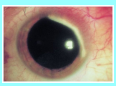
8.7.9. Immune-Mediated Diseases (IX): Scleritis

Pathogenesis

- immune-mediated (immune-complex) vasculitis
- underlying systemic immunologic disease
  - incidence
    - diffuse or nodular scleritis 1/3
    - necrotizing scleritis 2/3
  - associated diseases
    - 1/2
      - gout
    - infectious
      - syphilis
      - tuberculosis
      - herpes zoster
      - lyme disease
      - "cat-scratch" disease
      - leprosy
    - autoimmune connective tissue diseases
      - rheumatoid arthritis
      - systemic lupus erythematosus
      - seronegative spondyloarthropathies (eg, ankylosing spondylitis)
      - wegener granulomatosis
      - polyarteritis nodosa
      - giant cell arteritis
- 4-6 decades of life — exceedingly rare in children
- F>M
- 1/2 bilateral

Complications Of Scleritis

- peripheral keratitis — 37%
  - localized to the area of inflammation
- central stromal keratitis — rare
  - heavy vascularization and opacification if untreated
- sclerokeratitis
  - 
    Figure 7-26 (© 2020 American Academy of Ophthalmology)
  - fibrosis and lipid deposition of peripheral cornea and neighboring scleritis
  - area of involvement may gradually move centrally
  - accompanies herpes zoster scleritis
  - may also occur in rheumatic diseases
- scleral thinning — 33%
  - scleral translucency and/or thinning
  - only in the most severe forms of necrotizing disease/scleromalacia perforans
  - frank scleral defects
- uveitis — 30%
  - eyes with anterior scleritis — anterior uveitis
  - eyes with posterior scleritis — posterior uveitis
- glaucoma — 18%
- cataract — 7%

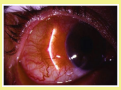
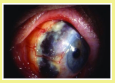
Laboratory Evaluation

- initial screening
  - complete blood count (CBC) with differential
  - erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP)
  - serum autoantibody screening (antinuclear antibodies, anti-DNA antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies)
  - urinalysis
  - serum uric acid test
  - syphilis serology
  - chest x-ray
  - sarcoidosis screening (serum angiotensin-converting enzyme and lysozyme)
- if systemic evaluation is initially negative, repeat the workup annually
- in conjunction with a rheumatologist or other internist

Management

- in general the treatment of scleritis is systemic
- topical corticosteroids
  - mild diffuse anterior or nodular scleritis
- oral NSAIDS
  - nonnecrotizing disease, especially diffuse disease
  - 600 mg of ibuprofen 3 times a day
- oral and/or high-dose (pulsed) intravenous corticosteroids
  - indications
    - severe nodular scleritis
    - necrotizing scleritis
    - sclerokeratitis
- systemic immunosuppressive therapy
  - methotrexate
  - cyclosporine
  - cyclophosphamide
  - if no therapeutic response is observed with corticosteroids
- TNF inhibitors such as infliximab
  - rheumatoid arthritis–associated scleritis
- subconjunctival corticosteroids
  - when systemic administration is contraindicated or not feasible
  - in nonnecrotizing scleritis
- antituberculosis and anti-pneumocystis coverage may be necessary for at-risk patients
- treatment failure
  - progression of disease to a more severe form (eg, nodular to necrotizing)
  - failure to achieve response to treatment after 2–3 weeks of therapy
  - an alternate therapeutic strategy will need to be instituted

Clinical Presentation

- gradual onset — over several days
- severe boring or piercing ocular pain
  - worsen at night
  - referred to other regions of the head or face
  - tender globe
- inflamed sclera — violaceous hue — best seen in natural sunlight
- inflamed vessels — crisscross pattern — adhere to the sclera — cannot be moved with a cotton-tipped applicator
- scleral edema — overlying episcleral edema
- types
  - *Diffuse versus nodular anterior scleritis*
    - Diffuse anterior scleritis
      - part of the anterior sclera (<1/2) — 60%
      - entire anterior segment — 40%
      -
        Figure 7-21 (© 2020 American Academy of Ophthalmology)
    - Nodular anterior scleritis — scleral nodule
      - deep red-purple color
      - immobile
      - separated from the overlying episcleral tissue
      - 
        Figure 7-22 (© 2020 American Academy of Ophthalmology)
  - *Necrotizing scleritis*
    - Necrotizing scleritis with inflammation
      - severe pain
      - localized patch of inflammation — edges more inflamed than center
      - avascular edematous patch of sclera — 25% — sclera may develop a blue-gray appearance
      - altered deep episcleral vessels — large anastomotic vessels circumscribe the involved area
      - may spread posteriorly to the equator and circumferentially
      -
        Figure 7-23 (© 2020 American Academy of Ophthalmology)
    - Necrotizing scleritis without inflammation (scleromalacia perforans)
      - long-standing rheumatoid arthritis
      - painless
      - signs of inflammation are minimal
      - sclera thins and the underlying dark uvea becomes visible
      - large abnormal blood vessels surround and cross the areas of scleral loss
      - spontaneous perforation is rare
      - may rupture with minimal trauma
      - bulging staphyloma — if intraocular pressure is elevated
      - 
        Figure 7-24 (© 2020 American Academy of Ophthalmology)
  - *Posterior scleritis*
    - no related systemic disease can be found in patients with posterior scleritis
    - can occur
      - in isolation — diagnosis can be missed in the absence of associated anterior scleritis
      - concomitantly with anterior scleritis
    - pain — pain may be referred to other parts of the head
    - vision loss
    - tenderness
    - retraction of the lower eyelid
    - proptosis
    - restricted motility
    - angle-closure glaucoma
    - choroidal folds
    - exudative retinal detachment
    - papilledema
    - thickened posterior sclera by echography, computed tomography, or magnetic resonance imaging

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