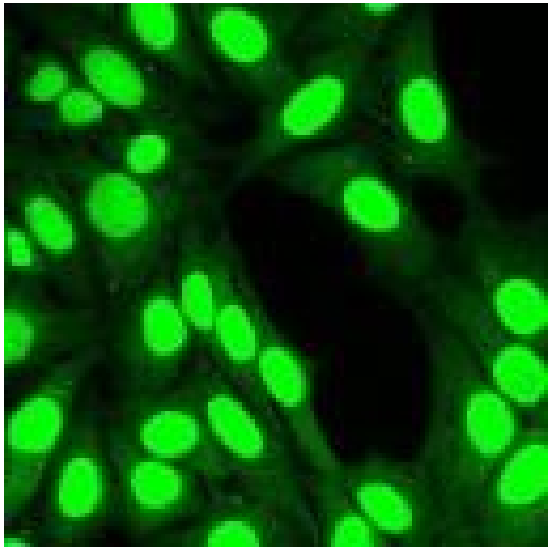
Autoimmune diseases

- Lupus erythematosus
- Scleroderma
- Dermatomyositis
- Overlap syndromes (e.g. Sharp syndrome)
- Vasculitis

Lupus erythematosus

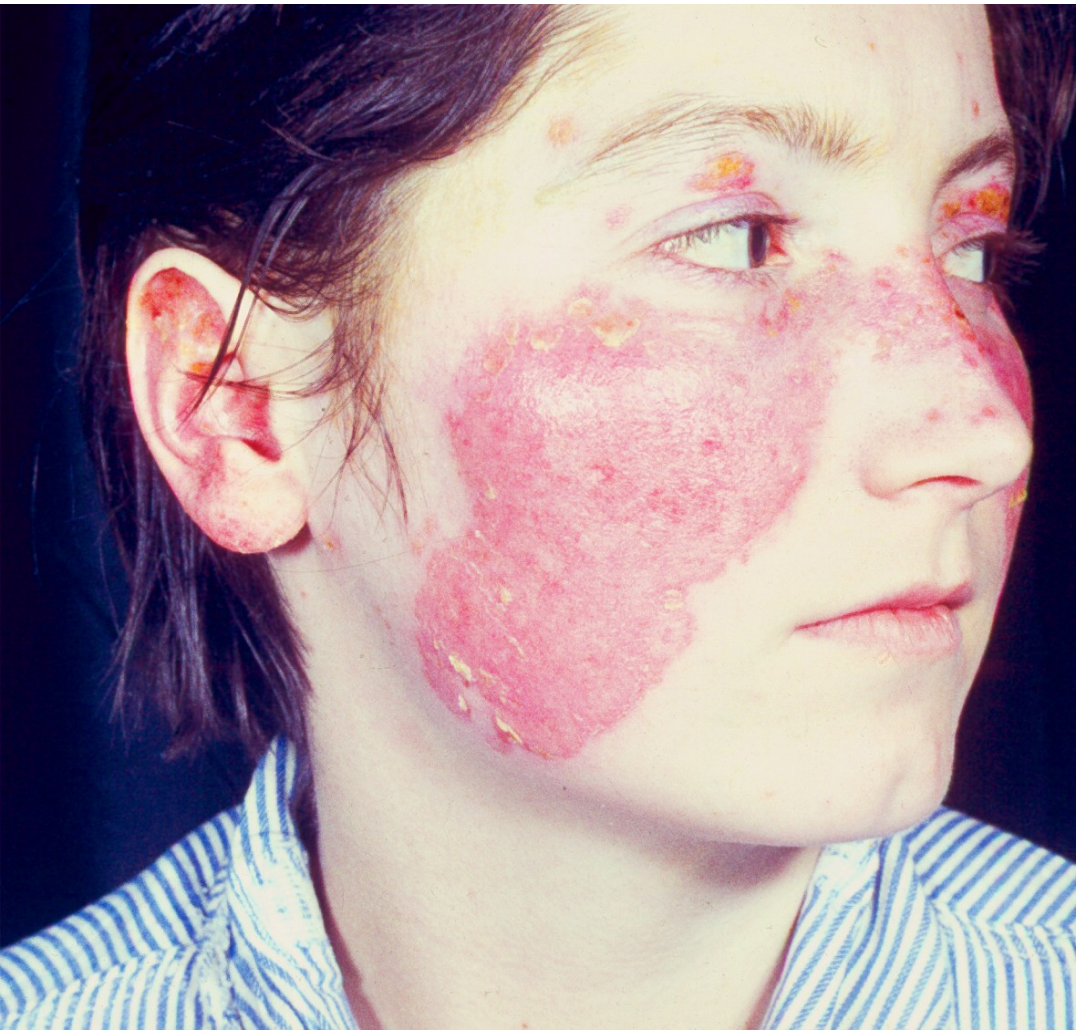
- ***systemic lupus erythematosus*** - SLE
- ***subacute cutaneous LE*** – SCLE
- ***chronic cutaneous LE*** – CCLE
  /chronic discoid LE – CDE/
- *LE in newborns*
- *Medicine induced (hydralazine, sulfonamides)*

