

Study of Patients with Pseudoexfoliation Syndrome Undergoing Manual Small Incision Cataract Surgery in a Teaching Hospital

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Purpose: To discuss preoperative, intraoperative and postoperative considerations and techniques for manual small incision cataract surgery in pseudoexfoliation (PXF) syndrome.

Materials and Methods: This prospective study was carried out in the ophthalmology department of a tertiary hospital from March 2019 to August 2019. Patients with cataract associated with PXF without glaucoma were included in this study. All patients underwent a complete ophthalmologic evaluation, including slit-lamp examination, tonometry, gonioscopy, and ophthalmoscopy before the surgery. Manual small incision cataract surgery (SICS) with posterior chamber intraocular lens were performed by a single surgeon who reported the intraoperative difficulties. Patients were followed up on day 1, day 7 and 1 month when they underwent visual acuity testing, IOP measurement and detailed slit-lamp examination.

Abstract

Results: Study included 25 eyes with PXF undergoing SICS. All cases were above 61 years of age, out of which 64% were females. Morphologically nuclear cataract (52%) was the most commonly observed. The most common intra-operative difficulty was poor pupillary dilation in 36% cases. One patient underwent anterior chamber intraocular lens (ACIOL) implantation and 1 patient underwent scleral fixated intraocular lens (SFIOL) implantation. There was no significant difference in the pre and post operative IOP, but post operative (1 month) intraocular pressure (IOP) was less than pre-op IOP ($p=0.4$). Post-operatively after 1 month 80% patients had best corrected visual acuity between 6/12 to 6/6. Majority of the patients (84%) had associated systemic illnesses like Diabetes Mellitus, Hypertension and Parkinsonism.

Conclusion: The present study highlights the importance of proper preoperative preparation, modification of the intraoperative techniques like sphincterotomy and close postoperative follow-up in cases of PXF syndrome undergoing manual SICS to achieve favorable visual outcomes.

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Keywords: Iridopathy, nuclear cataract, sphincterotomy, intra-ocular pressure, visual acuity.

Introduction

Pseudoexfoliation (PXF) syndrome is a systemic disorder, diagnosed by the appearance of a grayish white fibrillar material, also called PXF material, in abnormally high concentrations on the pupillary margin (Figure 1), lens surface, lens zonules, iris surface, corneal endothelium, trabecular meshwork, anterior hyaloid surface, intraocular lens (Figure 5) and on the posterior capsule.^{1,2} Fontana et al. in their study noted that the amyloid matter of PXF is also present in heart, lungs, liver, kidney, meninges and blood vessels which may explain why patients with ocular PXF may have a history of hypertension, abdominal aorta aneurysm, angina, cardiovascular disease and stroke.³ The reported prevalence in Indian population is 0.69-3.8%. Stein et al. in his study has mentioned prevalence of PXF in the rural population in central India as 0.95%.⁴ Age is the main risk factor for cataract progression, PXF represents an independent additional hazard for the development of nuclear sclerosis. The Framingham Eye study revealed an age related increase among non-glaucoma PXF individuals as 0.6% for ages 52 to 64 years, 2.6% for ages 65 to 74 years that rose to 5.0% for individuals aged 75 to 85 years.⁵

Although there is no established sex predilection for PXF, a female preponderance has been previously reported.⁶ The pathogenesis of PXF is multifactorial including geographical, environmental and genetic factors. There is association with mutations in the lysyl oxidase-like 1 gene (LOXL1) at the locus 15q22, which codes for elastic fiber components of extracellular matrix. For this reason, it has been suggested that PXF is a form of elastosis resulting from the overproduction of elastic microfibrillar components such as fibrillin-1.

The systemic origin of the disease explains why patients with unilateral involvement manifest PXF signs in the normal fellow eye at a later time, indicating that PXF is a generalized, bilateral disorder with a markedly asymmetric clinical presentation at onset. Patients with unilateral signs of the disease are usually younger than those with bilateral involvement.^{3,6,7}

PXF syndrome can cause changes in the eye other than glaucoma. PXF induced iridopathy and phacopathy with zonular instability makes routine cataract surgery a challenging task because of nondilating pupil, increased risk of zonular dialysis, inadequate size of Continuous Curvilinear Capsulorrhexis (CCC), associated corneal

endothelial dysfunction. A study found that such patients have 5 times greater likelihood of intraoperative complications in cataract surgery compared with normal cases like increased chances of Posterior capsule rent (PCR), Intra ocular lens (IOL) decentration (Figure 5), postoperative corneal edema etc. Recognition of this condition is hence very important before commencing surgery on such patients and take certain measures like use of soft shell technique, pupillary dilators, use of capsular tension ring (CTR), sphincterotomy which will lessen the rate of complications in pxf syndrome.⁸

Very few studies are available where manual SICS surgeries in PXF syndrome are discussed.

