Visual and Surgical Outcome of Phacoemulsification and Intraocular Lens (IOL) Implantation in Patients with Retinitis Pigmentosa

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ABSTRACT

BACKGROUND

Retinitis pigmentosa is a group of hereditary disorders characterised by difficulty of seeing at night, bony spicule pigmentation in mid retinal periphery, progressive visual field loss and abnormal electroretinogram (ERG) responses. Cataract surgery in patients with retinitis pigmentosa is associated with poor visual outcome with higher incidence of intraoperative and postoperative complications. This study was done to find the visual and surgical outcome of phacoemulsification and intraocular lens (IOL) implantation in patients with retinitis pigmentosa.

METHODS

This was a retrospective, non-comparative, observational study done at Regional Institute of Ophthalmology, RIMS Ranchi, India. Consecutive patients of retinitis pigmentosa who underwent phacoemulsification and IOL implantation between July 2015 to December 2019 were retrospectively analysed. Intraoperative and postoperative complications, visual and refractive outcomes were analysed.

RESULTS

Fifty six eyes of 48 patients (29 male, 19 female) with mean age 46.62 ± 8.45 years were included in study. Mean follow up was 17.92 ± 10.62 months. The types of cataract were posterior subcapsular in 30.35 %, cortical in 14.2 %, mixed in 51.8 % and nuclear sclerosis in 3.5 % of eyes. Intraoperative posterior capsular rent (PCR) was noted in 2 eyes (3.57 %) and zonular dehiscence in 3 eyes (35.35 %). Preoperative mean log minimum angle of resolution best corrected visual acuity (MARBCVA) was 1.45 ± 0.49 which improved to 1.24 ± 0.28 postoperatively at 4 weeks (P-value < 0.05). Postoperatively, vision improved in 46 (82.14 %) eyes, unchanged in 7 (12.5 %) eyes and worsened in 3 (5.35 %) eyes. Postoperative cystoid macular oedema (CME) was noted in 5 eyes (9.07 %). Significant posterior capsular opacification (PCO) was noted in 15 eyes (26.78 %) and anterior capsular phimosis was noted in 3 eyes (5.35 %).

CONCLUSIONS

Cataract surgery improves vision in most of the patients with retinitis pigmentosa. The incidence of PCR, zonular dehiscence, CME, PCO and anterior capsular phimosis is higher in these patients.

KEYWORDS

Retinitis Pigmentosa, Cataract, Capsular Phimosis

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BACKGROUND

Retinitis pigmentosa (RP) is a group of rare hereditary disorders that affects retinal photoreceptor and pigment epithelial function. It results in peripheral visual field loss and abnormal ERG responses.[1] Patients with retinitis pigmentosa typically had history of night blindness and presence of classical triad of bony spicules pigmentation on the retinal mid periphery, waxy optic disc pallor and arteriolar attenuation on fundus examination.

Cataract is a recognised complication of all types of retinitis pigmentosa and develops at relatively younger age, most common type being posterior subcapsular cataract (PSC).[1-3] Development of cataract further jeopardise patient vision by affecting central vision as well. Even lesser grade of cataract might cause significant visual disturbances. Also the incidence of glare is more in patients with retinitis pigmentosa who develops posterior subcapsular cataract.

Patients with retinitis pigmentosa are more prone to specific risk factors during cataract surgery. These include phototoxic retinal damage, post-operative cystoids macular oedema (CME), posterior capsular opacification (PCO) and anterior capsular phimosis.[4,5,6] These complications may result in poor visual outcome following cataract surgery. Also, in advanced retinitis pigmentosa, there is outer retinal atrophy of the macula resulting in very poor visual outcome. Also, there can be presence of pre-existing cystoid macular oedema even in less advanced stage of retinitis pigmentosa, which can result in sub optimal visual outcome.

Also, doing cataract surgery is more challenging in patients with retinitis pigmentosa due to diffuse zonular weakness.[7] So, risk of intraoperative complications such as posterior capsule rupture and zonular dehiscence are higher in patients with retinitis pigmentosa. Patients with zonular dehiscence will require implantation of capsular tension ring to stabilise the capsular bag and centration of intraocular lens. Also, there is risk of dislocation of intraocular lens and subluxation of IOL bag complex.[8]

Since limited data was available on outcome of phacoemulsication and IOL implantation in patients with retinitis pigmentosa, especially on this topic. So, this study was planned to assess the visual outcome and any intraoperative or postoperative complications of cataract surgery & IOL implantation in patients with retinitis pigmentosa.