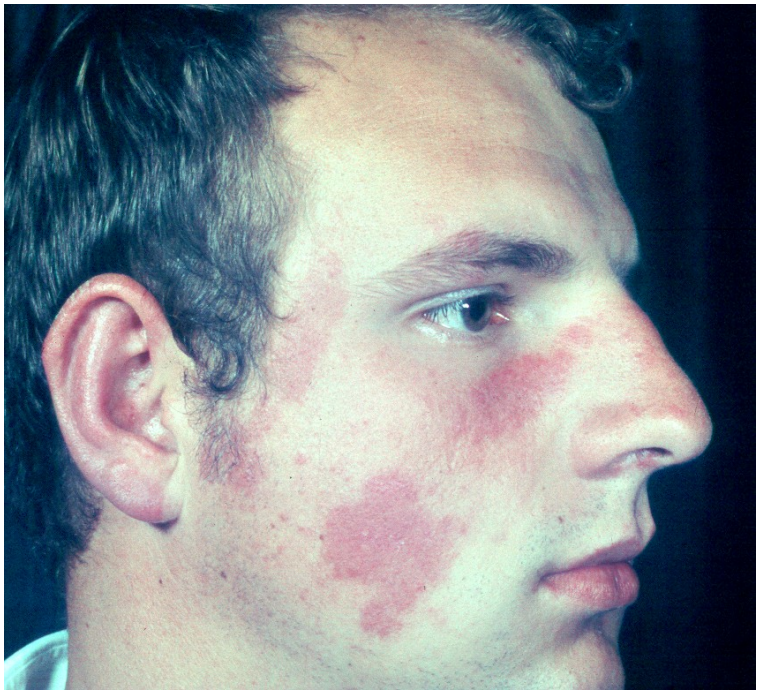
Systemic lupus erythematosus - SLE



- ARA criteria:
- Butterfly rash
- Discoid lesions
- photosensitivity
- Oral lesions
- arthritis
- serositis- pericarditis, pleuritis…
- neurologic diseases
- Kidney involvement (proteinuria 0,5g/d)
- Hematological abnormalities (leukopenia)
- Immunological abnormalities (LE cells,

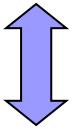
  ANA homogenous, periferal, dsDNA, Sm)



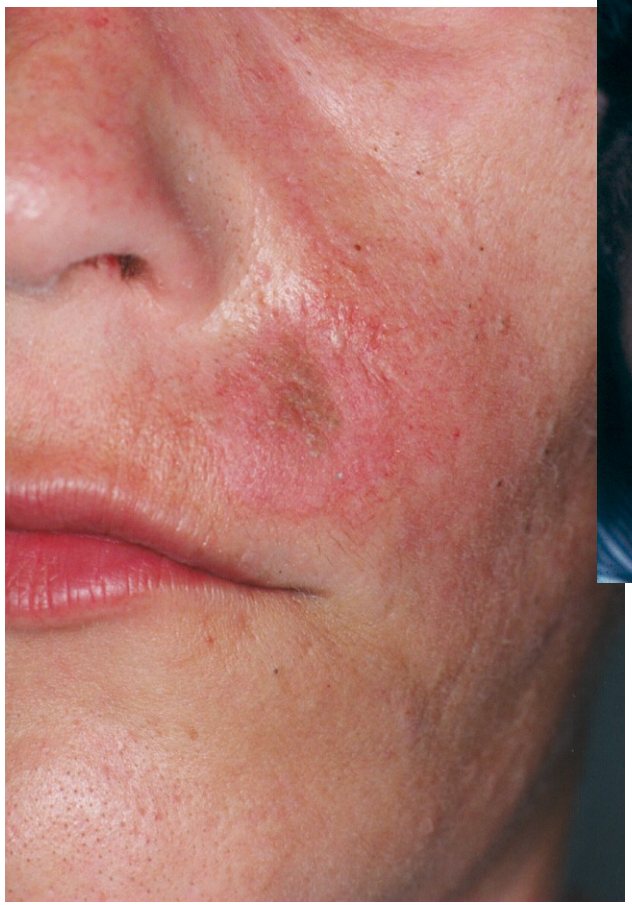


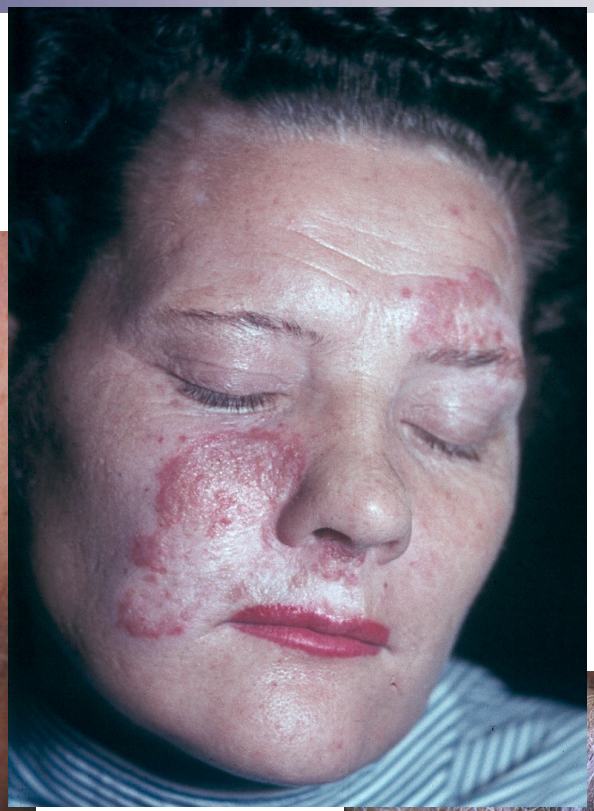
Chronic cutaneous LE - CCLE (CDE)

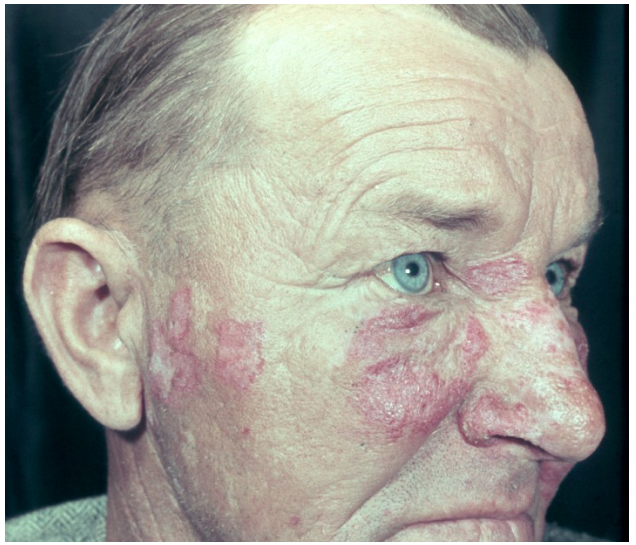
- Photosensitivity
- Discoid lesions - CDE
- Hypertrophic lesions - lupus tumidus
- Lupus paniculitis
- ANA only low titers, granular type
- Not systemic disease

