

*** Dermatomyo. → Pericardial ectrema
→ Telangiectasia

same as systemic sclerosis

Scleroderma

localized

fibrosing skin affect
children



- Skin only, no internal organ
- Morphea / Diffuse (generalized)
linear scleroderma / coup de sabre

Systemic

chronic / unknown etiology

Scleroderma + internal organ

limited cutaneous

- rapid progression

- Fingers - elbow - face

- Raynaud's precede
skin involvement

(critical ischemia)

- Pulmonary arterial HTN

- Calcinosis cutis

- Anti-centromere

diffuse cutaneous

slow

- Fingers - face - Trunk

- with skin

Pulmonary fibrosis

renal crisis (15%)

Anti-topo-iso-merase
(Scl-70) ¹

- F > M

- age (30-50)

- Blacks worse prognosis (diffuse form with Pulmonary fibrosis)

▷ Pathogenesis

Vascular injury (platelets & endothelial activation with tissue hypoxia)



Fibroblast activation & tissue fibrosis by collagen deposit

▷ Factors :

- genetics : non-mendelian pattern
- environmental : (bleomycin / pentazocine / cocaine)

▷ Clinical features

□ Raynaud's phenomenon

- Episodic vasoconstrictive in fingers & toe

- Triggered by
 cold
 stress
 vibration

- Stant pallor → cyanosis → erythema

vasoconstrictive ischemia reperfusion

Primary
 no autoimmune or CT disen
 more in females (menarche)

secondary
 other diseases - CT / hemato / endo
 occupational - vibrating
 β-Blockers
 anti-cancer
 (cisplatin / bleomycin)

Raynaud's Phenomenon

	Primary	Secondary
Sex	Female	Male and Female
Age of Onset	Menarche	Mid 20's or later
Finger Edema	No	Frequent
Periungual erythema	Rare	Frequent
Arthritis	No	Frequent
Nail fold capillaroscopy	Normal	Dilated tortuous capillaries
Autoantibodies	Absent No ANA	Present

[2] Skin

- Thickening ^{symmetrical} Bilateral

- fixed flexion contractures

- loss of creases on the dorsum of finger

- slow healing ulcer & infected

- Face ^{Taut & shiny}
loss of wrinkles
^{Microstomia}
^{Beaked nose}

- ^{Telangiectasia} ^{face} ^{Hand} ^{lips} ^{oral cavity}

- Calcinosis

[3] Lung (leading cause of death)

Interstitial lung disease



- cause restrictive lung with impaired gas exchange

- PFT = ↓ FVC ↓ DLCO
unaffected flow rate

- Risk factors → Male
severe GERD → African-American
diffuse form → Scl-70

- CT-scan: ground glass app.

Pulmonary HTN



- Diagnosed by right heart catheterization

← require for confirmation

- >25 mmHg at rest

- may lead to RHF

- Risk factors → limited form
severe Raynaud phenomenon → anti-centromere & RNA ab.
late age at disease onset

- PFT: normal FVC, ↓ DLCO

- X-ray: Pulmonary a. enlargement

- Echo: >40 mmHg

Aspiration pneumonitis

Restrictive (chest wall fibrosis)

4 GI

Abnormal motility

atrophy & fibrosis of smooth
muscles / intact mucosa / small-vessel
vasculopathy

- Symptoms
- GERD (heart burn/regurgitation / dysphagia)
 - Gastroparesis (early satiety / pain / distension)
 - Gastric antral vascular ectasia: (watermelon app. → unexplained anemia)
 - Malabsorption & chronic diarrhea ← due to bacterial overgrowth
 - Intestinal pseudo-obstruction (ilves)

5 Renal

scleroderma renal crisis

- within 4 years at onset
- Risk factors
 - Male / African-american
 - Diffuse form
 - auto. ab of RNA polymerase III
- present with abrupt onset of Malignant HTN.
- Urinalysis
 - proteinuria
 - Microscopic hematuria
- Thrombocytopenia
- MAHA
- Save lives → ACE inhibitors

6 MSS

- ✓ Arthralgia
- ✓ Tendon friction rubs
- ✓ inflammatory myositis
- ✓ joint contractures
- ✓ severe erosive asymmetrical polyarthritis
- ✓ Acro-osteolysis (Bone resorption in terminal phalanges)

▷ labs

- Normocytic anemia
- ESR normal

- ANA $\left\{ \begin{array}{l} \text{anti-centromere} \\ \text{anti-scl-70} \end{array} \right.$

▷ Treatment (based on symptoms)

- ① glucocorticoids \rightarrow \downarrow stiffness & pain
- ② cyclophosphamide \rightarrow \downarrow ILD
- ③ Mycophenolate (Methotrexate) \rightarrow improve skin induration
- ④ Colchicine \rightarrow treat calcinosis

(Raynaud)

- ✓ dress warm
- ✓ avoid β -Blockers
- ✓ CCB (nifedipine)
- ✓ phosphodiesterase inhibitors (sildenafil)
- ✓ serotonin reuptake inhibitors (fluoxetine)
- ✓ Topical NO
- ✓ IV PEG₂.

(PAH)

- phosphodiesterase type 5 inhibitors (sildenafil)
- endothelial-1 receptor antagonist (bosentan)
- Prostaglandin analogue
- O₂
- lung transplantation