7.4.5. Non-infectious Orbital Inflammations (II)

Polyarteritis nodosa
- necrosis of the muscularis layer
- neutrophils
- eosinophils
- small- & medium-sized arteries
- necrotizing vasculitis
- may affect orbital vessels but does not usually cause orbital disease
- retinal and choroidal infarction
- ophthalmic manifestations

IgG4 DISEASE
- multiorgan disease with mass-forming lesions
- dacryoadenitis
- xanthogranuloma
- orbital amyloidosis
- nonspecific orbital inflammation
- infiltration of IgG4-expressing plasma cells with inflammatory T lymphocytes in various organs
- elevation in the levels of serum IgG4 and an acute-phase response
- responds well to treatment with steroids

Sarcoidosis
- African or Scandinavian descent
- lungs are most commonly involved
- multisystem disease
- orbit
  - bilateral
  - lacrimal gland is most frequently affected
  - very rare
  - extraocular muscles and optic nerve
  - sinus involvement with associated lytic bone lesions can invade the adjacent orbit
  - without associated systemic disease!
  - isolated orbital involvement
  - orbital sarcoidosis
- histology
  - noncaseating collections of epithelioid histiocytes in a granulomatous pattern
  - mononuclear inflammation at the periphery of the granuloma
- diagnostic workup
  - 7% have clinically detectable enlargement of the lacrimal glands
  - positive in 80% of patients with systemic sarcoidosis
  - nonspecific
  - Gallium scanning of the lacrimal glands
  - lacrimal gland
  - biopsy
  - random conjunctival biopsies have a low yield
  - suspicious conjunctival lesion
  - hilar adenopathy
  - pulmonary infiltrates
  - chest radiography or CT
  - angiotensin-converting enzyme
  - serum lysozyme
  - serum calcium
  - bronchoscopy with washings and biopsy
- acute sarcoidosis
  - <2 years duration
  - frequently with associated anterior uveitis in young patients
  - Löfgren syndrome
    - acute iritis
    - bilateral hilar adenopathy
    - febrile arthropathy
    - erythema nodosum
  - responsive to systemic corticosteroids
  - good long-term prognosis
  - Heerfordt syndrome (uveoparotid fever)
    - uveitis
    - parotitis
    - facial nerve palsy
    - fever

Giant cell arteritis (GCA)
- affects the aorta and branches of the external and internal carotid arteries and vertebral arteries
  - orbital vessels are inflamed in GCA
    - generalized orbital ischemia is rare
  - usually spares the intracranial carotid branches
    - lack an elastic lamina
- symptoms
  - vision loss
    - central retinal artery occlusion
    - ischemic optic neuropathy
  - diplopia
    - ischemic dysfunction of cranial motor nerves
  - headache
  - scalp tenderness
  - jaw claudication
  - malaise
- diagnostic workup
  - elevated ESR
    - 90%
  - elevated CRP
  - elevated platelet count
  - temporal artery biopsy
    - provides a definitive diagnosis
    - bilateral biopsies are sometimes necessary
- treatment
  - ophthalmic emergency
    - timely treatment with corticosteroids usually prevents an attack in the second eye

Granulomatosis with polyangiitis (GPA)
- Wegener granulomatosis
- necrotizing granulomatous small-vessel vasculitis
  - ± giant cells
- features
  - lesions of the upper respiratory tract
    - sinus mucosal involvement with bone erosion
  - lesions of the lower respiratory tract
    - tracheobronchial necrotic lesions
    - cavitary lung lesions
  - necrotizing glomerulonephritis
- ocular disease
  - 1. orbit and nasolacrimal drainage system involvement by extension from the surrounding sinuses
    - Figure 4-9 (Courtesy of American Academy of Ophthalmology)
  - 2. isolated orbital involvement
    - unilateral or bilateral
    - may lack frank necrotizing vasculitis
      - in the absence of respiratory tract and renal findings, may be difficult to diagnose
  - 3. scleritis
- limited forms of the disease
  - absent renal component
  - isolated orbital involvement
- diagnostic workup
  - serum immunofluorescence
    - antineutrophil cytoplasmic antibody (ANCA)
      - C-ANCA
        - autoantibodies directed against proteinase-3
        - diffuse granular fluorescence within the cytoplasm
        - highly specific for GPA
        - may be negative early in the course of the disease
          - especially in the absence of multisystem involvement
        - can also be detected by ELISA
      - P-ANCA
        - fluorescence surrounding the nucleus
        - an artifact of ethanol fixation
          - can be caused by autoantibodies against many different target antigens
            - nonspecific
        - needs to be confirmed by ELISA for ANCA reacting with myeloperoxidase (MPO-ANCA)
          - MPO-ANCA testing has a high specificity for small-vessel vasculitis
      - absolute levels of ANCA do not define disease severity or activity
        - changing titers can give a general idea of disease activity
- treatment
  - immunosuppression
    - cyclophosphamide
    - treatment with corticosteroids alone is associated with a significantly higher rate of mortality
  - long-term treatment with TMP-SMX appears to suppress disease activity in some patients
  - may have a fulminant, life-threatening course