SYSTEMIC SCLEROSIS

Lidia Rudnicka

Scleroderma

LIMITED
morphea

SYSTEMIC
(systemic sclerosis)

limited

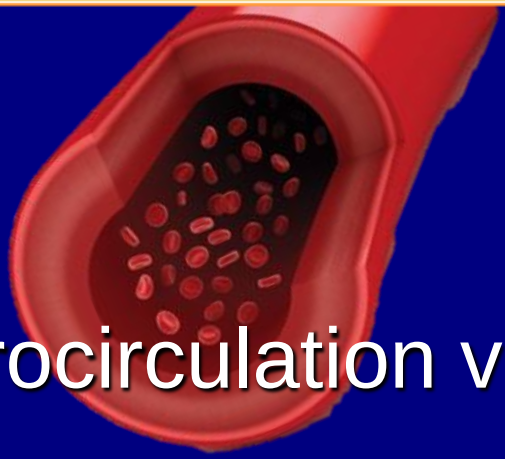
diffuse

Scleroderma

An autoimmune systemic disease occurring with the fibrosis of the skin and internal organs

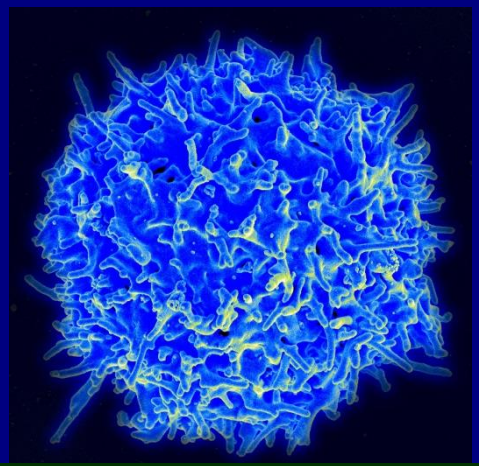
more common in women 9 : 1
onset: 20-40 years

SSc - pathogenesis



Microcirculation vessels

Fibroblasts



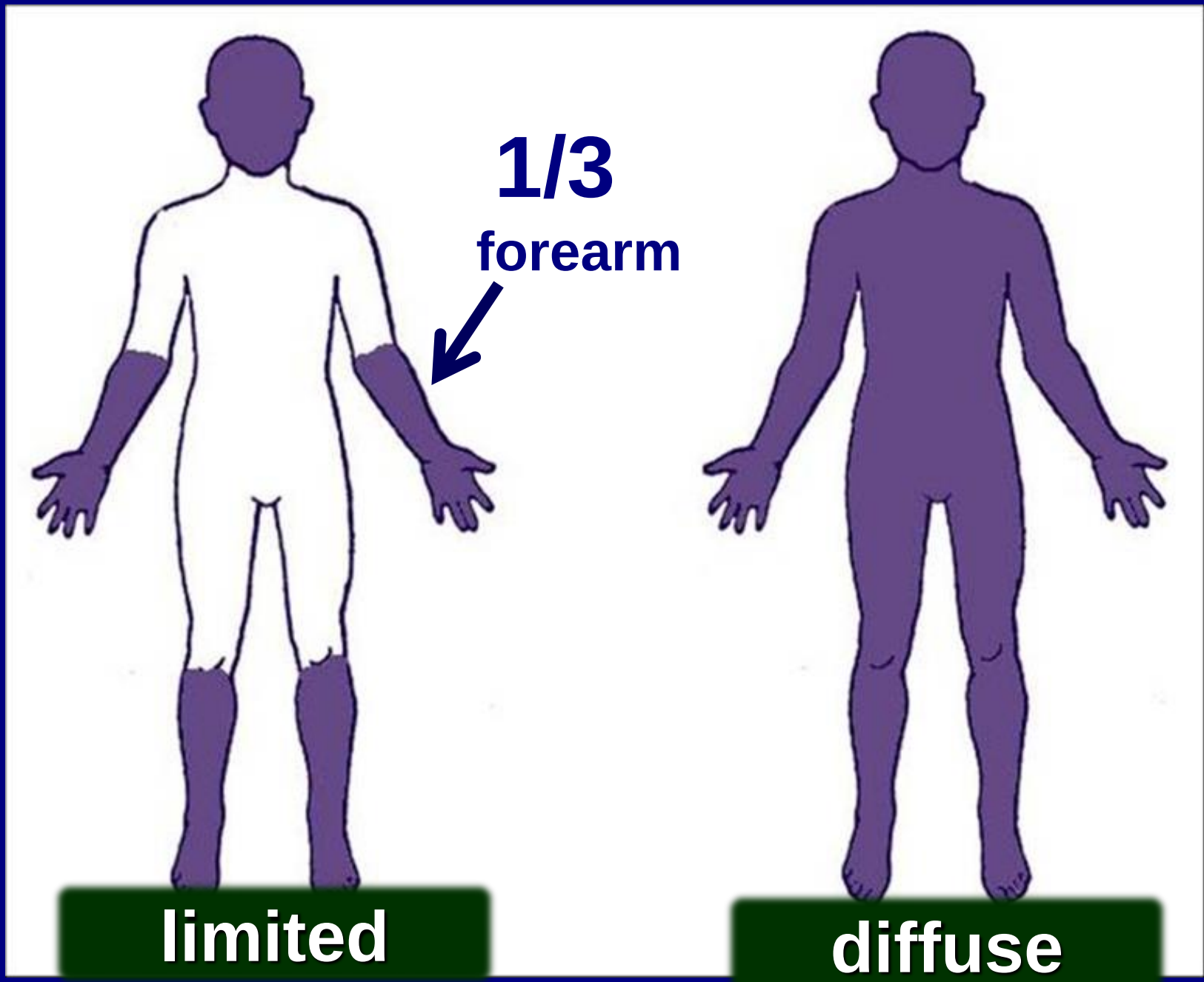
T and B lymphocytes

SSc – the most common manifestations

- Cutaneous hardening progressing inward (100%)
- Raynaud's phenomenon (96% of patients)
- Pulmonary fibrosis (approx. 60%)
- Dysphagia (approx. 60%)
- Arthralgia (approx. 30%)
- Renal involvement / scleroderma renal crisis
- Other organs

SSc - cutaneous manifestations

- finger hardening (sclerodactyly)
- peripheral ischemia
- mask-like face
- atrophy of alae nasi
- atrophy of rubor labiorum
- creases around lips (radiating perioral wrinkles)
- difficulty opening the mouth
- shortened frenulum of tongue


