Cataract surgery in sympathetic ophthalmia

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Purpose: To analyze the results of cataract surgery in patients with sympathetic ophthalmia.

Setting: Sankara Nethralaya, Medical Research Foundation, Chennai, India.

Methods: This study comprised 66 patients (132 eyes) with sympathetic ophthalmia seen at the uveitis referral clinic between January 1990 and July 2001; 42 eyes (31.8%) had cataract. Cataract surgery was performed in 17 sympathizing eyes and 1 exciting eye (17 patients). The records of these 18 eyes were retrospectively analyzed. Three eyes had extracapsular cataract extraction (ECCE) with intraocular lens (IOL) implantation, 6 eyes had ECCE without IOL implantation, and 9 eyes had phacoemulsification with IOL implantation. The mean follow-up was 28.7 months (range 3 to 60 months).

Results: The causes of sympathetic ophthalmia were penetrating trauma (n = 8 eyes), ocular surgery (n = 6), perforated corneal ulcer (n = 2), and cyclocryotherapy (n = 1). The most common cataract type, present in 7 eyes (38.8%), was mixed (posterior subcapsular and posterior polar). Visual acuity improved after surgery in 13 eyes (72.2%). The main factors impairing visual recovery were submacular scar and optic atrophy, which were sequelae of the sympathetic ophthalmia. Posterior capsule opacification was noted in 14 eyes (77.7%); it was visually significant in 6 eyes. There was no significant difference in postoperative inflammation or disease reactivation between the 3 types of surgery.

Conclusions: Cataract extraction in cases of sympathetic ophthalmia can be safely and successfully performed with vigilant preoperative and postoperative control of inflammation, careful surgical planning, and meticulous surgical technique. The final visual outcome, however, depends on the posterior segment complications of the disease.

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Sympathetic ophthalmia is a rare form of uveitis in which inflammation occurs in both eyes after nonsurgical penetrating trauma or after trauma from ocular surgery in 1 eye. The injured or operated eye is the exciting eye, and the noninjured fellow eye is the sympathizing eye. Sympathetic ophthalmia remains 1 of the most severe complications in ophthalmology. The incidence after nonsurgical trauma is 0.2%[1] and after surgical penetrating wounds, fewer than 10 cases per 100 000 population.[2] Sympathetic ophthalmia can occur as early as 9 to 10 days after the penetrating injury or as late as many decades after the initial trauma. However, in 80% of cases, it is clinically diagnosed within 3 months of injury to the exciting eye.[2]

The clinical features of sympathetic ophthalmia include granulomatous anterior uveitis or panuveitis, peripapillary choroiditis or exudative retinal detachment, and papillitis. The sequelae and complications of sympathetic ophthalmia that pose a major threat to vision include cataract, secondary glaucoma, optic atrophy, and significant chorioretinal scarring, which occurs in 25% to 30% of cases.[3]

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Although it is probably possible to prevent sympathetic ophthalmia by enucleating the exciting eye before the sympathizing eye becomes involved, high doses of systemic steroids and cytotoxic agents form the mainstay of therapy. In a study by Reynard and coauthors,[4] corticosteroid therapy was associated with a good visual outcome. However, the relapsing nature of the disease and its complications may result in a poor visual outcome.

Cataract in sympathetic ophthalmia often occurs secondary to recurrent anterior segment inflammation and long-term corticosteroid therapy.[4–6] The incidence of cataract in these high-risk uveitic eyes is not known. Cataract surgery may be required when cataract impairs vision or fundus examination.

The literature has few reports of the outcome of cataract surgery in patients with sympathetic ophthalmia.[7,8] In this retrospective case series, we evaluated the outcome of different types of cataract surgery in 18 eyes of 17 patients with a definite diagnosis of sympathetic ophthalmia.

Patients and Methods

Sixty-six patients (132 eyes; 66 sympathizing and 66 exciting) with sympathetic ophthalmia were seen at the uveitis referral clinic, Sankara Nethralaya, Medical Research Foundation, Chennai, India, between January 1990 and July 2001; 42 eyes (31.8%) had cataract. Cataract surgery with or without intraocular lens (IOL) implantation was performed in 17 sympathizing eyes and 1 exciting eye (17 patients) during this period.

