Sympathetic Ophthalmia Two Weeks After 23-Gauge Vitrectomy

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Abstract

Sympathetic Ophthalmia (SO) is a rare disease that presents as a bilateral, diffuse, granulomatous panuveitis. Sympathetic Ophthalmia is a clinical diagnosis with history of penetrating ocular injury in the inciting eye and presence of panuveitis in the sympathizing eye. Though early enucleation is believed to minimize the risk, there have been reports of SO even after enucleation of inciting eyes. The possible association between vitrectomy and SO has been initially proposed by Gass 9 and later studied extensively in a large cohort in the UK with an estimated SO risk of 1 in 799 vitrectomies 11.

There have been several case series and reports of SO following vitrectomy, however only three documented cases of SO following vitrectomy without use of silicone oil. These cases demonstrated an onset of SO ranging between 4 weeks to 2 months. We present a patient with SO in the sympathizing eye presenting 16 days after an uncomplicated 23-gauge (23G) sutureless pars plana vitrectomy (PPV) without the use of silicone oil.

Case Report

Sympathetic Ophthalmia (SO) is a rare disease that presents as a bilateral, diffuse, granulomatous panuveitis. Sympathetic Ophthalmia is a clinical diagnosis with history of penetrating ocular injury in the inciting eye and presence of panuveitis in the sympathizing eye. Though early enucleation is believed to minimize the risk, there have been reports of SO even after enucleation of inciting eyes. The possible association between vitrectomy and SO has been initially proposed by Gass [9] and later studied extensively in a large cohort in the United Kingdom with an estimated SO risk of 1 in 799 vitrectomies [11]. There have been several case series and reports of SO following vitrectomy, however only three documented cases of SO following vitrectomy without use of silicone oil with an onset of SO ranged between 4 weeks to 2 months. We present a patient with SO in the sympathizing eye presenting 16 days after an uncomplicated 23-gauge (23G) sutureless pars plana vitrectomy (PPV) without the use of silicone oil.

A 60-year-old Indian male presented with macula-off retinal detachment with multiple tears in his left eye (OS) and underwent 23G pars plana vitrectomy, endodiathermy, endodrainage, endolaser photocoagulation, and 15% octafluoropropane gas (C3F8) OS. His past medical history was significant
for thyroid cancer and total thyroidectomy and past ocular history was significant for cataract extraction and posterior chamber intraocular lens (PCIOL) implantation OS nine years ago with no history of intraocular trauma or surgery in the right eye (OD). His postoperative course was relatively unremarkable until he reported relative scotoma in the fellow eye (OD) on postoperative day 16. On examination, his visual acuity was hand-motion OD (from a baseline of 20/30-2) and counting-fingers (CF) OS from the postoperative gas bubble. Slit lamp exam (SLE) revealed no anterior chamber (AC) cells or flare OD, rare AC cell without flare OS and dilated fundus examination (DFE) showed serous retinal detachment of the macula with no disc edema, vascular sheathing, or vitritis OD (Fig. 1), and an attached retina with approximately 70% C3F8 gas fill OS. Optical coherence topography (OCT) confirmed a serous macular detachment with nasal pigment epithelial detachment (PED) (Figure 2a).

The patient was initially treated with acetazolamide (Diamox) for presumptive Central Serous Chorioretinopathy (CSR) with initial improvement in subretinal fluid (SRF) and visual acuity (OD: 20/200) (Figure 2b). However, two weeks later, the patient developed new bilateral granulomatous panuveitis with bilateral 1+ conjunctival injection, AC cell and flare, 2+ vitreous cells with haze OD greater than OS and disc edema OS. Quantiferon gold test was negative. No systemic associations were found, including sensorineural hearing loss, tinnitus, fever, headache, vertigo, vitiligo, poliosis or alopecia. In addition, there was no ocular depigmentation appreciated on exam. These findings along with a history of surgery preceding the uveitis suggests sympathetic ophthalmia and not Vogt-Koyanagi-Harada (VKH). He was diagnosed with Sympathetic Ophthalmia and he was started on high dose prednisone (80mg PO Daily). Peripheral retinal ischemia and a rhegmatogenous retinal detachment (RRD) developed in the right eye and so he was converted to intravenous pulse steroids for three days in order to quiet the inflamed eye prior to retinal detachment repair.

