

CREST Syndrome

A Variant of Scleroderma

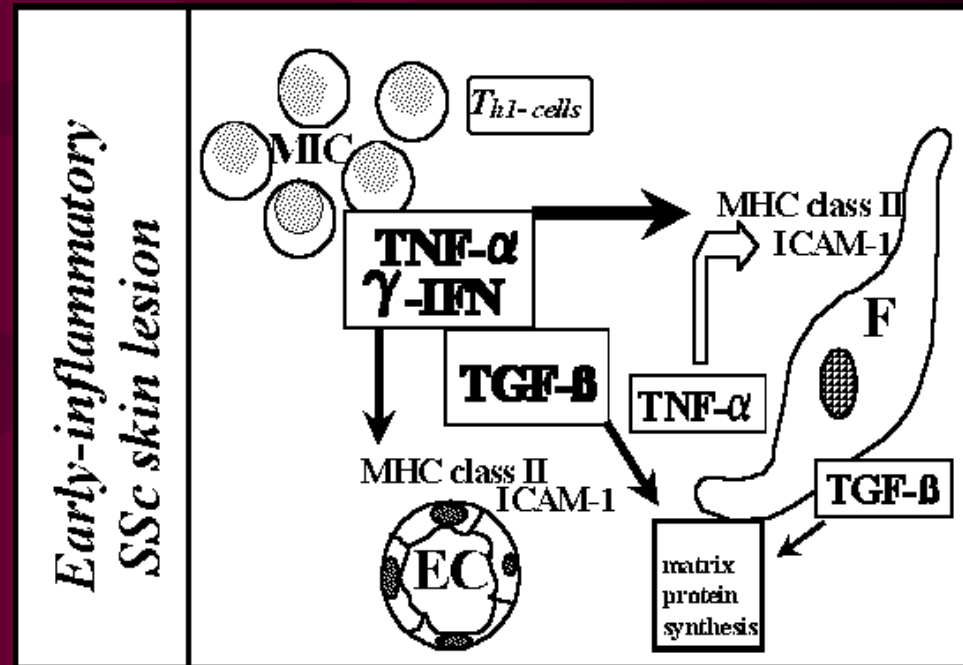
Dennis Timko

Scleroderma

- What is it?
 - “hard skin”
 - affects skin, joints, blood vessels, internal organs
- Epidemiology
 - ☺🔔☹(1:3)
- Pathogenesis
 - overproduction of collagen ?

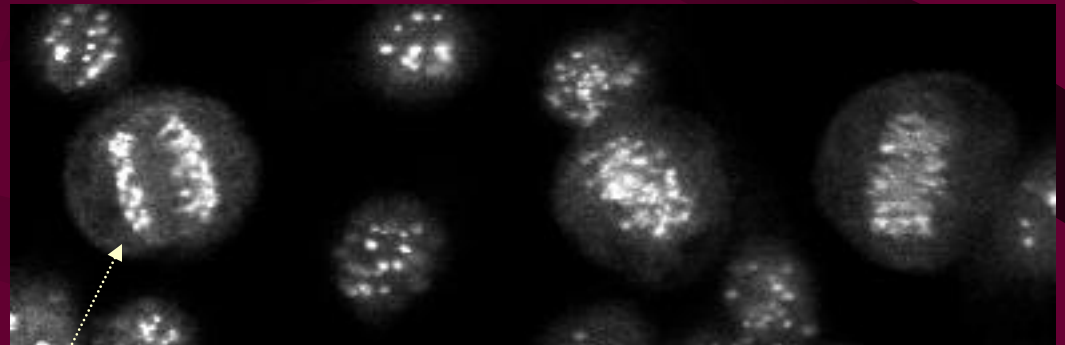
Pathogenesis of Scleroderma

- Over production of Collagen
 - Vascular disease
 - Autoimmune reaction
 - fibrotic reaction
 - Genetics ?



Diagnosis of Scleroderma

- May overlap with CTD
- History
- Physical Exam
- Lab Tests
 - ANA
 - anticentromere
 - topoisomerase I