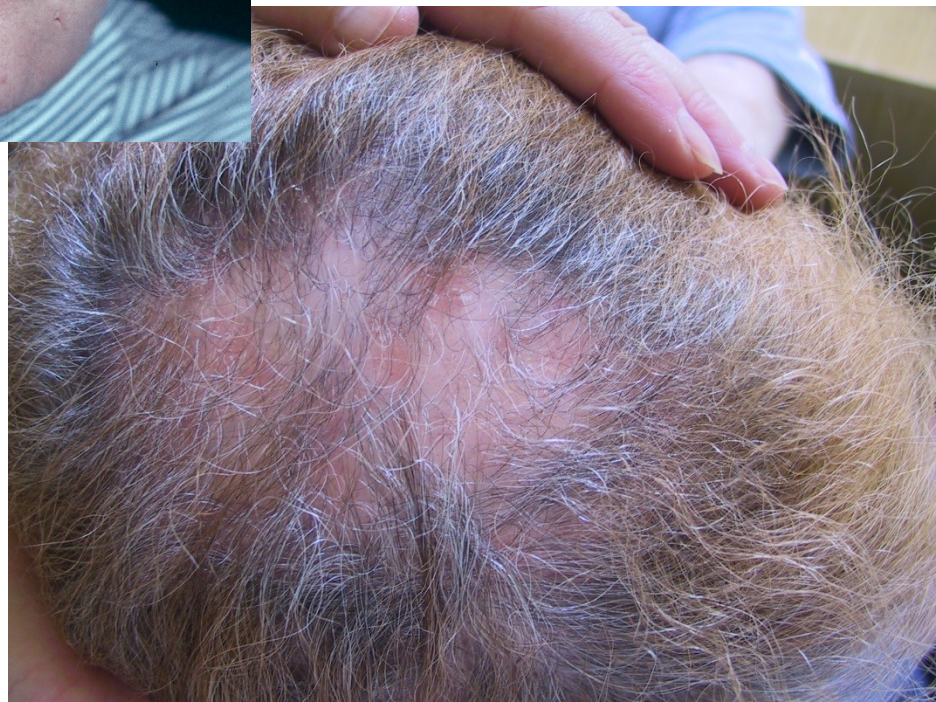


- Symptoms of SLE



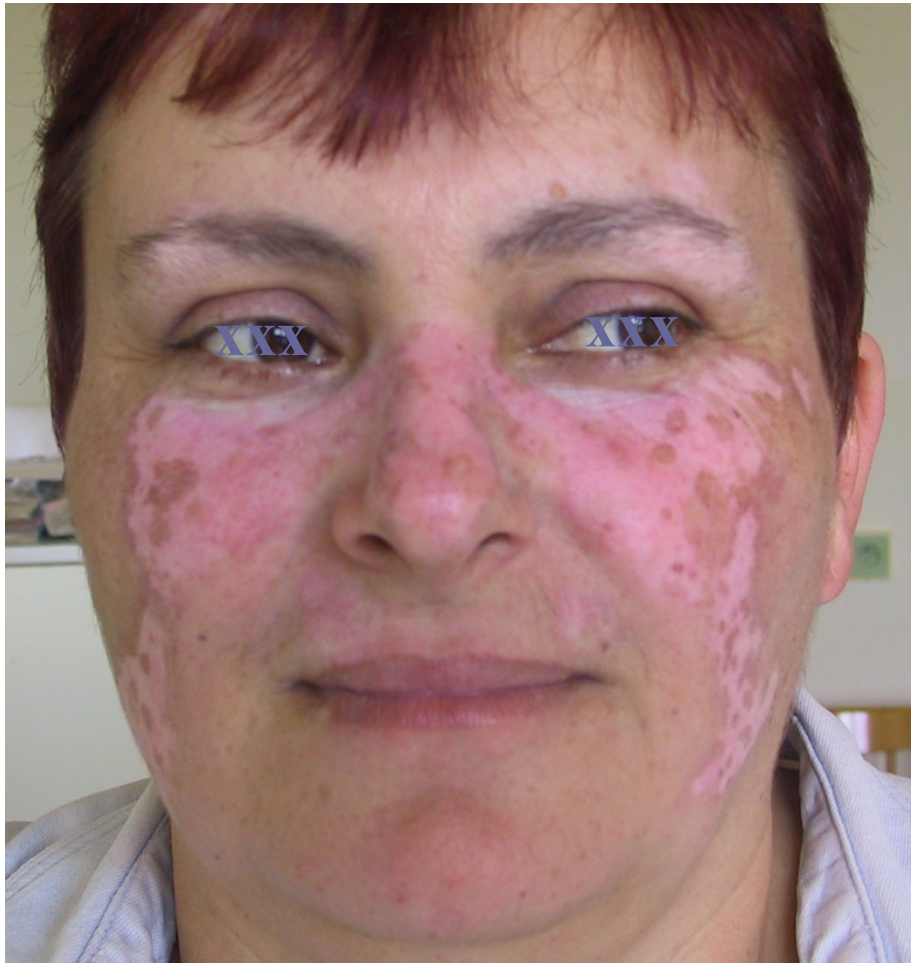


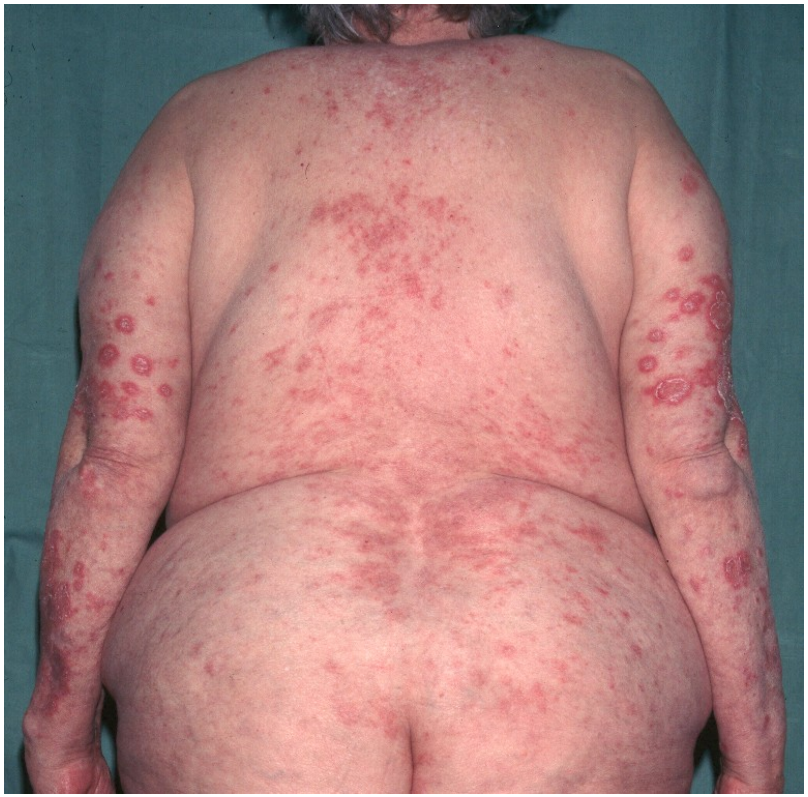




Subacute cutaneous lupus erythematosus - SCLE

- Annular lesions or papulosquamous lesions
- photosensitivity
- ANA, anti Ro/ SSA, La/SSB granular type
- Mild systemic disease (neurologic symptoms, myalgia, arthralgia, rarely nefritis)
- neonatal LE: newborns – mother with + anti SSA: cutaneous symptoms type SCLE a complete AV block, necessity of pacemaker!





Dg: histopathology, direct immunofluorescence (lupus band)

Lupus erythematosus

- UVA, UVB protection !!!
- antimalarials (chloroquine, hydroxychloroquine)

lower absorption of UV radiation

