Sympathetic Ophthalmia

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Overview

- Definition
  - Bilateral granulomatous panuveitis that develops after ocular surgery or penetrating trauma to one eye, causing disruption to the immune privilege of the eye
- Symptoms
  - Redness
  - Photophobia
  - Pain
  - Blurry vision
  - Floaters
- Laterality
  - Bilateral
- Course
  - Average time between injury/surgery to onset of SO: 2 weeks to 3 months (range: 5 days to 66 years).
    90% of cases manifest within 12 months of insult.
  - Onset can be insidious in the sympathizing, noninjured eye.
  - Severity of inflammation and its sequelae are wide-ranging, and relapsing nature of SO requires long-term monitoring.
  - 75% of patients retain ≥20/200.
- Age of onset
  - All ages affected
- Gender/race
  - Males more affected, likely due to higher risk of ocular injury
  - No racial predisposition

C. S. Foster et al. (eds.), *Uveitis*, https://doi.org/10.1007/978-3-030-52974-1_16

- Systemic association
  - Patients with SO are more likely to express HLA-DR4, HLA-DQw3, HLA-DRw53 (also seen in VKH).
  - VKH-like integumentary changes (poliosis and vitiligo) have been reported, but very uncommon.

Exam: Ocular

Anterior Segment

- Mild-to-severe anterior uveitis with mutton-fat precipitates.
- Corneal endothelium may decompensate with chronic inflammation → bullous keratopathy.
- Posterior synechiae.
- Secondary cataract is common.

Posterior Segment

- Mild-to-moderate vitritis
- Dalen-Fuchs nodules: multiple yellowish-white choroidal lesions in the periphery (also seen in VKH and sarcoidosis)
- Diffuse choroiditis
- Papillitis
- Exudative RD
- Subretinal fibrosis
- Retinochoroidal and optic atrophy

Exam: Systemic (Uncommon)

- Vitiligo
- Hearing dysfunction

Imaging

- OCT
  - Varied disruptions of the outer retinal segments
  - Subretinal fluid corresponding to exudative RD
  - Intraretinal edema and thickening
  - Diffuse choroidal thickening best seen on EDI-OCT: may be used to monitor disease and treatment response

- FA
  - Multiple hyperfluorescent leakage at the level of RPE during the venous phase that persist into the late phase
  - Dye pooling in subretinal spaces in severe cases
  - Areas of early blocked fluorescence corresponding to Dalen-Fuchs nodules
- ICG
  - Multiple hypofluorescent foci that become more prominent as angiography progresses
- B-scan
  - Marked choroidal thickening

Laboratory and Radiographic Testing

- HLA typing may help confirm diagnosis.
- Labs are done to rule out DDx:
  - ACE/lysozyme
  - PPD or QuantiFERON-Gold
  - FTA-ABS/RPR

Differential Diagnosis

- VKH (Table 16.1)
- Sarcoidosis
- Syphilis
- Tuberculosis
- Intraocular lymphoma

Table 16.1 Comparison of sympathetic ophthalmia (SO) and Vogt-Konayagi-Harada syndrome (VKH)

Characteristics	SO	VKH
Age	All ages	20–50 years
Racial predisposition	None	Asia and black
Penetrating injury	Always present	Absent
Skin changes	Uncommon	Common (60–90%)
CNS findings	Uncommon	Common (85%)
Hearing dysfunction	Uncommon	Common (75%)
Optic nerve inflammation	Occasional	Frequent
Exudative RD	Rare	Frequent
Choriocapillaris involvement	Usually absent	Frequent
CSF findings	Usually normal	Pleocytosis (84%)

Treatment

- Corticosteroids – both systemic and local – should be instituted as soon as possible.
- Steroid-sparing IMT should be started at time of diagnosis, as inflammation is certain to relapse upon steroid discontinuation.
  - Cyclosporine, azathioprine, mycophenolate mofetil, chlorambucil, cyclophosphamide, and infliximab have all shown efficacy.
- Fluocinolone acetonide (Retisert), if IMT not effective or not tolerated.
- Enucleation may lower chance of SO if done within 2 weeks of open globe injury; ineffective after development of autoimmune inflammation.

Referral/Comanagement

- Rheumatology