Case Report

Severe Sympathetic Ophthalmia Following Evisceration of a Perforated Staphylomatous Globe

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Abstract

Sympathetic ophthalmia (SO) is a rare bilateral panuveitis that follows a surgical or penetrating injury to one eye. The use of systemic corticosteroids and immunosuppressive drugs has greatly improved the outcome of this potentially blinding condition. It may, however, rarely run an aggressive course despite medical treatment with grave consequences. Herein, we report a severe case of SO in a 64-year-old man induced by the spontaneous perforation of a staphylomatous left eye. He presented with left panophthalmitis following a spontaneous perforation of the globe, and subsequently he had an evisceration. He, however, presented 18 days later with features of SO in the right eye. He was placed on systemic corticosteroid and immunosuppressive therapy but failed to respond to treatment and the eye eventually became phthisical.

Keywords: Evisceration, staphyloma, sympathetic ophthalmia

Introduction

Sympathetic ophthalmia (SO) is a rare, bilateral granulomatous uveitis that occurs after either surgical or accidental trauma to one eye.[1] Traditionally, accidental penetrating eye injury was considered the main risk of SO. Nowadays, ocular surgery, particularly vitrectomy for retinal detachment, is also recognised as a risk factor for SO.[2] Given its rare occurrence, the true incidence of SO is difficult to establish, and literature reports are variable, likely owing to the fact that the diagnosis of SO is based on clinical findings rather than on serological testing or histopathology.[3] Kilmartin *et al.*,[2] however, reported a minimum 1 year incidence of 0.03 in 100,000 persons in the United Kingdom and Republic of Ireland. The time from ocular injury to onset of SO varies greatly and has been reported between 1 week and 66 years.[4] The use of systemic corticosteroids and immunosuppressive drugs such as azathioprine and cyclosporin has transformed the outlook of this potentially blinding disorder, and good visual acuity (VA) in the sympathising eye can now be achieved.[3] We present, herein, a case of severe unilateral SO following evisceration of a blind staphylomatous fellow eye that had spontaneously perforated.

Case Report

A 69-year-old male Indian patient presented to Dr. D. Y. Patil Hospital and Research Centre in Navi Mumbai, India with pain and redness in the left eye (LE) of two months duration. He had lost vision in the eye at 4 years of age following blunt trauma to the eye. No ocular surgery was performed following the injury. On examination, the right eye (RE) had a best-corrected VA of 6/9 and was essentially normal. The LE had a VA of no perception of light, and there was an anterior staphyloma with corneal vascularisation. The posterior segment details could not be visualised, and the digital tension was high. He was started on topical antibiotics, cycloplegics and intraocular pressure lowering medication. On the subsequent follow-up 2 weeks later, he had developed a corneal perforation with pseudocornea formation in the LE. Ocular movements were restricted in all the directions of gaze. An ultrasound scan of the LE was taken, and this showed features of vitritis, total retinal

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detachment with choroidal effusion and thickening of the retina–choroid–scleral complex. A diagnosis of panophthalmitis was made, and he was commenced on intravenous antibiotics (cefotaxime 1 g daily and amikacin 250 mg daily) along with hourly topical antibiotic. A full blood count was taken which revealed an elevated neutrophil count. The erythrocyte sedimentation rate was also raised.

He then underwent a frill evisceration of the LE 72 h later. Under general anaesthesia, the eyelids were parted with a lid speculum and the cornea was excised. The intraocular contents were then removed completely using a scoop. The extraocular muscles were separated and the sclera was excised leaving a 3 mm frill around the optic nerve. The socket was then packed with gauze and a bandage applied. He was discharged on the fourth postoperative day. Histopathology report of the eviscerated eye showed polymorphonuclear cells and panuveal lymphocytic infiltration. On his next clinic visit 2 weeks after discharge, he complained of pain and glare in the RE of 4 days duration. Examination of the eye revealed the best-corrected VA had dropped to 6/60, and there was an anterior granulomatous uveitis with mutton fat keratic precipitates. On fundus examination, inferior and superior retinal detachments with shifting fluid and 360° shallow choroidal detachments were noted [Figure 1]. No retinal break was seen, and fundus findings were confirmed on B scan ultrasonography [Figure 2]. Systemic examination did not reveal any abnormalities. A chest X-ray and full blood count were taken, and mantoux test was conducted; all were within normal limits. Optical coherence tomography and fundus fluorescein angiography were ordered but were not taken due to financial constraints. A diagnosis of SO was made and he was placed on intramuscular methylprednisolone 1 g daily for 3 days followed by oral prednisolone at 60 mg daily and posterior sub-Tenon's triamcinolone injection 40 mg stat. Two weeks later, increasing intraocular inflammation and progression of the retinal detachments were noted. He then had intravenous methylprednisolone for 3 days followed by oral cyclophosphamide 100 mg daily. His clinical condition, however, did not improve and the eye subsequently became phthisical.