1/3
forearm
limited
diffuse

SSc - fingers

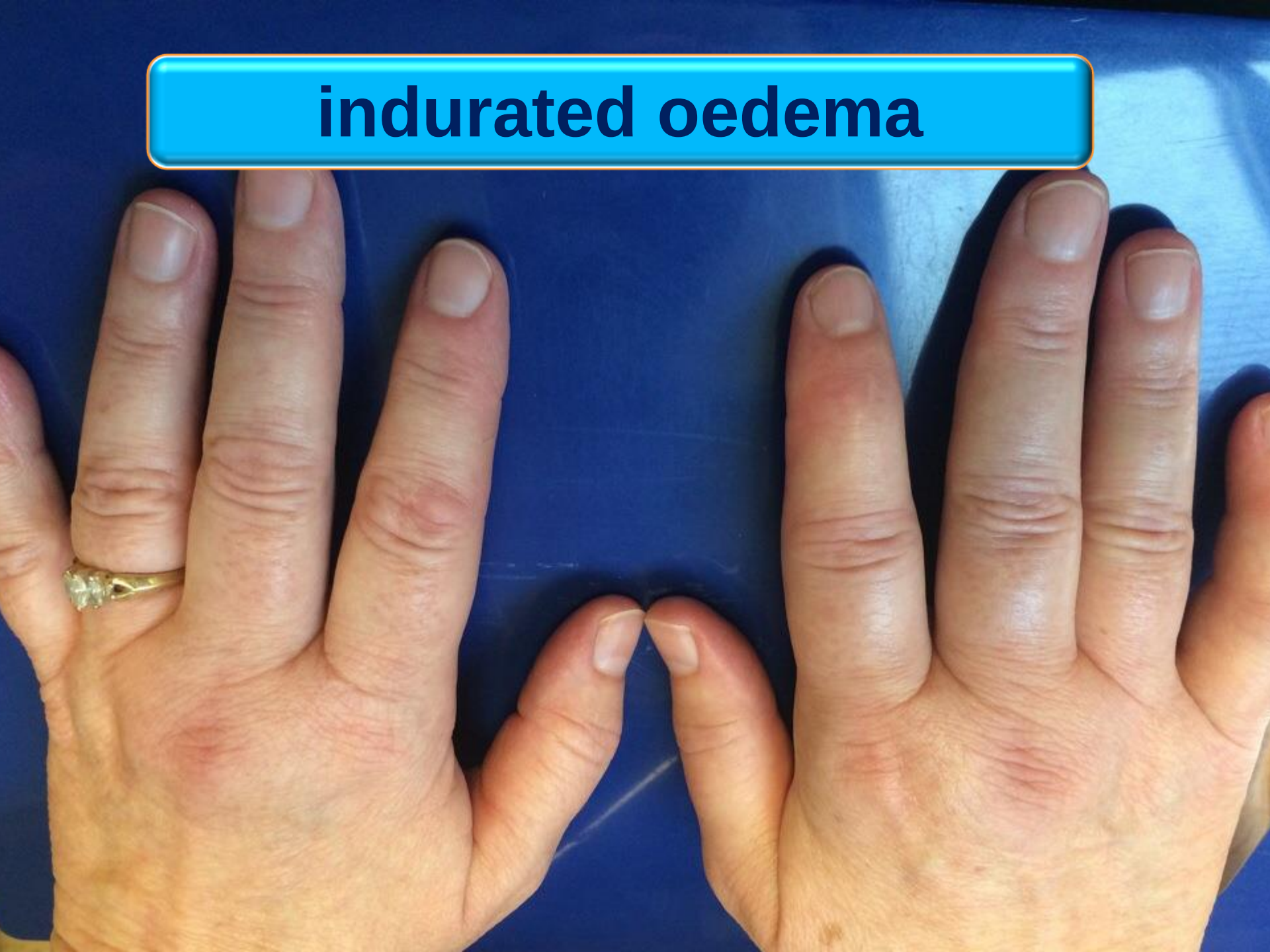
indurated oedema

induration and atrophy

erosion / deformity

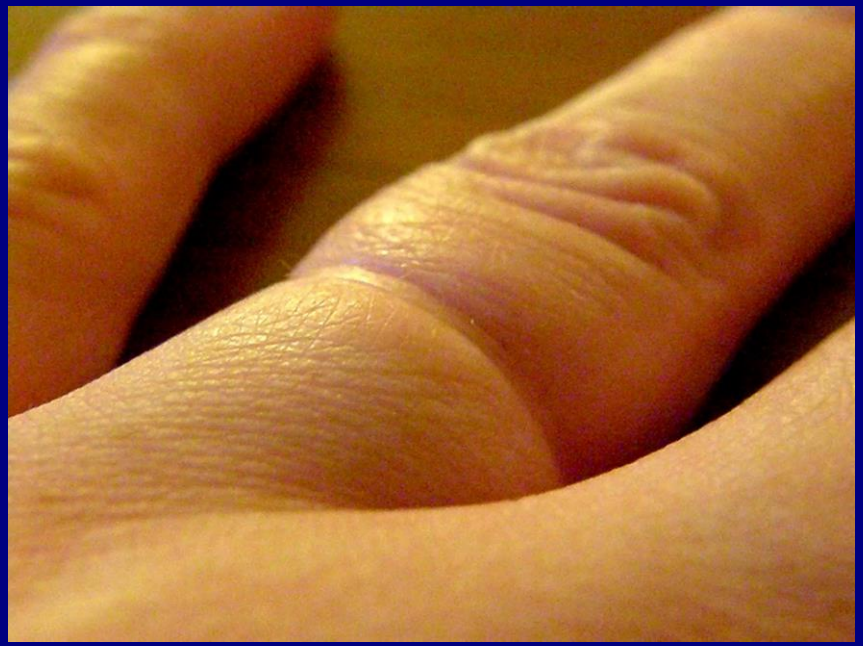
autoamputation

indurated oedema




indurated oedema

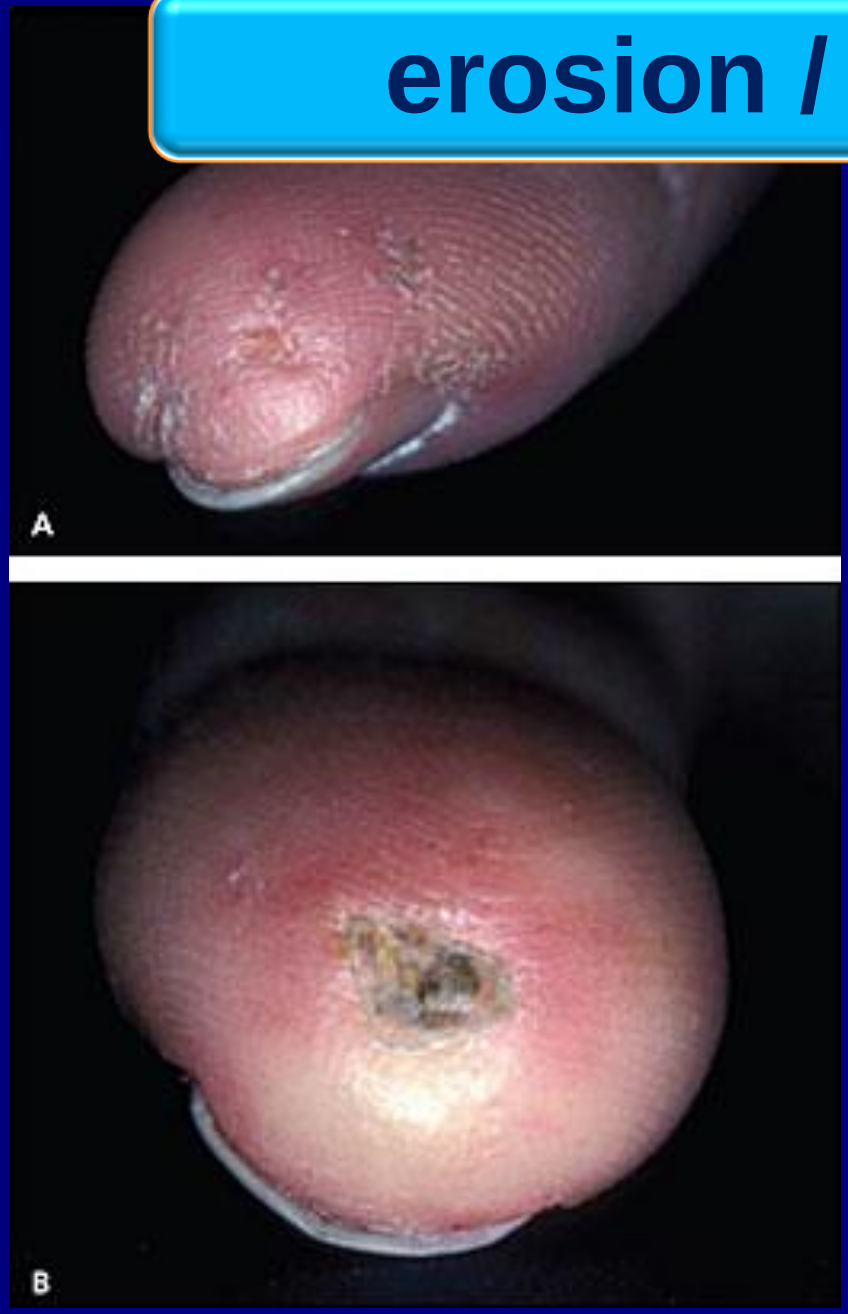
indurated oedema

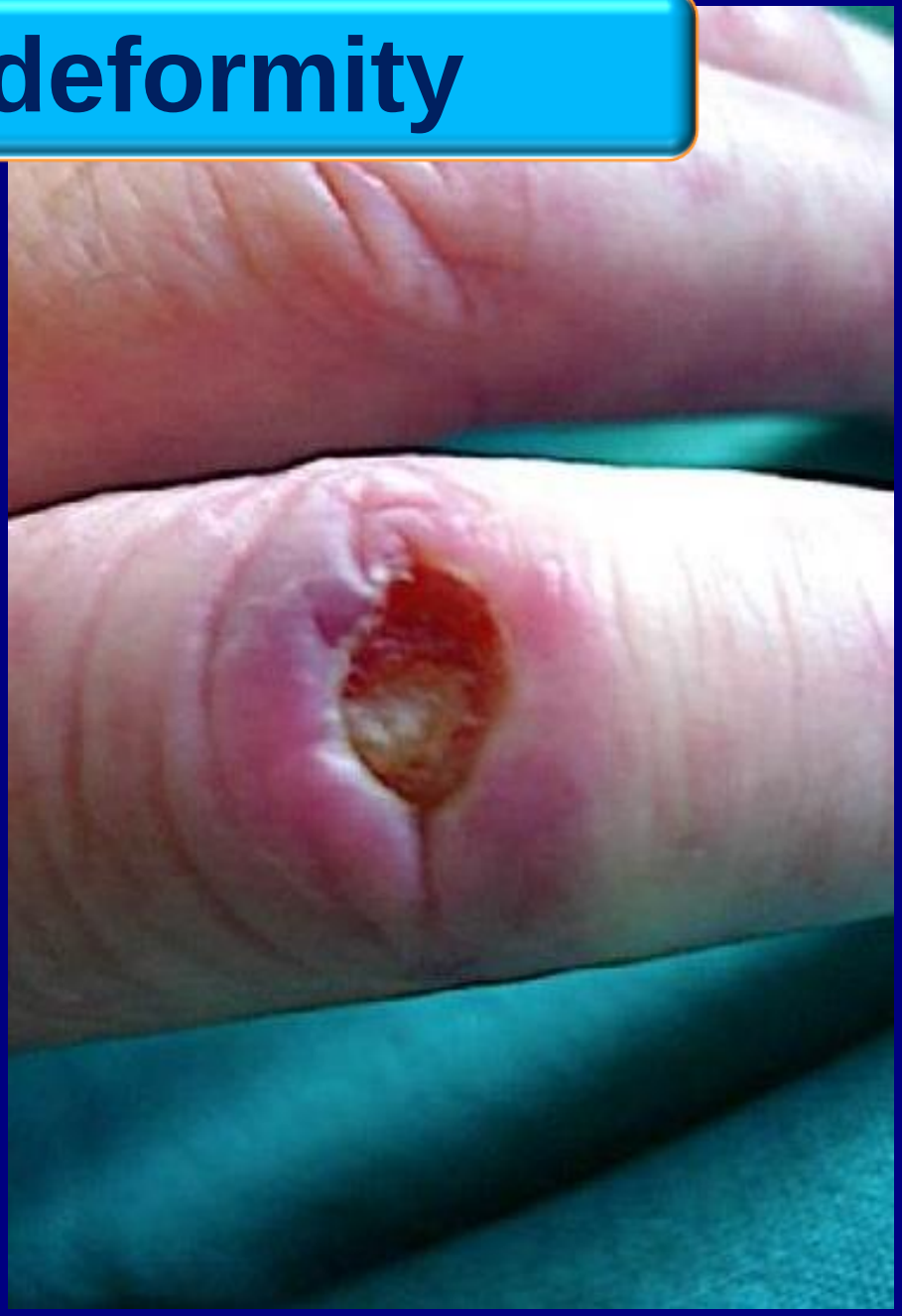



induration and atrophy


induration and atrophy

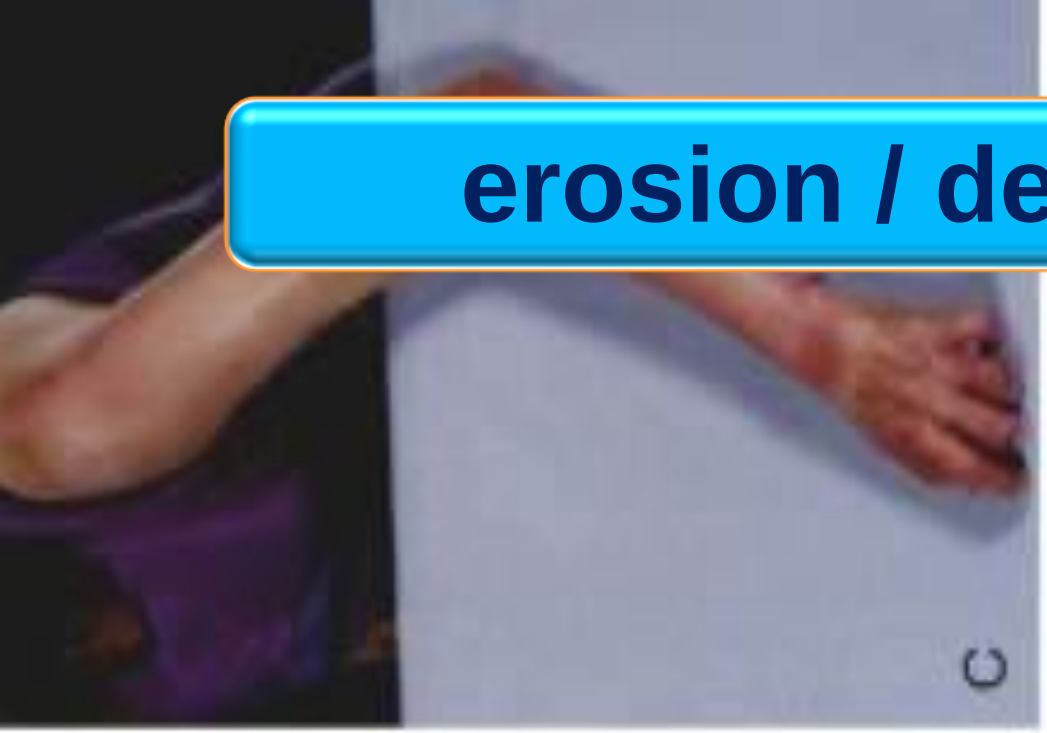
erosion / deformity

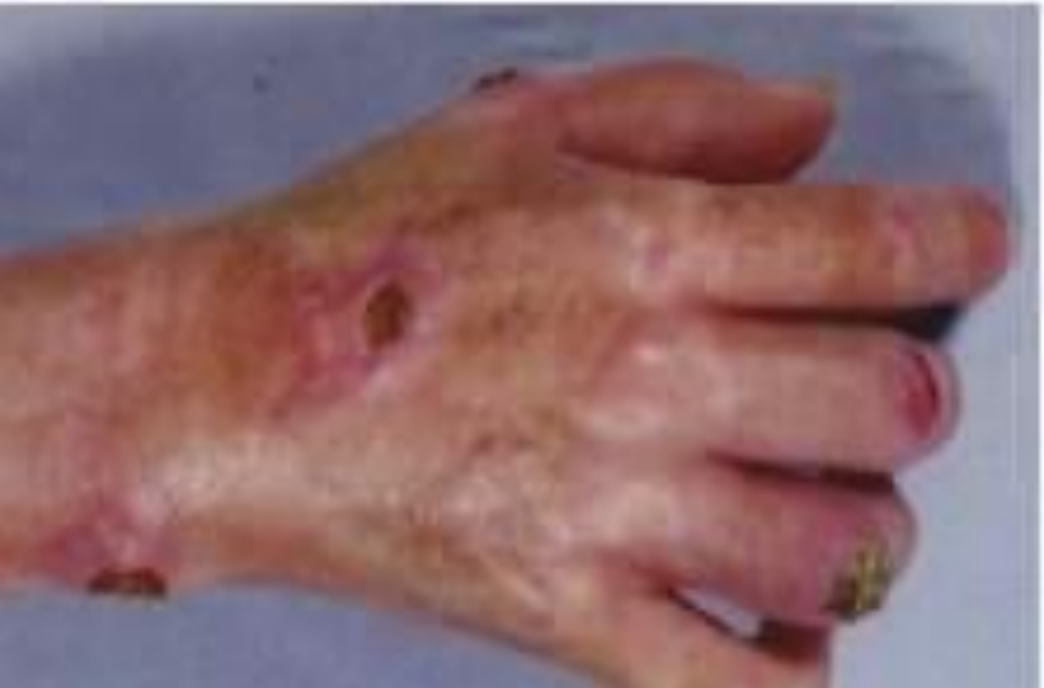


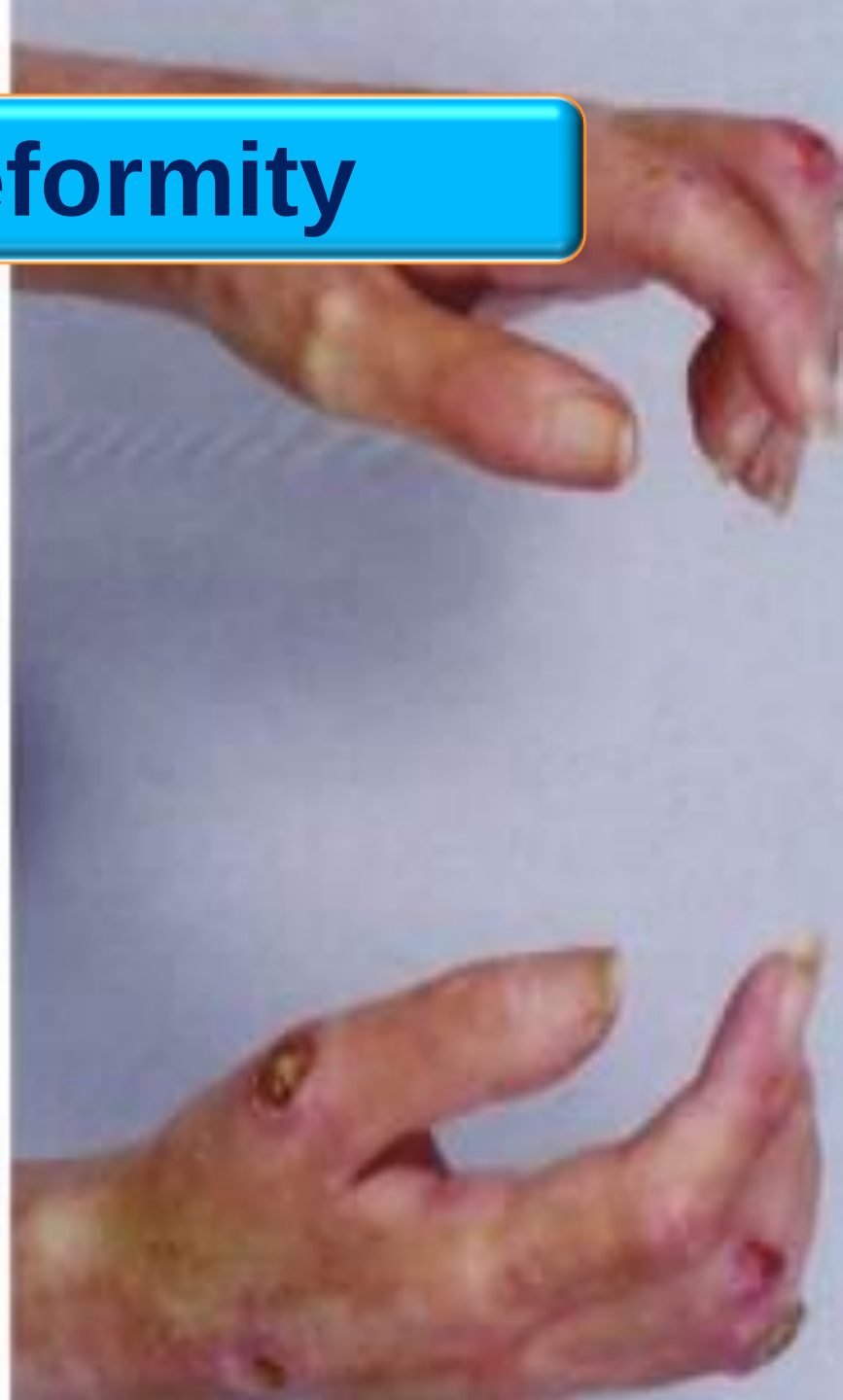



erosion / deformity

erosion / deformity

