EVALUATION OF OCULAR PROFILE OF PATIENTS WITH PSEUDOEXFOLIATION SYNDROME IN A TERTIARY EYE CARE CENTER IN WEST BENGAL

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ABSTRACT

BACKGROUND

Pseudoexfoliation Syndrome (PEX) is characterized by the deposition of a distinctive fibrillar material on the lens capsule, pupillary margin, iris, ciliary body and subconjunctival tissue and has also been identified in other parts of the body. PEX occurs worldwide and prevalence rates vary from 10 to 20% of the general population over the age of 60 years. Heightened awareness of this condition and its associated clinical signs are important in the detection and management of glaucoma, and preoperative determination of those patients at increased risk for surgical complications. The aim of the study was to evaluate the ocular profile of patients with Pseudoexfoliation Syndrome in a tertiary eye care centre in West Bengal and to assess surgical complications which may arise from Pseudoexfoliation Syndrome.

MATERIALS AND METHODS

This is institution based cross sectional study, conducted at the Regional Institute of Ophthalmology (RIO) OPD over a period of 1.5 years starting from February 2014 to July 2015. Fifty patients with the age between 20-80 years, attending RIO, OPD and diagnosed as having Pseudoexfoliation Syndrome were included in our study. Patients with other causes of secondary glaucoma and Fuchs Heterochromic Uveitis were excluded from our study

RESULTS

In our study we found results similar to other studies with respect to age distribution of pseudo exfoliation patients. Higher incidence was found in age group of patients more than 55 years. The PEX patients in our study had10.0%, 4.0%, 3.0% and 2.0% of pseudoexfoliation glaucoma (PXG), primary open angle glaucoma (POAG), primary angle closure glaucoma(PACG) and normal tension glaucoma (NTG) respectively. 81% eyes were non-glaucomatous. The mean IOP (mean ± s.d.) of the patients was 17.48 ± 2.58 mmHg with range 14 - 23 mmHg and the median was 17 mmHg. Surgical complication was observed in 22.22% of the cases, but this was not further analysed since very few patients (9 patients) underwent surgical procedures.

CONCLUSION

Pseudoexfoliation syndrome presents challenges that need careful preoperative planning and intraoperative care to ensure safe surgery and a successful postoperative outcome. It is associated with higher IOP, incidence of cataract, and poor vision. Further population based longitudinal studies are warranted to assess the prevalence of PEX and associated risk factors.

KEYWORDS

Pseudoexfoliation Syndrome, Pseudoexfoliation Glaucoma, Subluxation, Lens Capsule, Successful Postoperative Outcome.

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BACKGROUND

Pseudoexfoliation (PEX) syndrome is an age-related generalized fibrotic matrix process, which, is a risk factor for glaucoma (most often open-angle). The primary cause of chronic pressure elevation is local production of PEX material by trabecular meshwork cells.[1] It targets ocular tissues through the gradual deposition of fibrillary residue from the lens and iris pigment epithelium, mainly on the lens capsule, ciliary body, zonules, corneal endothelium and iris.[2] Two pathological manifestations of PEX, zonular weakness and poor pupillary dilatation, have been identified as the most significant risk factors for surgical complications. Intraoperatively, eyes with PEX are at greater risk for zonular dialysis, posterior capsule tear/rent, vitreous loss, and dropped nucleus or fragment; postoperatively, they have a higher incidence of inflammation in the form of increased aqueous flare and cell response, fibrin reaction, posterior synechiae, posterior capsule opacification, anterior capsule phimosis, and late intraocular lens (IOL) decentration and dislocation. Extraocular tissues involved include lung, skin, liver, heart, kidney, gallbladder, blood vessels, extraocular muscles, connective tissue in the orbit, and meninges. PEX

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is associated with transient ischaemic attacks, stroke, systemic hypertension and myocardial infarction.[3]

PEX occurs worldwide and prevalence rates vary from 10 to 20% of the general population over the age of 60 years.[4] Hospital-based studies from India have reported a prevalence rate between 1.8.[5] and 7.4%.[6] in adults over 45 years of female preponderance has been previously reported.[7] Pseudoexfoliation glaucoma (PXG) accounts for approximately 25% of all open angle glaucomas worldwide.[8] The prevalence of PXG as reported by population-based surveys from South India vary between 7.5 and 13%.[9,10] Unilateral PEX occurs in 48-76%.[11] of patients and converts to bilateral disease in up to 50% of patients within 5 to 10 years.[12]

Compared to POAG, PXG has a more aggressive clinical course and a worse prognosis. It is typically associated with higher mean IOP levels, greater diurnal pressure fluctuations, marked pressure spikes, higher frequency and severity of optic nerve damage, more rapid visual field loss, poorer response to medications, and a greater necessity for surgical intervention. In contrast to patients with POAG, patients with PXG are not abnormal steroid responders.

Heightened awareness of this condition and its associated clinical signs are important in the detection and management of glaucoma, and preoperative determination of those patients at increased risk for surgical complications. The chamber angle is often narrow, presumably as a result of anterior movement of the lens- iris diaphragm related to zonular weakness.

The pupil often dilates poorly. Phacodonesis and iridodonesis may occur due to zonular weakness, which may predispose to zonular dehiscence, vitreous loss and lens dislocation, during and after cataract surgery.

