SCLERODERMA 101

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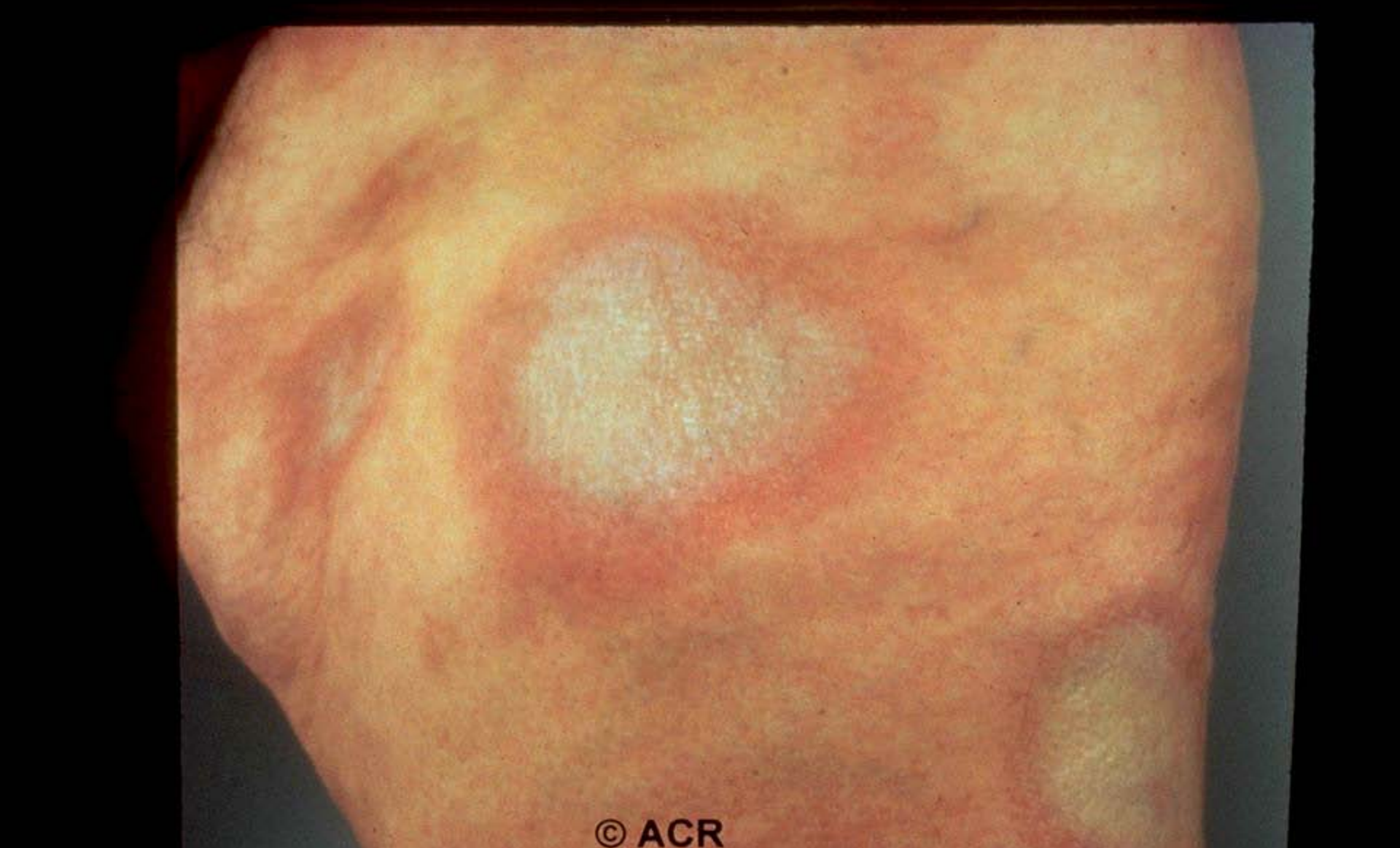
TYPES OF SCLERODERMA

