosis: salivary gland biopsy





focal lymphocytic infiltrate in the minor salivary glands on lip biopsy.

Causes of Salivary gland enlargement

Unilateral:

Bacterial infection

tumor

bilateral

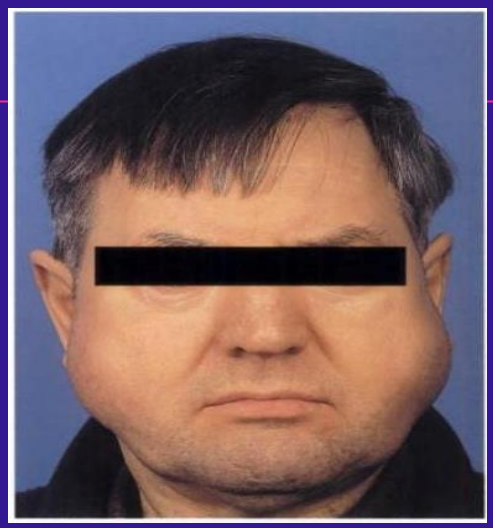
Viral mumps

sjogrens s

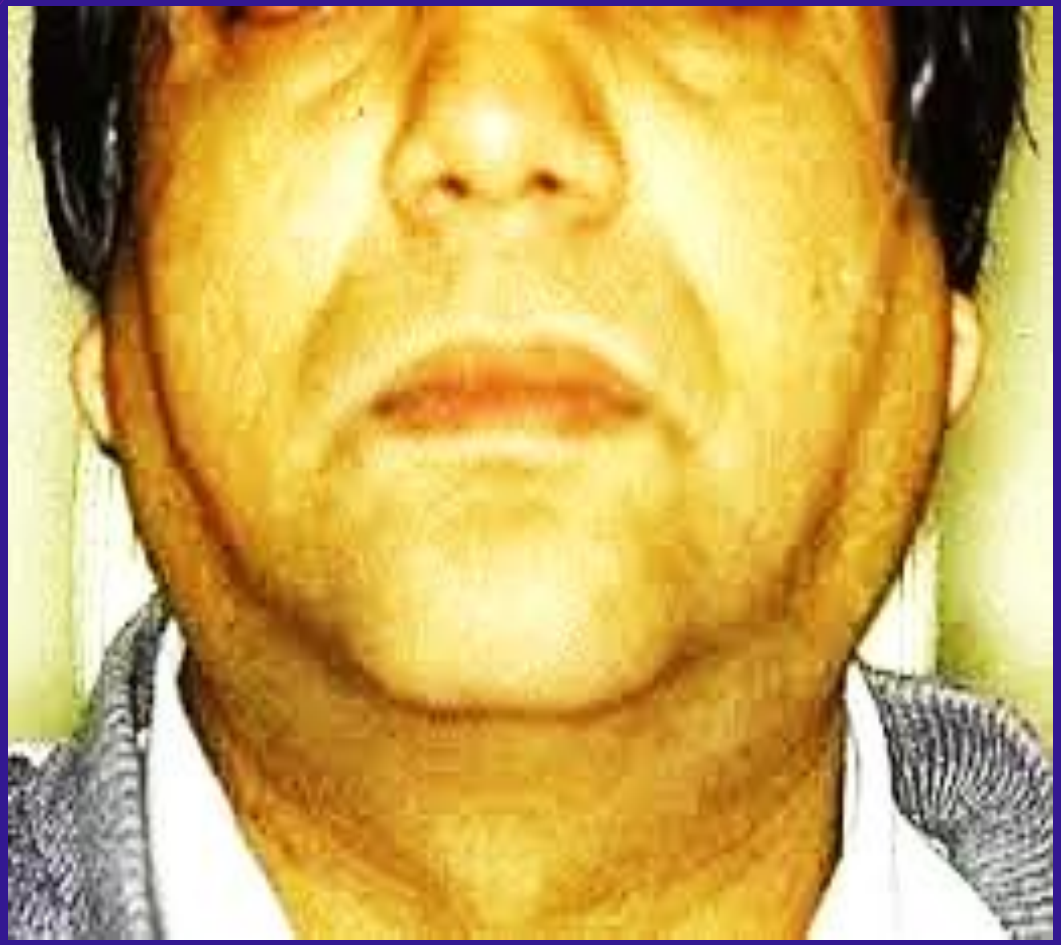
sarcoidosis

HIV

cirrhosis



What is the abnormality
Jaundice +bilateral salivary gland enlargment



Diagnosis

- symptoms of ocular dryness and/or oral dryness
- salivary gland swelling and biopsy
- and/or laboratory evidence of autoantibodies (anti-Ro/SSA and anti-La/SSB)

