

# Sympathetic Ophthalmitis: A Case Presentation and Review of the Literature

P. S. Mahar, Aimal Khan, Dilshad Laghari, Sadaf Ambreen

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See end of article for  
authors affiliations

Correspondence to:  
P. S. Mahar  
Isra Postgraduate Institute of  
Ophthalmology,  
Al-Ibrahim Eye Hospital, Malir,  
Karachi

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Sympathetic Ophthalmitis (SO) is a rare, bilateral granulomatous uveitis occurring after perforating eye injury or ocular surgical procedure to one eye. The pathophysiology of this entity is not clearly understood but an autoimmune hypersensitivity reaction against exposed ocular antigens in the injured eye is believed to be responsible for this disease.

In this article, we present a patient with clinical diagnosis of SO and review the literature.

**S**ympathetic Ophthalmitis (SO) is defined as a bilateral granulomatous uveitis of unknown etiology, occurring after penetrating trauma or intraocular surgery. It is believed to represent a form of autosensitization of ocular tissue following a perforating injury to one eye. Its exact incidence is not known but is thought to be 1.9% after trauma and 0.007% after intraocular surgery<sup>1</sup>.

SO is an uncommon disease due to improved surgical techniques employed in the repair of ocular injuries and early enucleation of the blind eye. The risk of developing SO in severely traumatized eyes with no visual potential that are not enucleated is exceedingly low<sup>2</sup>.

Numerous surgical procedures such as 23-gauge vitrectomy<sup>3</sup>, cataract extraction<sup>4</sup>, retinal detachment surgery<sup>5</sup>, penetrating keratoplasty<sup>6</sup> and trabeculectomy<sup>7</sup> have been complicated by SO.

We report a case of SO seen in our outpatient department (OPD) at Isra Postgraduate Institute of Ophthalmology / Al-Ibrahim Eye Hospital, Karachi.

## CASE REPORT

A 30 year old male, plumber by profession was seen in OPD with complaint of gradual painful loss of vision in his right eye of 10 days duration. About 25 days back, this patient sustained penetrating trauma to his left eye with a piece of wood. The patient did not have any significant medical and surgical history. On examination, his best corrected visual acuity (BCVA) was Hand movements (HM) in right eye and no perception of light (NPL) in left eye. His right eye, on biomicroscopic examination showed multiple mutton-fat keratic precipitates (KPs) with 2-3 plus cells and flare in the anterior chamber (Fig. 1 & 2). The pupil was fixed and dilated with multiple posterior synechie (PS) formation. The intraocular pressure (IOP) measured 12 mm Hg. The fundus examination revealed optic disc edema and serous retinal detachment (Fig. 3). The patient's left eye was pthysical with vertical full thickness corneal laceration (Fig. 4). The patient went under laboratory investigations of complete blood count (CBC), erythrocyte sedimentation rate (ESR), rapid plasma regain (RPR), venereal disease research

laboratory (VDRL), fluorescent treponemal antibody absorption test (FTA - ABS), toxoplasmosis IgG and IgM, angiotensin converting enzyme (ACE) and antinuclear antibody (ANA) test. Patient also had X-ray chest, B-Scan (Fig. 5), fundus fluorescein angiography (FFA), (Fig. 6) and optical coherence tomography (OCT). All laboratory tests including X-ray chest were within normal limits. A clinical diagnosis of SO was entertained and patient was commenced on tablet prednisolone 60 mg / day in divided doses (1 mg / kg body weight), prednisolone 1% drops (Predforte- Allergan, Pakistan), several times a day and atropine 1% twice a day (Ophth-Atropine - Ophth, Pakistan).

At two weeks patient's vision had improved to 6/24 with anterior chamber getting quite and reduction in the number of KPs. The optic disc margin, though still appeared blurred but swelling had subsided significantly. Subretinal serous fluid also had decreased with B-Scan appearing normal. Patient's systemic and topical treatment continued.

At four week follow up, patient's BCVA appeared stable at 6/18 on Snellen's quotations.

## DISCUSSION

The etiology of SO has not been completely understood, but the underlying pathophysiology is believed to be an autoimmune reaction against the exposed ocular antigens from the inciting eye<sup>8</sup>. The location of such antigens remains controversial and may be found in the uveal tissue, retina or choroidal melanocytes. The immunologic studies have shown CD4 helper and inducer T cells during the early phase of inflammation compared to infiltration by CD8 suppressor and cytotoxic T cells in the later stage. There are B lymphocytes also found in some patients<sup>9</sup>.

Lymphocytes from patients with SO were demonstrated to respond to several uveo-retinal antigens. Although no circulating antiretinal S-antigen antibodies were found, the serum from patients with SO showed antiretinal antibodies directed against the outer segment of photoreceptors and the Muller cells, when placed over normal human retinal tissue<sup>10</sup>.

It has also been hypothesized that a purulent infection within the eye would destroy the uveal tissue in such a way that SO would not develop. However some cases have been reported in eyes with endophthalmitis or fungal keratitis, indicating that the infection may not offer any prevention against development of SO<sup>11</sup>.

