Pseudoexfoliation Syndrome (PEX): Incidence of Glaucoma, Cataract and Related Surgical Complications

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ABSTRACT

Introduction: Pseudoexfoliation syndrome [PEX] is a relatively common but easily overlooked cause of chronic open angle glaucoma. Cataract extraction in the setting of PEX requires careful surgical evaluation and communication with the patient regarding increased surgical risks. Anticipating and managing the intra-operative surgical risks and possible postoperative issues associated with PEX pathology increases successful outcomes.

Material and Methods: The present study was done at tertiary care hospital in Western India over a period of 2 and half years. It was a prospective study which included 67 eyes of 48 patients with PEX, analysing complications and surgical outcomes in patients with PEX undergoing cataract surgery.

Results: Nuclear sclerosis grades 1 to 4 were present in 73.96% eyes of patients with PEX while 23.96% eyes had mature or hypermature cataracts. Intra-operative complications noted in this study included intra-operative floppy iris (IFI) (31.34%), posterior capsule tear (PCT) with vitreous loss was noted in 5 eyes (7.46%), zonular dialysis (ZD) was noted in 4 eyes (5.97%), out of vitreous loss occurred in 2 eyes, thus overall 7 eyes had vitreous loss (10.45%). Major late post-operative complications included raised IOP (5/67 eyes-7.46%), IOL decentration (5/67 eyes-7.46%) and posterior capsule opacity (PCO) which was seen in 28 eyes within 1 year follow-up (41.79%).

Conclusion: Our study highlights the importance of regular IOP monitoring and keeping a vigil for the signs of PEX in the elderly. Further larger and extensive studies need to be carried out for statistical analysis of PEX related manifestation and surgical outcomes.

Keywords: Pseudoexfoliation Syndrome, PEX, Glaucoma, Cataract, Prognosis, Surgical Outcome

INTRODUCTION

Pseudoexfoliation syndrome [PEX] is an age-related disease in which abnormal fibrillar extracellular material is produced and accumulates in many ocular tissues. PEX is a relatively common but easily overlooked cause of chronic open angle glaucoma. When PEX is accompanied by glaucoma, the terminology preferred is Pseudoexfoliation Glaucoma [PEG] or "Capsular Glaucoma". The PEX has to be differentiated from true exfoliation, pigmentary glaucoma and primary amyloidosis.[1,2]

Glaucoma occurs more commonly in eyes with PEX than in those without it; 50% of patients will develop glaucoma at 10 years.[2,3] Patients with PEX are also predisposed to develop angle-closure glaucoma. Further, glaucoma in PEX has a more serious clinical course and worse prognosis than primary open-angle glaucoma. There is increasing evidence for an etiological association of XFS with cataract formation, and possibly with retinal vein occlusion.[5]

Pseudoexfoliation (PEX) syndrome is frequently associated with open angle glaucoma, melanin dispersion, poor pupillary dilatation, and may present with a specific type of PEX keratopathy. These features in combination with zonular instability predispose eyes with PEX to complications during cataract extraction.[6]

However, PXF is a systemic condition with PXF material present throughout the body. The presence of PXF material has been weakly associated with cardiovascular, cerebrovascular morbidity and sensorineural hearing loss. Unlike PXF and glaucoma, a direct cause-effect relationship remains to be clearly established between the presence of PXF material and the aforementioned systemic associations.[7]

Even though this is a systemic disorder, many studies have demonstrated either predominately unilateral or bilateral involvement depending on the patient demographics. In general, patients with bilateral involvement tend to be older.[7]

Initial management is medical, but frequently surgical intervention is necessary because PXF glaucoma is more aggressive than primary open angle glaucoma. Cataract extraction in the setting of PXF requires careful surgical evaluation and communication with the patient regarding increased surgical risks.

Study was done with the aim of anticipating and managing the intra-operative surgical risks and possible postoperative issues associated with PXF pathology increases successful outcomes.

MATERIAL AND METHODS

The present study was done at the department of Ophthalmology at a Tertiary Hospital over a period of two and half years in Western India. It was a prospective study which included 67 eyes of 48 patients for analyzing prognosis and surgical outcomes in patients with Pseudoexfoliation syndrome.

Inclusion Criteria were age > 40 year, signs of PEX either unilateral and/or bilateral presentation in all patients presenting in OPD during the period.

