Review article

Sympathetic ophthalmia: Enucleation or evisceration?

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Abstract

The purpose of this review is to find out the risk of sympathetic ophthalmia after evisceration and enucleation particularly in a case of penetrating ocular (non-surgical or surgical) injury and to know about the cosmetic outcome of evisceration and enucleation. Ophthalmologists around the world, whenever dealing with a case of penetrating ocular (non-surgical or surgical) injury, might thus be helped in deciding whether to keep the traumatized eye or remove it. And if they finally decide to remove it, they will be able to choose between evisceration and enucleation for the best cosmetic outcome, and to minimize the risk of sympathetic ophthalmia.

The results of the review are the following. Enucleation is not found to be the best protective measure against development of sympathetic ophthalmia, as it was thought to be, since there are ample reports of sympathetic ophthalmia occurring not just after evisceration but also after enucleation. The cosmetic outcome of evisceration as opposed to enucleation is found to be better.

Key words: sympathetic ophthalmia, enucleation, evisceration

Introduction

Whenever an ophthalmologist in any part of the world is confronted with a case of ocular trauma he or she starts to worry about the possibility of sympathetic Ophthalmia (SO) in the future. So, he/she is confronted with many issues. If the trauma is so bad that the globe is extensively injured, without any useful vision and also without the visual potential, we definitely would like to remove that eye. But then the question arises as to whether to go for evisceration or enucleation. One might worry that evisceration, although an easy procedure with probable better cosmetic outcome, will not prevent the patient from developing SO.

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The other issue is whether we would prefer to save the eye with all the new available surgical techniques even if the visual outcome is not optimal, without the fear of SO once again. The next issue is if we finally decide to enucleate the eye, would we hurry up and just do it as soon as possible and not really pass the 'safety period' (Reudemann,1964) of the first two weeks from the day of trauma. Well, the answers are not straightforward.

SO is a bilateral simultaneous granulomatous panuveitis that occurs within a variable period of time after a penetrating injury or manipulation of one eye that generally involves incarceration of some uveal tissue. The traumatized eye is called the exciting eye and the contra-lateral eye is called the sympathizing eye (Ramadan et al 1996). Eighty percent of SO occurs from two weeks to three months after ocular injury and ninety percent of cases occur within one year (Ramadan et al 1996).The shortest interval of SO after trauma

reported is 10 days (Stafford 1965) and the longest interval reported is 66 years (Zaharia et al 1987).

It was postulated from the rabbit model that the presentation of uveo-retinal antigen to the lymphatic system via the conjunctiva is responded by development of SO. However, no anti-retinal S antigen was detected in patients with SO (Rao et al 1983).

SO is now labeled as an autoimmune disorder associated with HLA DRB1 04 and DQA1 03 genotypes in the white population, similar to that of the Japanese people (Kilmartin et al 2001). Although the disease has been described since the age of Hippocrates (Albert, 1989), its low incidence, varying from 0.03/100, 000 cases of ocular surgeries and ocular trauma (Kilmartin et al 2000) to less than 10/100 000 cases of surgical penetrating wounds (Marak 1979) and the possibility of its occurrence several decades after injury, any control study on it is very difficult or almost impossible. All the same, the literature has reports of various individuals that have made attempts to conduct retrospective case studies and long-term follow-up studies since the beginning of the 20th century. We have attempted to find answers to our queries from within these studies. The following has been revealed after an analysis of the relevant literature.

In almost all the reported case series of SO, the disease occurred after penetrating ocular trauma or various ocular surgical procedures.

Among the surgical procedures, the reported ones are cataract surgery, iridencleisis, iridectomy, trephine, cyclotherapy, trabeculectomy, vitrectomy, retinal detachment repair, laser cyclophocoagulation, laser photocoagulation, local irradiation and keratectomy (Lewis et al 1978; Ramadan et al 1996; Lam et al 1992; Bechrakis et al 1994). Infact several recent studies have shown that the vitreo-retinal surgery is a major factor for the development of SO (Kilmartin et al 2000; Sisk et al 2008; Ozbek et al 2010).

What about the occurrence of SO after enucleation or evisceration of the eye with ocular trauma, accidental or surgical? Yes, there are reports on SO after both kinds of surgeries.

Now, talking about evisceration leading to SO, Ruedemann's literature (1963 and 1964) has the following information. Forty-seven cases of SO after evisceration were reported before 1917. Besides four cases which were operated within 14 days of the trauma, the rest were operated from 14 days to years after the injury. Follow-up on 506 cases of evisceration (1947 - 1962) for at least three months showed not a single case of SO. More than one half of the eviscerations were performed after more than 14 days of injury.

Von Graefe's series of 240 cases, Beren et al's series of 230 cases and Paulard's series of 190 cases of evisceration had no SO in Ruedemann's report (1963). Ruedemann's speculation (1964) was that the presence of low-grade uveitis in the uninjured eye, prior to the use of the slit-lamp, could have been missed. Or in other words, the onset of SO could have occurred even before evisceration was performed, since all the cases were of trauma.