Localized versus Systemic

Two Kinds of Scleroderma

- **Localized** Scleroderma
  - Morphea
  - Linear Scleroderma
  - En coupe de sabre
  - Combinations
- **Systemic** Scleroderma (SSc, Systemic sclerosis)
  - Limited cutaneous (lcSSc)
  - Diffuse cutaneous (dcSSc)

MORPHEA



LOCALIZED SCLERODERMA (not Systemic)

- Involves the skin and underlying tissue – usually does not involve internal organs
- Linear Scleroderma –a line of thickened skin down one leg or arm
- Scleroderma en coup de sabre (cut of the saber) a line of thickened skin involving the scalp, forehead and the face
- Morphea:
  - Patches of thickened skin anywhere on the body
  - Can be a single patch or multiple patches

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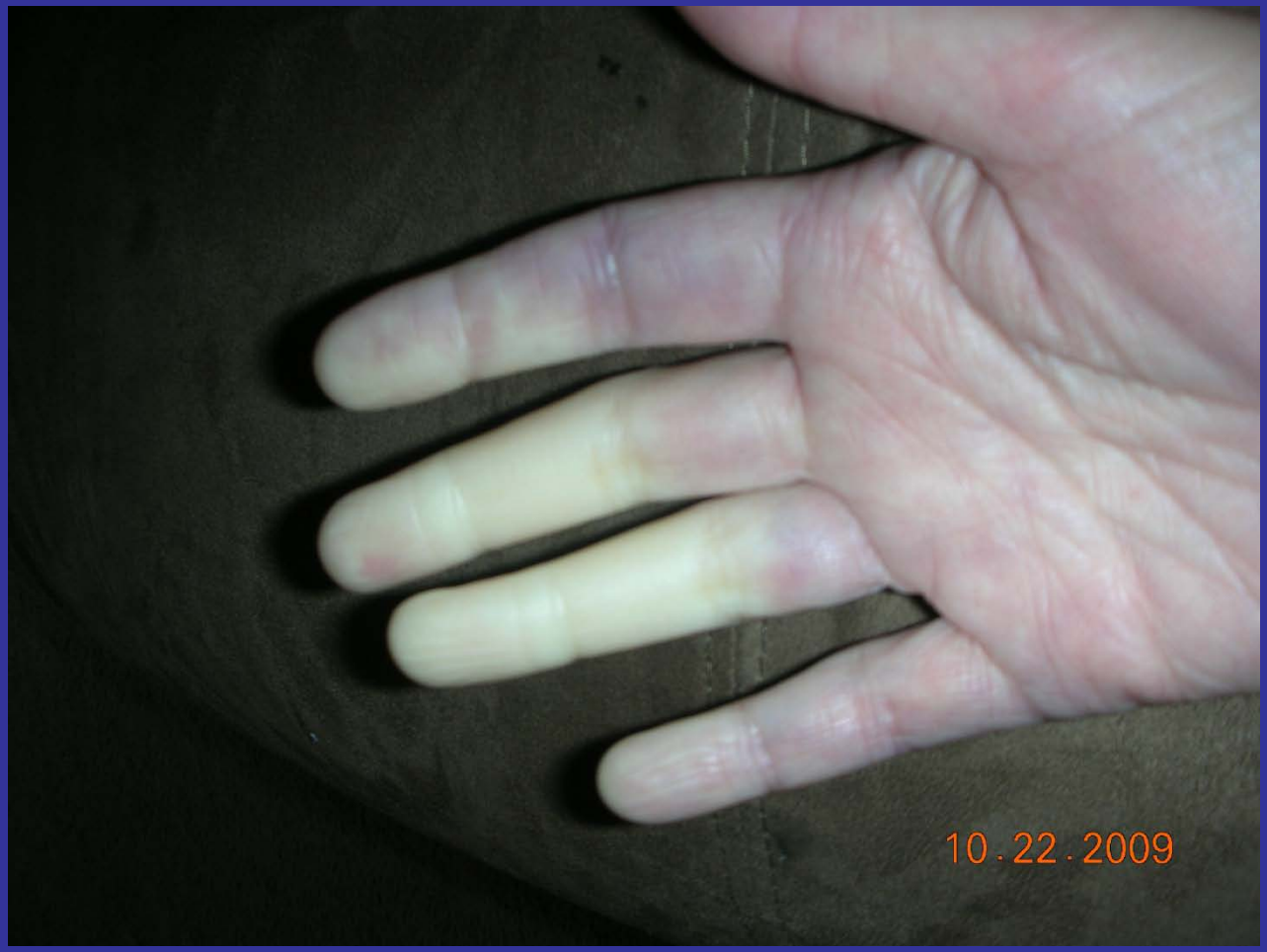
Systemic Sclerosis (SSc)