Exclusion Criteria were prior history of any ocular surgery, uveitis, keratopathy, complicated cataract, trauma to eye, mental disorders, patient not willing to be a part of study, intraocular tumors and true exfoliation syndrome.

Each patient was followed up for a period of one year. Examination of each patient included complete ophthalmic

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examination along with Goldmann applanation tonometry, gonioscopy. Automated perimetry was advised and performed if needed as per the clinical situation.

Cataract surgery was advised if there was advanced cataract or in patients with poor vision documented due to cataract. Pre-operative dilatation was done with standard (tropicamide 0.8%, phenylephrine 5% and flurbiprofen 0.03%) drops were instilled every 15 – 20 minutes one hour prior to surgery to maintain pupillary dilatation, pupil was considered small or non-dilating if it failed to dilate beyond 6mm on slit lamp examination.

All patients with PEX and Cataract underwent Phacoemulsification with Intraocular lens implantation (until unless precluded due to intra-operative complication). Meticulous surgery was performed and intra-operative complications managed accordingly. Position and type of IOL implantation was noted.

Patients diagnosed with glaucoma were classified into open or closed angle depending on gonioscopy findings. Patients found to have glaucoma were managed accordingly.Patients with PEG were followed-up initially weekly for a month and then three monthly.On each visit patients underwent slit lamp examination, IOP check-up with applanation tonometry and fundus examination; gonioscopy and perimetry was performed initially and then at 6 month interval.

RESULTS

Our study shows that those with PEX were diagnosed clinically with Glaucoma or PEG in about 10% of patients (Table 1). The incidence of dense grades of cataract (NS3, NS4 and Mature/Hypermature) was found to be around 54% (Table 2). Surgical intervention for cataract was needed in about 67% of patients with PEG. The preferred procedure used was Phacoemulsification. Trabeculectomy was performed where clinically significant (Table 3).

Nearly half of those undergoing surgery in PEX reported complications. The most common complication noted was floppy iris syndrome, about 1/3 of patients. The rate of vitreous loss was also significant, being around 10% (Table 4). The most common type of IOL was in the bag, followed by in sulcus. Two patients were left aphakic, as the post operative inflammation was anticipated to be more (Table 5).

Glaucoma	No. of patients	Percentage (%)
Present	5	10.42
Absent	43	89.58
Total	48	100

Table-1: Incidence of glaucoma in patients with PEX

Grades of Cataract	No. of eyes	Percentage (%)
NS1-NS2	42	43.75
NS3	13	13.54
NS4	16	16.67
Mature and Hypermature	23	23.96
Pre-existing PCO	02	2.08
Total	96	100

Table-2: Status of Lens/IOL in patients with PEX

Management	No. of eyes	Percentage (%)
Medical therapy	1	11.11
Laser iridotomy	2	22.22
Trabeculectomy (with Cataract extraction)	6	66.67
Total	9	100

Table-3: Management of Glaucoma

Complication	No. of eyes	Percentage (%)
Intra-operative floppy iris syndrome (IFI)	21	31.34
Posterior capsule tear (PCT)	5	7.46
Zonular dialysis (ZD)	4	5.97
Vitreous loss (VL)	7	10.45
No complication	37	55.22
(Note- More than one complication occurred in few patients. Hence the above result.)		

Table-4: Intra-operative complications during cataract surgery

IOL implantation	No. of eyes	Percentage (%)
In the Bag	61	91.04
PCIOL on sulcus	03	4.48
ACIOL	01	1.49
Aphakia	02	2.99
Total	67	100

Table-5: IOL implantation

DISCUSSION

PEX is accompanied by glaucoma, the terminology preferred is Pseudoexfoliation Glaucoma [PEG] or “Capsular Glaucoma”. The prevalence of PEX in glaucoma patients, particularly open angle glaucoma is significantly higher than in age-matched non-glaucomatous populations. Between 3 and 47% of open angle glaucoma cases are exfoliative glaucoma. About 10-20% of newly discovered PEX patients also have glaucoma or elevated IOP found at the same time and about 50% are ultimately diagnosed with glaucoma. This indicates that PEX is a warning sign that glaucoma may develop. Though PEX is more common in women, the risk of glaucoma is however higher in males.[1,2]

In this study we found that incidence of glaucoma (Table 1) in PEX was 10.42%, which is in accordance to most of the published reports.[8,9]

