9.6.13. Posterior Uveitis (VI): White Dot Syndromes (II)

Serpiginous choroiditis

- epidemiology
  - M=F
  - 2nd-7th decades
- pathogenesis
  - unknown
  - immune-mediated occlusive vasculitis
  - microorganisms may be a potential trigger
  - reported to occur in patients with
    - Crohn disease
    - sarcoidosis
    - PAN
- clinical presentation
  - symptoms
    - painless
    - paracentral scotomata
    - decreased vision
  - bilateral asymmetric
  - no AC reaction
  - minimal vitreous reaction
  - active areas
    - pseudopodial/geographic gray-white lesions at the level of the RPE projecting from the optic nerve
    - progressive centrifugal extension
    - disease activity is confined to the leading edge of the advancing lesion
    - less common
      - macular or peripheral lesions may present without parapapillary involvement
    - ← shallow subretinal fluid
    - ← vascular sheathing
    - ← RPE detachment
    - ← neovascularization of the disc
  - late findings
    - atrophy of the choriocapillaris, RPE, and retina
    - extensive RPE hypopigmentation and subretinal fibrosis
    - at the border of an old scar
    - CNV
      - ≤25%
- natural course
  - chronic progressive
    - new lesions & recurrent attacks are typical
  - final visual acuity of 20/200 - counting fingers
    - ≤39%
- paraclinical evaluation
  - FA
    - early phase
      - blockage of the choroidal flush
    - late phase
      - staining of the active edge of the lesion
      - early hyperfluorescence with late leakage is indicative of CNV
  - ICG angiography
    - hypofluorescence throughout all phases of the study for both acute and old lesions
    - may reveal more extensive involvement than FA or clinical examination
    - CNV
      - localized hyperfluorescence during the middle to late phases
      - useful in distinguishing active lesions from CNV
  - FAF imaging
    - sensitive for detecting RPE damage and monitoring disease course
    - hyperfluorescence highlights active disease
    - hypoautofluorescence corresponds closely to regressed disease
  - OCT imaging
    - outer retinal reflectivity and disruption
      - active disease
      - atrophic changes in the retina and RPE
      - regressed disease
- serpiginous-like choroiditis
  - Mycobacterium tuberculosis
    - can cause inflammation that simulates typical serpiginous choroiditis
    - patients exposed to active pulmonary TB
      - countries where TB is endemic
    - tuberculin skin testing
    - interferon-gamma release assay
    - chest X-ray often appears normal
    - lesions appear and progress in multiple areas
      - unilateral
      - involve posterior pole, mid-periphery, and periphery
      - usually sparing the juxtapapillary area until late in the disease
      - more prominent vitreous cellular reaction
    - responds to anti-TB treatment
      - complete control may take months
      - ← corticosteroids
  - herpetic choroiditis
  - syphilitic choroiditis
- treatment
  - systemic, periocular, and intravitreal corticosteroids
    - for active lesions
      - particularly lesions threatening the fovea
    - corticosteroids alone are ineffective
    - patients require prolonged anti-inflammatory therapy
  - systemic IMT
    - cyclosporine monotherapy
      - may induce rapid remission of acute disease
      - prolonged therapy is required
      - disease recurrence is frequently observed as drugs are tapered
    - triple therapy with prednisone, cyclosporine, and azathioprine
    - cyclophosphamide
    - chlorambucil
      - cytotoxic therapy
      - induce long, drug-free remissions
      - patients intolerant of systemic therapy
  - intravitreal fluocinolone acetonide implant
  - CNV treatment
    - intravitreal anti-VEGF drugs
    - focal laser photocoagulation
    - photodynamic therapy
- references

Figure 6-32 (© 2020 American Academy of Ophthalmology)

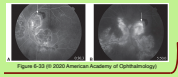

Figure 6-33 (© 2020 American Academy of Ophthalmology)

