Pars Planitis

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Overview

- Definition
  - A subset of intermediate uveitis that is not associated with a systemic disease or infection and characterized by the presence of snowballs in the vitreous and snowbanks along the inferior pars plana and retinal periphery
  - After idiopathic and JIA, it is the third most common etiology of pediatric uveitis
- Symptoms
  - Floaters
  - Blurry vision
- Laterality
  - >75% bilateral, but can be asymmetric
- Course
  - Gradual onset
  - Chronic with low incidence of remission
- Age of onset
  - Most commonly childhood to adolescence, but also older adults in their 40s
- Gender/race
  - Equal gender distribution in pediatric population; slight female preponderance in older cases
  - Caucasians
- Systemic association
  - By definition, pars planitis is not associated with any systemic disease

C. S. Foster et al. (eds.), *Uveitis*, https://doi.org/10.1007/978-3-030-52974-1_11

Exam: Ocular

Anterior Segment

- Mild to moderate AC inflammation
- Band keratopathy
- Keratic precipitates
- Posterior synechiae
- Peripheral corneal endotheliopathy
- Cataracts

Posterior Segment

- Vitreous haze and cells
- Snowballs/snowbanks (65–98%): must indent sclera inferiorly
- Retinal vasculitis (17–90%)
- Optic nerve leakage (70%)
- CME and ERM common
- Retinal and optic disc neovascularization
- Vasoproliferative tumor
- Retinoschisis (children only)
- Rhegmatogenous/tractional/exudative RD
- Glaucoma requiring surgery is rare

Exam: Systemic

- No systemic findings in true pars planitis, but must keep in mind of DDx (below) and ask pertinent questions

Imaging

- OCT
  - CME
  - ERM
- FA
  - Retinal vasculitis (may be only visible on wide-field FA)
  - Optic nerve leakage
  - Macular leakage

Laboratory and Radiographic Testing

Pars planitis is a diagnosis of exclusion, so systemic diseases must be ruled out:

- Multiple sclerosis: MRI brain/spine, HLA-DR15, and DR2
- Sarcoidosis: ACE, lysozyme, CT chest, Gallium scan
- Syphilis: RPR, FTA-ABS
- Lyme and Bartonella serologies
- Tuberculosis: chest radiograph, PPD, QuantiFERON
- Intraocular lymphoma: MRI brain, diagnostic vitrectomy

Differential Diagnosis

- Multiple sclerosis
- Sarcoidosis
- Syphilis
- Lyme disease
- Cat scratch disease
- Tuberculosis
- Whipple's disease
- Intraocular lymphoma (elderly patients)
- Other conditions that can mimic intermediate uveitis
  - Anterior uveitis with spill-over cells: FHI, HLA-B27
  - Mild vitritis with subtle chorioretinitis: Behcet's, VKH, Eales disease
  - Severe vitritis with visually obscured chorioretinitis: toxoplasmosis, toxocariasis, ARN, endophthalmitis

Treatment

- Treatment is indicated when there is reduced vision, significant vitreous opacities, macular edema, or retinal vasculitis
- Periocular corticosteroid injection (triamcinolone 40 mg/1 ml) with topical steroids if AC inflammation is also present; supplement with oral corticosteroids if necessary
- After three recurrences in the affected eye(s), or in presence of steroid-induced OHTN/glaucoma, or if inflammation is refractory to corticosteroids, treatment should be escalated. Four options:
  1. Peripheral retinal cryopexy or indirect laser photocoagulation to the snowbanks of the inferior pars plana
     - Induces regression of vitreous base neovascularization and consequently stabilizes inflammation

2. Steroid-sparing IMT
   - When disease is bilateral, and/or there is significant retinal vasculitis
   - Anti-metabolites and cyclosporine are first line
   - Biologics including adalimumab, infliximab, and tocilizumab are all effective
     - Keep in mind of association of pars planitis and intermediate uveitis with MS, as TNF-alpha inhibitors – and perhaps tocilizumab as well – can trigger or unmask demyelination
     - Must conduct careful neuro-ROS and inquire about family history of MS and other demyelinating disorders
     - When in doubt, consider neurology consult and MRI
   - Preferred over PPV/cryo/laser (see below) if disease is bilateral or if there is significant retinal vasculitis
3. Pars plana vitrectomy with cryopexy/endolaser
   - When disease is unilateral or asymmetric bilateral, or when there are visually significant vitreous opacities or concurrent VR complications (VH, ERM, TRD)
   - In our experience, this approach alone rarely results in disease quiescence in the presence of retinal vasculitis, but may reduce IMT burden
   - Stronger anti-inflammatory effects than cryopexy/laser photocoagulation along, by way of removing the vitreous which serves as a cytokine scaffold
   - Cryopexy is favored over endolaser in phakic patients to avoid peripheral instrument-crystalline lens touch
4. Fluocinolone acetonide 0.59 mg implant (Retisert)
   - If all else fail, but cataract is a guarantee and glaucoma requiring surgery is likely

Referral/Co-management

- Rheumatology