The Challenges in the Management of Sympathetic Ophthalmia: A Case Study

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Introduction

Sympathetic ophthalmia (SO) is a rare, bilateral, granulomatous uveitis caused by exposure of previously immune-privileged ocular antigens from trauma or surgery with a subsequent bilateral autoimmune reaction to this tissue. A prospective study in Europe revealed incidence of SO with 0.03 over 100,000 while others reported incidence of 0.2-0.5% following ocular injury and 0.01% following ocular surgery.[1,2] Hence, it interests us to report a case of SO as junior ophthalmologist might miss it due to its rarity. The first line treatment for SO is aggressive use of corticosteroid or immunosuppressive therapy for steroid non-responder.[3] However, the relapsing nature of the disease and the side effects patient suffered from high dose corticosteroid therapy are where the challenges lie in managing patient with this condition.

Objectives

To report a case of post traumatic sympathetic ophthalmia.

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Case report

A 48-year-old gentleman with underlying diabetes mellitus, hypertension and dyslipidemia allegedly fall at his backyard face down and thudded into the ground on 20th December 2020. The patient complained of left eye acute painful loss of vision immediately post fall. Examination showed left eye visual acuity 3/60 with pinhole 6/18 and left eye reverse relative afferent pupillary defect was negative. His left eye pupil was eccentric towards 10-12 o'clock with hyphema level of 1.8 mm (Figure 1). There was subconjunctiva hemorrhage with chemosis. The intraocular pressure was 9 mmHg. The left eye fundus view was not appreciated, however ultrasound B scan revealed clear vitreous and flat retina. There was regular globe outline with no intraocular foreign body seen on computed tomography scan. With suspicion of globe rupture, he underwent examination under general anesthesia which revealed left eye traumatic scleral rupture with uveal prolapse and hyphema secondary to blunt trauma. Subsequently, patient underwent left eye scleral suturing, uveal excision, anterior chamber washout and intravitreal antibiotics injection. The patient was discharged well with topical prednisolone 1% and moxifloxacin 0.5% every 2-hours tapered over 3 weeks. Left eye vision postoperatively was counting fingers, ultrasound B scan showed clear vitreous with flat retina.

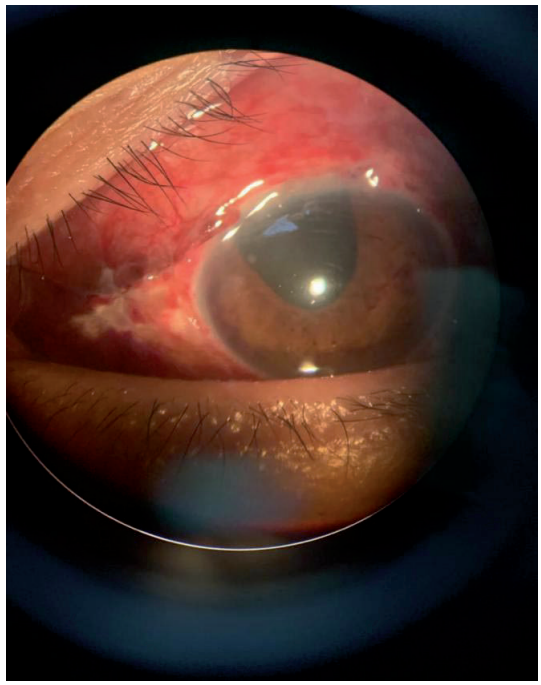

Figure 1

Patient presented to us again, one month post trauma with blurring of vision and redness over the right eye. Ophthalmic assessment revealed right eye vision dropped from 6/6 to 6/30. There was bilateral anterior uveitis with keratic precipitate and multifocal serous retinal detachment on fundus examination. (Figure 2, Figure 3). OCT demonstrated bacillary detachment with subretinal fluid, undulation of RPE and thick choroid (Figure 4, Figure 5). He was diagnosed to have right eye SO and was immediately commenced on high dose intravenous methylprednisolone 1 g OD for 3 days once systemic workup such as full blood count, ESR, chest X-ray, Mantoux, VDRL, urine analysis were negative to rule out tuberculosis, syphilis, and urinary tract infection. Then, he was maintained on oral prednisolone 60 mg OD (1 mg/kg) and topical prednisolone 1% every 2 hours over both eyes.

Unfortunately, our patient developed steroid-induced glaucoma with intraocular pressure in the 30 s with cup disc ratio of 0.3 despite on maximal medical antiglaucoma therapy. Patient was co-managed with the glaucoma team. Hence, steroid-sparing immunomodulating agents were initiated to taper off the systemic steroid fast. He was able to achieve remission with resolution of bilateral exudative retinal detachment and uveitis on mycophenolate mofetil 1 g BD and cyclosporin 150 mg OD over 5 months. During follow up, his right vision was 6/9 and left vision was counting fingers attributed by steroid induced posterior subcapsular cataract and traumatic cataract. Both fundi were flat with intraocular pressure of 18 mmHg bilaterally. He did not require glaucoma filtration surgery in the end. One year after his traumatic injury, he successfully underwent left eye phacoemulsification achieving final visual acuity of 6/9 without reactivation SO.