METHODS

This was a retrospective observational study done at Regional Institute of Ophthalmology, RIMS Ranchi, India. All consecutive patients with retinitis pigmentosa who underwent phacoemulsification and intraocular lens (IOL) implantation between July 2015 to December 2019 were included in the study.

Diagnosis of retinitis pigmentosa was based on history of night blindness and presence of classical triad of bony spicules pigmentation, waxy optic disc pallor and arteriolar attenuation on fundus examination. The indications for cataract surgery in patients with retinitis pigmentosa was loss of central vision due to lenticular opacity. Patients with minimum follow up of 6 months were included in study. Patients with history of uncontrolled diabetes, active uveitis, retinal vascular occlusion disease, pseudoexfoliation syndrome, traumatic cataract or any prior ocular surgery were excluded from study.

All patients underwent a thorough preoperative ocular examination including visual acuity, intraocular pressure (IOP) by applanation tonometer and slit lamp examination. The visual acuity was measured by using Snellen visual acuity chart at 6 metre and converted into logarithm of minimum angle of resolution (log MAR) values for statistical analysis. Grading of cataract was done according to lens opacification classification system III. (LOCSIII)

Fundus examination was done after pupillary dilatation with indirect ophthalmoscopy. Pre-operative optical coherence tomography (OCT) of the macula was done in selected cases where media clarity allowed the good scan. This was done to better prognosticate the visual outcome. Automated visual field and electroretinogram were also done in selected cases.

Informed consent was taken from each patient. They were informed that only central vision will improve after surgery to certain extent and peripheral vision as well as dim vision will remain same. Also, the risk of intraoperative and postoperative complications were explained in detail.

Biometry was performed by immersion ultrasound A-scan and manual keratometer. Intraocular lens power was calculated by using SRK-T formula. Preoperatively all patients were treated with topical moxifloxacin 0.5 % 3 times a day (TDS) and nepafenac 0.3 % TDS two days before surgery.

Surgical Procedure

All surgeries were done by single surgeon (SK) under topical or peribulbar anaesthesia. Two side ports were made and anterior chamber was filled with cohesive viscoelastic. Main incisions were made with 2.8 mm keratome and capsulorhexis of at least 5 to 5.5 mm were made. Nuclear emulsification was done with direct chop technique. Cortical aspirations were done with bimanual irrigation – aspiration canula and hydrophobic acrylic IOL were implanted in the capsular bag.

Postoperatively all patients were prescribed topical prednisolone acetate 1 % 4 - 6 times a day which was tapered off in 4 - 6 weeks, moxifloxacin 0.5 % four times a day (QID) for 10 days and nepafenac 0.3 % TDS for two months. Postoperative follow up examination was done at day 1, weekly for first month, monthly for next 3 months and after that at every 3 months. Visual acuity, IOP, slit lamp examination and fundus examination was done at every follow up visits. Refraction was done in all cases at 3 - 4 weeks. Optical coherence tomography of macula was performed at 1 and 3 months.

Statistical Analysis

All the data was entered on Microsoft Excel sheet and analysed using SPSS 21.0 Package Statistical Package for the

Social Sciences (SPSS Inc. Chicago, USA). The mean age, visual acuity and axial length were calculated using Microsoft Excel. Paired t-test was used to compare between preoperative and postoperative best corrected visual acuity (BCVA). A P-value < 0.05 was considered to be statistically significant.

RESULTS

A total of fifty six eyes of 48 patients with mean age 46.62 ± 8.48 (range 38 - 57 years) were included in study, out of which 29 (60.41 %) were males and 19 (39.58 %) were females (Table 1). The mean follow-up was 17.92 ± 10.62 months (range 6 - 36 months). The types of cataract were posterior subcapsular in 17 eyes (30.35 %), mixed in 29 eyes (51.8 %), cortical cataract in 8 eyes (14.2 %) and nuclear sclerosis in 2 eyes (3.5 %). The mean axial length was 22.86 ± 0.87 mm (range 21 - 25 mm) and mean IOL power was 21.06 ± 0.35 D. (range 18 - 24).