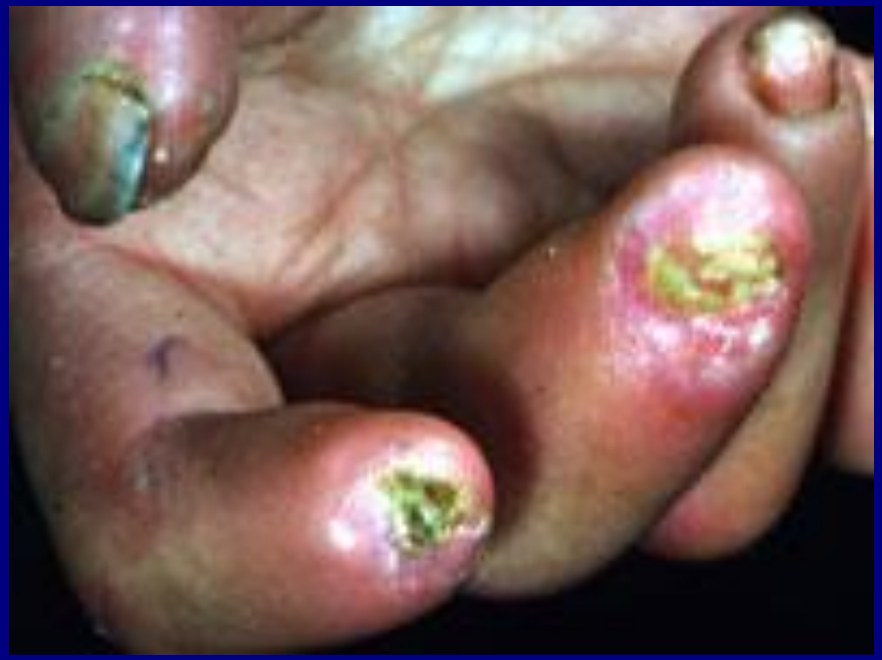


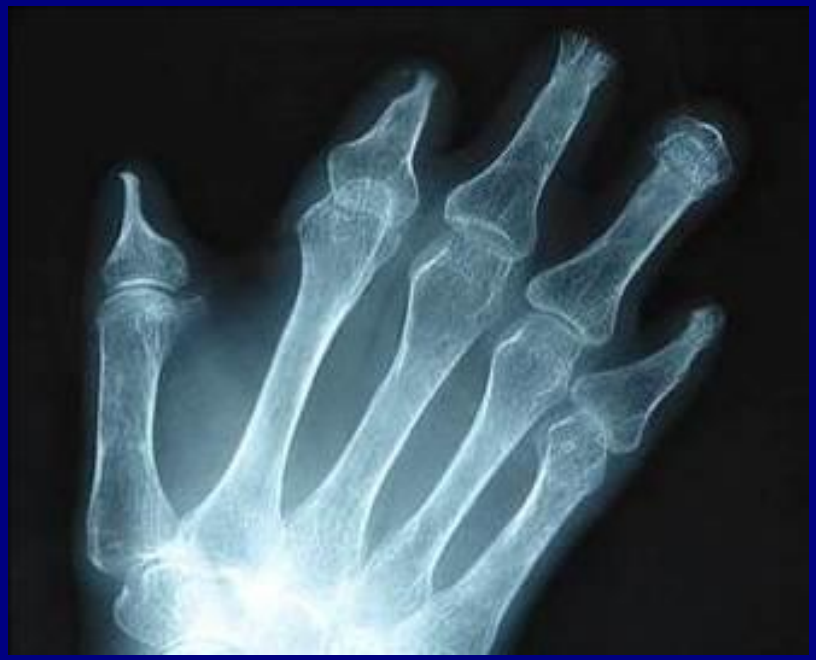




Necrosis / autoamputation

autoamputation





Maria Rivelli

SCLERODERMA FOUNDATION

SUPPORT · EDUCATION · RESEARCH


massive skin indurations

CREST syndrome

a specific subtype of *limited systemic sclerosis*

Calcinosis

Raynaud

Esophagus

Sclerodactyly

Telangiectasia

Immunological markers

- **ANA**: 95% of patient**s**
- Anti-DNA topoisomerase I antibodies (**Scl 70**) – a marker of a more severe course (dSSc)
- Anti-centromere antibodies (**ACA**) – a marker of a less severe course (ISSc)