stabilization of lysosomal membranes (antiinflammatory effect)

mild immunosuppressive effect

Adverse effects:

retinopathy

GIT problems, bone marrow depression, hepatotoxicity, rashes

Therapy:

- corticosteroids
- immunosuppressants (cyclophosphamide)
- NSAID (DMARDs)
- thalidomide
- plasmaferesis, puls therapy atd.

Systemic sclerosis

- Disturbance of synthesis of collagen

  autoimmune disease: T ly ->IL-2,3,6 ->

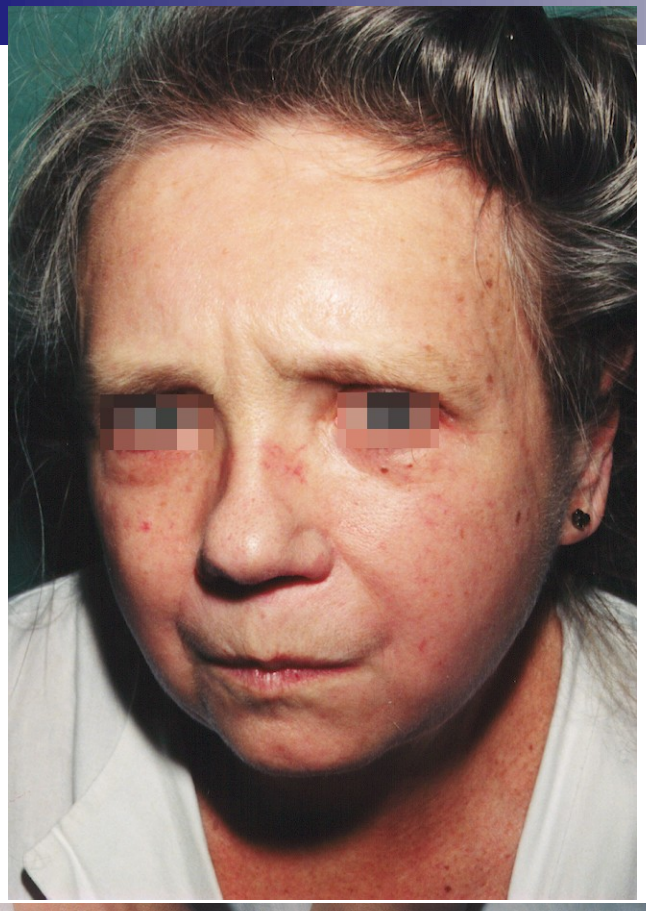
  fibroblasts -> overproduction of collagen V -> fibrosis
- Immunologic abnormalities –

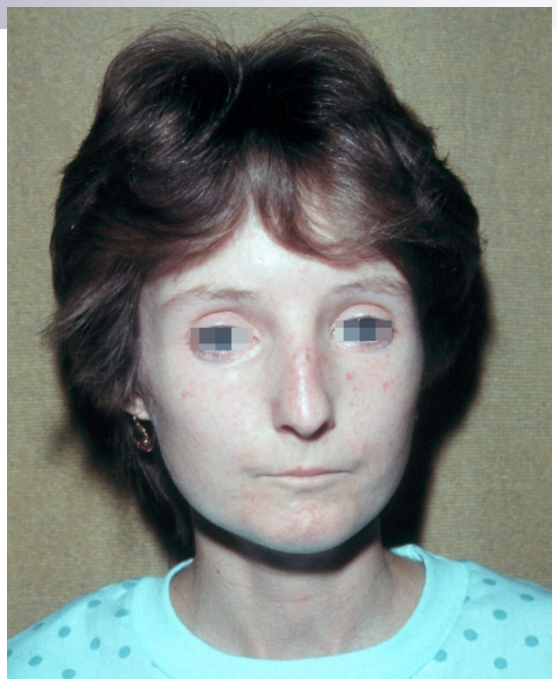
  antibodies (anti Scl70, ANA , anti -centromeras)

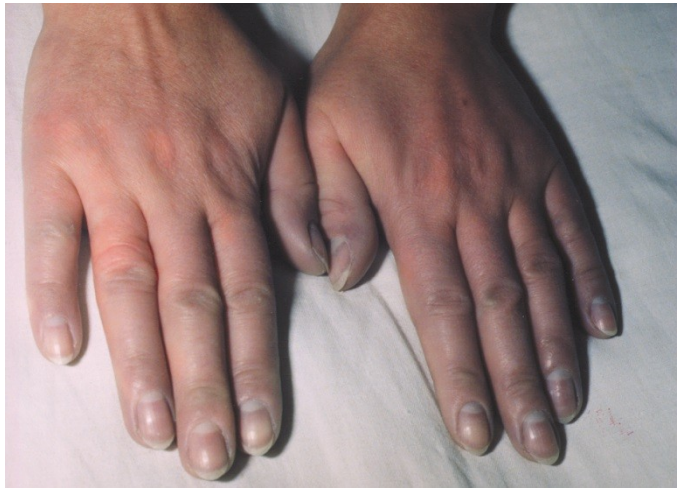
  defect of immune cells
- Vessel changes – involvement of endothelium

