9.6.9. Posterior Uveitis (II): Collagen Vascular Diseases (II): Systemic Lupus Erythematosus

ocular manifestations

lupus retinopathy

- 50%
- eyelid lesions (discoid lupus erythematosus)
- 20% secondary Sjögren syndrome
- scleral inflammatory disease
- neuro-ophthalmic lesions
  - cranial nerve palsies
  - optic neuropathy
  - retrochiasmal and cerebral visual disorders
- uveitis
  - rare
  - anterior or intermediate uveitis, are not commonly associated with SLE
- cotton-wool spots with or without intraretinal hemorrhages
  - independently of hypertension

Figure 6-16 (© 2020 American Academy of Ophthalmology)

- severe retinal vascular occlusive disease (arterial and venous thrombosis)
  - retinal nonperfusion and ischemia
  - secondary retinal neovascularization
    - vitreous hemorrhage

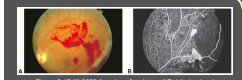

Figure 6-17 (© 2020 American Academy of Ophthalmology)

- an important marker of systemic disease activity
  - 3% with mild disease
  - 29% with more active disease
- associated with
  - CNS lupus
  - antiphospholipid antibodies
    - retinal vascular thrombosis
    - hypercoagulable state

lupus choroidopathy

- serous elevations of the retina, retinal pigment epithelium (RPE), or both
- choroidal infarction
- choroidal neovascularization (CNV)

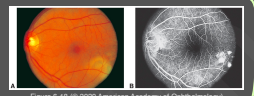

Figure 6-18 (© 2020 American Academy of Ophthalmology)

- arteriolar narrowing
- disc edema

treatment

control of the underlying disease

- NSAIDs
- corticosteroids
  - need monitoring for cataracts and glaucoma
- IMT
- plasmapheresis
- systemic antihypertensive medications
- patients with severe vaso-occlusive disease or antiphospholipid antibodies
  - antiplatelet therapy
  - systemic anticoagulation
- hydroxychloroquine
  - to ameliorate skin, joint, and constitutional symptoms and prevent relapses
  - may help prevent nephritis and ANA-mediated thrombosis
  - need monitoring for retinal toxicity

proliferative retinopathy

- panretinal photocoagulation

vitreous hemorrhage

- vitrectomy surgery

epidemiology

- women of childbearing age
- higher incidence among African Americans and Hispanics in US

pathogenesis

- autoimmune disorder characterized by
  - B-lymphocyte hyperactivity
    - polyclonal B-lymphocyte activation
  - hypergammaglobulinemia
  - autoantibody formation
    - antinuclear antibodies (ANA)
    - anti-ssDNA and anti-dsDNA
    - antibodies to cytoplasmic components (anti-Sm, anti-Ro, and anti-La)
    - antiphospholipid antibodies
      - antiphospholipid antibody syndrome (APS)
        - in association with SLE and other rheumatic diseases
        - venous or arterial thrombosis
          - deep venous thrombosis is the most common type of thrombosis
  - T-lymphocyte autoreactivity with immune complex deposition
- association with HLA-DR2 and DR3

systemic manifestations

- acute cutaneous diseases
  - 70%–80%
  - malar rash
  - discoid lupus
  - photosensitivity
  - mucosal lesions
- arthritis 80%–85%
- renal disease 50%–75%
- Raynaud phenomenon 30%–50%
- neurologic involvement 35%
- cardiac, pulmonary, hepatic, and hematologic abnormalities
- SLICC criteria for diagnosis of SLE See Table 9-1

lab testing

- ANA testing in patients with no systemic or ocular findings to suggest SLE has a very high false-positive rate
- ANA testing should be limited to patients
  - with signs or symptoms suggestive of SLE
  - who have juvenile idiopathic arthritis

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