- **Anti-RNA polymerase-III** antibodies – most commonly in dSSc with renal involvement
- **Anti-fibrillarin** antibodies (anti-U3RNP) – more commonly with internal organ involvement

Immunological markers - overlap syndromes

- **Anti-PM-Scl** antibodies – the marker of systemic sclerosis overlaping with dermatomyositis = scleromyositis
- **Anti-U1RNP** antibodies - the marker of MCTD (mixed connective tissue disease)

2013 ACR/EULAR Criteria for the Classification of Systemic Sclerosis

- Skin hardening on the hands (9 points)
- Sclerodactyly (2-4)
- Erosive lesions/ulceration of fingers (2-3)
- Telangiectasia (2)
- Abnormal capillaroscopy (2)
- Pulmonary fibrosis / PAH (2)
- Raynaud's phenomenon (3)
- SSc immunological markers (3)

≥9

Systemic sclerosis - treatment

- Vasodilators
  - sildenafil
  - alprostadil
  - nifedipine
- Immunosuppressive drugs
  - cyclophosphamide
  - mycophenolate mofetil
  - methotrexate

The main aim of treatment: to stop the progression of the disease

Prognosis

5-year survival rate

- 1950: 50%
- Currently: > 90%

20-year survival rate

- Currently: > 80%

Pons-Estel et al. 2009

Scleroderma ?