Management

- General principles
- Xerostomia.
- Stimulation of existing salivary flow

Cevimeline(Cevimeline is a cholinergic agonist)

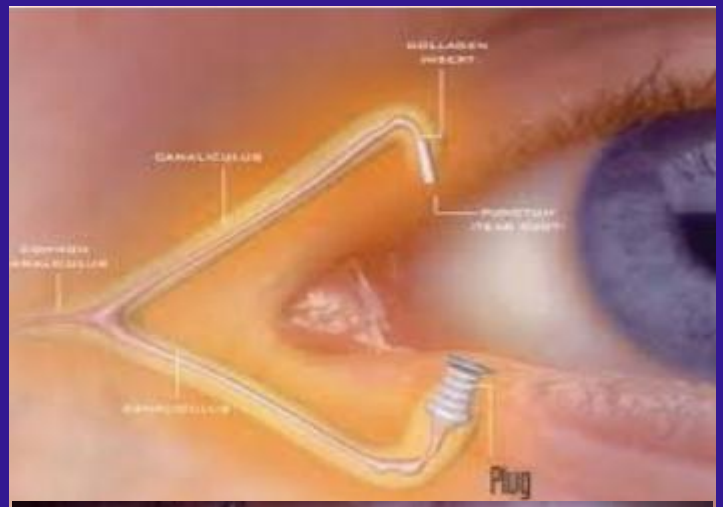
- Replacement of oral secretions .
- Dental caries prevention.

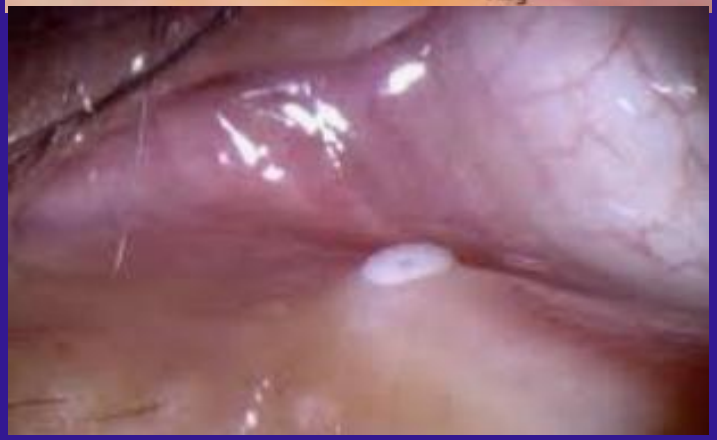
(dry eyes)

- Artificial tears and ocular lubricants

- Topical cyclosporine

Conservation of tears (Punctal occlusion)





TREATMENT OF EXTRAGLANDULAR MANIFESTATIONS

- corticosteroids and, if so, other immunosuppressive drugs such as hydroxyl chloroquine,MTX etc.

