

Initial Presentation of Pseudoexfoliation, At a Tertiary Care Hospital

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ABSTRACT

OBJECTIVE: To evaluate the clinical presentation management and visual prognosis in patients with pseudoexfoliative glaucoma in eastern Odisha.

MATERIAL AND METHOD: A hospital based prospective study included 1253 patients with pseudoexfoliative material in one or more of the anterior segment structures along with IOP >21 mm of Hg and/or glaucomatous optic disc changes were selected from the ophthalmology OPD of SCB Medical college from April 2015 to March 2018.

RESULT: Incidence of pseudoexfoliation was found to be 1.3% among >40 years of OPD patients, 8.9% of pseudoexfoliation had glaucoma.

CONCLUSION: After medical and surgical management most of the patients showed visual improvement with >6/60 in 70% of cases except those who had poor visual acuity at presentation because of advanced glaucomatous damage

KEY WORDS: Pseudoexfoliation, pseudoexfoliative glaucoma, Glaucomatous optic atrophy.

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I. INTRODUCTION:

Pseudoexfoliative material, the presence of which in the eye is termed as pseudoexfoliative syndrome (PXS), is a common cause of secondary open angle glaucoma worldwide. Pseudoexfoliation should be differentiated from true capsular exfoliation, which occurs due to chronic infrared exposure in glassblowers. Pseudoexfoliation is a greyish white fibrillary amyloid like material; it may derive from abnormal extracellular matrix metabolism in ocular and other tissue. The material deposited on various ocular structures like lens capsule, zonular fibers, iris, trabecular meshwork, and conjunctiva. Also Pseudoexfoliative material has been found in the skin and visceral organs, leading to the concept of Pseudoexfoliation syndrome as the ocular manifestation of systemic disorder. Open angle glaucoma associated with PXF (sometimes called capsular glaucoma) is conventionally due to elevated IOP, likely mechanism includes trabecular obstruction by PXF and liberated iris pigments with secondary degenerative outflow dysfunction. A less common mechanism of glaucoma in PXF includes acute or chronic angle closure glaucoma like zonular weakness causing anterior movement of lens, increased adhesiveness of iris to lens due to exfoliative material, sphincter muscle degeneration and uveitis. Pseudoexfoliative glaucoma prognosis is often worse than POAG; the IOP is often higher and may exhibit marked fluctuation. Severe damage may present at diagnosis or can develop rapidly.

Medical treatment is similar to POAG, but failure is more common. Laser trabeculoplasty in Pseudoexfoliative glaucoma (PXG) is probably more effective than POAG. Phacoemulsification alone may significantly lower the IOP, though it may give better result with combined trabeculectomy. Filtration surgery in PXG has similar success rate to POAG. Early recognition and appropriate management are essential for good outcomes. As Pseudoexfoliation glaucoma is especially challenging to control, patients may require aggressive treatment and frequent, close follow up. Keeping in mind, we studied Pseudoexfoliation glaucoma cases in our department and discussed their presentation, clinical feature, management, and visual prognosis.

II. MATERIALS AND METHODS:

All the patients for the study of Pseudoexfoliative glaucoma were selected among those, presenting to the Regional Institute of Ophthalmology, SCB Medical college, Cuttack in the time period of April 2015 - March 2018.

All patients of age >40 years presenting to the OPD and with Pseudoexfoliative glaucoma were included in the study.

Patients with Pseudoexfoliative material in one or more of anterior segment structures along with IOP>21 mm Hg and/or glaucomatous optic disc changes (vertical cup:disc \geq 0.7:1 or cup asymmetry \geq 0.2:1 between both eyes) were taken as Pseudoexfoliative glaucoma cases.

EXCLUSION CRITERIA:

1. Patients not given consent for the study.
 2. Patients lost to follow up within six months.
 3. Patients with history of trauma.
 4. Previous history of ocular surgery .
 5. Presence of diabetes mellitus, history of corticosteroid use, uveitis or other causes of secondary glaucoma.
- Patients selected for study were evaluated clinically after taking written informed consent and a detailed relevant clinical history was taken under following schedule.

CASE SHEET:

1. Case number.
2. Personal history of patient, name, age, sex, address, socioeconomic status, habitat, occupation,
3. Detailed history of present, past illness and associated disorder.
4. Family history.
4. Treatment history.
5. General and systemic examination.
6. Local examination of eyes.
7. Slit lamp examination for further evaluation of ocular structure.
8. Retinoscopy.
9. Fundoscopy by direct, indirect ophthalmoscopy, 90D examination in slit lamp.
10. Visual acuity for DV, NV, BCVA.

