

## COLLAGEN VASCULAR DISORDERS

### I. Lupus erythematosus

#### A. Discoid LE (DLE)

1. pink, violaceous well defined plaques in sun exposed distribution
2. scarring, including alopecia
3. follicular plugging, atrophy
4. ANA-negative
5. no systemic manifestations
6. Treatment:
  - a steroids (systemic, topical)
  - b antimalarials
  - c sunscreens

#### B. Systemic LE (SLE)

1. autoimmune antibody/antigen complex deposition with Type III inflammation
2. drug induced
  - a hydralazine
  - b procainamide
  - c isoniazid INH
3. 4 or more ARA criteria for diagnosis:
  - a malar ("butterfly") rash with sun exposure
  - b discoid lesions
  - c photosensitivity

- d oral ulcers
- e arthritis
- f serositis (pleuritis, pericarditis)
- g renal disease
- h neurologic sx's (seizures, neuropathy, psychosis)
- i hematologic (anemia, pancytopenia)
- j immunologic (anti-DNA ab's; LE prep; false + RPR) ANA

4. Treatment:

- a steroids
- b immunosuppressants
- c antimalarials
- d sunscreens

C. Subacute LE (SCLE)

1. ANA negative lupus
2. +SSA, SSB antibodies (anti Ro, anti La)
3. less cardiac, renal involvement compared to SLE
4. small, annular red plaques in sun exposed areas
5. Neonatal lupus:
  - a (infants born to mothers with SCLE)
  - b 3<sup>o</sup> heart block
  - c discoid lesions
  - d anemia, "HELP" syndrome (Hepatic Enzymes Low Platelets)
  - e usually resolves within 6 months, but heart block permanent

## II. Dermatomyositis

- A. Inflammatory disorder of proximal muscles, skin
- B. Associated with internal malignancy 25 to 50% in patients over 55
- C. Skin:
  - 1. "heliotrope" rash
  - 2. Gottron's papules
  - 3. periungual telangiectases
- D. Muscle: limb/girdle weakness with abnormal EMG findings
- E. Laboratory:
  - 1. elevated CPK, Aldolase
  - 2. + ANA
  - 3. ESR elevation
- F. Treatment
  - 1. treat underlying malignancy
  - 2. steroids

## III. Scleroderma

- A. Morphea (localized)
  - 1. atrophic plaques without systemic involvement
- B. CREST Syndrome
  - 1. Calcinosis cutis
  - 2. Raynaud's disease
  - 3. Esophageal strictures

4. Sclerodactyly

5. Telangiectases

C. PSS (Progressive Systemic Sclerosis)

1. proximal sclerosis

2. pulmonary fibrosis

3. renal disease

4. cardiac (restrictive cardiomyopathy)

D. Treatment:

1. steroids

2. d-penicillamine