The records of the patients having cataract surgery were retrospectively reviewed with regard to duration of sympathetic ophthalmia, preoperative medications, interval between the last inflammatory episode and surgery, duration of postoperative follow-up, preoperative and postoperative visual acuities, intraoperative and postoperative complications, and postoperative fundus findings. Patients who did not have a minimum follow-up of 3 months were excluded from the study.

All 17 patients were diagnosed with sympathetic ophthalmia based on a history of penetrating ocular trauma or ocular surgery in 1 eye and clinical features of sympathetic ophthalmia in the other eye. Clinical features included granulomatous anterior uveitis or panuveitis, peripapillary choroiditis or exudative retinal detachment, and papillitis. To document thickening of the choroid, ultrasonography during the acute phase of the clinical presentation was done in all 17 sympathizing eyes. All patients also had a complete systemic examination to rule out systemic disease such as tuberculosis and sarcoidosis. The cause of sympathetic ophthalmia included penetrating ocular trauma in 8 cases, ocular surgery in 6 cases, perforated corneal ulcer in 2 cases, and cyclocryotherapy[9] in 1 case. Demographic characteristics such as the age and sex of the patients were noted.

All patients had a complete ophthalmic examination preoperatively that included Snellen best corrected visual acuity (BCVA), anterior segment evaluation with a Haag-Streit slitlamp (1.6 mm × 1.0 mm beam) to note the condition of the anterior chamber and the presence of vitreous inflammation, extent of posterior synechias, pupil dilation, type of cataract, intraocular pressure (IOP) by applanation tonometry, and fundus evaluation by indirect ophthalmoscopy. Ultrasonography was done to assess the posterior segment in eyes in which dense cataract precluded view of the fundus.

All eyes had to be free of inflammation for at least 3 months before surgery. Control of inflammation was achieved with systemic, periocular, and topical steroids and with immunosuppressive agents when required. The absence of anterior chamber cells was confirmed at the last preoperative visit. Systemic oral prednisolone, 1 mg/kg body weight, was administered to all patients 4 days before surgery and was gradually tapered over 4 to 6 weeks postoperatively depending on the degree of ocular inflammation. Two days before surgery, 12 eyes had posterior sub-Tenon's injection of 20 mg triamcinolone acetonide using the Smith and Nozik technique.[10] The other 6 eyes did not receive periocular steroids as they were diagnosed with steroid-responsive glaucoma. All patients were operated on under cover of preoperative antibiotics, and all received peribulbar anesthesia.

The operated eyes were divided into 3 groups based on type of cataract surgery. Group 1 had conventional extracapsular cataract extraction (ECCE) with single-piece poly(methyl methacrylate) (PMMA) posterior chamber IOL (CeeOn® 724B, Pharmacia) implantation (n = 3). Group 2 had conventional ECCE without IOL implantation (n = 6). Group 3 had phacoemulsification with posterior chamber IOL implantation (n = 9). In the phacoemulsification group, 4 eyes had implantation of a single-piece PMMA IOL (Slim Plant LX10BD, Alcon) and 5 eyes, of a hydrophilic heparin-surface-modified PMMA IOL (CeeOn 812C, Pharmacia).

Surgical Technique

In Groups 1 and 2, conventional ECCE was performed with a routine corneoscleral section. Posterior synechias were released by synechiolysis with an iris spatula under cover of hydroxypropyl methylcellulose. Pupils were dilated by sphincterotomy, sector or keyhole iridectomy, or stretching with hooks. A can-opener anterior capsulotomy was created and the lens delivered using a bimanual technique. Meticulous cortical cleanup was followed by IOL implantation in Group 1 and no IOL implantation in Group 2. The corneoscleral wound was sutured with 10-0 nylon.