SPECIAL INVESTIGATION:

1. Evaluation for dry eye.
2. CCT central corneal thickness.
3. Gonioscopy by Zeiss four mirror gonioscope.
4. OCT or optical coherence tomography.
5. Visual field testing using Humphrey perimeter.

MANAGEMENT:

MEDICAL MANAGEMENT: Patients with IOP less than 30 mm of Hg at presentation were given monotherapy preferably with prostaglandin analogue +/- beta blocker/carbonic anhydrase inhibitor /alpha agonist. Patients with IOP>30mm of Hg at presentation or who did not show response to monotherapy after 3 weeks , were treated with additional drops or fixed drug combination. Those patients with IOP>40 mm of Hg at presentation were managed with oral acetazolamide tablets /oral glycerol/ i.v.mannitol along with topical drops. Patients who showed good response with IOP<21 mm of Hg continued with same , those with still elevated IOP at 6 weeks of follow up were planned for surgery.

SURGICAL MANAGEMENT:

1. Trabeculectomy/cataract extraction with IOL implantation
 2. Laser peripheral iridotomy.
- Follow up was done in all cases at 3 weeks, 6 weeks, 3 months, 6 months, with BCVA and IOP measurement and fundoscopy.

III. RESULTS:

Total OPD attendance was 168945 out of which 96054 were above 40 yrs of age. 1253 number of patients had Pseudoexfoliation syndrome. 203 patients had Pseudoexfoliative glaucoma. After taking exclusion criteria , 112 cases were considered for this study.

Table No 1: Prevalence and Incidence of PXG

Total OPD cases of age >40yrs	No. of patients with PXS	Percentage(%)	No of patients with PXG	Percentage(%)
96054	1253	1.3	112	8.9

Table 1 shows prevalence is 1.3% and incidence 8.9%. The comparative study shows a wide range of variation in prevalence might be due to racial, genetic, and/ or geographical differences worldwide.

Table No 2: Age distribution of PXG.

Age group (in years)	No of cases n=112	Percentage
41--50	15	13.3
51--60	26	23.3
61--70	35	31.3
>70	36	32.1

In this present study,13.3% patients were within 41—50 years of age . The incidens gradually increased within 51—60 years, 31.3 cases within 61—70 years and 32.1 % cases were with more than 70 years of age. In the present study, 63 patients were male and 49 female with Male : Female ratio 1.29: 1.

Table No 3: Symptoms of PXG:

Symptoms	No of patients(n=112)	Percentage
Diminution of vision	112	100
Pain in eye	16	14.3
Redness	12	10.7
Headache	10	8.9
Lid swelling	4	3.6
Nausea, vomiting	2	1.8

In all patients, the main symptom was defective vision either for near or distance rest of the symptoms like pain redness due to acute elevation of IOP, symptoms like nausea vomiting were mainly associated with angle closure.

Table No 4: Best corrected visual acuity in PXG at presentation and after 3 months

BCVA	At presentation no of eyes(n=140)	Post treatment after 3 months(n==140)
≥6/60	64(45.7%)	98(10%)
<6/60-2/60	46(32.9%)	22(15.7%)
Counting finger at 1 meter	15(10.7%)	7(5%)
Hand movement	7(5%)	5(3.6%)6
PL/PR+	6(4.3%)	6(4.3%)
PL/PR --	2(1.4%)	2(1.4%)

Visual acuity was ≥ 6/60 in 64 (45.7%) eyes,5/60 to 2/60 in 46 (32.9%) eyes. Visual acuity was CF at meter in 15(10.7%) ,HM in 7(5%),PL in 6(4.3%) eyes

Table no 5: IOP at present and after 3 week, six weeks, three months and six month

Duration	3 rd week		6 th week		3 rd month		6 th month	
	≤21	>21	≤21	>21	≤21	>21	≥21	>21
IOPin mm of Hg								
No of eyes	93	47	103	37	135	5	135	5

IOP was between 22-25 mm of Hg in 46(32.9%) eyes, 26-30 mm of Hg in 53(37.9%),31-35mm of Hg in 24(17.1%),36-40mm of Hg in 8(5.7%),41-45mm of Hg in 6(4.3%) and 46-50 in 3(2.1%)eyes. Mean IOP was 28.73 ± 27.04 mm of Hg.

Table No 6: Visual field changes at presentation.

