Systemic Sclerosis

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- Chronic multisystem disorder characterized by….
- Abnormality of skin n visceral organs due to.. Excessive accumulation of connective tissue
- Etiology : Unknown

Classification:

- Systemic Sclerosis
  - Limited Cutaneous ds.
  - Diffuse Cutaneous ds.
  - Sine Scleroderma
  - Overlap Syndrome
  - Undiff. CTD
- Localised Scleroderma
  - Morphea
  - Linear Scleroderma
  - En coup de sabre
- Scleroderma-like dr.
  - Chemical induced

e.g. Bleomycin and Penta -zocine therapy, venyl chloride exposure

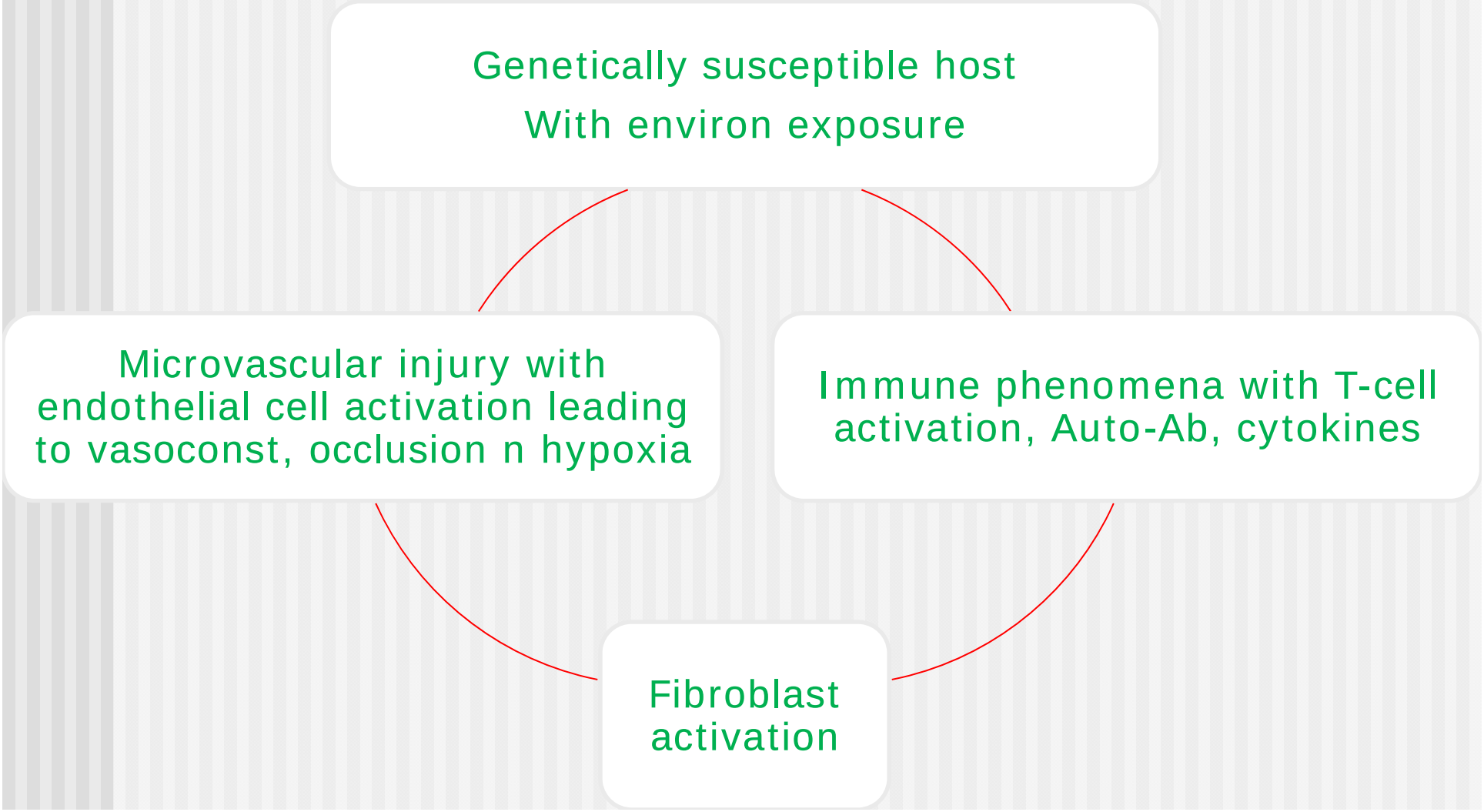
  - Others

e.g. Eosino. Fascitis, EMS, Amyloidosis, GVHD in BM transplnt recepients, digital sclerosis in DM etc.

