Birdshot Retinochoroidopathy

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Overview

- Definition:
  - Birdshot retinochoroidopathy (BSRC) is a clinically distinct, uncommon form of posterior uveitis, characterized by vitritis, retinal vasculitis, and multiple, bilateral, hypopigmented, postequatorial inflammatory lesions at the level of RPE and choroid
  - Patient's visual complaints are often out of proportion to the measured visual acuity, and fundus findings can be extremely subtle in early disease stage
- Symptoms
  - Floaters
  - Photophobia
  - Photopsia
  - Glare
  - Some degree of nyctalopia
  - Reduced contrast sensitivity
  - Blue-yellow dyschromatopsia and other color vision disturbances
- Laterality
  - Bilateral
- Course
  - Chronic, characterized by multiple exacerbations and remissions
  - Self-limited in only 20% of cases
- Age of onset
  - 40–50 years
- Gender/race
  - F:M = 3:1
  - Almost exclusively Caucasian, with a higher incidence in those of northern European descent

C. S. Foster et al. (eds.), *Uveitis*, https://doi.org/10.1007/978-3-030-52974-1_15

- Systemic association
  - HLA-A29 association is perhaps most well known in ophthalmology, if not all of medicine
  - Though not as prevalent as HLA-A29, HLA-B44 is found in 50% of BSRC patients
  - Systemic hypertension appears to be more prevalent in BSRC population

Exam: Ocular

Anterior Segment

- Quiet eye without conjunctival injection or ciliary flush
- Mild nongranulomatous anterior uveitis (≤1+ cells) without synechia
- +/− fine keratic precipitates
- Normal IOP

Posterior Segment

- BSRC lesions
  - Multiple, cream-colored, choroidal lesions with indistinct borders, the long axis of which is radial to the optic disc scattered throughout the postequatorial retina
  - Round to ovoid in shape, varying in size from 50 to 1500 μm
  - More easily visualized clustered around the optic disc, in the inferonasal quadrant
  - With time, lesions may become confluent, producing large areas of geographic depigmentation
- Mild-to-moderate vitritis (≤2+ haze)
- Retinal vasculitis (predominantly phlebitis) typically in the absence of visible vascular sheathing
- Retinal arteriolar attenuation and vascular tortuosity may be seen
- CME is the most common structural complication of BSRC (cumulative incidence approaching 84% over 5 years) and is the most frequent cause of reduced central VA
- ERM is common
- CNV and retinal NV leading to NV are relatively rare (<10%)
- Optic nerve head swelling and nerve fiber layer hemorrhages

Imaging

- FA
  - Macular leakage
  - Vasculitis (predominantly phlebitis)
  - Generalized abnormal hyperfluorescence

- ICG
  - Hypofluorescent spots and fuzzy vessels in *active* disease
  - Hypofluorescence often not seen in treated or burn-out disease
  - May or may not correspond to fundus lesions seen on microscopy
- Fundus autofluorescence
  - Abnormal in 80% of patients
  - Peripapillary, macular, or extramacular hypoautofluorescence
- OCT
  - CME and ERM
  - IS/OS attenuation
  - EDI-OCT may show choroidal lesions otherwise not apparent on ophthalmoscopy
- ERG
  - Early (inner retinopathy): loss of oscillatory potentials and b-wave amplitude
  - Late (outer retinopathy): reduction of photopic b-wave amplitude and progressive prolongation of 30 Hz implicit time (IT), used for follow up the effect of treatment
- Blue-on-yellow perimetry (SITA-SWAP)
  - Generalized constriction of the peripheral visual field
  - Central and paracentral scotomata
  - Enlargement of the blind spot
  - Abnormalities on visual field testing may occur even among minimally symptomatic patients with good VA

Laboratory Testing

- HLA-A29 phenotype
  - Present in 7–8% of general population
  - 95–100% of BSRC patients
  - Confers 50–224 times risk of developing BSRC
- Labs to rule out the masqueraders (see below)

Differential Diagnosis

- Sarcoidosis
- Intraocular lymphoma
- VKH
- Tuberculosis
- Syphilis
- POHS
- White dot syndromes: MCP, PIC, MEWDS, APMPPE

Treatment

- Periocular and systemic corticosteroids have inconsistent treatment efficacy and provide only short-term reduction in vitritis and CME, and thus should be used only for acute exacerbations
- A variety of immunomodulatory agents including methotrexate, mycophenolate, cyclosporine, azathioprine, cyclophosphamide, chlorambucil, intravenous immunoglobulin (IVIg), TNF-alpha inhibitors, and anti-IL6 receptor have been employed as part of a steroid-sparing strategy in the treatment of BSRC
- We typically are following this order of therapy in the majority of our BSRC patients:
  1. Combination therapy of mycophenolate mofetil (1–3 g/day) and cyclosporine (100–300 mg/day)
  2. TNF-alpha inhibitors: infliximab 5–10 mg/kg administered every 4 weeks is very effective
  3. Fluocinolone acetonide implant:
     Higher likelihood of concurrent or subsequent glaucoma incisional surgery in BSRC patients

Referral/Comanagement

- Rheumatology