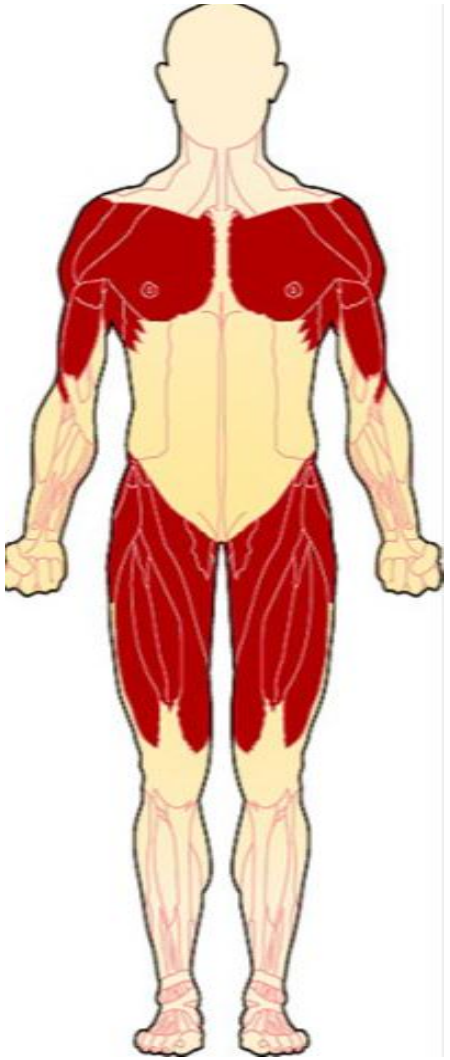
Polymyositis and dermatomyositis

Polymyositis and dermatomyositis

- connective tissue disorders characterized by muscle weakness and inflammation .
- Proximal muscle weakness +high CPK

  +Myopathic EMG+

inflammatory myopathy in muscle biopsy



- Proximal muscle weakness +pain.
- high CPK.
- Myopathic EMG+.
- inflammatory myopathy in muscle biopsy.

- Other systemic autoimmune diseases such as SLE can also cause myositis

Polymyositis

- The typical presentation is with <u>symmetrical proximal muscle weakness</u>.
- Patients has difficulty rising from a chair, climbing stairs and lifting in combination with muscle pain.

Systemic features

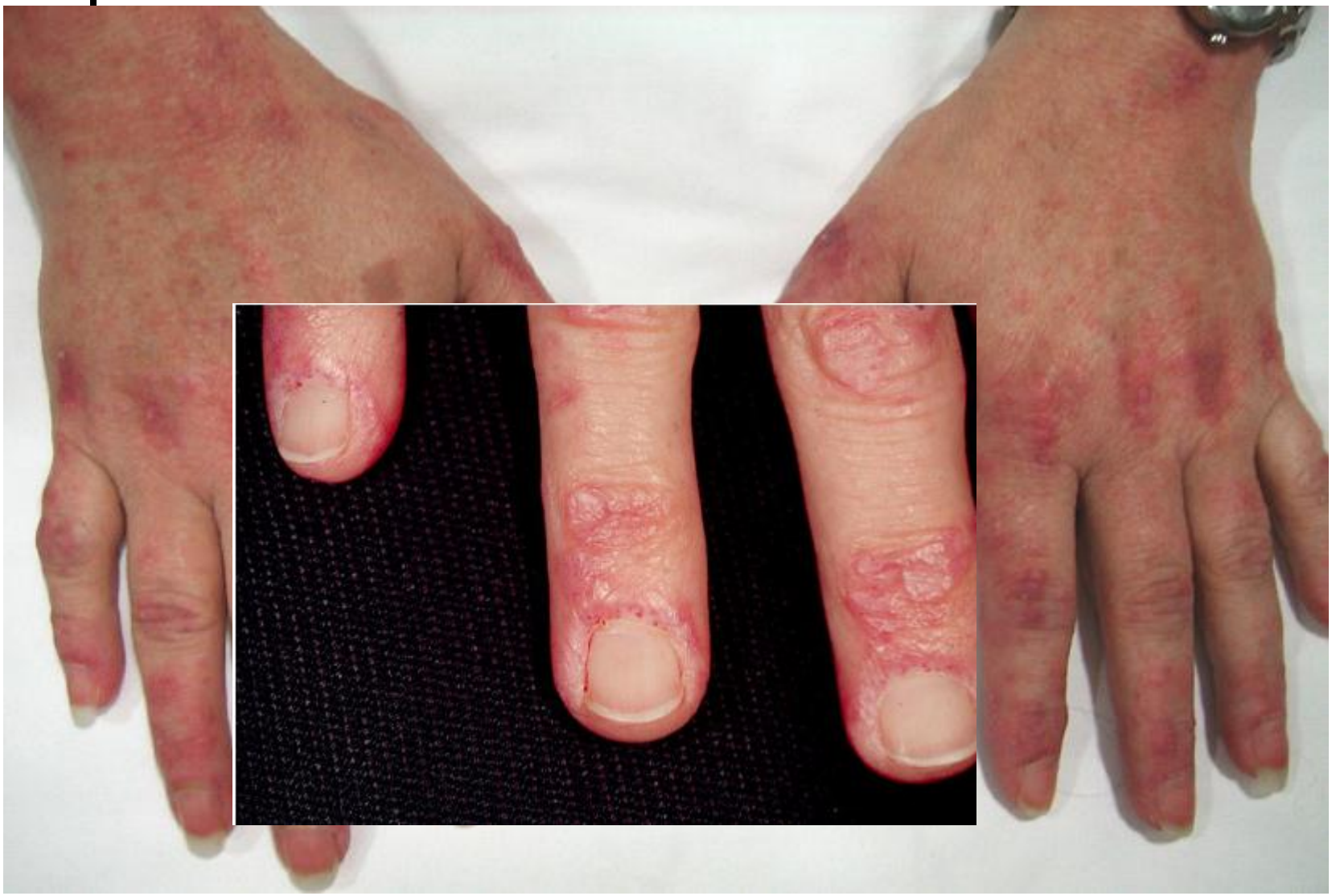
- Fever, weight loss and fatigue.
- Dysphagia.
- Interstitial lung disease.
- presence of antisynthetase (e.g. anti-Jo1) antibodies

