Sympathetic Ophthalmia

Abstract

Background: Sympathetic ophthalmia (SO) is a rare but serious bilateral inflammatory condition of the eyes that usually follows penetrating injury to one eye. When diagnosed and managed early and appropriately, there is a good chance of controlling the inflammation and retaining useful vision. Till this time there is no case report of SO on literatures from Ethiopia making this the first.

Case presentation: A 24-year-old male patient presented to Jimma ophthalmology department with left eye pain and reduced vision of three months duration. He lost his right eye vision four months back from trauma. On examination, he was having non-seeing shrunk right eye with corneoscleral scar. The left eye was having ciliary flush, hazy cornea with large keratic precipitates (KPs), turbid aqueous and irregular pupil with posterior synechiae. Vision in the left eye was hand motion. The patient was diagnosed with sympathetic ophthalmia (OU) plus phthisis bulbi (OD) and admitted. Treatment was started with steroids (both topical and systemic), and cycloplegia for which he responded well but later on he developed refractory glaucoma and secondary cataract and operated.

Conclusion: This is the first case of SO reported in literatures in Ethiopia. As corticosteroids are the mainstay of treatment for SO, blinding complications from their long term use has to be anticipated and managed accordingly without delay.

Keywords
Sympathetic ophthalmia; Cataract; Glaucoma; Jimma; Ethiopia

Background

Sympathetic ophthalmia (SO) is a rare bilateral, diffuse granulomatous panuveitis that occurs following penetrating trauma or intraocular surgery to one eye, the exciting eye. The fellow non-traumatized sympathizing eye also shows similar inflammatory response usually with mutton-fat KPs suggesting involvement of autoimmune response [1]. Though the time from ocular injury to onset of SO was said to vary greatly, ranging from a few days to fifty years with 90% of the cases occurring within 1 year [2-5], recent series showed that only one-third of patients developed SO within 3 months and fewer than 50% did so within 1 year of injury [6].

It has been speculated that an infectious agent or a bacterial antigen may precipitate an immune response resulting in the development of SO; but instigating organisms were not identified and the precise etiology of SO remains unknown to this time [5,7].

The prevalence of SO is difficult to measure because it has always been a relatively rare disease plus as a result of improvements in modern surgical and medical treatments, it has now become even more uncommon. It accounted for about 0.3% of uveitis and its 1-year incidence was calculated to be a minimum of 0.03/100,000 population [8]. In a retrospective analysis of 2,340 cases of open-globe injuries in Iran, 0.08% (only two cases) was diagnosed with sympathetic ophthalmia from 1998 to 2003 [9].

Patients with SO typically present with asymmetric bilateral panuveitis with more severe inflammation in the exciting eye than in the sympathizing eye at least initially. Signs and symptoms in the sympathizing eye vary in their severity and onset ranging from mild problem in near vision, photophobia and slight redness to very severe granulomatous anterior uveitis [5].

Significant improvements in the management of ocular trauma combined with the advent of immune-modulators (including corticosteroids) and the use of antibiotics has led to a dramatic decrease in the incidence of sympathetic ophthalmia. However intraocular surgery is now considered the major risk factor particularly vitreoretinal surgery [10,11].

Case Presentation

A 24-year-old male patient from Southwest Ethiopia presented to Jimma hospital eye department in March 2009 with pain and loss of right eye (OD) vision following trauma from a flying stone four months back. He was also having severe left eye (OS) pain associated with progressive reduction of vision, photophobia, redness and headache since three months. On examination, vision was no light perception (NLP) in OD and hand motion in OS. Intraocular pressure (IOP) was hypotonous in OD and 22mmHg in OS. The right eye was having ciliary flush, hazy cornea, diffuse large KPs, shallow anterior chamber (A/C), irregular nonreactive pupil and contracted eyeball. In the left eye he was having ciliary flush, hazy cornea with endothelial dusting and many large KPs, A/C was deep, turbid aqueous with 4+ cells, sluggishly reactive pupil and limited lens and posterior segment visualization because of media haziness.

He gave history of occasional headache but no history of allergy, hearing disturbances or other symptoms and his blood
pressure was 110/70mmHg, laboratory investigations were as follows: haemoglobin 13.2gm/dl, WBC 4500/mm³, erythrocyte sedimentation rate 18mm/hr, fasting blood sugar 78mg/dl, VDRL was nonreactive and chest x-ray showed no cardiopulmonary pathology.

With the diagnoses of sympathetic ophthalmia (OU) and traumatic phthisis bulbi (OD), the patient was admitted, started on prednisolone tablets 1mg/kg PO daily, dexamethasone 0.1% eye drop 2hrly, atropine 1% eye drop tid for both eyes. Two weeks later the right eye remained painful, vision in the left eye improved to 6/60. Enucleation of the right eye was done and the patient continued to take prednisolone PO, tapering dose, and dexamethasone and atropine topically. Three months later the inflammation in the left eye improved remarkably and vision improved to 6/36, IOP was 16.4mmHg, there was posterior synechia.