This study is done to evaluate the intra-operative complications of manual SICS with posterior chamber IOL (PCIOL) implantation in patients with PXF syndrome without glaucoma, their effect on post operative visual acuity and different approaches to minimize the likelihood of such complications.

Materials and Methods

This prospective, study was carried out in the ophthalmology department of a tertiary hospital from March 2019 to August 2019.

Inclusion Criteria: Patients of cataract associated with PXF.

Exclusion Criteria: Patients with a history of miotic use, traumatic cataract, complicated cataract, PXF glaucoma, high myopia, previous ocular surgery.

This study adhered to the Helsinki Declaration of 1975, as revised in 2013. Institutional ethics committee clearance was obtained before the start of the study (IEC/338/19).

A written and informed consent was obtained from all patients after explaining the procedure and associated risk. Patients were admitted 1 day prior to surgery and detailed history was taken. Best corrected visual acuity (BCVA) was measured using Snellen's visual acuity chart. IOP was measured by non contact tonometer. Gonioscopy was done for angle evaluation using by goldmann 4 mirror gonioscope and graded by Shaffer's classification. Detailed slit lamp examination was done and PXF deposits were looked for on the cornea, iris and pupillary margin and after dilatation, on the anterior capsule of the lens (Figure 2). Cataract was graded using "lens opacity classification system (LOCS II)." Preoperatively Ofloxacin eyedrops (0.3%) were instilled 2 hourly. Flurbiprofen (0.003%) drops were instilled 3 times in morning at half hourly interval. Pupils were dilated with Phenylephrine (5%)+ Tropicamide (0.8%) eye drops and 2% Homatropine eyedrops. Pupillary diameter after dilatation was measured and graded as poor (<5 mm), fair (5-7 mm) and good (>7 mm or more).

All patients were operated by the same surgeon with more than 5 years' experience using a technique of SICS with PCIOL implantation. Peribulbar block with 3 ml of 2% Xylocaine with adrenaline 1:200000 and 1 ml of 0.5% Bupivacaine with 150 units/ml of Hyaluronidase. Eye to be operated was painted with Povidine-iodine (7.5%). For SICS, fornix base conjunctival flap was made, straight scleral incision was made measuring 6mm with back cuts, 1 to 1.5 mm away from limbus superiorly using bard

parker knife with 11 no. blade and sclerocorneal tunnel was constructed with crescent knife. Side port entry was made by 15° side port blade, trypan blue dye (0.06%) was injected intracamerally for 20seconds to stain the anterior capsule. Soft shell technique for viscoelastic use was employed (first dispersive viscoelastic – hydroxypropyl methylcellulose (HPMC)- injected towards undersurface of cornea followed by injection of cohesive viscoelastic-sodium hyaluronate - for flattening the anterior lens capsule). Gentle CCC aimed at 6 mm was done using the needle cystitome. Hydrodissection was performed to separate cortex from nucleus. Nucleus was prolapsed into the anterior chamber and then delivered by visco expression. Irrigation and aspiration was done with Simcoe's two way irrigation and aspiration cannula. Rigid, single piece, biconvex, polymethylmethacrylate (PMMA) PCIOLs with optic diameter of 5.50 mm was used in all except in 1 case where 6mm PMMA lens was implanted using Kelman McPherson forceps. Visco wash was done. Intracameral moxifloxacin (0.5%) injected and side port was hydrated. Subconjunctival injection of 1ml dexamethasone + 1ml gentamicin given and eye padded.

Modifications in the technique like sphinchterotomies (Figure 3), conversion of CCC to can-opener with



Figure 1: Showing fibrillar material on the pupillary margin in PXF syndrome



Figure 2: Distribution of PXF material on anterior lens capsule seen after dilatation of pupil.



Figure 3: Sphincterotomies with in the bag IOL



Figure 4: Primary ACIOL with PI in a case of PXF cataract.



Figure 5: Fibrillar material on IOL in a post-op PXF patient [1,2]

implantation of 6mm IOL, use of Y pushers, automated anterior vitrectomy, implantation of anterior chamber IOL (ACIOL) (Figure 4) or sclera fixated IOL (SFIOL) were made in cases where intra-operative complications were noted.

Post-operatively, patients were put on topical antibiotics (gatifloxacin 0.3%) and steroids (prednisolone 1%) which was tapered over 4-6 weeks depending upon the post-operative inflammation.

Minimum Follow Up: post-operative day 1, day 7 and 1 month. On every follow-up BCVA and IOP was noted and detailed slit lamp and fundus examination was done and findings were noted.

Statistical Analysis

Mean and SD was calculated for quantitative characters, and frequency and percentages were obtained for qualitative characters. Paired t test was applied to check the significant difference in preoperative and postoperative mean IOP.