Dermatomyositis

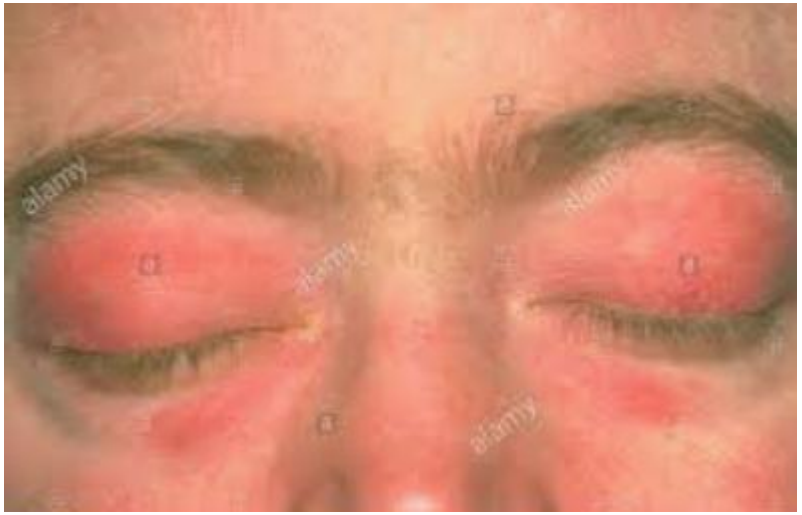
- polymyositis, but occur **in combination with characteristic cutaneous manifestations.**

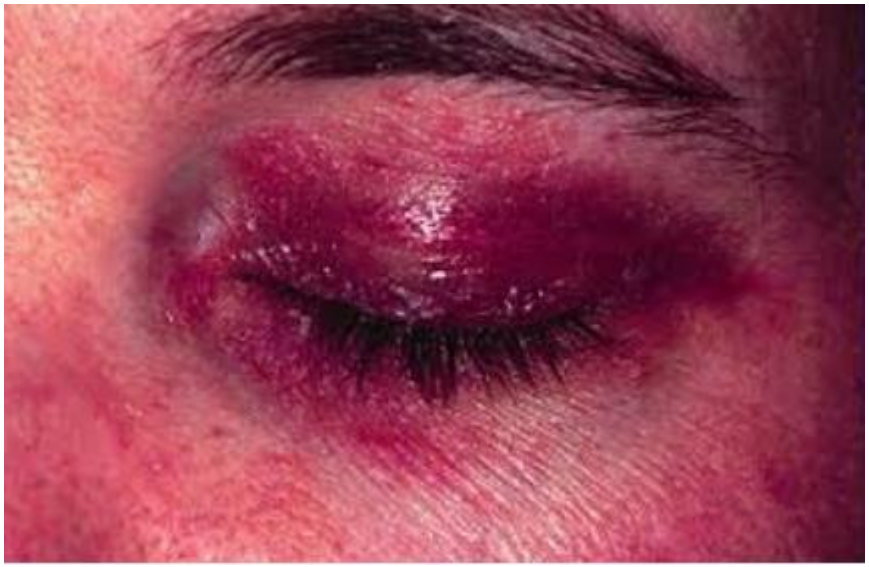
1. Gottron's papules are scaly erythematous/violaceous plaques or papules occurring over the extensor surfaces of the proximal and distal interphalangeal joints.