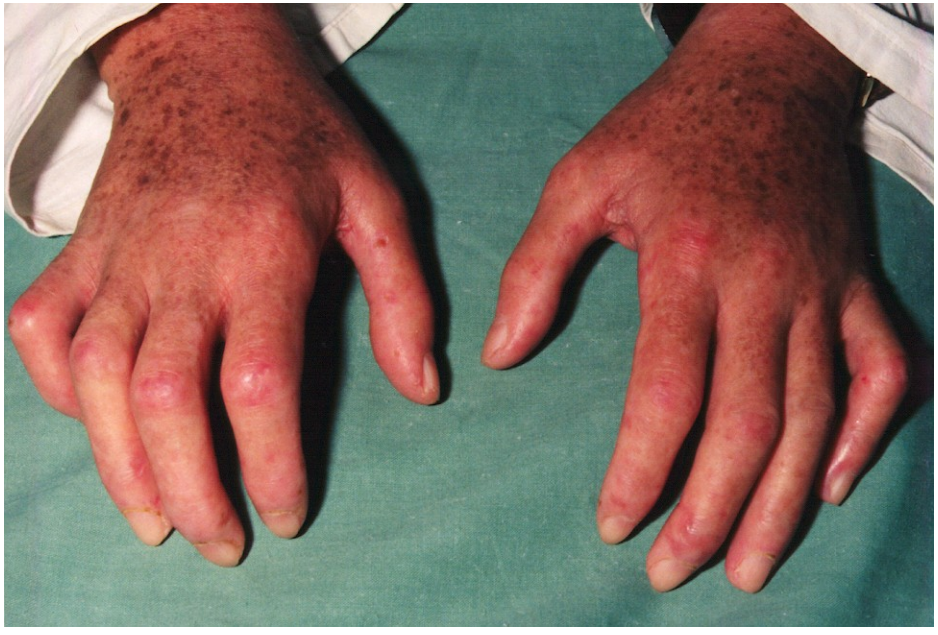
SS – types:

- Limited (acrosclerosis, sclerodactyly)
  Raynaud‘ s phenomenon
  hard skin of fingers, sclerodactyly
  ulcerations on distal digits
  hypomimia, microstomia
- Diffuse
- CREST syndrome









Systemic sclerosis

ARA criteria:

- proximal scleroderma
- Bilateral lung intersticial fibrosis
- Loss of tissue in distal digits (fingertips)
- Sclerodactyly, thickening and swelling of the fingertips

Other signs:

- Raynaud‘s phenomenon
- Esophageal changes
- Renal involvement - nephrosclerosis
- Pulmonary hypertensis
- pericardial effusion, myocardial fibrosis

CREST syndrome

- C alcinosis
- R aynaud‘s phenomenon
- E sophageal changes
- S cleroderma
- T eleangiectasias
- anti centromere antibody

Scleroderma

Therapy:

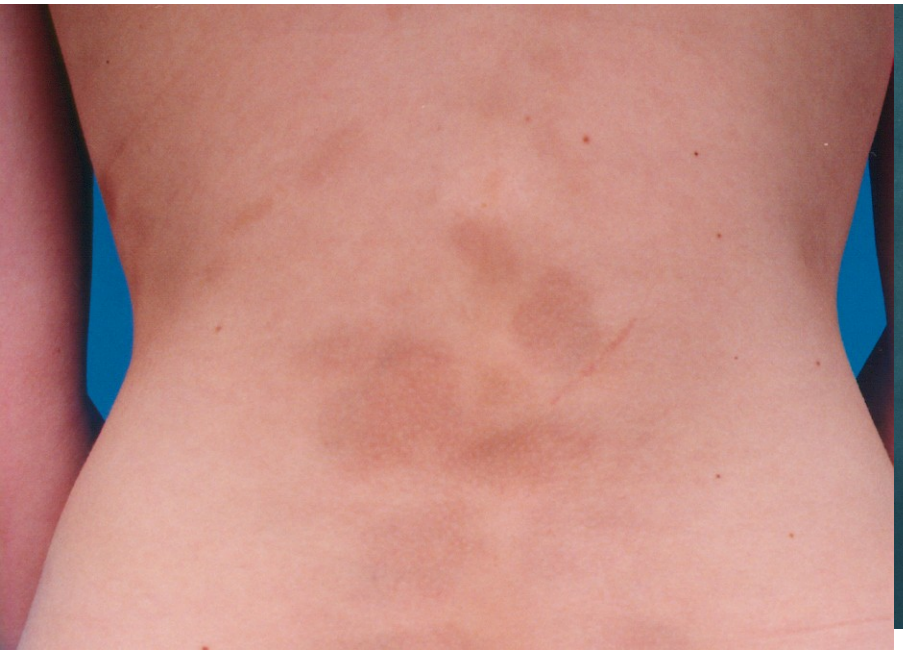
- corticosteroids
- immunosuppressants /Aza, CF, MTX/
- d-penicilamine
  - lower synthesis of collagen
  - but: frequent adverse effects!
- PUVA, UVA-1, bath PUVA
- IFN γ – lower synthesis of collagen