In Group 3, phacoemulsification was performed through a 5.0 mm scleral tunnel into the anterior chamber. Synechio-

lysis was done using an iris spatula under cover of sodium hyaluronate 1%. In eyes with small pupils, the pupil was stretched manually with Kuglen hooks or self-retaining iris hooks were used. The nucleus was sculpted and emulsified using a divide-and-conquer technique. After cortical aspiration with a mechanical irrigation/aspiration device, a posterior chamber IOL was implanted in the capsular bag.

Postoperatively, patients were treated with topical and systemic steroids that were tapered over 4 to 6 weeks. Patients who took immunosuppressives preoperatively were maintained on them postoperatively. The frequency of the topical steroids and dosage of the systemic steroids were altered depending on the postoperative inflammatory response.

Patients were examined 1 and 3 days postoperatively and then 6 weeks, 3 months, and 6 months later. The postoperative visits at 1 and 3 days included evaluation of the cataract section, corneal status, anterior chamber inflammation, pupil, IOL location, visual acuity, and IOP as well as the presence of an IOL or posterior synechias, IOL deposits, and posterior capsule opacification (PCO). At subsequent postoperative visits, a complete ophthalmic examination was done that included Snellen BCVA, slitlamp evaluation of the anterior segment, applanation tonometry, and detailed fundus evaluation by indirect ophthalmoscopy.

Using the above data, the results of the cataract surgery were analyzed. The main parameters analyzed were visual outcome, incidence of complications, and reactivation of the sympathetic ophthalmia.

Results

Preoperative Data

The causes of sympathetic ophthalmia were penetrating trauma (n = 8), ocular surgery (n = 6), perforated corneal ulcer (n = 2), and cyclocryotherapy (n = 1). The most common cataract type was mixed (posterior subcapsular and posterior polar), which was present in 7 eyes (38.9%).

Eleven men (64.7%) and 6 women (35.3%) (9 right eyes, 9 left eyes) had cataract surgery performed by 1 of 2 surgeons. One patient had cataract surgery in both the exciting eye and the sympathizing eye. The mean age of the patients at the time of surgery was 47.8 years (range 8 to 69 years). The mean time from the onset of sympathetic ophthalmia to surgery was 11.3 months (range 3 to 37 months).

In 10 eyes (55.6%), the pupil diameter was 5.0 to 6.0 mm. In 8 eyes (44.4%), 4-quadrant posterior synechias resulted in a pupil diameter of 3.0 mm. Secondary glaucoma was diagnosed preoperatively in 4 eyes and was successfully controlled with antiglaucoma

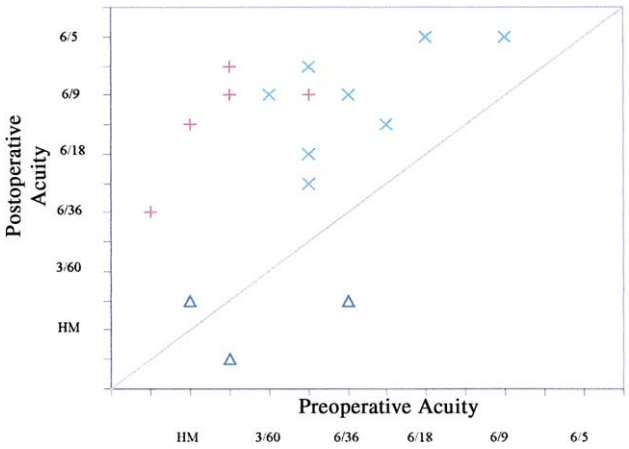

Figure 1. (Ganesh) Comparison of preoperative and postoperative visual outcomes (+ = ECCE with no IOL implantation; × = phacoemulsification and IOL implantation; Δ = ECCE and IOL implantation).

medications. In 13 eyes, chorioretinal scarring and subretinal gliotic alterations suggestive of the remission phase of sympathetic ophthalmia were noted. The remaining 5 eyes had an ultrasound evaluation as the fundus could not be visualized because of dense cataract.

Intraoperative Data

In eyes with small pupils, synechiolysis was done with an iris spatula under cover of viscoelastic material in 11 eyes. Iris hooks were used in 3 eyes to stretch and dilate the pupil. No significant intraoperative complications were encountered.