Gottren’s sign

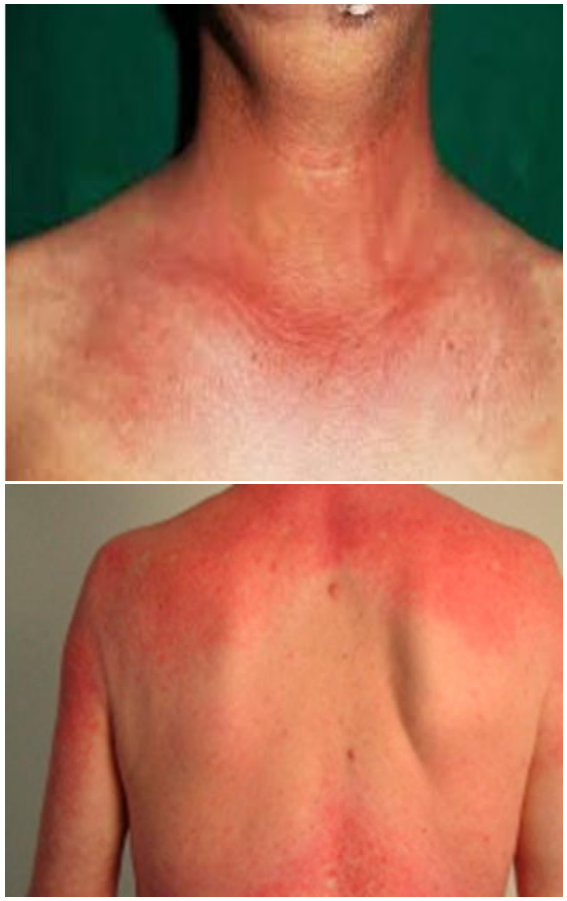


- Red purplish rash around eye.
- Heliotrope sign.





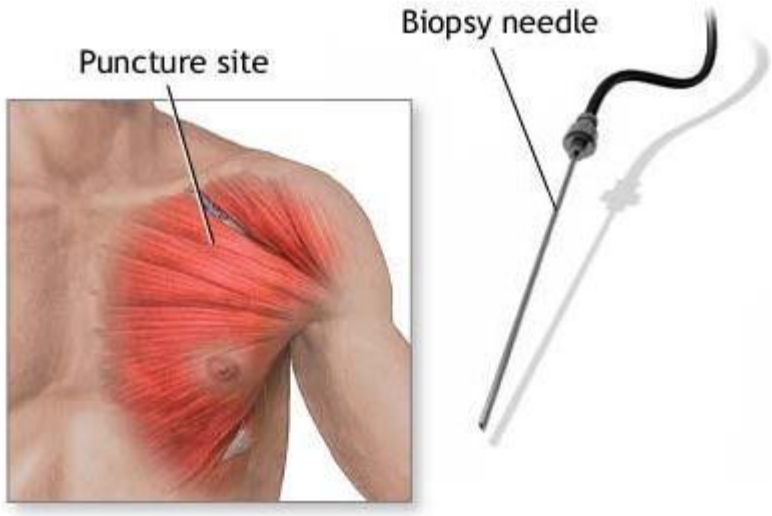
Similar rashes occur on the upper back, chest and shoulders **('shawl' distribution).**