Discussion

SO is a clinical diagnosis based on the development of a bilateral panuveitis (when both eyes are present) following penetrating injury, and rarely intraocular surgery, to one eye.[5] This ocular disease is probably the best known and the most classic model of an autoimmune-associated disease occurring in humans.[3] Current evidence suggests a role for immune dysregulation as a primary etiological mechanism. There appears to be a cell-mediated immune response directed against ocular self-antigens found on photoreceptors, the retinal pigment epithelium and/or choroidal melanocytes.[3] In this current case, the SO may have been triggered by the left corneal perforation. Dada *et al.*[6] reported a case of SO associated with antecedent adherent leucoma following a perforated corneal ulcer 15 years earlier. With corneal perforation, the intraocular uveo-retinal antigens are able to gain access to the regional lymph nodes via the conjunctival lymphatics. This may cause a sensitising reaction to these antigens and set up a delayed T cell hypersensitivity, which may be responsible for the disease process.[7]

SO may also rarely occur following an evisceration as a result of uveal tissue remaining in scleral emissary channels.[8] We, however, do not think that this was the cause in our case as the sensitising reaction most likely occurred prior to the evisceration. Griepentrog *et al.*[9] had earlier reported a case of SO following evisceration of a blind, painful, posttraumatic, glaucomatous eye.

The differential diagnosis of SO includes all diseases that can present as panuveitis, particularly Vogt–Koyanagi–Harada (VKH) syndrome. However, the patients with VKH have systemic involvement and no history of ocular injury or surgery.[3] Tuberculosis, sarcoidosis, and syphilis are

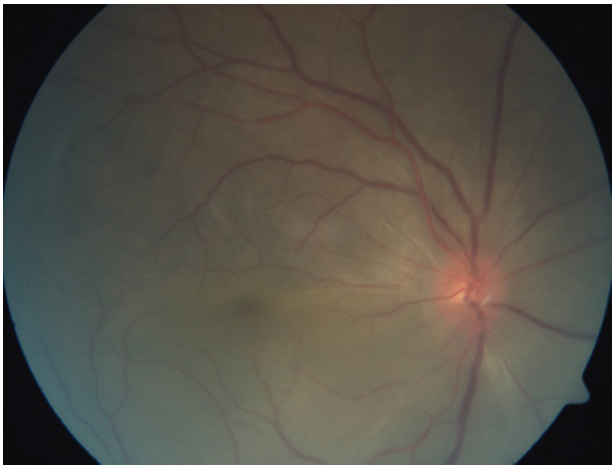

Figure 1: Fundus picture of right eye showing serous retinal detachment involving the macula

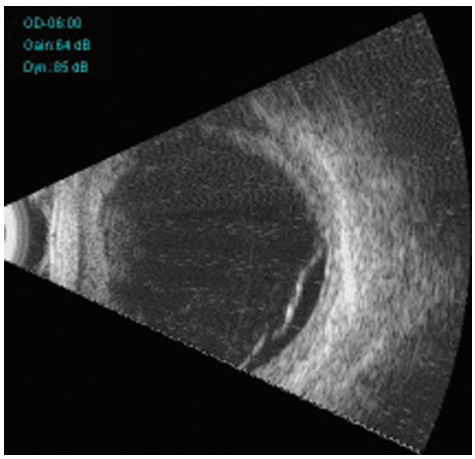

Figure 2: B scan of the right eye showing retinal detachment

usually accompanied by constitutional signs and the symptoms of the underlying systemic disease.[3] The history of trauma and spontaneous globe perforation and absence of systemic abnormalities made any of these disorders unlikely in this case. Surgery prior to disease onset appears to play an important role in the prevention of SO.[3] In our case, SO developed despite evisceration suggesting that sensitisation to uveal antigens had already occurred. SO can be effectively treated, and visual prognosis is good with prompt and adequate immunosuppression.[3] Our case, however, did not respond to medical treatment with the eye becoming phthisical. Similarly, Matysik-Woźniak *et al.*[10] reported a severe case of SO that failed to respond to intensive pharmacological treatment. In conclusion, SO may rarely occur following a spontaneous globe perforation in a staphylomatous eye. Prompt surgical removal of the eye should, therefore, be considered in such cases to reduce the risk of this potentially blinding condition. A high index of suspicion must also be maintained whenever inflammation occurs in the fellow eye of a staphylomatous eye that has suffered spontaneous perforation, as delay in diagnosis and treatment may have disastrous consequences.

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Conflicts of interest

There are no conflicts of interest.

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