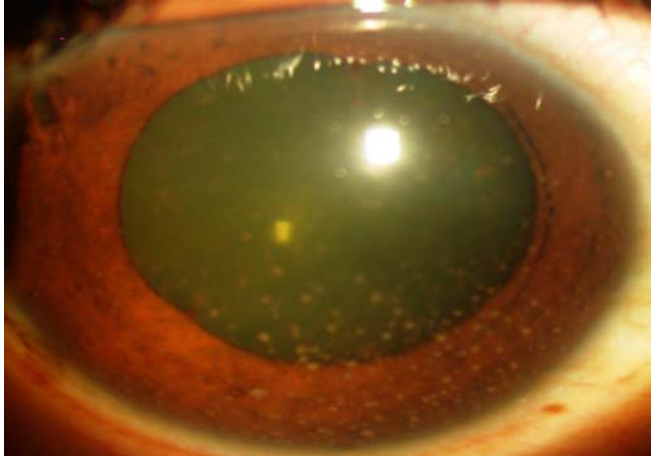
The SO can occur between two weeks and three months after an ocular injury, although it can develop as early as several days and as late as 50 years, majority of cases present within first three months. Classically, the inflammation is granulomatous with multiple mutton-fat KPs adhered to corneal endothelium. The iris can be thick and sticky with PS formation. The IOP can be normal or fluctuating upwards or downwards due to the inflammatory involvement of ciliary body and trabecular meshwork. The vitreous is usually infiltrated with moderate to severe cellular reaction. The fundus can show swollen optic disc and multiple yellow-white lesions in the periphery, corresponding to the presence of Dalen-Fuchs nodules, which may not be seen in almost 50% of the cases. Serous retinal detachment or macular edema may be present with subretinal neovascularization.

On fundus fluorescein angiography (FFA), the optic nerve head shows hyperemia and dye leakage more pronounced in the late frames. There are multiple hyperfluorescent areas of choroidal leakage corresponding to the presence of Dalen-Fuchs nodules. The less common appearance on FFA is that of early hyperfluorescent lesions with staining in the late phase. This type of picture is thought to be related to whether the Dalen-Fuchs nodules have an intact or disrupted over lying retinal pigment epithelium (RPE)<sup>12</sup>.

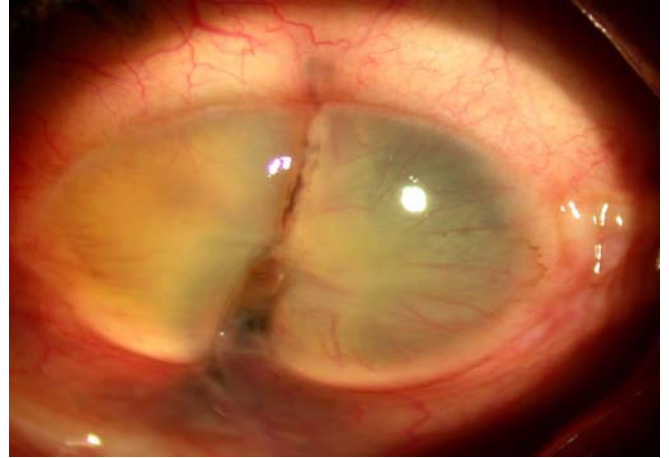
Extra ocular findings such as pleocytosis of cerebro-spinal fluid, hearing loss, alopecia, poliosis and vitiligo have been reported with SO, although these findings are more common in Vogt Koyanagi Harada (VKH) disease.

The sequelae of the ocular inflammation include secondary glaucoma, cataract, optic atrophy, retinal detachment with subretinal fibrosis and choroidal atrophy.

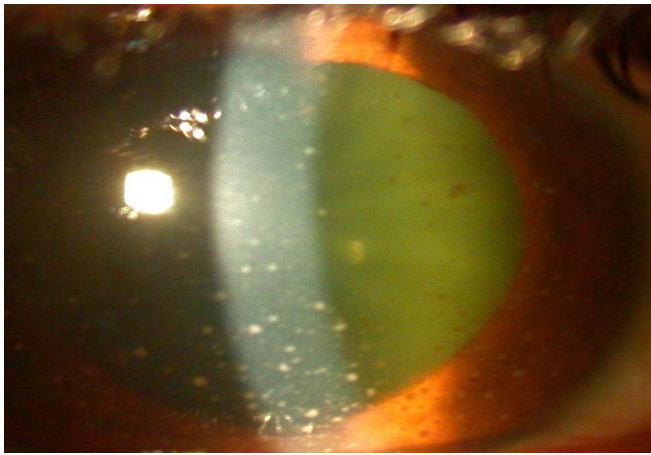
SO is characterized by a diffuse granulomatous, non-necrotizing inflammation involving entire uveal tract. The choroid is thickened with lymphocytic infiltration along with the presence of eosinophils and plasma cells. Typically, the choriocapillaris is spared. The Dalen-Fuchs nodules representing migrated and transformed RPE cells are typical but not pathognomonic and may be present in other disease such as: VKH syndrome. These nodules are collection of epitheloid histocytes and lymphocytes, present between RPE and Bruch's membrane<sup>13,14</sup>.