Epidemiology n Etiopathogenesis:

- Women:Men = 3:1
- Some Infectious agent (? CMV, ? Parvo B 19) inducing autoimmune phenomena
- Genetic susceptibility…î RR in 1° relatives, HLA DQA_2 asso.
- Environ factors… Coal n Gold miners
  Vinyl chloride exposure
  Bleomycin n Pentazocine t/t

Etiopathogenesis:



Etiolpathogenesis:

- Skin→ T-cell infiltration in dermis n loss of dermal appendages, tethering of dermis to subcutaneous tissue bcs of collagen bands
- GIT→ ↑ collagen in serosa, submoucosa n lamina propria, secondary mucosal atrophy
- Lungs n Pleura→ ILD, bullous emphysema, PAH, Pleural fibrosis
- Musculo-skeletal→ perivascular lymphocytic infilt^{n}, fibrosis, MNCs infilt^{n} in synovium

Etiolpathogenesis:

- Heart→ myocardial fiber degeneration, interstitial fibrosis
- Kidney→ intimal hyperplasia of interlobular arteries, fibrinoid necrosis of afferent arterioles

Clinical manifestations:

- Raynaud's phenomena:
  - First symptom, MC symptom following skin thickening
  - Exposure to cold, vibration, emotional stress
  - h/o digital pallor most reliable
- Skin changes:
  - Swelling of digits→ thickening of overlying skin→ thinned-out atrophic skin, flexion contractures, resorption of phalanges

Clinical manifestations:

- Skin changes:
  - Hyperpigmentation of skin with perifollicilar sparing (Salt-Pepper appearance)
  - Coarse n Dry skin→ loss of sebaceous n sweat glands
  - Calcific deposits (subcutaneous)

Clinical manifestations:

GIT: Microstomia, pinched nose appearance

- Esophagus→ GERD, Esophagitis, Esophageal dysmotility
- Stomach→ GERD, Gastroparesis, Vascular ectasia (Watermelon stomach)
- Small bowel→ Malabs synd secondary to Bact Overgrowth synd, Peudo-obstruction/ paralytic ileus
- Large bowel→ Constipation, Anal sphincter incontinence
- SSc Sine scleroderma

Clinical manifestations:

- Pulmonary:
  - Leading cuase of death in SSc
  - ILD, Pulm vasculitis, PAH
  - Pleural fibrosis
- Cardiac:
  - Pericarditis with or without effusion
  - Condution blocks/ arrhythmia
  - Heart failure (Restrictive CMP, cor-pulm.)

Clinical manifestations:

- Renal:
  - renal crisis (malig. HTN)

- Musculoskeletal:
  - Carpal tunnel synd
  - Disuse muscle atrophy
  - Myositis (Overlap synd)

Lab investigations

- Î ESR
- Anemia of chronic disease
- Megaloblastic anemia (Malabs synd)
- Anti-RNA polymerase
- Anti-Th RNP, Anti-U_3 RNP

Diagnosis:

- ACR diagnostic criteria (M+2m)
  - For clinical studies only
  - Sensitivity=97% ; Specificity=98%
  - Major criteria→ Sclerodematous changes of skin proximal to digits with involv. of face or trunk in a symm pattern
  - Minor criteria→ Sclerodactyly, digital pitting scars, loss of digital pulp on volar aspect, bibasilar pulm fibrosis

Differential Diagnoses:

- Raynaud's phenomena
  - Thoracic outlet synd (cervical rib, scalenus)
  - Cryoglobulinemia, Hep B,C; PNH
  - Vinyl chloride exposure

- Skin changes n visceral involvement
  - Scleredema
  - Eosin fascitis, EMS
  - amyloidosis

Treatment:

- Symptomatic treatment→
  - GERD, Gastroparesis
  - Raynauds ds (æ blockers, ARBs, Ca- channel blockers, warmth, occlusive dressing)
  - Dry eyes (methylmethacrylate eye drops)
  - Dry mouth (pilocarpine)
- Supportive therapy→
  - Renal replacement therapy
  - CHF, Anti-HTN medication

- Supportive therapy:
  - domicilliary O2 therapy, polyvalent pneumococcal vaccine n yearly influenza vaccine (ILD)
  - Anticoagulants n Ca-channel blockers (PAH)

- Rehabilitative therapy:
  - Muscle strength building exercises, maintenance of range of motion at joints

Treatment:

- Antiplatelet therapy to all patients
- Sterois→ high dose for myosistis, alvoelitis and pericarditis
  → low-dose short-course for digital edema or NSAID refractory joint pains
  - Renal crisis asso with use of Steroids
- Immunosuppressants n antifibrotic agents either not effective or unacceptable S/Es