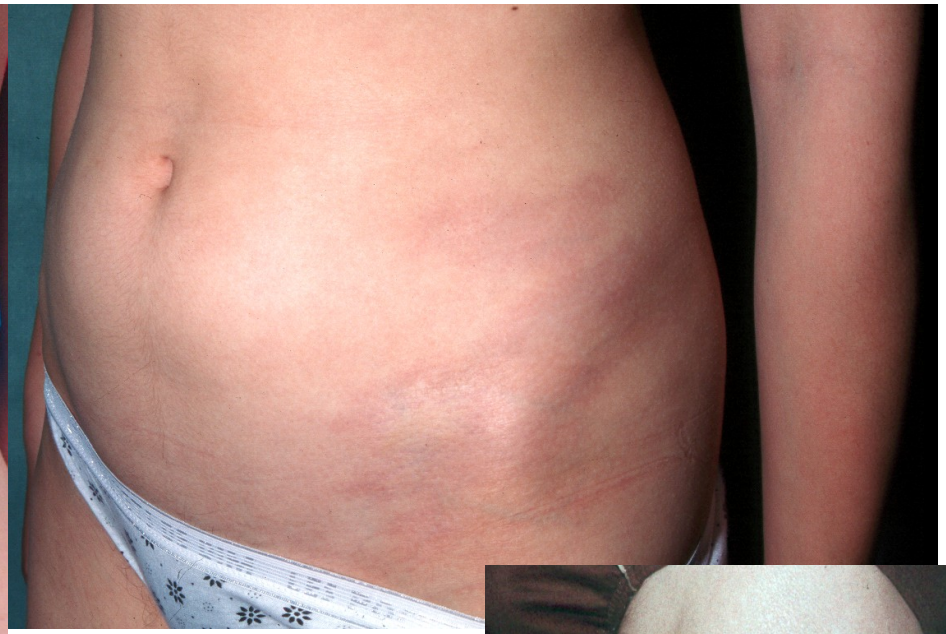
- vasoactive and rheologic medication (pentoxiphylline, prostaglandins)
- calcium antagonists – nifedipin

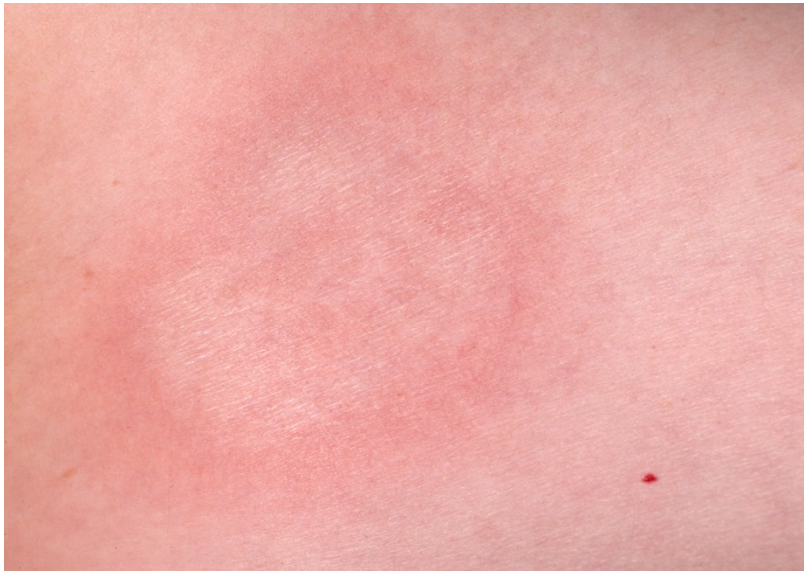
- prokinetics, antacids
- physical therapy

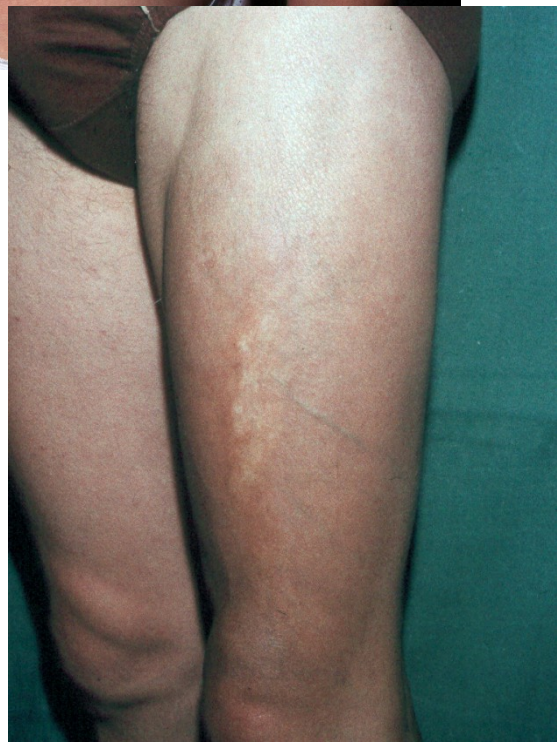
Localised scleroderma

- Morphea /isolated patches of hardened skin/
- guttate
- linear
- generalised (pansclerotic morphea)
- subcutaneous – eosinophilic fasciitis
- ANA – only low titers
- No systemic involvement