SYSTEMIC SCLEROSIS

Profound skin involvement + internal organs

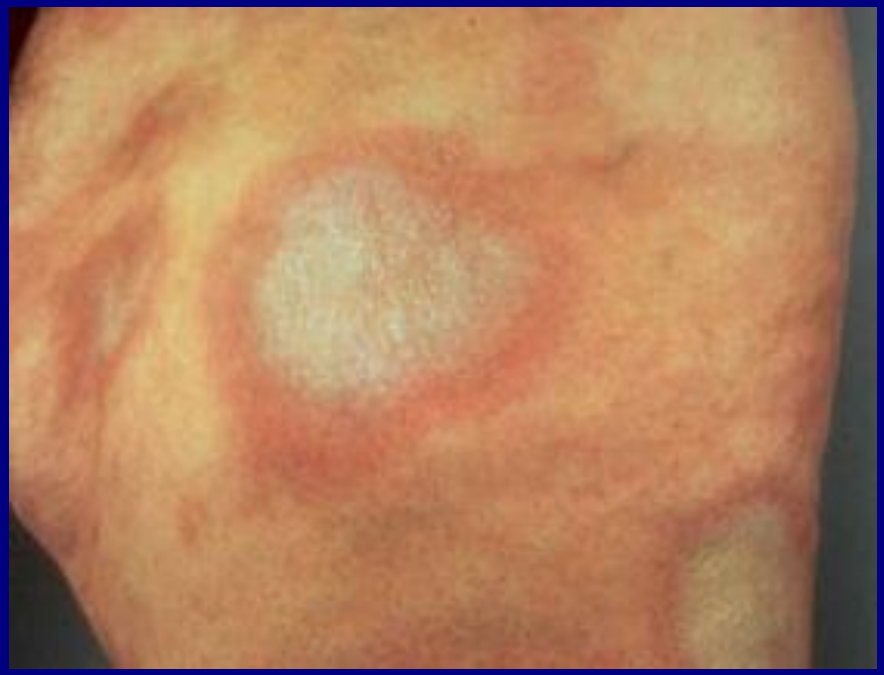
MORPHEA

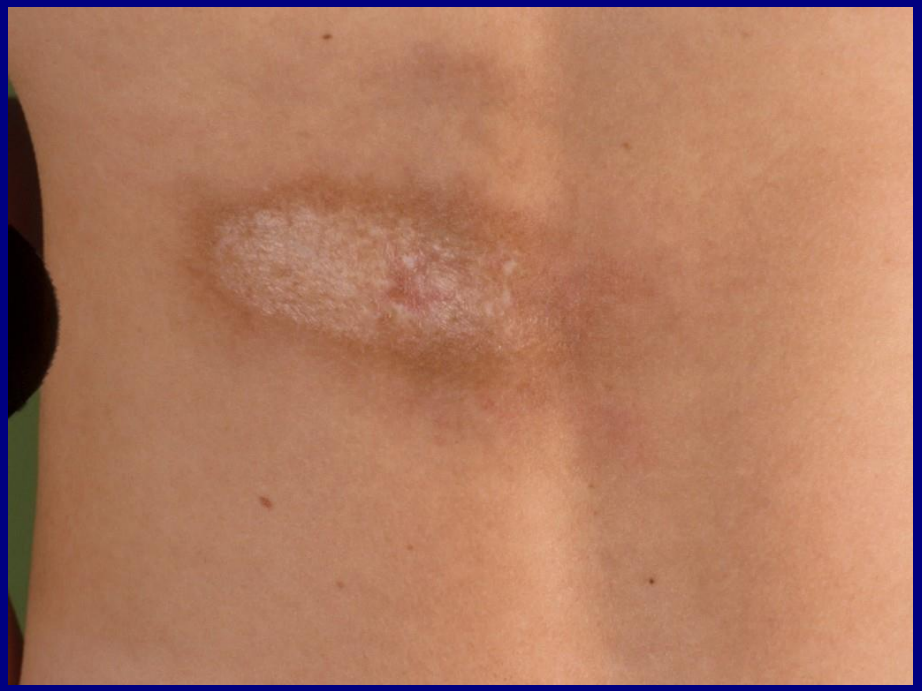
Skin only

Morphea - clinical presentation

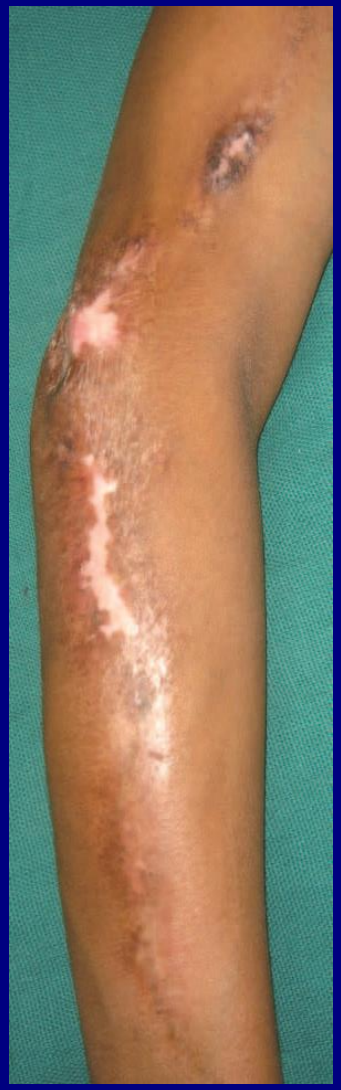
- plaque
- generalized
- linear
- *en coup de sabre*
- facial hemiatrophy *(hemiatrophia faciei)*