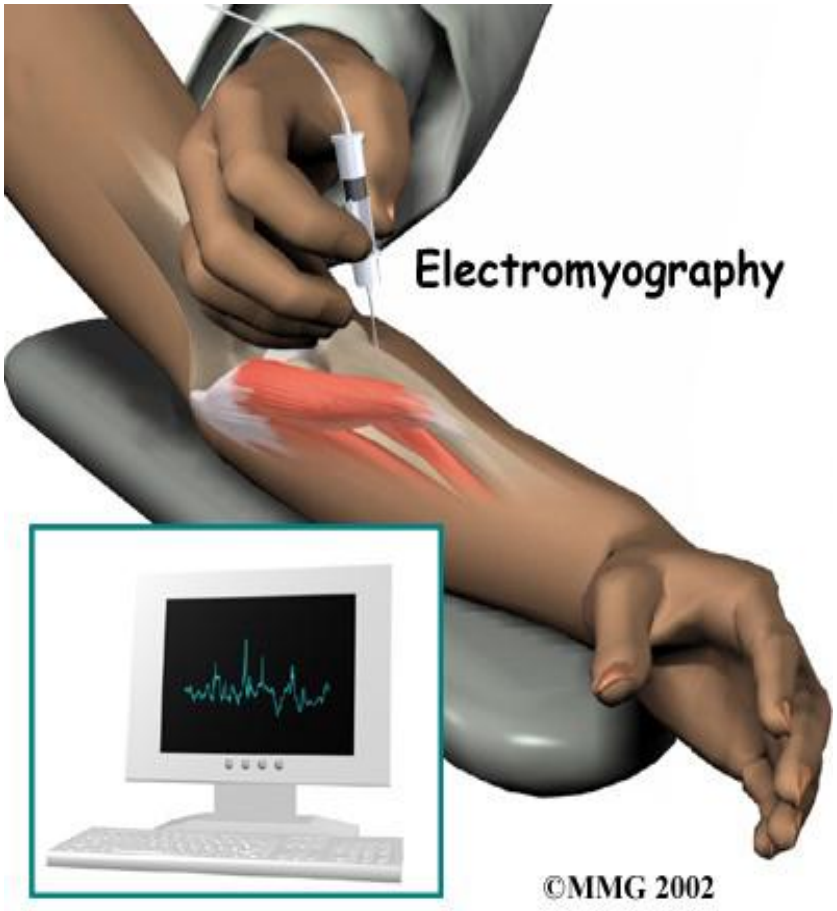


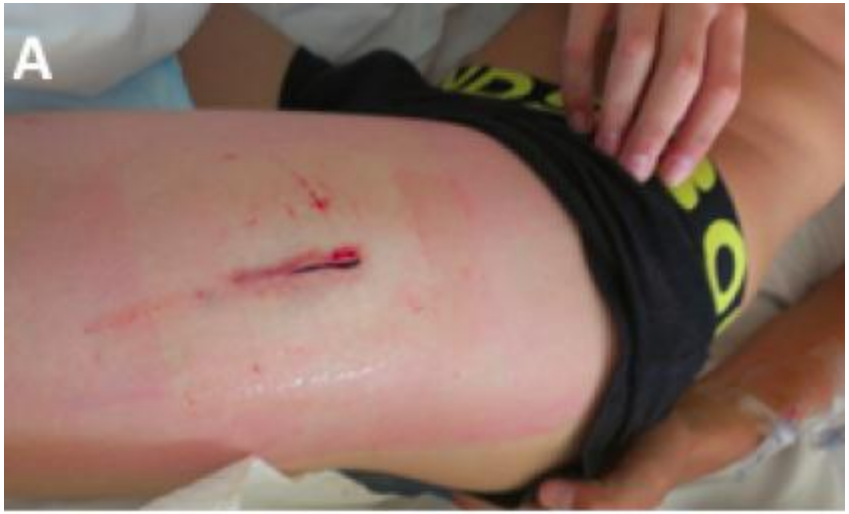
Shawl sign
erythematous rash on upper part of the body

Polymyositis / Dermatomyositis

- **Diagnostic criteria**
  1. Proximal muscle weakness
  2. Elevated serum CK
  3. Myopathic changes on EMG
  4. Muscle biopsy demonstrating lymphocytic inflammation
  5. Dermatomyositis: Skin rash as well as criteria above