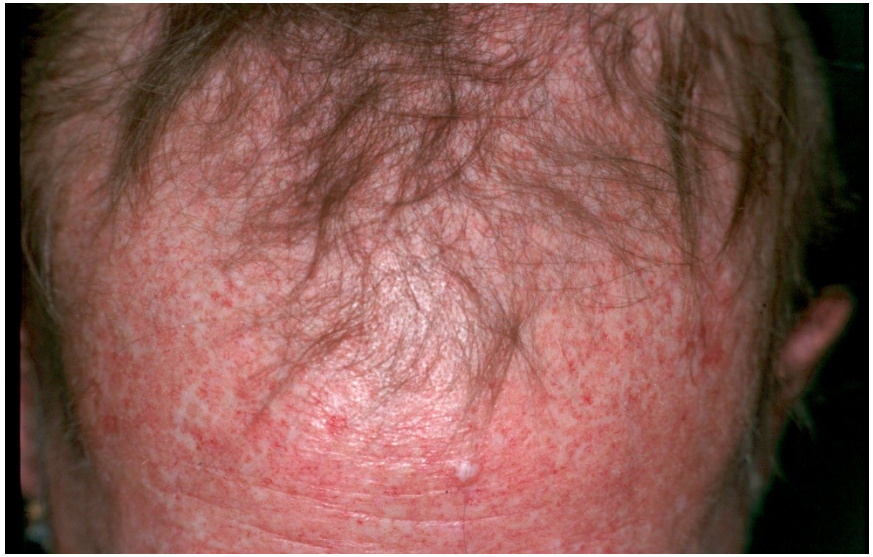
Localised scleroderma

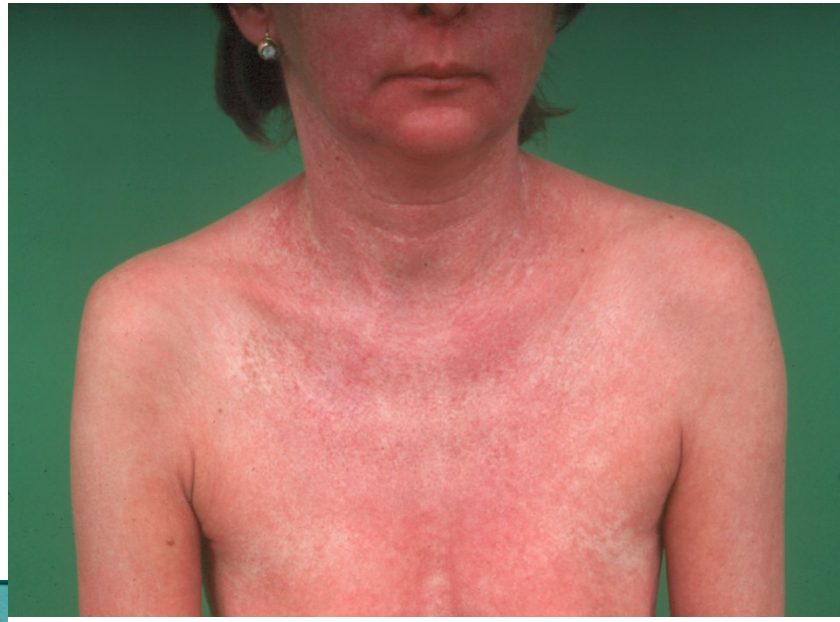
Therapy:

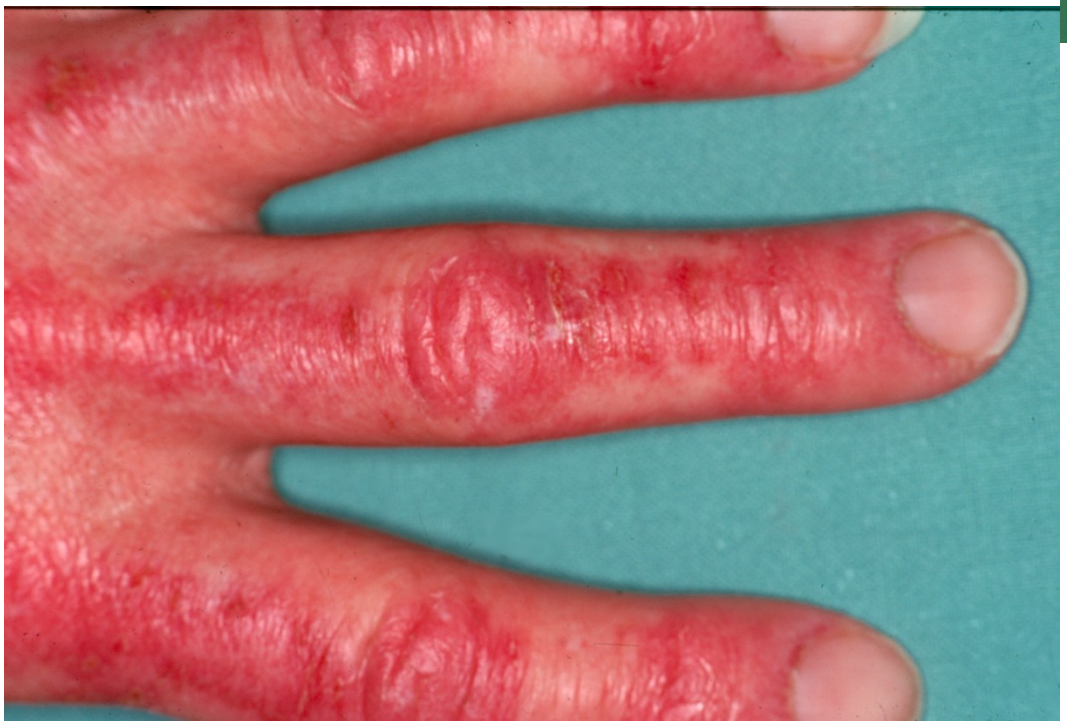
- Local CS, masage with Heparoid ung
- Analogs of vit. D3 - calcipotriol
- Local PUVA
- P-PNC, penicilamine
- Antimalarials
- PUVA
- retinoids