Grades of visual field defect	No of eyes n=64	percentage
Normal	23	35.93
Mild	26	40.62
Moderate	15	23.43
Severe	—	-

Visual field examination could be done in 64 eyes with presenting visual acuity 6/60 or better. Visual field was normal in 23 eyes(35.93%). Rest of the patients had visual field defects.40.62% had mild and 23.43% had moderate. Severe field defect was absent because all of them had visual acuity less than 6/60

Table 7: Signs of pseudoexfoliative glaucoma.

SIGNS		NO OF EYES (N=140)	PERCENTAGE
Corcumcornealcongessin		29	20.7
Corneal oedema		17	12.1
PXF material lining corneal endothelium		23	16.4
Anterior	Shallow	16	11.4

chamber depth	Normal	124	88.6
Iris	Peripupillary Iris atrophy	56	40
	Posterior synechia	2	1.4
Pupil	Size	Normal	133
		Dilated	7
	Mydriasis	Good	53
		Poor	87
	PXF Material around Pupil	102	72.9
Lens	Pseudoexfoliation of lens	Peripheral Ring	27
		3-Ring Sign	98
	Cataract	122	87.1
	Subluxation	2	1.4

Table no 8: Fundoscopy finding in PXG

Findings	Total no of eyes n=113	Percentage
Media	Clear	18
	Hazy	95
C:D ratio	<0.7:1	12
	0.7:1 - 0.8:1	78
	>0.8:1	23

Table no 9: Mean RNFL thickness at presentation.

Grades	No of eyes (n =78)	Percentage
Within normal limit	37	47.4
Border line	28	35.9
Outside normal limit	13	16.7

In 78 eyes, mean RNFL thickness could be measured and in rest 62 eyes it was not possible due to hazy media.

Table no 10: Efficacy of medical management at 6 weeks of follow up.

Response	No of eyes (n=140)	Percentage
Good	103	73.6
Poor	37	26.4

Table no 11: Surgical intervention in PXG.

Surgery	No of eyes (n= 106)	Percentage
Trabeculectomy	2	1.9
Lens extraction	67	63.2
Trabeculectomy and lens extraction	35	33
Laser PI	2	1.9

IV. DISCUSSION:

The present study included presentation, clinical features, management and prognosis of Pseudoexfoliation glaucoma. Prevalence of PXS was found to be 1.3%^{1,2} among >40 years of patients. Incidence was 8.9%. Most of the patients were above the age of 70 years. According to Irfan Shafiqet al³ highest prevalence was in age group >70 years. Mean age of patients in present study was 62.5± 18.84 years^{4,5}. The more number of male cases might be due to our male dominated society and negligence towards female health care^{3,4,5}. In present study 84 (75%) patients had unilateral and 28 (25%) bilateral involvement. In 124 eyes (88.6%) angle was open and 16 (11.4%) eyes were occludable angle. If the pigmented trabecular meshwork was not visible in at least 180- 270 degree of the angle circumference without indentation or manipulation, then the angle was called occludable. Also patients with shallow anterior chamber (PACD < 1/4th corneal thickness by Van Herick method) were taken into this category. The main symptoms in all patients were defective vision either for near or for distance. Rest of the symptoms like pain in eye associated with acute elevation of IOP. In present study most of the cases presented with diminution of vision only (100%) which suggests the silent nature of the disease which might be a cause for late presentation of PXG patients.

Normal AC was present in 124(88.6%) eyes and shallow AC in 11.4% eyes⁶. Peripupillary iris atrophy was found in 40% eyes. Good mydriasis found in 37.9% eyes, poor in 62.1%, might be due to atrophy and degeneration of the iris muscle cells. PXF material was deposited around pupil in 72.9 eyes on corneal endothelium 16.4% cases, over anterior capsule of lens and pupillary border in 89.3% and 72.9% eyes respectively. In 25.7% eyes PXF material deposits observed in angle of anterior chamber by gonioscopy. Ravi Thomas (2002) found exfoliative material present mainly on anterior lens capsule⁷. In contrary to this Alan P Rotchford et al (2003) found PXF deposits mainly at pupillary margin. In present study PXF deposits found mainly over lens capsule. So it is ideal to dilate the pupil to avoid missing of cases with PXS.