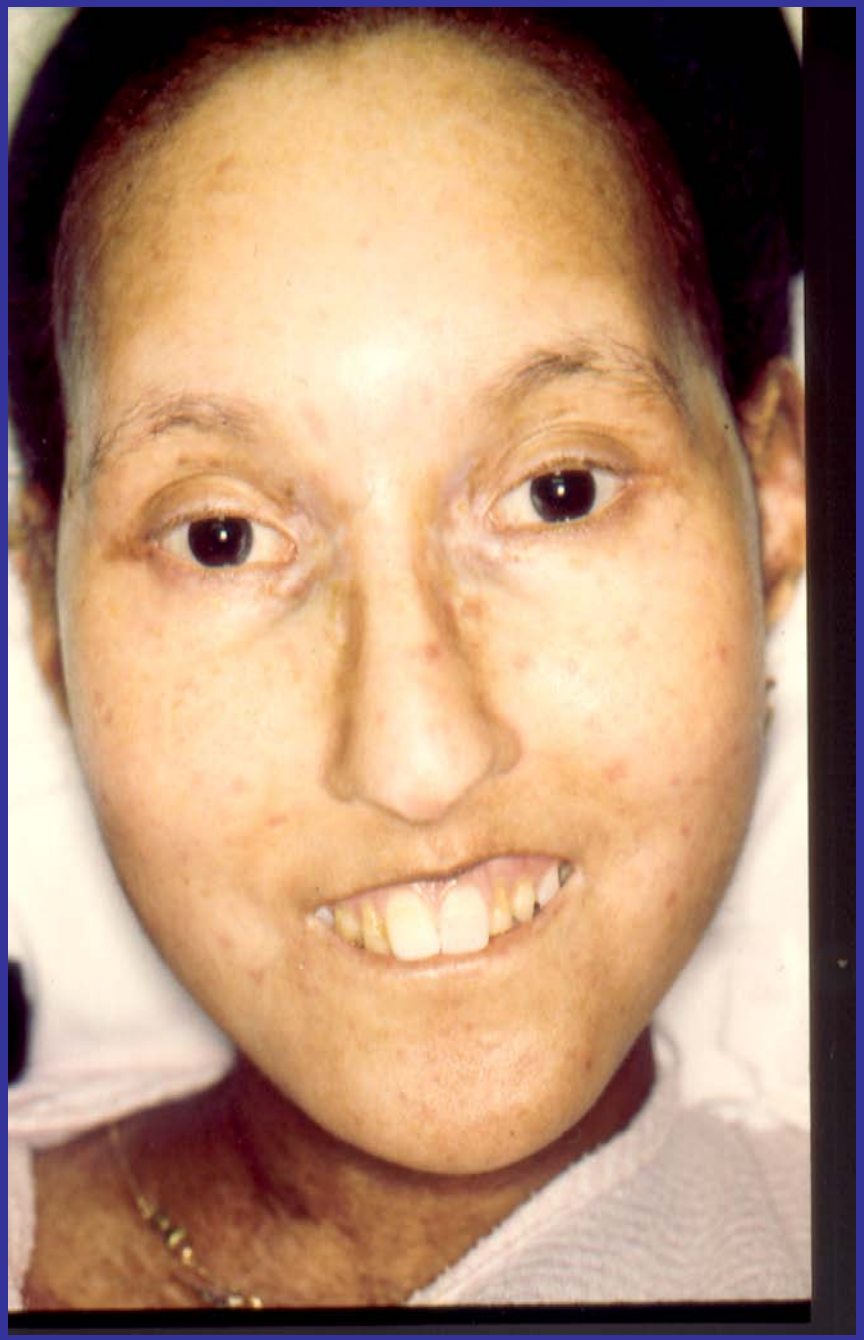
Limited versus Diffuse Disease

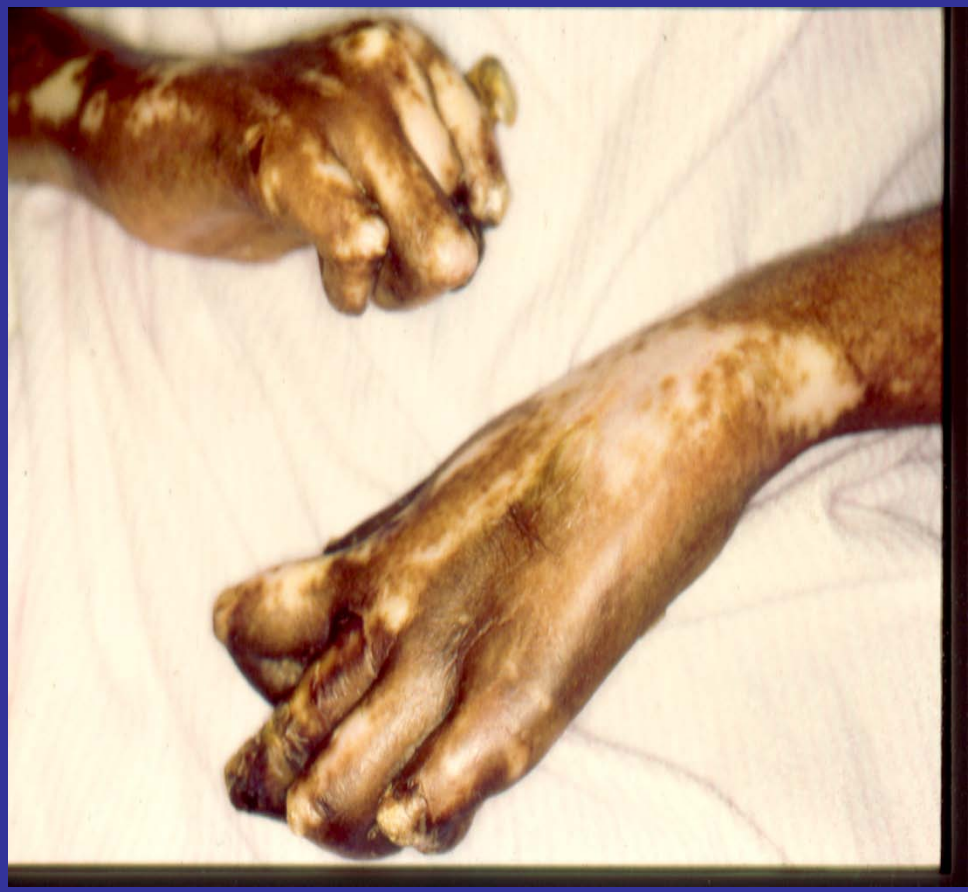
RAYNAUD'S PHENOMENON:
Pallor Phase



RAYNAUD'S PHENOMENON:
Pallor and Cyanosis

TWO FACES OF Systemic SCLERODERMA (SSc)