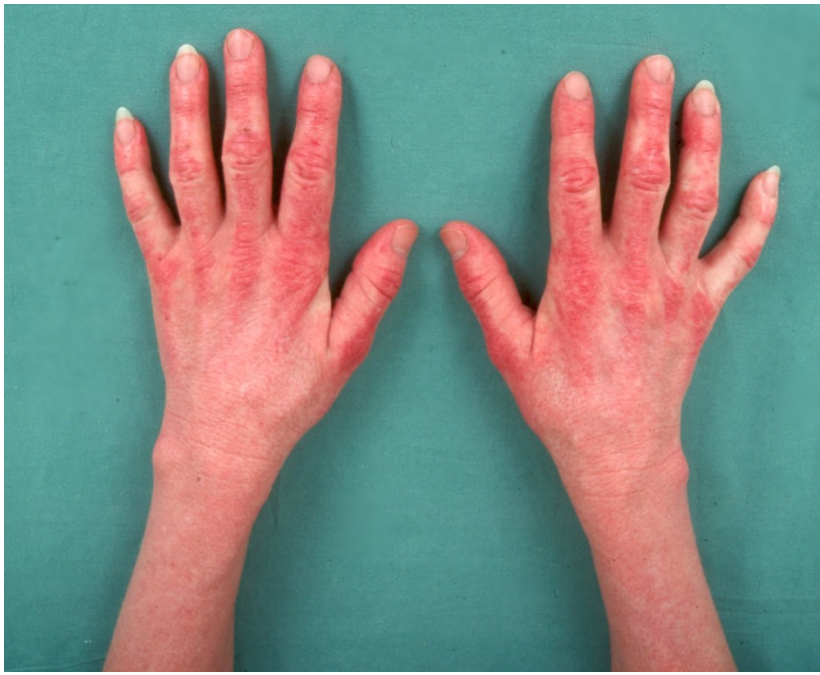
Dermatomyositis

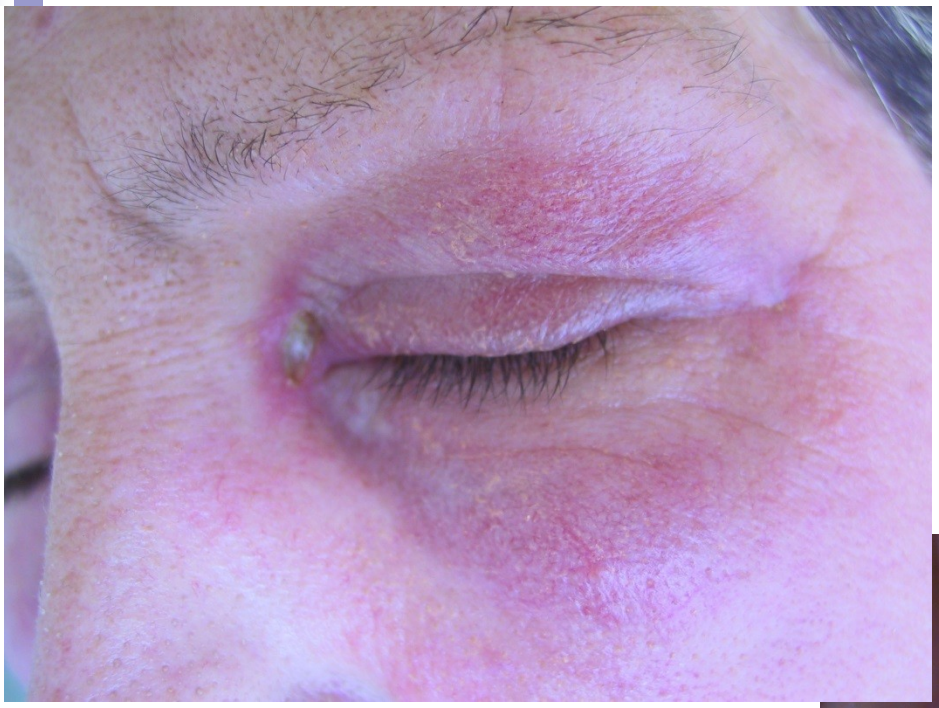
- **Juvenile type –** association with infections
- **Adult type - ** association with tumours (paraneoplasia)
- heliotropic rash, Gottron‘s sign, poikilodermatitis, erytematous lesions
- EMG
- ANA, anti Jo-1 / histidyl –t-RNA synthetase/ PM>DM
  anti Mi-2 / nucleoproteins 30-240 kDa/ DM
- CK, LDH, GGT, ALD, AST, ALT, myoglobin
- Histopathology – muscle, skin

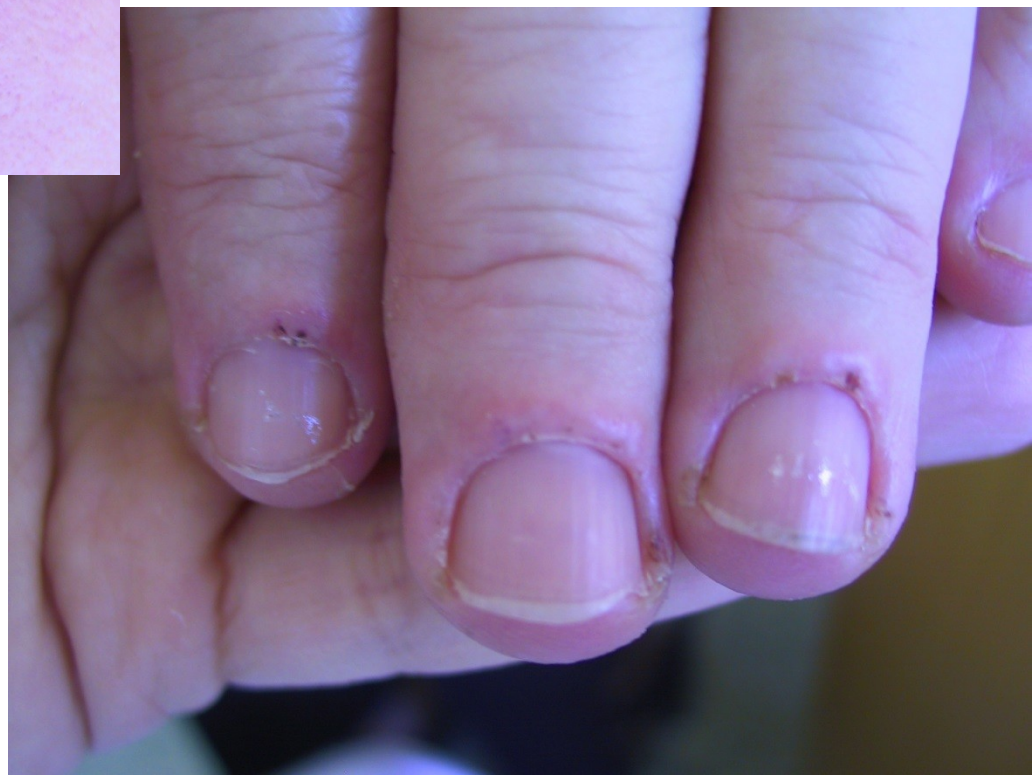












Dermatomyositis

- criteria ARA: 1) symetric proximal muscle weakness
  2) elevated levels of CK, LD or myoglobin
  3) emg signs of myopathy
  4) muscle biopsy with features of inflammatory myopathy
  5) typical skin lesions of DM

- definite dg DM : at least 3 from the first, 4 + 5
- supposed: at least 2 from the first, 4 + 5
- autoAb: ANA posit in 50%,
  others:: anti Mi-2 /nucleoproteins 30-240 kDa / 10-30%
  anti Jo-1 / histidyl-tRNA syntetase/ 15-40%

Dermatomyositis - therapy

- Corticosteroids - inicial and long term dose
- Immunosuppressants - MTX, azathioprine, cyclosporine
- Antimalarials – only in skin involvement

- IVIG
- rituximab

Overlap syndromes

- MCTD – Sharp´s syndrom

  S Scl, PM/DM sometimes RA
- Scleromyositis SScl/PM
- SScl/SLE

- SScl/RA
- SLE/PM