Six months later patient came complaining reduced vision and pain of a week duration. On examination his vision (OS) was counting fingers, IOP 37.2mmHg, hazy cornea, few KPs, multiple posterior synechiae with inflammatory pupillary membrane and iris bombe. Gonioscopy showed appositional angle closure. YAG laser iridotomy was done, angle opened and IOP decreased to 20mmHg. Later patient developed pupillary membrane for which membranectomy and synechiolysis were done. Patient continued applying dexamethasone drops topically and prednisolone PO from time to time and vision improved to 6/36. IOP maintained between 14-17mmHg for over a year.

Two years later patient presented complaining poor vision and on examination his vision was CF at 2 meters, deep A/C, irregular pupil and dense PSC lens opacity. Ocular ultrasound showed no pathology in vitreous or retina. With the assessment of posterior subcapsular cataract (steroid induced), cataract extraction (SICS) was done and +21 PC-IOL was inserted. Two weeks postoperatively vision improved to 6/60, funduscopy showed syneretic vitreous with suspended fibrous strands, 0.5 C/D ratio and chorioretinal thinning. Patient continued applying dexamethasone drops topically and prednisolone PO from time to time and vision improved to 6/36. IOP maintained between 14-17mmHg for over a year.

Discussion

Sympathetic ophthalmia, also known as sympathetic uveitis, is thought to be a delayed hypersensitivity immune response to uveal antigens. It may manifest with a spectrum of clinical findings ranging from mild posterior uveitis to severe bilateral granulomatous panuveitis [2,12]. Following injury from either accident or surgery, a variable period of time passes before a sight threatening inflammation develops in both eyes. In this particular case typical features occurred one month following the trauma. Both eyes showed intense inflammatory reaction with large KPs and turbid aquesous.

The etiology and pathophysiology of the disease is still unclear but is largely thought to be autoimmune in nature and that the injured eye and the contralateral eye demonstrate similar pathology suggesting involvement of an autoimmune response [5].

Until recently accidental penetrating ocular trauma was the classic most common precipitating event for SO. These days ocular surgery particularly vitreoretinal surgery has emerged as major risk for the development of SO. Non-perforating ocular procedures like irradiation and laser therapies have also been associated with SO [13]. Improved access to emergency surgical care following penetrating ocular traumas and improved microsurgical techniques have undoubtedly influenced this etiologic shifts from penetrating injuries to surgical traumas [5].

Recent studies also showed that SO can follow endophthalmitis though uveal antigenicity markedly decreases by purulent intraocular infection [12]. There has also been long speculation that an infectious agent may be required for the development of SO. However, no organism has ever been consistently isolated from eyes with SO, and the disease has never been incited in animal models following injection of an infective agent [14].

In this particular patient onset was insidious with recurrent periods of exacerbation. Classically patients present with bilateral anterior uveitis associated with mutton-fat KPs and moderate to severe vitritis, choroiditis, and papillitis [5]. As in this case, SO runs a chronic course with a marked tendency toward relapses. Finally the disease may culminate in a phthisical and blind eye.

Diagnosis of SO is primarily based on patient history, clinical findings and a history of ocular trauma or surgery. However in about 20% of cases diagnosis is confirmed based on histologic findings [15]. In vague cases, other causes of granulomatous uveitis, such as Vogt-Koyanagi-Harada disease, sarcoidosis, tuberculosis and syphilis should be considered and ruled out with appropriate diagnostic methods.

SO can be prevented before its occurrence or can be treated once diagnosed. Enucleation, the only indisputable way of preventing SO is generally recommended within two weeks post-injury [16,17]. However, it appears that preference for evisceration over enucleation is currently increasing with advancements in technique and greater perceived benefits. A retrospective study in South Africa from 1995 to 2004 showed that no cases of SO were found in 491 primary eviscerations and 11 secondary eviscerations [18]. Taken with superior functional and cosmetic outcomes, evisceration appears to be favored nowadays. Both enucleation and evisceration however seem unnecessary these days due to the relatively better availability and use of potent immunosuppressive and immunomodulatory drugs which effectively control intraocular inflammation and also improve visual outcome if initiated promptly.

After onset of SO corticosteroid therapy that targets inflammation systemically is considered the mainstay of treatment [5]. However the long-term use of corticosteroids has been a problem as they are associated with the development of cataract and glaucoma, plus major systemic adverse effects such as diabetes mellitus, adrenal insufficiency, arterial hypertension and osteoporosis [19]. This case had developed dense posterior...
subcapsular cataract (operated) and steroid-induced glaucoma (medically controlled). SO patients who had been on systemic corticosteroids need strict follow ups and systemic evaluations so that early identification and appropriate treatment of both ocular and systemic long term complications is possible.

Acknowledgement

I am very grateful to my patient for agreeing and allowing me to report this curious and rare case on literatures. I would also like to thank my wife Dr. Selamawit Amare for her immeasurable assistance in reviewing and editing the case before submission for publication.

References