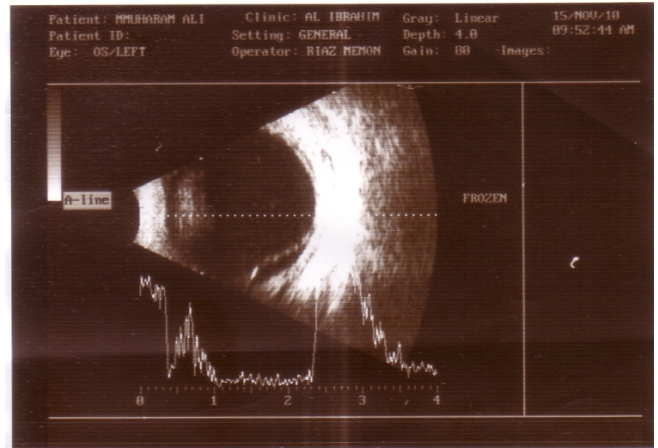
**Fig. 1.** Multiple mutton-fat keratic precipitates in right eye



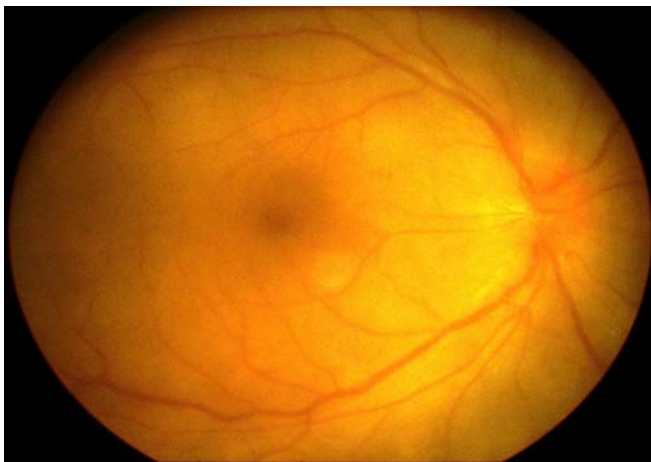
**Fig. 4.** Severe lacerated cornea with pthysical left eye



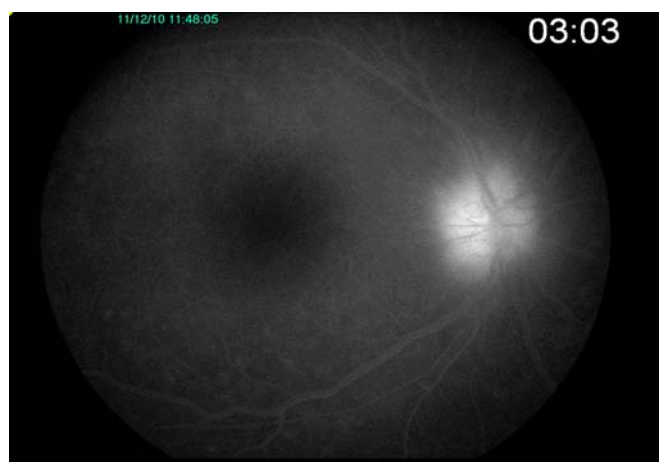
**Fig. 2.** Multiple mutton-fat keratic precipitates in right eye seen with slit beam



**Fig. 5.** Serous retinal detachment in right eye on Ultrasonic B-scan



**Fig. 3.** Swollen optic disc and serous retinal detachment in right eye



**Fig. 6.** Hyperfluorescent disc in late venous phase in right eye

It is important to rule out the other causes of granulomatous uveitis before a diagnosis of SO can be entertained. Although diagnosis of SO is clinical, histopathology can be confirmatory. Autoimmune disease like VKH, sarcoidosis, and multifocal choroiditis can have similar presentation. Intraocular lymphoma and bilateral phacoanaphylaxis can also have a similar picture. Infections like tuberculosis and syphilis should always be excluded.

## CONCLUSION

SO is a rare but a significant complication of penetrating ocular injury. In addition to systemic and intravitreal steroid therapy, immunosuppressive drugs also play a significant role in the medical management of this disease. The patient's medical treatment needs to be carefully monitored to reduce any side effects and improve visual prognosis.

### Author's affiliation

P. S. Mahar  
Isra Postgraduate Institute  
of Ophthalmology  
Karachi

Aimal Khan  
Isra Postgraduate Institute  
of Ophthalmology  
Karachi

Dilshad Laghari  
Isra Postgraduate Institute  
of Ophthalmology  
Karachi

Sadaf Ambreen  
Isra Postgraduate Institute  
of Ophthalmology  
Karachi

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