He had mentioned about Morax's report on a review of SO in France during World War I. It showed 37 cases of SO after enucleation and just 2 cases after evisceration, total number of cases not being mentioned. He also mentioned about Rimpler's report on 10 cases of SO in 1900 after enucleation of 260 of traumatic eyes as opposed to two after evisceration of 149 eyes.

So, Ruedemann gave an opinion that by utilization of modern therapy and surgical techniques, the evisceration procedure is acceptable except in the presence of deep chronic inflammation, in known intraocular malignancy and when a pathological specimen of the whole eye may be useful for diagnosis and treatment.

Green (1972) reported four cases of SO after evisceration. In one of the cases, evisceration was performed one week after the trauma. In two, it

was as late as 24 to 25 days. The fourth one was a case of a painful blind eye due to an unknown reason with no history of preceding ocular trauma. But as Migliori (2002) pointed out, Green's manuscript did not mention the total number of cases out of which four ended up SO. So, the calculation of the relative risk of SO following evisceration was not possible.

Levine et al (1999) had two components in their study. In the first one they studied 90 cases of evisceration. Out of 51 cases that could be followed up, none had clinical evidence of SO. The time of evisceration from the date of trauma was as early as two days and as late as 20years. The follow-up period after evisceration ranged from three months to 15 years. About 80 % of cases were followed up for more than one year.

The second component was a survey sent to the American Society of Ophthalmic Plastic and Reconstructive Surgery, the Uveitis Society and the Eastern Ophthalmic Pathology Society. 841 eviscerations were reported. Five cases of SO (two non-trauma cases and three trauma cases) were recalled by survey correspondents, but unfortunately none had documented evidence.

One interesting case report on SO after evisceration is as follows. One man, who had lost vision in his right eye after sustaining a penetrating injury to it several years ago had to undergo evisceration of that same eye at the age of 75 years because of the development of NVG. Surprisingly, diffused uveitis consistent with SO appeared in his good eye on the fourteenth post-operative week. The eviscerated right eye on HPE did not show any signs of acute or chronic inflammation (Griepentrog et al 2005).

Gurdal C et al (2002) followed up 183 eviscerations and 34 enucleation cases on penetrating eye injury for the mean period of 10.3 ± 6.6 years. 69 cases had early (within two weeks of trauma) and 148 had late surgical intervention (evisceration or enucleation). They reported not a single case of SO in either group.

Toit et al (2008) in their retrospective case study reported not a single case of sympathetic ophthalmia out of 502 cases of evisceration following penetrating ocular trauma. (In their study, other three cases of penetrating ocular trauma that underwent enucleation also did not develop SO.) But the follow-up period for only 12.6 % of patients was for one year or more. Androudi et al (2010) reported a case of post-operative endophthalmitis which developed SO two weeks following evisceration and five months after the occurence of endophthalmitis.

The rest of the literature which were reviewed dealt with enucleation procedures. Although we were able to study a number of reports on SO after enucleation, none of them give the time interval between the time of ocular injury and the procedure. The reports in favor and against enucleation are as follows. Winter (1955) reported seven cases of SO occurring after enucleation of traumatic eyes. Jennings and Tessler (1989) had one case of SO after enucleation performed three weeks from the day of trauma. Bellan (1999) reported a case of SO which happened despite the performance of enucleation less than five days after the trauma. The inflammation took a course of relapses and remission with preservation of vision (20/40) till the patient died of other systemic conditions. Ravin (2002) wrote on James Thurber, the writer and cartoonist, who suffered from SO despite the enucleation of his injured left eye. But the time lapse between the penetrating injury and the enucleation is unknown.

Now, should the exciting eye be enucleated once the sympathetic ophthalmia occurs? Let's review the relevant articles and try to find find out. Lubin et al (1980) after performing a retrospective clinic-pathologic review of 105 cases of SO concluded that enucleation within two weeks of the onset of sympathetic inflammation improved visual outcome. Lubin and Albert (1980) wrote that with the more surgical procedures performed in an attempt to save a badly injured eye, the number of cases of SO diagnosed clinically was increasing.

Reynard and Riffenbergh (1983) stated that early enucleation not only improved visual acuity, but also is associated with a relatively benign clinical course of the disease. The term 'benign' is not explained in detail in the article.

However, Irvine (1940), after doing a clinical review of 63 cases of SO, thought that enucleation of the exciting eye is not beneficial in terms of improving visual acuity in the sympathizing eye. Winter (1955) stated that enucleation of the exciting eye should not be performed except on a blind or nearly blind-eye, because of an appreciable error in the clinical diagnosis of SO. In one series on enucleation, out of seven eyes (0.7 %) suspected of having SO in trauma cases, only one was found to have SO on histopathological study. Lim (1976) gave his opinion that enucleation should not be performed because of a suspected presence of SO.

In Mackley and Azar's (1978) series of 17 cases of SO after surgical trauma or accidental injury, even after the performance of enucleation plus steroid treatment in 16 cases, relapses occurred in 60 % of the eyes and 70 % of the eyes had complications. Here, relapse meant some type of flare-up of symptoms when an attempt was made to discontinue corticosteroid treatment. 'Relapse' then might be equivalent to the term 'chronic'.