A fluorescein angiography (FA) performed prior to starting intravenous (IV) steroids showed notable hyperfluorescence of the optic nerves in both eyes and significant inferotemporal nonperfusion with pruning of vessels temporally OD (Figure 3). A combined cataract extraction, PCIOL insertion and retinal detachment repair with PPV, endolaser, air-fluid exchange and C3F8 gas was performed on the right eye approximately 6 weeks after the left eye retinal detachment repair was performed.
Figure 1: a) Color fundus photograph of the right eye b) red free of the right eye show hypopigmented/yellowish areas c) Fluorescein angiogram in the early phase demonstrates patchy hypofluorescence early and d) late leakage and pool corresponding to areas of yellow hypopigmentation.

Figure 2: a) Optical coherence topography of the right eye demonstrating a large macular serous detachment. b) Optical coherence topography of the right eye demonstrating improvement in macular serous detachment after Acetezolamide.

Figure 3: a) Color fundus photograph of the right eye with inferotemporal RRD macula off. b) Color fundus photograph of the left eye with macula attached and periphery with laser scars more evident temporally. Mid-periphery chorioretinal scar from drainage and superior C3F8 bubble with 40% fill. c) Fluorescein angiography of the right eye late images demonstrating disc hyperfluorescene, temporal nonperfusion and vascular staining and leakage in the area of the retinal detachment. Mild macular hyperfluorescene is present. D) FA of the left eye, late images demonstrating disc staining, and no other abnormal hyperfluorescence other than staining of postsurgical laser scars.

Over the next year, the patient was managed at the National Institute of Health (NIH) with systemic immune suppression, topical and local anti-inflammatory treatment to the left eye. At the one year follow-up, the patient’s vision is OD: 20/200 and OS: 20/25-1. An OCT OD shows an epiretinal membrane (ERM) with loss of foveal contour, mild intraretinal edema and significant irregularity of the retinal pigment epithelium (RPE) (figure 4).

Figure 4. OCT right eye, one year after initiation of treatment for panuveitis from SO. ERM without significant inner retinal wrinkling, loss of foveal contour, mild intraretinal edema, significant RPE irregularity.

Sympathetic Ophthalmia has been associated with specific major histocompatibility complex (MHC) and patients are likely to express human leukocyte antigen (HLA) DR4 (closely related to HLA-DQw3 and HLA-DRw53) phenotype [2]. These specific marker associations suggest a role for immune dysregulation, increased susceptibility, and increased severity associated with pathogenesis [2].
Although the etiology is not clearly understood, it encompasses autoimmunity and cell-mediated immune mechanisms. It has been postulated to be a delayed-hypersensitivity reaction [2,4]. Interestingly our patient was positive for HLA-DR4.

The eye is considered an immune privileged site and SO requires sequestered ocular antigens to be exposed systemically, likely from a scleral perforation, whether surgical or traumatic. Though a link between SO and vitrectomy was made in the 1980s, only a few cases of SO have been reporter following uncomplicated vitrectomy without the use of silicone oil or without antecedent trauma or endophthalmitis. In most cases the average time of onset of SO was 2 months to one year [13]. There are only two cases of SO [1,13] that occurred 5 weeks to 2 months after small incision PPV without the use of silicone oil for tamponade. To the best of our knowledge, our case presented here represents the earliest case of SO following 23G sutureless vitrectomy for RD without silicone oil and antecedent trauma with an onset at 16 days following surgery. Sympathetic Ophthalmia should be considered in cases of new onset posterior uveitis soon after vitrectomy.

Declarations

**Ethics approval and consent to participate**

Ethics and Institutional Review Board approval and consent was obtained by the George Washington University Office of Human Research.

**Consent for publication**

Consent for publication was obtained from the all study subjects and participants.

**Availability of data and material**

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

**Competing interests**

The authors declare that they have no competing interests.

Each of the authors involved in this study have no proprietary interest.

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**Authors’ contributions**

All of the authors contributed to examining, collecting and analyzing the patient data found in the manuscript. All author contributed to writing the manuscript. All authors read and approved the final manuscript.

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**Abbreviations**

SO = sympathetic ophthalmia

23G = 23 gauge

PPV = pars plana vitrectomy

OS = left eye

OD = right eye

C3F8 = octafluoropropane gas

PCIOL = posterior chamber intraocular lens

SLE = slit lamp exam

AC = anterior chamber

DFE = dilated fundus examination

OCT = Optical coherence topography

PED = pigment epithelial detachment

CSR = central serous chorioretinopathy

SRF = subretinal fluid

VKH = Vogt-Koyanagi-Harada

FA = fluorescein angiography

IV = intravenous

RRD = rhegmatogenous retinal detachment

NIH = National Institute of Health
RPE = retinal pigment epithelium

MHC = major histocompatibility complex

HLA = human leukocyte antigen

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Figures
Figure 1

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Figure 4

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