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

- epidemiology
  - healthy young adults
    - 2nd-3rd decade
  - M = F
    - similar to serpiginous & AIRPE
    - unlike other WDSs
  - influenza-like prodrome (50%)
- pathogenesis
  - genetic predisposition
    - HLA-B7
    - HLA-DR2
  - immune-mediated vascular alteration
    - choroidal/choriocapillaris perfusion abnormalities with secondary involvement of the RPE/photoreceptors
  - noninfectious systemic associations
    - erythema nodosum
    - cerebral vasculitis
      - potentially life-threatening
      - should undergo urgent neurologic evaluation
    - scleritis and episcleritis
    - GPA
    - PAN
    - sarcoidosis
    - ulcerative colitis
  - infectious systemic associations
    - group A streptococcal infection
    - adenovirus type 5 infection
    - tuberculosis
    - Lyme disease
    - mumps
    - hepatitis B vaccination
- clinical presentation
  - natural course
    - sudden onset
    - resolve over a period of 2–6 weeks
    - new peripheral lesions may appear in a linear/radial array over the next 3 weeks
    - fellow eye becomes involved within days to weeks
      - bilateral
  - symptoms
    - vision loss
      - central and paracentral scotomata
      - photopsias may precede loss of vision
  - multiple large, flat, yellow-white placoid lesions at the level of the RPE
    - 1–2 disc areas
    - throughout the posterior pole to the equator
    - resolve over a period of 2–6 weeks
      - permanent, well-defined RPE alterations (depigmentation and pigment clumping)
      - rapid evolution of the pigmentary changes, often over days, is a typical feature
  - ← papillitis
  - ← mild vitritis
  - atypical findings
    - retinal vasculitis
    - retinal vascular occlusive disease
    - retinal neovascularization
    - exudative retinal detachment
    - minimal or no anterior uveitis
    - CME
      - uncommon
- lab evaluation
  - FA
    - acute phase
      - early hypofluorescence
      - typically more numerous than on funduscopy
      - late hyperfluorescent staining
    - subacute phase
      - increased central hyperfluorescence with late staining
    - resolution phase
      - transmission defects
  - ICG angiography
    - choroidal hypofluorescence
      - typically more numerous than on funduscopy
      - become smaller in the inactive stages
    - hypervisualization of the underlying choroidal vessels in both the acute and inactive stages
  - FAF imaging
    - RPE alterations
      - appear during recovery well after the choroid is affected
      - are fewer in number
  - OCT
    - acute lesions
      - hyperreflectivity of the outer retinal layers
      - subretinal or intraretinal fluid
    - resolving lesions
      - outer retinal and photoreceptor loss
- differential diagnosis
  - choroidal metastasis
  - viral retinitis
  - sarcoidosis
  - VKH syndrome
  - pneumocystis choroiditis
  - serpiginous choroiditis
    - insidious and progressive
    - APMPPE is acute, usually nonrecurring disease!
- treatment
  - systemic corticosteroids
    - may not alter visual outcome
    - some authorities advocate treatment in patients with
      - extensive macular involvement
      - associated CNS vasculitis
  - visual acuity returns to ≥20/40 within 6 months
    - 20% are left with residual visual dysfunction
  - risk factors for loss of vision
    - foveal involvement at presentation
    - older age
      - APMPPE is a disease of the young
    - unilateral disease
      - APMPPE is bilateral
    - longer interval between initial and fellow eye involvement
      - Usually days–weeks
    - recurrence
      - APMPPE is usually non-recurrent

Figure 6-27 (© 2020 American Academy of Ophthalmology)

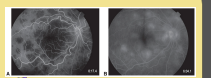

Figure 6-28 (© 2020 American Academy of Ophthalmology)

Figure 6-29 (© 2020 American Academy of Ophthalmology)

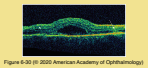

Figure 6-30 (© 2020 American Academy of Ophthalmology)

Ophthalmology Mind Maps/Arman Mashayekhi, MD