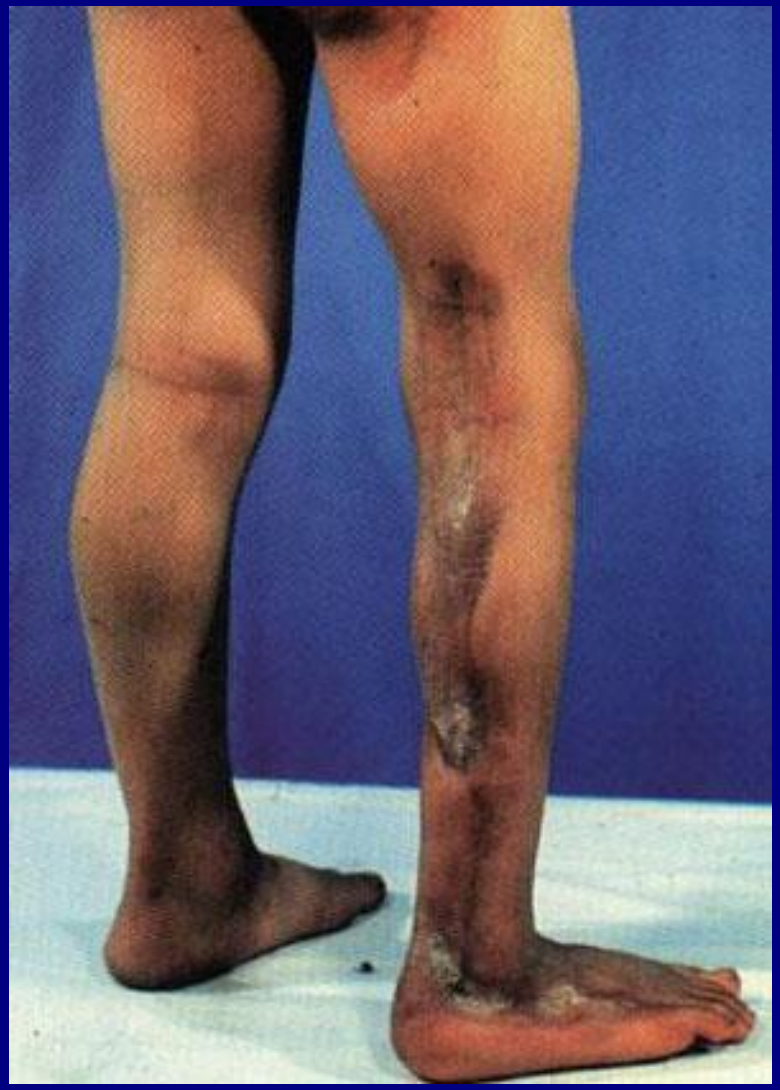
Morphea – plaque type

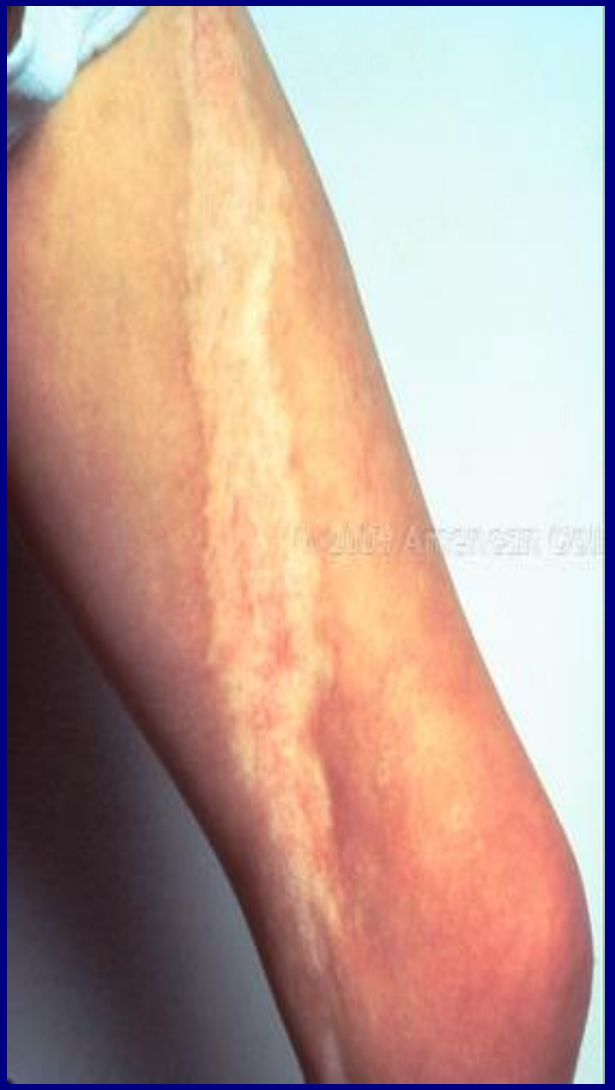




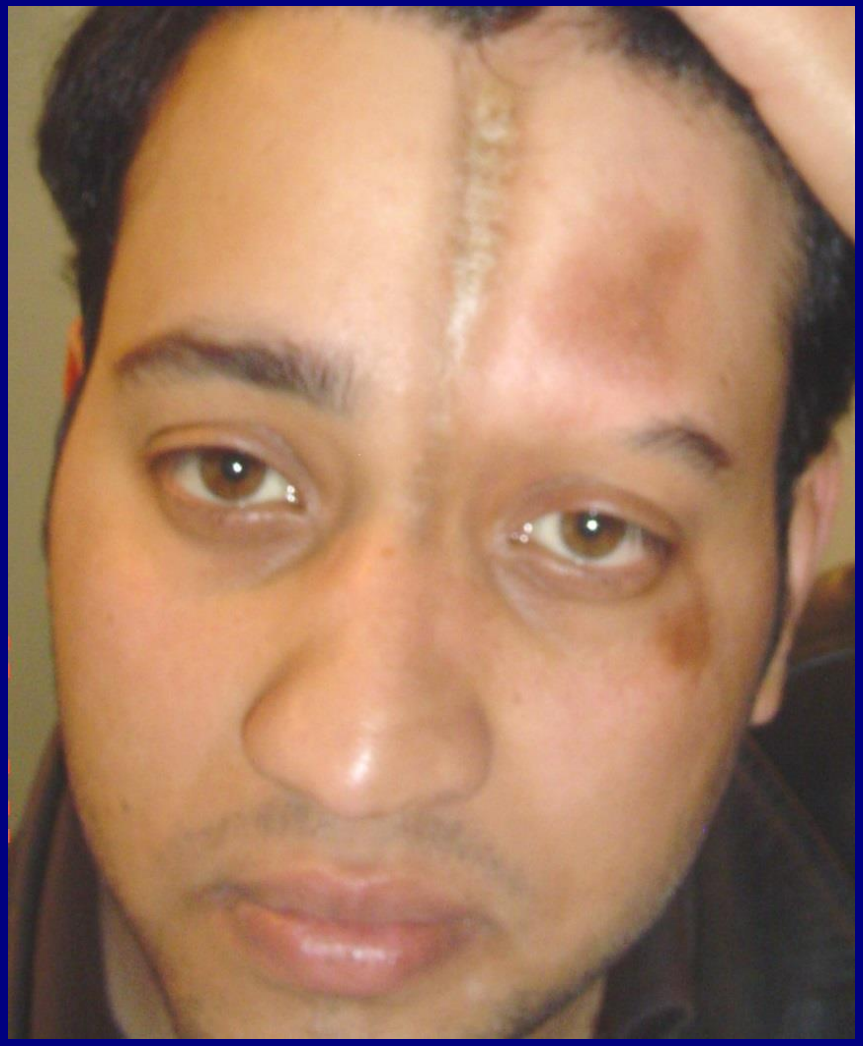
Morphea - linear

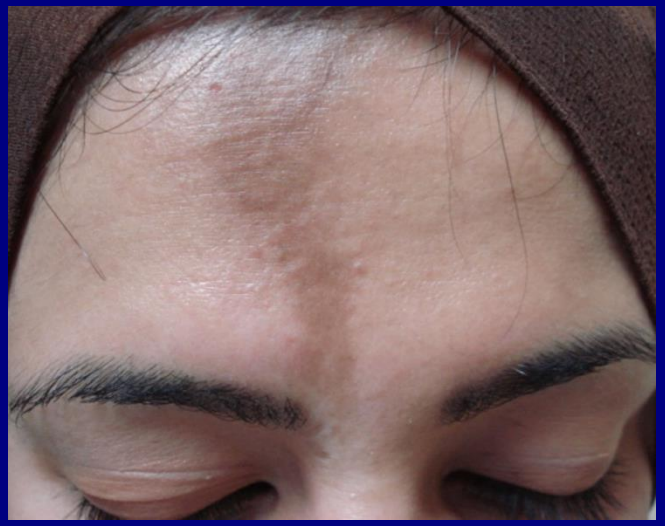




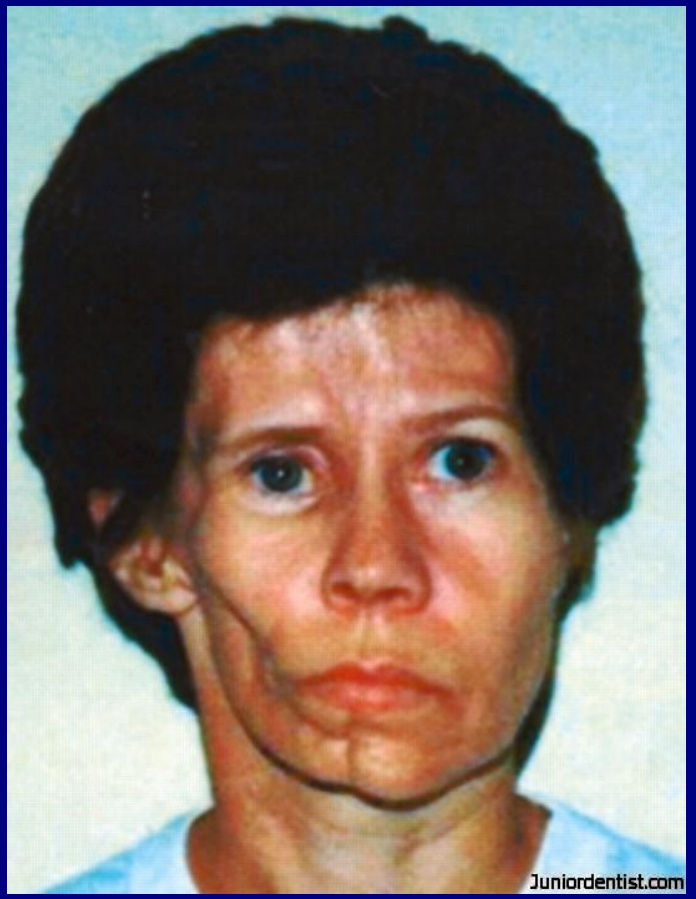


Morphea - *en coup de sabre*





Morphea - facial hemiatrophy *(hemiatrophia faciei)*



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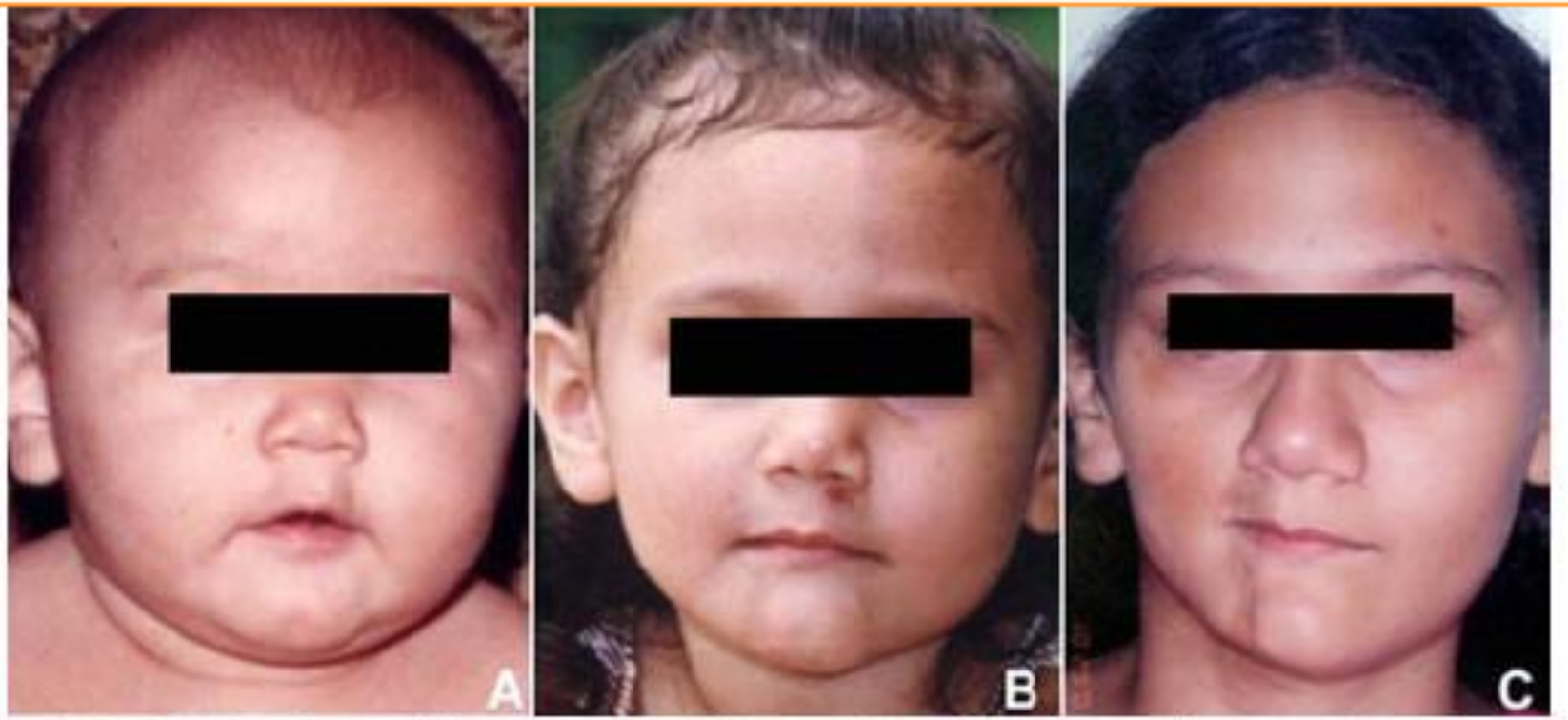
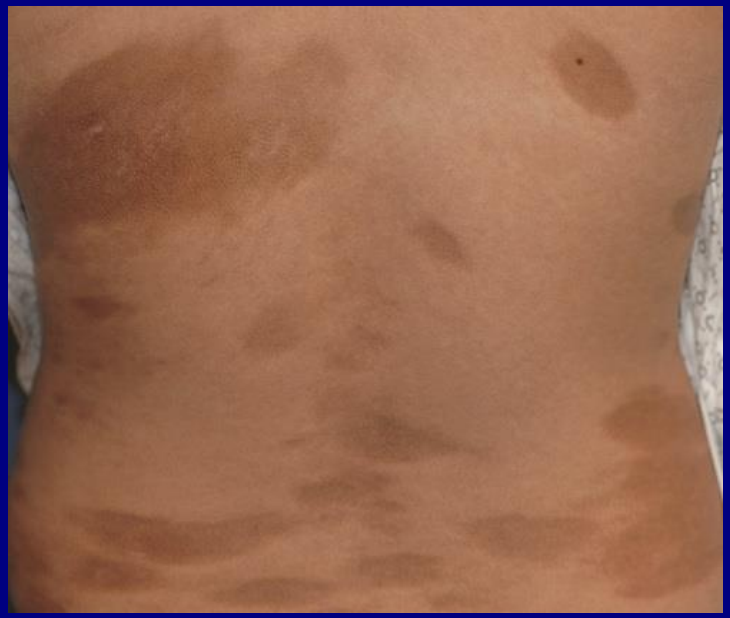
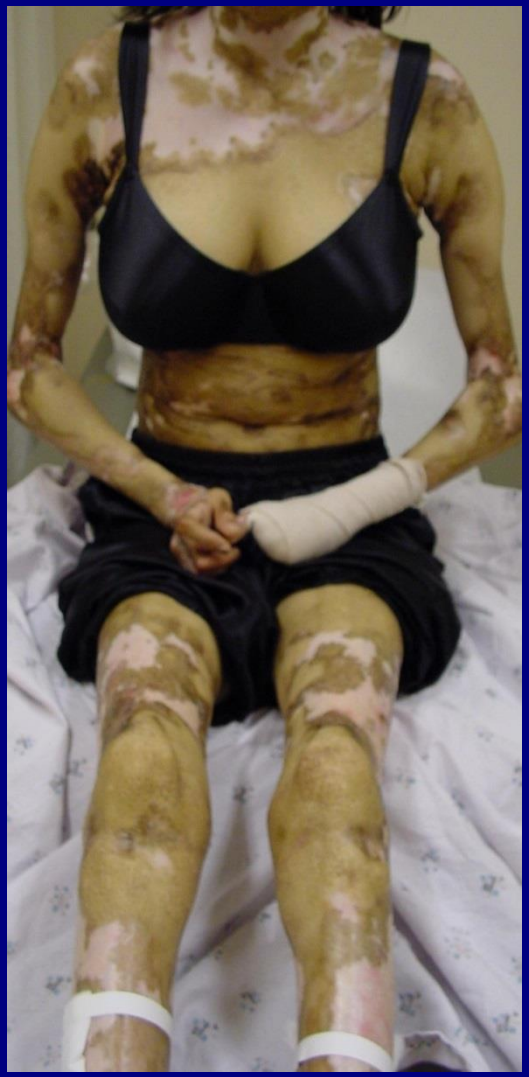


Fig. 3. A. Patient with 2 years and 8 month, without syndrome alterations; B. Patient with 5 years old with mild enophthalmic signs and facial atrophy in right side; C. With 11 years old, the exophthalmia is evident, 'coup de sabre' scar in parasinfisis region and mid right maxillary lip featuring the hemifacial atrophy.

Morphea – generalized type





Morphea – therapy

- Topical
  - Glicorticosteroids
  - Vitamin D3 analogues
- Systemic
  - Methotrexate
  - Mycophenolate mofetil
  - PUVA