Results

Total 25 phakic eyes of 25 patients diagnosed as PXF were evaluated during the study period. All patients were >61 years old, out of which 52% patients were in between 61 to 70 years of age. Further, 9 patients were males (36%) and 16 were females (64%) (Table 1).

Out of 25 patients distribution of PXF - 14 patients (56%) had PXF in both eyes, 9 patients (36%) had unilateral PXF and 2 patients (8%) had pseudophakia in one eye due to which laterality could not be commented upon.

Distribution of Morphological types of cataract with PXF - Nuclear cataract (52%) was the most commonly observed. 20% cases were Mature cataract (n=5) and 12% (n=3) had Hyper-mature cataract (Table 2).

Table 3 shows distribution of fibrillar PXF material on ocular structures.

Pre operative maximum pupillary dilatation observed in our study was poor in 12% cases (n=3), fair in 76% cases (n=19) and good in 12% cases (n=3) (Table 4).

The intraoperative difficulties observed were as follows:

The most common Intra-operative difficulty was poor pupillary dilation, which was managed by sphincterotomy and the use of 'y' pushers or during the surgery.

One patient had rhexis extension, which was managed by conversion of CCC to can-opener technique with 6mm PCIOL implantation.

Two patients had iridodialysis due to poorly constructed tunnel with premature entry with inadequate tunnel incision. Iridodialysis was small and present superiorly at 12'o' clock which was covered by eyelid hence they were asymptomatic.

Posterior capsular rupture occurred in 2 patients, for which anterior vitrectomy was performed.

Out of these 2, 1 patient underwent primary ACIOL implantation with peripheral iridectomy (PI), while the other patient had PCR with Zonular dialysis, this patient underwent SFIOL implantation (Table 5).

Comparison of pre-operative and post-operative IOP:

IOP was measured pre-operatively and post-operatively on

day 1, day 7 and 1month (Table 6).

There was no significant difference in the pre and post operative IOP, but post operative (1 month) IOP was less than pre-op IOP (p=0.4).

Comparison of pre-operative and post-operative BCVA-

The mean Pre-operative vision was counting fingers at 3 meters. On post-operative day 1 and day 7, 19 patients had BCVA between 6/12 to 6/6. After 1 month mean post operative vision was 6/9.5 (Table 7).

Systemic associations seen in PXF syndrome patients-

Diabetes, hypertension and ischemic heart disease(IHD) were seen in patients with PXF. One patient was a known case of parkinsonism who was operated under General Anesthesia (Table 8).

Table 1: Age and sex distribution of study patients

	Sex		No. of patients with pseudoexfoliation
	Males	Females	
51-60	0	0	0
61-70	4	9	13
71-80	4	5	9
81 and above	1	2	3

Table 2: Type of cataract

Type of Cataract	No. of eyes with pseudoexfoliation
Hyperature cataract	3
Mature cataract	5
Cortical + nuclear	2
Posterior sub capsular + nuclear	1
Posterior polar + nuclear	1
Nuclear cataract	13

Table 3: Distribution of fibrillar material in ocular structures

Distribution of fibrillar material in ocular structures	Number of eyes	Percentage (%)
Pupillary margin	5	20
Iris	6	24
Lens	6	24
Iris, papillary margin and lens	8	32

Table 4: Maximum pupillary dilatation observed

Pupillary dilatation	No. of eyes with pseudoexfoliation
<5 mm (poor)	3
5 to 7mm (fair)	19
>7 mm (good)	3

Table 5: Intra-operative difficulties

Intra-operative difficulties	No. of eyes	Percentage (%)
Poor pupillary dilatation	9	36
Rhexis extension	1	4
Posterior capsular rupture	3	12
Iridodialysis	2	8
Zonular dialysis	1	4

Table 6: Range of Pre-operative and post-operative intra-ocular pressure (IOP)

IOP (mmHg)	Pre-operative	Post operative		
		Day 1	Day 7	1 month
<10	3	5	3	4
11-15	11	12	12	14
16-20	11	8	10	7

Table 7: Comparison of range of Pre-operative and post operative best corrected visual acuity (BCVA)

BCVA	Pre-operative	Post-operative		
		Day 1	Day 7	1 month
6/6-6/12	0	19	19	20 (80%)
6/18-6/36	8	6	6	5 (20%)
6/60 or less	17	0	0	0

Table 8: Associated systemic illness

Associated systemic illness	No. of patients
Hypertension (HTN)	6
Diabetes Mellitus (DM)	7
HTN+DM	4
Ischemic heart disease (IHD)	3
Parkinsonism	1
No systemic illness	4

Discussion

PXF syndrome is an age related disorder. PXF syndrome uncommonly presents before age 60 years and increases in frequency of onset with age.