PEX glaucoma is thought to arise secondary to congestion of the trabecular meshwork by pseudoexfoliative material. PEX glaucoma is the most common form of secondary open angle glaucoma. Less commonly the mechanism of glaucoma is a consequence of angle closure associated with zonular laxity. In general, PEX-associated glaucoma has a more aggressive course compared with primary open angle glaucoma. The mean IOP is also higher in affected PEX glaucoma individuals compared with primary open angle glaucoma patients with IOPs that also fluctuate more widely.[4,11] This study showed that 60% of patient had open angle glaucoma while 40% patient had closed angles. This is in accordance with the literature.[12,13]

Nine eyes of five patients had glaucomatous changes; out of these six eyes (66.67%) underwent trabeculectomy combined with cataract extraction in the same procedure. As all these patients also had advanced cataract; in two eyes (22.22%) a prophylactic laser peripheral iridotomy was done and one eye was put on medical therapy consisting of beta-blockers. In all

patients intraocular pressure was maintained at the end of one year follow-up.

A higher percentage of nuclear opacities have been observed in PEX eyes and the affected eye of individuals with uni-ocular PEX often demonstrate the more advanced cataract. The occurrence of cataract may simply be a consequence of the increased age of individuals with PEX as no causal link has been established.[15]

In this study we found that there was a high incidence of some or other degree of cataract in patients presenting with PEX. Nuclear sclerosis grades 1 to 4 were present in 73.96% eyes of patients with PEX while 23.96% eyes had mature or hypermature cataracts; showing that a majority of patients with PEX present with higher grades of cataract than in general population.[16]

The presence of PEX material poses unique challenges for cataract surgery including corneal endotheliopathy, poor mydriasis, floppy iris and tendency for iris prolapse into the cataract incision, zonular instability and lens subluxation, increased risk for vitreous loss, heightened postoperative inflammation, postoperative IOP elevation, and late intraocular lens implant decentration and prolapse into the posterior segment.[13-16]

Intra-operative complications noted in this study included (Table 4); intra-operative floppy iris (IFI) which occurred in 21 eyes out of 67 operated (31.34%), posterior capsule tear (PCT) with vitreous loss was noted in 5 eyes (7.46%), zonular dialysis (ZD) was noted in 4 eyes (5.97%), out of these four; vitreous loss occurred in 2 eyes, thus overall 7 eyes had vitreous loss (10.45%). All four eyes with ZD had implantation of capsular tension ring (CTR) to prevent collapse of bag and to avoid further complications, as all the ZD were less than or equal to 4 clock hours. Few eyes had one or more intra-operative complications. Thirty-seven eyes (55.22%) had no intra-operative complication.

Intraocular lens implantation in this study was as per table 5. These results are in accordance with other ones in regard to higher rates of intra-ocular complication and their consequences and suggests that the surgeon has to be extra cautious while operating on PEX patients and should keep instruments ready for adequate intra-operative dilatation, CTR, vitrectomy machine and stand-by ACIOLs or Iris Claw lenses.

A higher rate of post-operative complication than in general population was also seen in this study. Early post-operative complications include inflammation and striate keratopathy. Post-operative inflammation occurred in 37 eyes (55.22%) and this was the most important factor which delays immediate post-operative visual recovery. Striate keratopathy occurred in 25 eyes (37.31%) and was more common in eyes with decompensated corneas pre-operatively. Major late post-operative complications included raised IOP (5/67 eyes-7.46%), IOL decentration (5/67 eyes-7.46%) and posterior capsule opacity (PCO) viz. secondary cataract which was seen in 28 eyes within 1 year follow-up (41.79%). These results were similar to those reported by Puska et al and other studies.[17,18]

Further larger and extensive studies need to be carried out for statistical analysis of PEX related manifestation and surgical outcomes, especially with long term follow-up for the position of the lens and optic nerve status.

CONCLUSION

The present study highlights the importance of diligently following patients with Pseudoexfoliation syndrome, both for glaucoma as well as lenticular changes. Many patients ultimately require surgical management for glaucoma. A higher percentage of nuclear opacities have been observed in PEX eyes and the affected eye of individuals with PEX often demonstrates the advanced cataract. As PEX is associated with increased risk of complication during and after cataract surgery, there is a need to be prepared and counsel the patients accordingly. Longer follow-up and extensive studies are recommended to assess the outcomes in later life of the patient and their effect on life-style.

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