Postoperative Data

The mean postoperative follow-up was 28.7 months (range 3 to 60 months). One eye in Group 2 and 1 eye in Group 3 were lost to follow-up after 6 weeks and were excluded from the study.

Figure 1 compares the preoperative and postoperative visual outcomes. At the final follow-up, 13 eyes (72.2%) had better visual acuity than preoperatively; 12 eyes (66.6%) had an improvement of 2 or more Snellen lines, and 10 eyes (55.5%) achieved a BCVA of 20/40 or better. None of the 3 eyes in Group 1 had improved vision postoperatively; 1 eye developed a thick inflammatory membrane behind the IOL that required multiple neodymium:YAG (Nd:YAG) capsulotomies, and 2 eyes developed a severe submacular scar that impaired visual improvement.

Two eyes developed a fibrinous reaction in the immediate postoperative period that resolved with aggressive

Table 1. Additional complications.

Complication	Eyes
Secondary glaucoma	6
Submacular scar	5
Disc pallor	4
Cystoid macular edema	2
IOL synechias	1
Pupillary capture	1
Vitreous hemorrhage	1

IOL = intraocular lens

oral and topical steroid treatment. Of the 14 eyes (87.5%) that developed posterior capsule fibrosis, 6 required an Nd:YAG capsulotomy as the fibrosis was visually significant.

The sympathetic ophthalmia reactivated in 6 eyes (3 in Group 1 and 3 in Group 3) postoperatively. In most of these eyes, reactivation occurred more than once.

Secondary glaucoma was noted preoperatively in 4 eyes and postoperatively in 1 eye. The glaucoma was managed with medication alone.

One eye developed vitreous hemorrhage as a result of proliferative diabetic retinopathy in the sympathizing eye. The hemorrhage was managed by vitreous surgery. Table 1 shows the other postoperative complications.

Discussion

Sympathetic ophthalmia remains a serious eye disease with poor visual outcomes. Prophylaxis might include enucleation of the exciting eye before the autoimmune response develops. The complications of sympathetic ophthalmia include cataract, glaucoma, exudative retinal detachment, and optic neuritis.[2]

Treatment of sympathetic ophthalmia requires high doses of oral or pulse steroid therapy followed by long-term low-dose systemic steroid therapy. This is often supplemented with topical or periocular steroids until the inflammation resolves. In many cases, immunosuppressive medications are used as steroid-sparing agents.

Cataract, a frequent complication of sympathetic ophthalmia, results from the chronic recurrent nature of the disease process or from long-term steroid therapy. The main issues with cataract surgery in any uveitic eye are adequate control of preoperative and postoperative inflammation, intraoperative management of the pupil, and choice of IOL implantation as these high-risk eyes are prone to complications that result from reactivation of inflammation after breakdown of the blood–aqueous barrier (BAB). There are no reliable data on the safety of cataract surgery in patients with sympathetic ophthalmia. Although few, recent articles on cataract extraction in uveitis[4,11–17] report that cataract surgery and IOL implantation are safe in eyes with uveitis. A Medline search revealed no major study on cataract surgery in sympathetic ophthalmia except isolated case reports on cataract extraction in sympathizing eyes,[7,8] probably because of the rarity of the disease.

Our retrospective study is probably the largest series of cataract surgeries in patients with sympathetic ophthalmia. It comprised 66 sympathizing eyes, 42 (63.6%) of which had significant cataract. In the 18 eyes (17 sympathizing, 1 exciting) that had cataract surgery, mixed cataract (posterior subcapsular cataract and posterior polar) was most common. However, we found no other study with which to compare the incidence and type of cataract with our results. The high incidence of cataract in our series was probably the result of 3 factors: Ours is a tertiary referral center, high-doses of steroids were frequently used, and there was a high rate of disease reactivation.