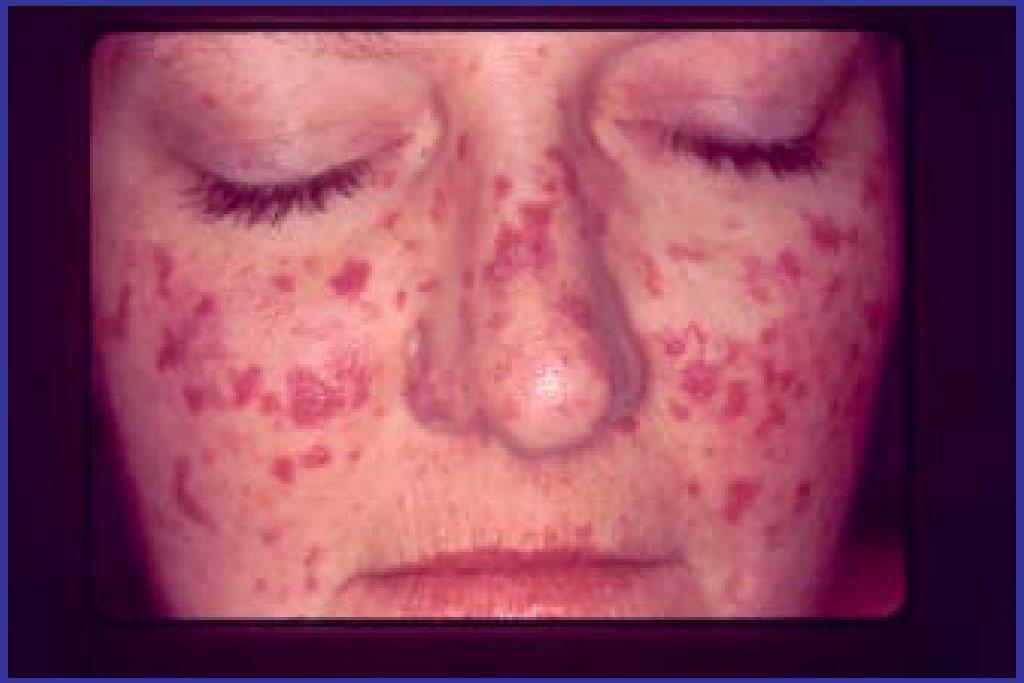


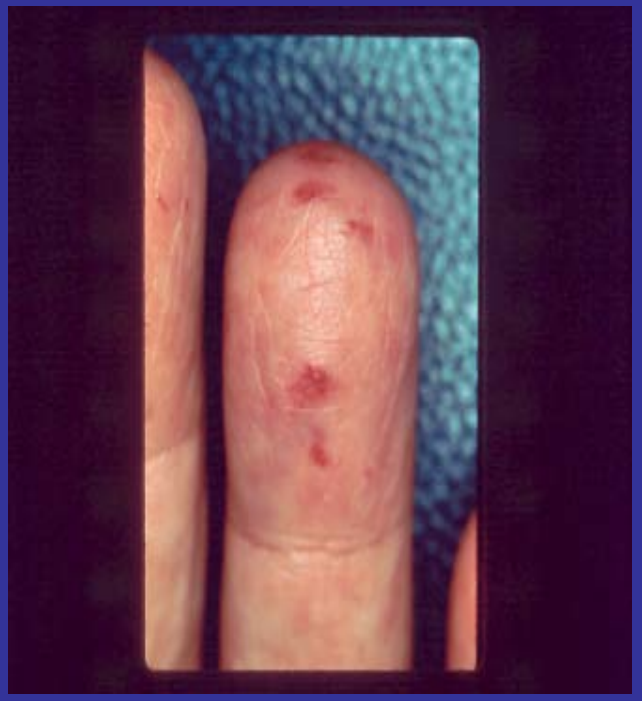


Diffuse Scleroderma

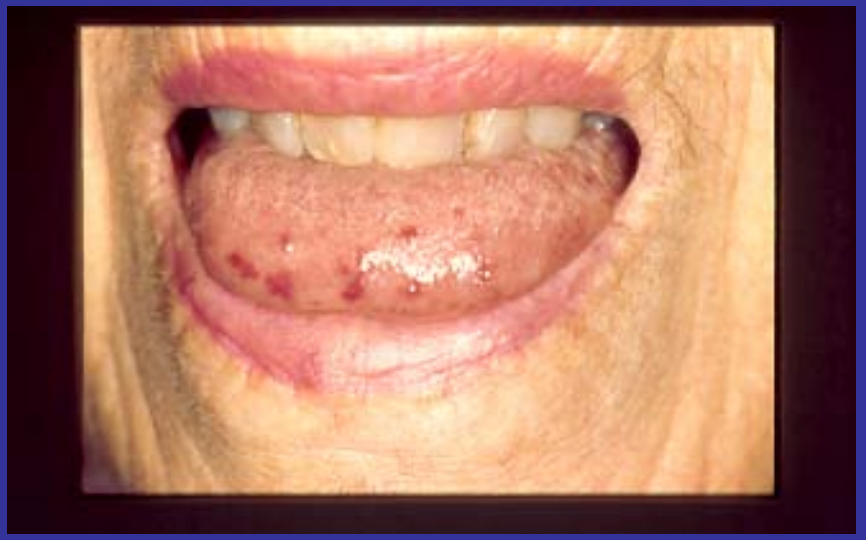
Diffuse Cutaneous SSc

- Skin thickening upper arms, thighs, trunk in addition to hands and distal extremities
- 40% of SSc cases
- More likely to be associated with progressive lung fibrosis, and/or scleroderma renal crisis, and/or significant GI disease, and/or cardiac disease
- Each patient is different





Telangiectasia of Limited SSc



Limited Cutaneous SSc

- Skin thickening confined to distal extremities (both forms can involve the face)
- Usually has more mild internal organ disease
- Associated with anticentromere antibodies
- Sometimes called '**CREST**' for *C*alcinosis, *R*aynaud's, *E*sophageal Dysmotility, *S*clerodactyly, and *T*elangiectasias.

ORGAN INVOLVEMENT in SSc

- Raynaud's phenomenon and digital ulcers
- Pulmonary fibrosis
- Pulmonary hypertension
- Cardiac involvement
- Gastroesophageal (GI) problems
- Joint problems
- Other

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The Causes of SSc

What we know

&

What we don’t know

The Causes of SSc

What we know:

- Some genes increase susceptibility – but are not enough
- The Immune system is activated
- Small blood vessels start to narrow
- Deep skin cells (fibroblasts) increase collagen production

What we don't know:

- The external trigger that starts the process
- The key points in the process that can be interrupted to stop the disease