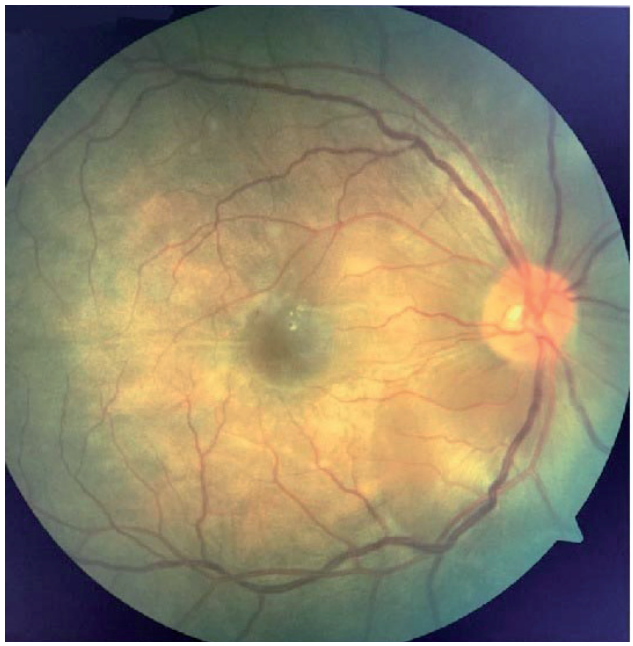

Figure 2

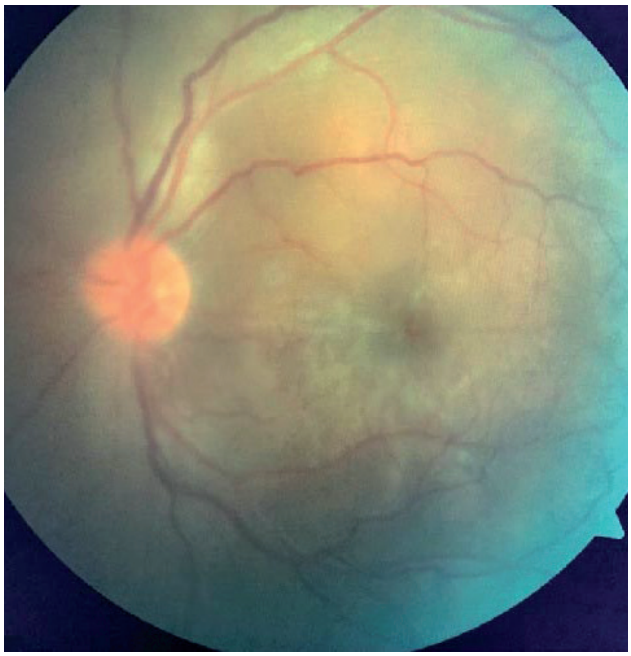

Figure 3

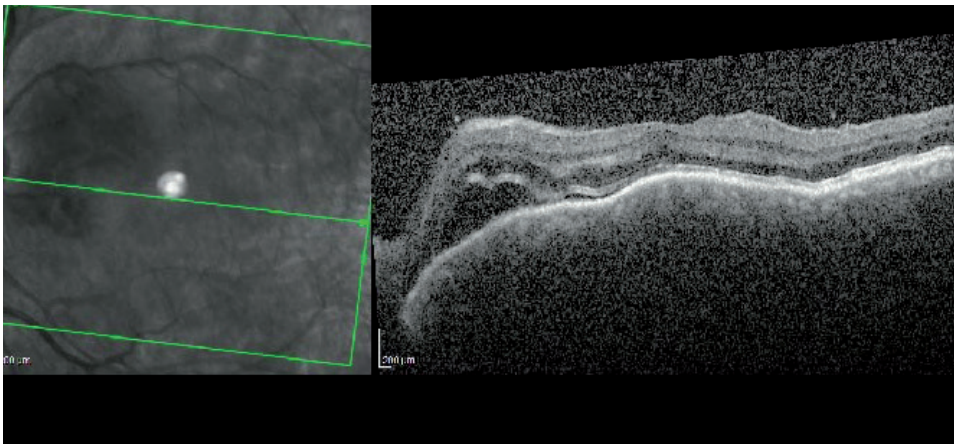

Figure 4

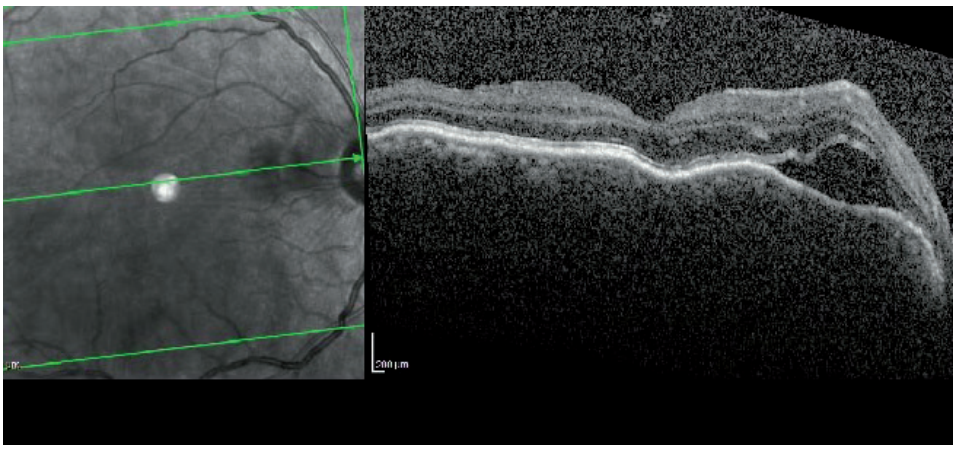

Figure 5

Discussion

The management of SO is often very challenging. It is a clinical diagnosis largely based on clinical findings and presentations, in addition to multimodal imaging which can often offer further insight in aiding diagnosis, monitoring for progression and treatment response. It has been accepted that the pathophysiology of sympathetic ophthalmia is T-cell mediated hypersensitivity reaction and the antigen responsible are Mart-1 melanoma and retinal S-antigen.[2] Lymphatic system was also found to expedite the sensitization through exposure of uveal tissue following injury to conjunctival lymphatics.[2]

Our case emphasizes the importance of early recognition and prompt immunosuppressive therapy in management of SO, which by and large provides satisfactory visual outcome. Following trauma, the patient sustained loss of iris tissue with prolapsed uveal subconjunctivally at the injured eye. Hence, we postulated the cause of SO in this case is due to the antigen sensitization from the uveal tissue via conjunctiva lymphatics. Vogt-Koyanagi-Harada disease was excluded due to the history of prior ocular injury and absence of systemic signs and symptoms such as hearing dysfunction, meningism and skin manifestation although they share strikingly similar presentations. Other differential diagnoses

such as syphilis and tuberculosis were unlikely but excluded prior to commencement of intensive corticosteroid therapy.

Systemic steroid therapy and steroid-sparing agents are the mainstay therapy for the management of SO. Damico et al. recommended intravenous pulse steroid therapy (methylprednisolone 1.0 g/day for 3 days) followed by oral prednisolone for severe cases.[4] Vote et al. suggested a management algorithm with initial intensive high dose oral steroid 50-75 mg daily followed by tapering steroid down to 10 mg daily over a course of 2-3 months period according to clinical response.