Systemic Sclerosis (SSc)

Clinical Hallmarks → skin thickening

Skin thickening + 1 more for diagnosis of SSc:

(Fingers)
- Sclerodactyly (involving fingers)
- Digital pitting
- **Raynaud** (pretty much required)
  - Without Raynaud, it is not SSc. But another scleroderma-like condition is possible.
  - If only symptom is Raynaud, look for nailfold capillary destruction and dilated capillary loops.

(Lungs)
- Interstitial lung disease
- Pulmonary Hypertension

(GI)
- GERD & esophageal dysmobility
- Small bowel psudoobstruction
- Malabsorption from bacterial overgrowth

(others)
- Calcinosis
- Kidney disease
- Inflammatory arthritis, DIP joints and wrists

Scleroderma-like conditions:
- Eosinophilic fasciitis (edema of proximal extremities)
- Nephrogenic Systemic Fibrosis (gadolinium)
- Sclerederma (diabetics)
- Scleromyxedema (red-yellow papules, seen in Multiple Myeloma/AL amyloidosis)
- Chronic GVHD (Lichen planus-like skin lesions, seen after HSCT)

Diffuse vs Limited
- Limited only has cutaneous involvement distal to elbows/knees. Diffuse can go everywhere. (Both can involve the face).
- Limited does not cause scleroderma renal crisis.
- Diffuse does not have the CREST syndrome.
- Serology → ANA for both. "centromere" for limited. "Scl-70" for diffuse.
- Lung → Pulm HTN for Limited. "clean lungs". Other one is the other one.

Lung disease
- Primary cause of morbidity and mortality

Screen with :
- High-res CT and PFTs (including DLCO)
- Echocardiogram (baseline and annual)

Kidney disease
- 50% will have mild proteinuria, elevated plasma creatinine concentration, and HTN
- Most do not progress to AKI or CKD

Treatment
- Treat organ specific manifestations. No overall disease modifying agent.
- Raynauds → amlodipine, sildenafil, nitroglycerin paste
- Alveolitis/ILD → mycophenolate (1st line). Otherwise, cyclophosphamide.
- GI / motility → PPI for GERD; promotility agents (metoclopramide)
- Bacterial overgrowth → broad spectrum antibiotics
- Renal crisis → ACE-I. Only if suspecting renal crisis. Continue even with renal failure. Avoid primary prevention → ace-I may precipitate renal crisis.
- **AVOID glucocorticoids**. Can precipitate normotensive renal crisis (AKI without HTN).

Garg's Simple Medicine
10/05/2019