Visual outcome after surgery are shown in Table 2 and Table 3. Following surgery, BCVA improved in 46 (82.14 %) eyes, unchanged in 7 (12.5 %) eyes and worsened in 3 (5.35 %) eyes. 44 patients (91.66 %) had improvement in symptoms of glare and haloes. The mean preoperative BCVA was 1.45 ± 0.49 logMAR (range 1.0 - 2.0) which improved significantly to 1.24 ± 0.28 log MAR (range 0.6 - 2.0). This improvement in vision was statistically highly significant (P-value = 0.012).

Various intraoperative and postoperative complications are shown in Table 4. Intraoperatively, posterior capsular rent occurred in two eyes (3.57 %) and zonular dehiscence was noted in three eyes (5.35 %).

Cystoid macular oedema developed in 5 eyes (9.07 %) on OCT and fluorescein angiography. Fifteen eyes (26.78 %) developed posterior capsular opacification requiring neodymium-doped yttrium aluminum garnet (ND: YAG) capsulotomy. Mean period of development of posterior capsular opacification was 10.85 months. Anterior capsular phimosis occurred in three eyes (5.35 %), managed by ND: YAG anterior capsulotomy. None of the eyes had IOL dislocation or late subluxation of IOL bag complex.

Parameters	Values
Age (Mean ± SD) (yrs.)	46.62 ± 8.45
Male	29 (60.41 %)
Female	19 (39.58 %)
Axial length (mm)	22.86 ± 0.87
LogMARBCVA (Mean ± SD)	1.45 ± 0.49
Types of Cataract	
1. Post-subcapsular	17 (30.35 %)
2. Cortical	08 (14.20 %)
3. Nuclear	02 (3.5 %)
4. Mixed	29 (51.80 %)

Table 1. Baseline Demographic and Preoperative Data of Patients

Visual Acuity	N (%)
Improved	46 (82.14 %)
Unchanged	07 (12.5 %)
Worsened	03 (5.35 %)

Table 2. Visual Outcome Following Surgery

Variables	Pre-Operative (Mean ± SD)	Post-Operative (Mean ± SD)	t-Value [95 % CI]	P-Value
Visual acuity (BCVA) in logMAR	1.45 ± 0.49	1.24 ± 0.28	2.55	0.012*

Table 3. Comparison of Visual Acuity Pre- and Post-Surgery in Terms of BCVA

*Indicates statistically significant difference at P < 0.05

Complications	N (%)
Posterior capsular rent	02 (3.57 %)
Zonular dehiscence	03 (5.35 %)
Posterior capsular opacification	15 (26.78 %)
Anterior capsular phimosis	03 (5.35 %)
Cystoid macular oedema	09 (16.07 %)

Table 4. Intraoperative and Postoperative Complications

DISCUSSION

Cataract is common ocular complication in patients with retinitis pigmentosa which affects the central vision. Cataract surgery in patients with retinitis pigmentosa may have poor visual outcome due to various intraoperative and postoperative complications.

Cataract has been observed to develop at a relatively younger age in patients with retinitis pigmentosa, with a mean age at surgery of 47 - 58 years in various studies.[5,6,9,10] In our study, mean age at surgery was 46.62 years which was similar to other studies.

Previous studies have reported higher incidence of postoperative cystoid macular oedema, posterior capsular opacification and anterior capsular phimosis in patients with retinitis pigmentosa.[4,5,9]

Higher incidence of posterior capsular rupture has been noticed in previous study.[10,7] High risk of posterior capsule rupture could be because of lax posterior capsule due to zonular weakness. In our study also posterior capsular rupture occurred in 3.5 % of eyes. Cases with posterior capsular rupture were managed by doing two port limbal anterior vitrectomy. Also, in cases with posterior capsular rupture, three-piece hydrophobic acrylic IOL were implanted in the ciliary sulcus and optic capture was done in anterior capsulorhexis margin for better centration of IOL.