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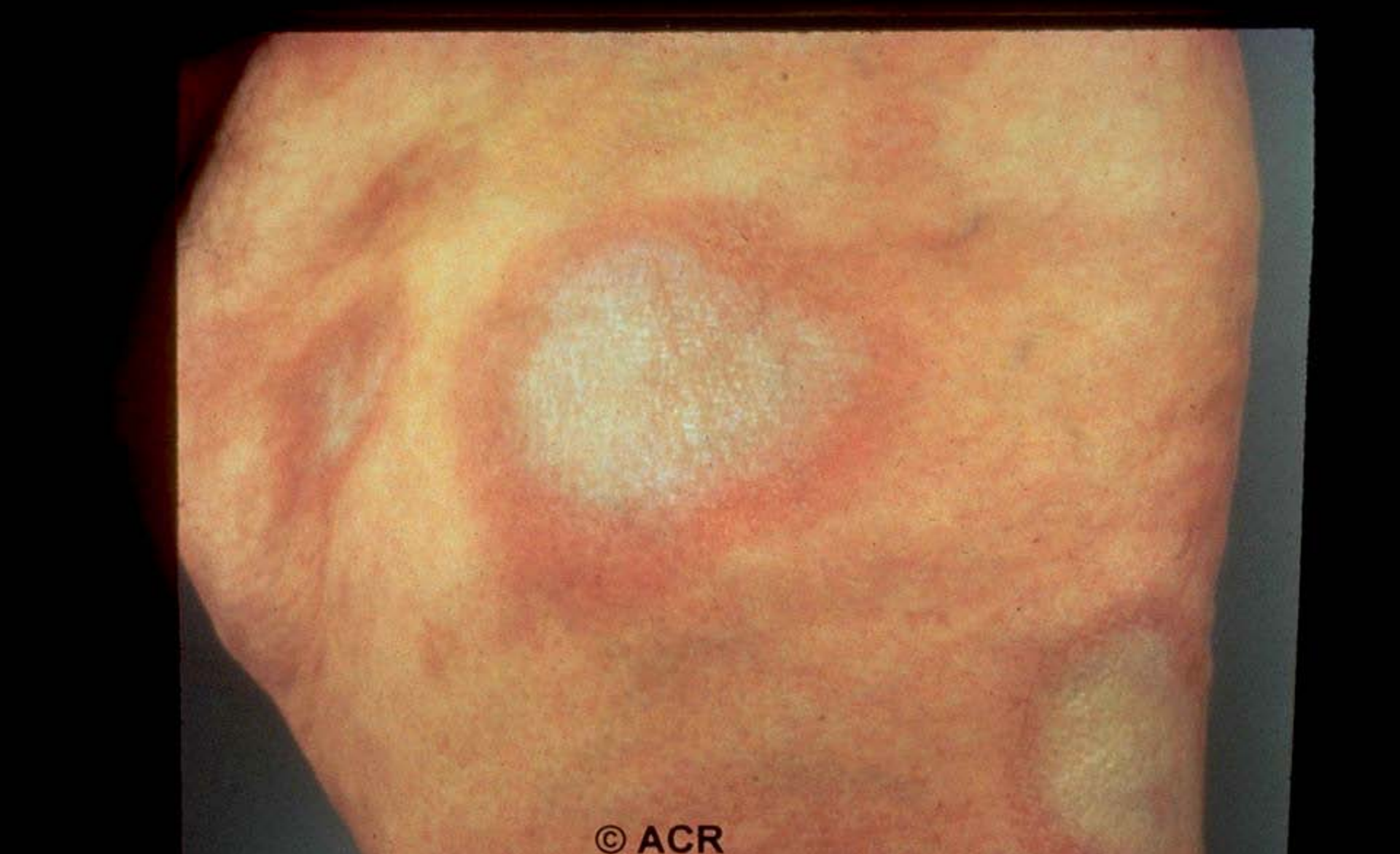
THE TREATMENT OF Scleroderma

Treatment of MORPHEA



The Treatment of Localized Scleroderma

- Is it active?
  - Intralesional steroids
  - Methotrexate
  - Other immunosuppressive medications
  - Risk versus benefit ratio


RISKS >
BENEFITS

RISKS = BENEFITS

RISKS << BENEFITS

TREATMENT OF Systemic Scleroderma

Therapy depends on the organs involved

What are the treatment goals?

- **Skin → decrease thickening (fibrosis**)
- Tendons and joints → decrease inflammation
- Raynaud's phenomenon → improve circulation
- Digital ulcers → improve circulation
- Pulmonary fibrosis → decrease scar tissue (fibrosis)
- Pulmonary hypertension → improve circulation
- Gastrointestinal involvement – esophagus, stomach, bowel → decrease acid, improve motion, treat bacterial overgrowth
- Kidney involvement → control blood pressure

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All SSc Patients are Different

Each person is likely to have a different combination of problems

So each person may be on a different combination of medications

Some may be on little or no medicine

Others are on ‘tons’ of medicine

WHAT YOU CAN DO

- Learn about the disease
- Keep track of your test results
- Ask questions
- Participate in trials

QUESTIONS, CONCERNS, ISSUES