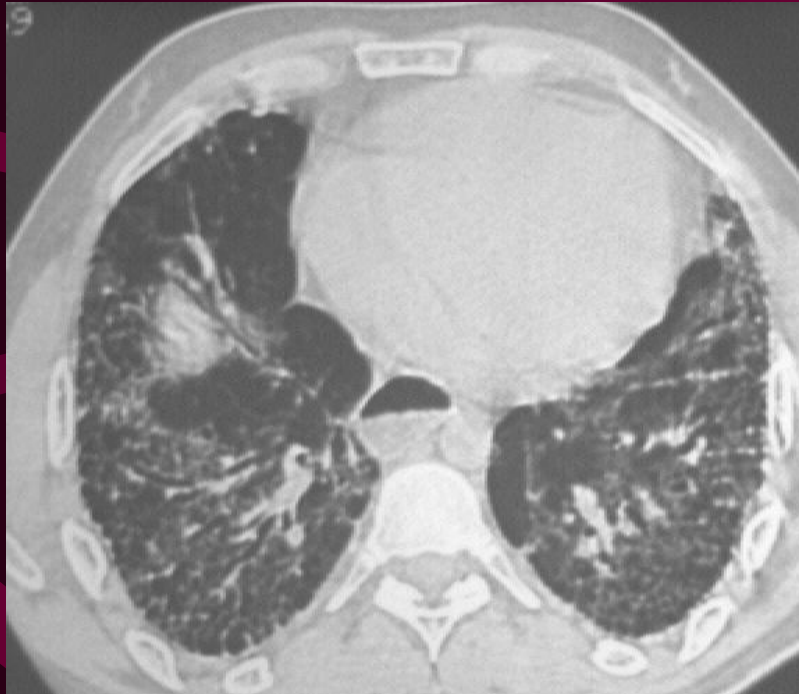
Scleroderma Types

- Systemic Scleroderma (SSc) (Generalized)
 - CREST
 - Limited: Hands, face, & neck
 - Diffuse: Wrists & Elbows
- Localized
 - linear
 - morphea
 - generalized morphea
 - en coup de sabre

Diffuse vs Limited

- 1st 2-4 years
- Skin above the wrists
- Scleroderma *sans* Scleroderma
 - internal organs fibrosis
 - no skin tightening
- Milder form
- Pitting digital ulcers
- Raynauds
- Esophageal problems
- Slow onset
- Good prognosis

Diffuse vs Limited



Localized Scleroderma

- **Morphea**
 - hard, oval shaped patches w/ purplish ring
 - improves over time
- **Generalized Morphea**
 - larger patches
- **Linear**
 - line of thickened skin and deeper layers
 - can effect bones, muscles & joints
 - younger age
- **en coup de sabre** : “*stroke of a sword*”

Morphea vs Linear



Morphea



CREST Syndrome

- **C**alcinosis
- **R**aynaud's phenomenon
- **E**sophageal dysfunction
- **S**clerodactyly
- **T**elangiectasia

CREST Types

- Pure CREST
 - 2 or more symptoms
 - Not limited or diffuse form
- Plus CREST
 - Crest symptoms with other form of Scleroderma

Calcinosis

- Calcium deposits in skin
- Elbows, knees, fingers
- Potential infection



Raynaud's Phenomenon

- 90% affected
- induced by cold
- bilateral
- white to blue to red
- ↓ perfusion pressure
- ↓ capillary radius
- ↑ blood viscosity

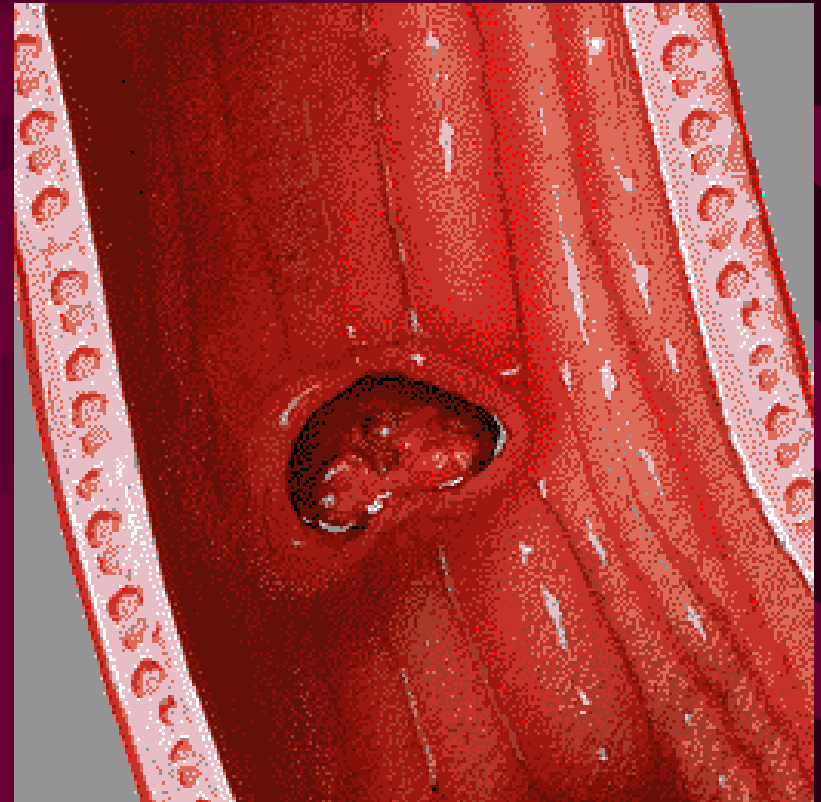


Raynaud's Phenomenon



Esophageal Dysfunction

- Hard to swallow due to internal scarring
- 50% of patients
- lower 2/3 of esophagus
- Wide esophagus
 - heartburn
 - scarring
 - inflammation



Sclerodactyly

- Tightening of skin on digits
- ↑ collagen production
- Shiny appearance
- Difficult to bend



Sclerodactyly



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Telangiectasia

- Dilation of small blood vessels
- Form red spots
- Not harmful
- On face, hands, & mouth behind lips



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Treatment

- No cure, treat symptoms
- PT & ROM exercises
- Medications
 - Nsaids
 - Skin moisturizers
 - Abx
 - Vasodilators
- Treat the individual patient

Summary

- Connective Tissue & Autoimmune disease
- 2 main types
 - Systemic (Adults)
 - Localized (Children)
- CREST is Systemic limited form
- More common in Females
- No genetic etiology
- No cure right now