In our series, penetrating trauma was the most common cause of sympathetic ophthalmia followed by surgical trauma. This is comparable to the results in previous studies that report a high incidence of sympathetic ophthalmia after penetrating and surgical trauma.[2] The female:male ratio was 6:11 in our study, which compares favorably with the ratio in previous studies that describe a higher incidence of sympathetic ophthalmia in men, especially after eye injury.[2]

In our study, we could not determine the exact mean age at which cataract developed as some patients had cataract at presentation. However, most patients were in the fifth decade of life at the time of surgery, with a mean age of 47.8 years.

The reported risk factors for cataract in any eye with uveitis include chronic recurrent inflammation, particularly recurrent anterior segment inflammation; long duration of disease; and steroid therapy for more than 6 months.[5,6,11] Similar risk factors accounted for the cataracts in our series.

Management of visually significant cataracts in patients with sympathetic ophthalmia requires careful sur-

gical planning and handling of ocular tissues during surgery as well as meticulous preoperative, intraoperative, and postoperative control of inflammation. The surgical plan and intraoperative management depend on preoperative anterior segment findings; that is, type of cataract, pupil diameter, extent of posterior synechias, and presence of pupillary membrane. Eight eyes (44.4%) with 4-quadrant posterior synechias had a pupil diameter of approximately 3.0 mm. Although this limited access to the cataract, the small pupils were successfully managed with synechiolysis alone; iris-stretching maneuvers; or self-retaining, flexible iris hooks.

Good preoperative control of inflammation and meticulous surgical technique led to few complications during surgery.

Posterior capsule opacification was the most common cause of postoperative visual impairment after uneventful cataract surgery, occurring in all 3 groups (14 eyes, 77.7%). Visually significant PCO requiring an Nd:YAG capsulotomy was present in 6 eyes (33.3%). A primary posterior curvilinear capsulotomy can be done in these high-risk eyes.

At the final follow-up, 13 eyes (72.2%) had improved visual acuity, 12 (66.6%) by 2 or more Snellen lines, and 10 eyes (55.5%) achieved a BCVA of 20/40 or better. All eyes in Groups 2 and 3 had improved vision; however, no eye in Group 1 did as a result of a thick inflammatory membrane behind the IOL requiring multiple Nd:YAG capsulotomies or a significant submacular scar. As observed in several recent studies of cataract surgery in uveitis,[15–17] as well as in our study, the key to a good visual outcome after cataract surgery is strict preoperative and postoperative control of inflammation along with meticulous surgical planning and technique. The causes of poor visual recovery in our study were the result of preexisting ocular pathologies involving the retina or optic nerve that were sequelae of sympathetic ophthalmia. Hence, the final visual outcome depends on the posterior segment complications of sympathetic ophthalmia. Disease reactivation occurred equally in all 3 groups.

The results in our study indicate that phacoemulsification has advantages in the management of complicated cataract in sympathetic ophthalmia. Phacoemulsification causes less BAB breakdown, and creation of a continuous curvilinear capsulorhexis ensures in-the-bag IOL placement. Twelve eyes, 9 having phacoemulsification and 3 having manual ECCE, had IOL implantation. Highly biocompatible IOLs, such as those with heparin surface modification, were used in the phacoemulsification group. No eye that had phacoemulsification developed IOL synechias. However, after manual ECCE, 1 eye developed IOL synechias; 1 eye, pupillary capture of the IOL optic; and 1 eye, a thick inflammatory membrane behind the IOL requiring multiple Nd:YAG capsulotomies. Thus, we believe that phacoemulsification with in-the-bag IOL is probably a better surgical option than manual ECCE with IOL implantation in these eyes.

Conclusion

Although our study was nonrandomized and retrospective, with all the inherent drawbacks, it still is probably the largest analyzing the results of cataract surgery in cases of sympathetic ophthalmia. Our results indicate that many patients with uveitis, particularly sympathetic ophthalmia, are young and that IOL implantation is the best means of visual rehabilitation.

Although sympathetic ophthalmia is a rare disease with severe complications, cataract surgery, especially phacoemulsification with in-the-bag IOL implantation, is safe and gives good results with vigilant preoperative and postoperative care and careful, meticulous intraoperative techniques. A larger randomized prospective multicenter clinical trial is required to develop definite guidelines and conclusions.

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