Open muscle biopsy

Management

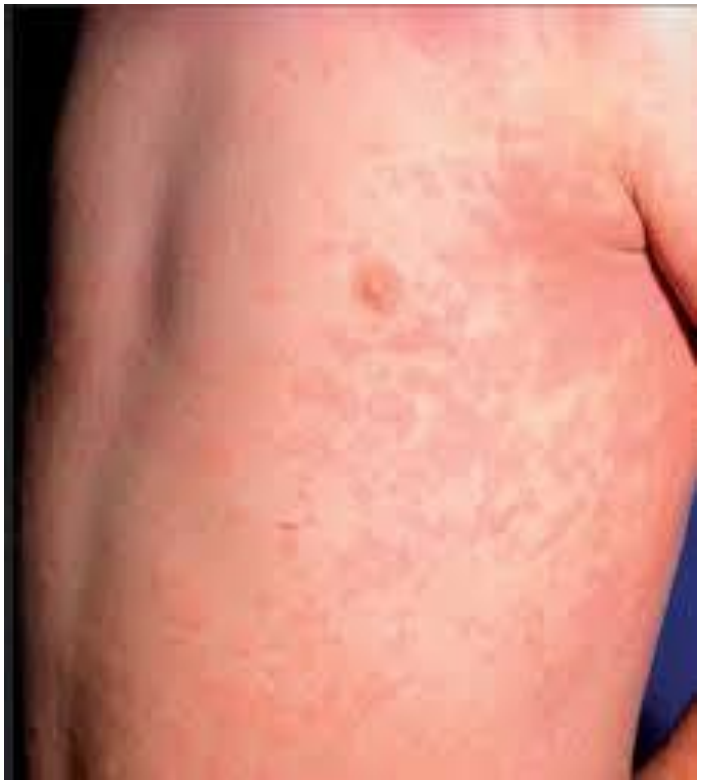
- Corticosteroids.
- Methotrexate.Azathioprine.
- Iv.Ig(iv immunoglobulin).
- Rituximab.
- physiotherapy.
- Beware malignancy as per pt S\S.

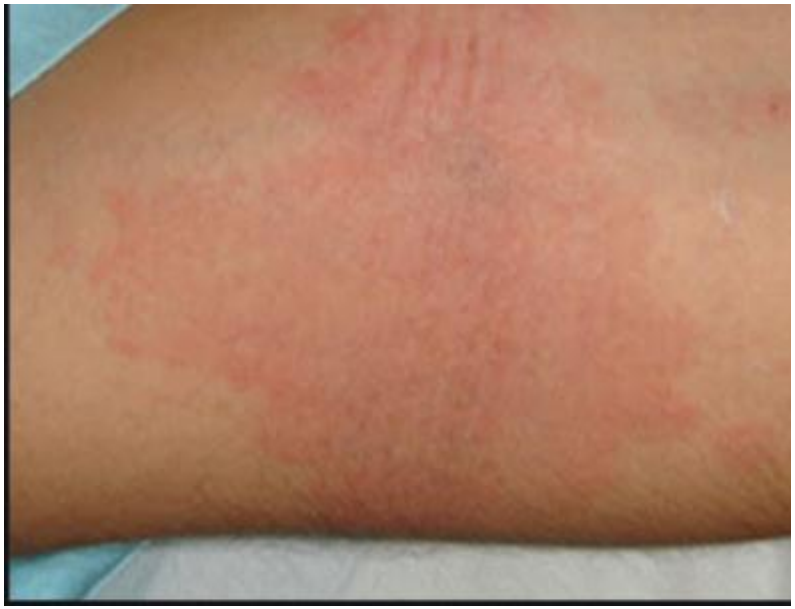
Adult onset still’s disease

- Arthritis, fever, skin rash.
- Splenomegaly ,hepatomegaly,lymphadenopathy.
- May sore throat ,abdominal pain

high spiking fevers, salmon-colored rash that comes and goes, and arthritis.

Negative ANAand negative RH factor.





diagnosis

- Typical presentation.
- Acute phase reactants esr)is high.
- High ferrettin level.
- Negative ANA.negative Rh factor

Treatment

- Most patients respond to steroids
- Otherwise refferal to rheumatologist for immunosuppressive rx or biological rx.