Mean central corneal thickness (CCT) in present study was $520.36 \pm 27.04 \mu\text{m}$. In Tomaszewski BT et al 2014⁸ study, mean CCT in eyes with PXG was thinner ($508.2 \pm 32.6 \mu\text{m}$) than eyes with PXS syndrome without glaucoma ($529.7 \mu\text{m} \pm 30.3 \mu\text{m}$) and control group ($527.7 \mu\text{m} \pm \mu\text{m}$). Fundus view was not possible in 27 eyes having very hazy media due to lenticular opacity and / or corneal oedema. C:D ratio was between 0.7:1 to 0.8:1 in 78 (69%) of eyes. Gonioscopy was not possible in 15 eyes due to corneal oedema. PAS was present in 1.65 eyes. DARK proposed that angle closure in PXS might result from the development of iridocapsular adhesion and subsequent pupillary block. Other possible mechanism includes anterior lens movement resulting from weak zonules.

Visual field examination could be done in 64 eyes with VA 6/60 or better. Visual field was normal in 23 eyes (35.93%) Rest of the patients had visual field 40.62% mild, 23.43% moderate visual field defects. Severe field defect was absent because all of them had visual acuity $< 6/60$. B.E. Stephan (1999), et al⁵ study shows 11.3% cases had normal visual field. Mean RNFL thickness was within normal limits in 47.4% eyes, borderline in 35.9% and outside normal limit in 16.7% cases.

All PXG cases initially managed medically. Laser PI was done in 2 eyes (1.9%) with pupillary block and then managed with topical steroids for 2 weeks and then continued topical antiglaucoma medication.

Patients who showed good response with IOP ≤ 21 mm of Hg by topical drops continued the same and those with still elevated IOP at 6 weeks of follow up were planned for surgical management. Medical management showed good response in 103 eyes (73.6%) but poor response in 37 eyes (26.4%) at 6 weeks. Those cases are planned for trabeculectomy alone or combined trabeculectomy with cataract extraction and IOL implantation.

Historically PXG cases are difficult to manage either medically or surgically, as most of the patients were with cataract, combined trabeculectomy and cataract surgery is the preferred procedure. According to W.E Gillies (1973) and Goder J (1988) lens extraction plays an important role in reduction of IOP. Combined procedure is also the procedure of choice in patients with uncontrolled IOP. A prospective study by Konstas et al 1997⁹ shows a higher success rate after trabeculectomy among exfoliative glaucoma eyes (mean untreated postoperative IOP of 11.8 ± 4.4 mm Hg) compared with the response in those with primary open angle glaucoma (mean untreated postoperative IOP of 15.0 ± 4.6 mm Hg) at 6 months follow up.

Major surgical complication encountered, out of 106 eyes in this study was damage to sphincter pupillae in 23 eyes (21.7%) due to poor mydriasis. Posterior capsular rupture and zonular dialysis found in 12 cases (11.35%) and 11 eyes (10.4%) respectively. Vitreous prolapse was there in 7 eyes (6.6%) and iridodialysis in 2 eyes (1.9%).

The major complications during postoperative follow up includes corneal haze in 38 (35.9%) eyes, subluxation of IOL 8 eyes (7.5%), uveitis in 5 (4.7%), shallow AC in 6 eyes (5.7%), hyphaema in 4 (3.7%), Hypotony in 3 (2.8%), CME in 4 (3.7%) eyes. Similar complications were reported by Scoroli et al 1998¹⁰ who stated that postoperative complication in PXG cases were five times that without PXS.

During follow up at 6 months most of the cases showed lowering of IOP below 21 mm of Hg with appropriate medical and surgical management, except 5 cases. Those patients with still elevated IOP at 6 months, were planned for glaucoma valve surgery.

After medical and surgical management, most of the patients showed visual improvement with more than 6/60 in 70% cases (at presentation it was 45.7%). Most of the patients showed improvement except those who had poor visual acuity at presentation, because advanced glaucomatous damage had already occurred in them,

V. CONCLUSION:

Blindness is a major problem in India for which glaucoma is an important contributing factor. Damage caused by glaucoma is irreversible, but it can be prevented. So early diagnosis and proper timely intervention are the key factors to halt the disease progression.

As PXG has mostly incipient course, by meticulous examination by slit lamp before and after and pupillary dilation, large number of cases can be diagnosed early. PXG shows rapid progression and poor response to treatment. Appropriate medical and surgical treatment can control disease progression.

PXS has increased prevalence of vascular disorder, hearing loss and Alzheimer disease, so patients with PXS should be evaluated for those systemic associations.

ABBREVIATIONS:

AC - anterior chamber
CCT – central corneal thickness
CME - cystoid macular edema
HM – hand movement
IOP – intra ocular pressure
OCT – ocular coherence test
PGA- prostaglandin analogue
PI – peripheral iridotomy
PXG – pseudo exfoliation glaucoma
PXS - pseudo exfoliation syndrome

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