[5] Systemic adverse effect must be anticipated with intensive corticosteroid therapy especially in patient with multiple comorbidity such as our case. In fact, our patient developed hyperglycemia with the highest random blood glucose level of 22 mmol/l during treatment which required co-management with physician. One of the greatest challenges in this case is the relapsing nature of his disease during steroid tapering to minimize the adverse effect while maintaining the control of his ocular inflammation. Mycophenolate mofetil and cyclosporin were included in his treatment regime whilst steroid was tapered off and our patient responded well to them. Regular reviews showed that the patient was able to achieve remission with good visual outcome. In SO, immunosuppressive therapy is always indicated and should be initiated alongside systemic steroid to control the inflammation as well as minimize steroid related adverse effects.

Our patient developed steroid-induced cataract and glaucoma as the sequela of his treatment. Preoperative counseling for cataract or glaucoma surgeries is crucial in ensuring patient understands the general operative risks as well as the visual prognosis. It has been suggested that surgery to be done after the eye has been quiescent for at least 3 months which will significantly reduce the risk of postoperative cystoid macular oedema.[6] Agrawal et al. suggested glaucoma surgery is preferably done after cataract surgery because bleb failure rate is high with drainage of post cataract surgery inflammatory exudate through a healing bleb.[6] However, our patient did not require glaucoma filtration surgery in the end and has achieved good visual outcome post phacoemulsification.

Enucleation prior to the disease onset in SO prevention has slowly fallen out of favor as patient generally able to achieve good visual outcome with immunosuppressive therapy. Ying et al. reported that enucleation does not eliminate the risk of SO completely.[7] In addition to enucleation, prompt surgical intervention without delay, early recognition with effective control of inflammation also plays a crucial role in the management of SO. In our case, prophylactic enucleation was not considered as his traumatic eye still has good visual potential upon presentation.

Conclusion

The key to manage SO is early recognition by retaining an index of suspicion in patients with history of ocular trauma or surgery as well as prompt immunosuppressive therapy. A good visual outcome is attainable by fine-tuning the balance between the control of ocular inflammation and complication of the immunosuppressive therapy.

Conflicts of Interest

We declare that there are no conflicts of interest.

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