In this study, age group of patients diagnosed with PXF was in the range of 61-90 years. Majority of study cases (n=13) were in the age group of 61 – 70 years (52%). Study included 3 cases (12%) above 81 years of age. Sufi et al. in their study of 1117 patients with PXF syndrome ranging in age from 49 years to 89 years, noted that mean age of patients with PXF as 72.61 years.⁹

Of the 25 patients in present study, 9 patients were males (36%) and 16 were females (64%). Pranathi et al. in their study of 52 patients noted 53.8% males and 46.2% females.⁹

In this study 14 patients (56%) had bilateral PXF and 9 patients (36%) had unilateral PXF which is similar to the study done by Hegde et al. in which 6% patients had bilateral PXF and 36% had unilateral PXF¹⁰. Clinically, unilateral involvement is often a precursor to bilateral, progression to bilaterality was reported in upto 50% cases within 5 to 10 years after diagnosis.

In this study 52% cases had nuclear cataract where as hyperature cataract was observed in 3 and mature cataract in 5 eyes. Joshi et al. in his study noticed 25% patients having nuclear cataract, 98% hyperature and 50% mature cataract¹. Pre operative maximum pupillary dilatation observed in this study was poor in 12% cases (n=3), fair in 76% cases (n=19) and good in 12% cases (n=3). Naik et al. in their study noted 13% cases with poor papillary dilatation, 67% with fair and 30% cases had good pre-operative pupillary dilatation.¹¹

Routinely in PXF cases deposits of white material is seen on the anterior lens surface. The classic picture in PXF consists of 3 distinct zones on the anterior surface of the lens: a central disc, a granular peripheral zone and an intermediate clear zone created by the iris rubbing exfoliation material from the lens surface during its normal excursions.

The most common intraoperative difficulty observed in 36% cases in this study was intra operative miosis (n=9). Sphincterotomy was done in 6 eyes and 3 eyes needed 'y' pushers to dilate the pupils during surgery. Srivastava et al. in their study also adopted sphincterotomy as commonest measure to facilitate nucleus management.¹²

Various other studies have shown use of intracameral lidocaine and epinephrine along with viscoelastics to produce a sufficient pupil size. Viscomydriasis was attempted in all patients of this study apart from sphincterotomies done in 6 eyes and y pushers used in 3 eyes. If the pupil still needs further dilatation use of iris hooks, Malyugin rings are found to be effective but were not used in this study.

In SICS, to bring the nucleus in anterior chamber becomes difficult with small pupil. Also small pupil results into small CCC. The CCC should be atleast 5.5 to cover the peripheral part of the IOL as small CCC can lead to capsulophimosis which may lead to spontaneous lens subluxation. Attempted nucleus prolapsed through small pupil results in extension of CCC posteriorly leading to PCR.

In this study 2 patients had PCR out of which 1 underwent anterior vitrectomy with peripheral iridotomy followed by primary ACIOL implantation. In study done by Islam et al. 14 cases out of 3334 patients had posterior capsular rent which was managed similarly.¹³

Other patient having PCR with zonular dialysis underwent secondary SFIOL. In the study done by Sastry et al. 4 patients had zonular dialysis intraoperatively out of which 2 cases required SFIOL.¹⁴ Eight percent cases in present study had iridodialysis (n=2). In study done by Bangal et al. they had observed iridodialysis in 4% cases as a result of intraoperative manipulation of intraocular tissue.¹⁵

This study showed no significant difference in the pre (mean=14.23) and post operative IOP on day 1 (mean=13.45), day 7 (mean=13.47) and 1 month (mean=13.73). Sastry et al. in their study on cataract surgery outcome in patients with non-glaucomatous pseudoexfoliation noted mean POD1 IOP as 26.23+ 11.40.¹⁴

On 1 month follow-up in this study, 20 pts had BCVA between 6/12 to 6/6 and 5 had BCVA between 6/18 to 6/36, which is similar to the findings noted by Pranathi et al. in their study.⁸

Systemic illnesses seen in the patients in present study were diabetes, hypertension, and ischemic heart disease. In the Blue Mountain Eye Study (Australia), PXF correlated positively with a history of hypertension, angina, myocardial infarction or stroke and parkinsonism suggestive of vascular effects of the disease.¹⁶

The association of PXF with systemic disorders is supported by UTAH project on PXF syndrome.¹⁶

Conclusion

PXF syndrome patients when presented for cataract surgery posed certain specific challenges. Thorough pre-operative evaluation for signs of PXF syndrome helped in diagnosis. Intra-operative modifications like soft shell technique and sphincterotomies helped in giving optimum visual outcome in majority of study cases. Manual SICS offers a good solution to cataract patients having PXF syndrome. Immediate post operatively patient needs to be evaluated for increased inflammation and raised IOP while IOL decentration remains a concern in late postoperative period.

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