Chan et al (1995), after their retrospective review of 32 cases of SO, wrote that enucleation of the exciting eye appeared not to have a significant association with visual acuity. Bilyk and Jury (2000) pointed out that enucleation is not indicated once SO is diagnosed since the visual prognosis is not improved and in some cases the exciting eye may become the better-seeing side. They also said that the risk of missing intraocular malignancy is higher than the potential danger of SO when considering evisceration. They, however, concluded that that the potential risk of SO after evisceration is still unknown to us and that without having this knowledge, one effectively cannot persuade or dissuade patients from undergoing evisceration.

Chu and Foster (2002) also gave their opinion that the decision to enucleate the exciting eye should be made cautiously since there might be no benefit at all in the control of inflammation of the sympathizing eye and also since the exciting eye may ultimately become the better seeing eye.

Zhang et al (2009), after performing a retrospective case study on globe injury cases, did not recommend a prophylactic enucleation after open globe injury. Savar et al (2009) were in favor of primary repair of open globe injuries of eye rather than primary enucleation. In there retrospective study of 660 eyes with open globe injury, only two cases out of 605 repairs developed SO.

The following few articles tell us about the world's changing opinion about evisceration. Eighty-five board-certified ocularists of the United States responded to a survey on outcomes of enucleation and evisceration. Eighty-percent of them reported that evisceration is superior to enucleation in terms of best ocular motility and best overall cosmesis. When they were asked which procedure would be their first choice for a patient who required removal of an eye, ninety-two percent stated evisceration (Timothy et al 2003).

Migliori (2002) after reviewing several retrospective studies concluded that the definitive risk of SO after evisceration is still inconclusive, but that most of the surgeons recognize better motility and cosmetic appearance after eviscreation than after enucleation, and finally said that evisceration may be more acceptable to patients since only the diseased part of the eye is removed.

Hansen (1999) reported that over the last 20 years there has been a change in choice of operation from enucleation to evisceration. In this respect, Genevois et al aslo found an increased proportion of evisceration in ten years from 1990 to 2000. Gurdal et al (2002) reported an increased rate of complications with enucleation as opposed with evisceration. The authors concluded that evisceration is the choice of treatment in traumatized eyes.

Lui (2005), from his prospective study done to compare the implant extrusion rates and postoperative pain after evisceration and enucleation, found that post-enucleation pain appears more severe than post-evisceration pain.

Tawfik et al (2007) found a low overall complication rate and successful retention of the primary implants in cases of evisceration with endophthalmitis or panophthlamitis. Tari et al (2009) from their prospective study of patients after evisceration with scleral quadrisection and allopalstic implantation and those after enucleation with hydroxyapatite implantation, conclude that this particular evisceration method can be an effective substitute for a more extensive procedure such as enucleation in terms of providing rapidity, ease and better implant excursion, and cost effectivity.

Before the discovery of corticosteroids, the treatment of SO was very difficult, and the disease invariably made the patients blind. However, the treatment outcome has become more favorable nowadays than in the past because of the use of corticosteroids along with newer immunomodulators (Zhang et al 2009; Castiblanco and Adelman 2009).

Summary

We found literature with evidence of SO occurring in cases of penetrating ocular injury (accidental or surgical) despite the performance of enucleation, while on the other hand, there are reported cases of penetrating ocular injury which did not end up with SO despite undergoing evisceration instead.

We now know that there is enough literature that supports or disapproves early enucleation of the exciting eye. However, one issue is clear - that although enucleation has always been regarded as the classic and only-known protection of SO in trauma cases, it is not the absolute protection of SO.

It is difficult to comment on whether or not prophylactic enucleation or evisceration is more protective if performed within a safety period, based on the available literature. Another point is that enucleation of the exciting eye is not the curative treatment of SO. Therefore, in any situation, it is unjustified to perform it on any eye with penetrating injury with visual potential either to decrease inflammation or to improve the visual acuity in the sympathizing eye.

In a case of ocular trauma where the eye is severely damaged to the extent that it cannot be salvaged by repair and that the eye has no perception of light, then only we should consider a destructive surgery in that eye. And in my opinion, unless the scleral shell is badly mutilated, we should perform evisceration since the patient's aesthetic concerns have also to be considered, because we have found from this review that evisceration is superior to enucleation in terms of best ocular motility, best overall cosmesis, and complications.

Since SO is now proven to be an autoimmune genetic disorder, its occurrence, early development, and severity do not solely depend upon penetrating ocular injury, but also depend upon the individual's genetics.

We must also keep in mind that the treatment modality of SO in this 21st century has become quite advanced.

Finally, we must say that until the advent of an extraordinary procedure that will make cosmetic outcome of enucleation as good as or even better than that of evisceration, any ophthalmologist who is concerned about the potential danger of SO as well as about the outlook of the patient, will have to make his or her own judgment and decision keeping in mind the fact that he or she is neither condemning the patient with SO nor absolutely protecting him or her from it with either procedure.

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