Also in patients with retinitis pigmentosa there was diffuse zonular weakness. Exact pathogenesis of zonular weakness in patients with retinitis pigmentosa was not known. This could be because of damage to the zonules by toxic substances and cytokines by degenerating retina. In a study by Dikopf MS et al. incidence of zonular insufficiency was found in 18.8 % of cases.[7]

In our study, zonular dehiscence was noted intraoperatively in 5.35 % of eyes. Cases with zonular dehiscence were managed by implantation of capsular tension ring in the bag. Capsular tension ring helped by expanding the capsular bag and evenly distributing zonular tension. It also helped in reducing the risk of anterior capsular contraction. But advanced zonulopathy will require implantation of modified capsular tension ring such as Cionni ring which has to be sutured to the sclera.

Postoperatively, vision improved in 82.14 % patients in our study which was comparable to previous study.[11,12] In 7.5 % of cases visual acuity did not improve after surgery. This was mainly because of cystoid macular oedema and

outer retinal atrophy of macula especially in cases with advanced retinitis pigmentosa. In 5.35 % of cases, visual acuity worsened after surgery. This was due to development of cystoids macular oedema. More than 91 % of patients had improvement in their visual symptoms like glare and haloes following surgery. Postoperative cystoid macular oedema was vision threatening complication resulting in suboptimal visual outcome. The incidence of cystoid macular oedema has been reported ranging 10 - 15 % in various studies.[4-6,10] Use of nonsteroidal anti-inflammatory drugs (NSAIDs) eye drops are recommended pre and post operatively to prevent cystoid macular oedema.

In our study, cystoid macular oedema developed in 9.07 % of eyes. Use of preoperative and postoperative topical nepafenac might had resulted in slightly lower incidence of cystoid macular oedema in our series.[13]

Posterior capsular opacification was another very common complication after cataract surgery in patient with retinitis pigmentosa. Exact pathogenesis of high incidence of posterior capsular opacification was not known. It could be because of increased cellular proliferation due to mediators released by the neurosensory retina. Posterior capsular opacification requiring Nd: YAG capsulotomy was observed in 26.78 % eyes in our study which was much less as compare to previous study.[5,6,9,10,7]

In a study by Dikopf MS et al. incidence of posterior capsular opacification who underwent Nd: YAG capsulotomy was 52.5 %. Lower incidence of posterior capsular opacification in our study could be due to better IOL material and square edge design.[14] Also, in this series all eyes underwent phacoemulsification with capsular bag IOL implantation.

Patients with retinitis pigmentosa are also more susceptible to anterior capsular contraction.[5,6] High incidence of anterior capsular phimosis in retinitis pigmentosa could be due to generalised zonular weakness. In our study, 3 eyes (5.35 %) developed anterior capsular phimosis. All eyes with anterior capsular phimosis were managed by Nd: YAG anterior capsulotomy.[15] Creation of large size capsulorhexis was important in prevention of anterior capsular phimosis.

Dislocation of intraocular lens was also reported in patients with retinitis pigmentosa.[8] This was especially common with silicone lenses. In our study, none of the eyes had dislocation of lens, and we have implanted hydrophobic acrylic lenses in all patients. Also in this study, none of the eyes had late subluxation of IOL bag complex. All three eyes in which capsular tension ring was implanted did well and had well centred IOL.

Preoperative counselling was also very important in these patients. They should be given realistic expectation and must be explained that only central vision will improve after surgery and peripheral as well as night vision will remain same. Also increased risk of intraoperative and postoperative complications were explained in detail.

Preoperative optical coherence tomography of the macula should be done in all cases. This helped in detection of outer retinal atrophy and cystoid macular oedema. Eyes with outer retinal atrophy of the macula had suboptimal visual outcome.

The strength of our study was that it had sufficient number of patients from single centre and all cases were operated by single surgeon. The major drawback of this study was that it was a retrospective and also optical coherence tomography of the macula could not be done in all cases. So, the exact incidence of cystoid macular oedema could be higher than what was noted in this study.

CONCLUSIONS

Cataract surgery improves vision in most of the patients with retinitis pigmentosa. The postoperative incidence of posterior capsular opacification, cystoid macular oedema and anterior capsular phimosis is higher in patients with retinitis pigmentosa. Large size capsulorhexis and acrylic lenses are recommended to avoid complications.

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