PEX is rarely seen in persons younger than 50 years and occurs most commonly in individuals older than 70 years. The prevailing presence of elastic fiber epitopes, mainly elastic microfibrillar components (elastin, vitronectin, amyloid P, fibrillin-1, LTBP-1), has led to the current theory explaining PEX as a type of elastosis, affecting especially elastic microfibrils.[13] The lOP is often higher with greater diurnal fluctuations than in POAG and the overall prognosis is worse.[14] In a series by Kozart and Yanoff, pseudoexfoliation syndrome was 3 times more common in women than in men.[15] Loss of zonular support for the lens or posterior chamber intraocular lenses (IOLs), must be factored into the pre- and postoperative management of persons undergoing cataract surgery.[16] Early diffuse corneal endothelial decompensation explained by a damaged and numerically reduced endothelium.[17] PEX endotheliopathy leads to an increased incidence of bullous keratopathy.[18] Endothelial cell density is significantly decreased, and pleomorphism and polymegathism of cells are increased in PEX eyes, particularly when intraocular pressure is high.[19] Also, the corneal endothelium in eyes with PXF is vulnerable to cataract surgery, careful surgical procedures are necessary.[20]

Some authors have found that pseudoexfoliation is linked with Alzheimer disease, senile dementia, cerebral atrophy, chronic cerebral ischemia, stroke, transient ischemic attacks, heart disease, and hearing loss.[21]

A high incidence of narrow or occludable angles in eyes with pseudoexfoliation has been reported. The main risk factor for glaucoma seems to be the degree of chamber-angle pigmentation.[22] Elevated IOP leads to development of glaucoma in about 50% of patients. PEX subjects may be at greater risk of RNFL thinning and may be evaluated using OCT.[23]

Cataracts have been reported to be more common in PXE patients.[24,25] and PXE has also been shown to be associated with zonulysis during cataract surgery as well as postoperative lens dislocation.[26]

Aims and Objectives

The aims of the study were to evaluate the ocular profile of patients with Pseudoexfoliation Syndrome in a tertiary eye care centre in West Bengal and to assess surgical complications which may arise from Pseudoexfoliation Syndrome.

MATERIALS AND METHODS

This was an institution based cross sectional study, conducted at the Regional Institute of Ophthalmology (RIO) outpatient department, over a period of 1.5 years starting from February 2014 to July 2015. Fifty patients with the age between 20-80 years, attending RIO, outpatient department and diagnosed as having Pseudoexfoliation Syndrome were included in our study. Patients with other causes of secondary glaucoma and Fuchs Heterochromic Uveitis were excluded from our study.

We defined PXG as IOP >21 mmHg in PEX eyes with optic disc changes (CDR ≥ 0.7). Glaucomatous optic neuropathy was diagnosed with two or more of the following features: vertical CDR ≥ 0.7, asymmetry in CDR > 0.2 between the two eyes, characteristic glaucomatous excavation of the neuroretinal rim and typical wedge nerve fiber layer defects.

All patients underwent a comprehensive ophthalmological assessment including visual acuity assessment, Goldmann applanation tonometry, slit-lamp examination before and after dilation of pupils, and dilated fundus examination. The diurnal variation of tension (DVT) was performed. The mean IOP of a particular eye was calculated from two recordings of IOP during the DVT. A difference in mean IOP between the PEX eye and the fellow non-PEX eye of >4 mmHg was considered clinically significant.

Statistical Analysis was performed with help of Epi Info (TM) 3.5.3 which is a trademark of the Centers for Disease Control and Prevention (CDC). Using this software, basic cross-tabulation and frequency distributions were prepared. x^2 test was used to test the association between different study variables under study. Corrected x^2 test was used in case of any one of cell frequency was found less than 5 in the bi variate frequency distribution. Test of proportion (Z-test) was used to test the significant difference between two proportions. t-test was used to test the significant difference

between means. p≤0.05 was considered statistically significant.

RESULTS AND ANALYSIS

Age (in Years)	Number	Percentage
45-54	08	16
55-64	18	36
65-74	17	34
≥75	07	14
Total	**50**	**100**
Table 1. Age Distribution		

The mean age (mean ± s.d.) of the patients was 64.28 ± 9.31 years with range 45 – 80 years and the median age was 64.0 years. Most of the patients (84.0%) were with age >54 years which was significantly higher (Z=5.65; p=0.0001). Only 16.0% of the patients were with age ≤54 years.

Sex	Number	Percentage
Male	31	62
Female	19	38
Total	**50**	**100**
Male: Female	1.6:1	
Table 2. Gender Distribution		

Proportion of males (62.0%) was significantly higher than that of females (38.0%) (Z=3.39; p=0.0007). Ratio of male and female was 1.6:1.

Laterality of Disease	Number	Percentage
Both Eyes	27	54.0
Left Eye	8	16.0
Left Eye	8	16.0
Right Eye	15	30
Total	**50**	**100**
Table 3. Laterality of Disease		

Most of the patients (54.0%) had disease in both eyes followed by right eye (30.0%) which was significant (Z=3.43; p=0.0006). 16.0% of the patients had disease in left eye.

Gonioscopy	Number	Percentage
Open	92	92
Occludable	8	8
Total	**100**	**100**
Table 4. Gonioscopy		

Gonioscopy of 92.0% of the eyes showed open angle and rest 8.0% had occludable angle. (Z=11.87; p=0.00001). The mean IOP (mean ± s.d.) of the patients was 17.48 ± 2.58 mmHg with range 14 - 23 mmHg and the median was 17 mmHg.

Cup-Disc Ratio	Number	Percentage
0.2	6	6
0.3	20	20
0.4	27	27
0.5	20	20
0.6	6	6
≥0.7	9	9
Hazy media	12	12
Total	**100**	**100**
Table 5. Cup-Disc Ratio		

Cup-Disc ratio of 27% of the patients was 0.4 followed by 0.3 and 0.5 (20%) but it was not statistically significant (Z=1.31; p=0.19). 12% of the patients had hazy media.

Eye	Number	Percentage
Normal	81	81
NTG	2	2
PACG	3	3
POAG	4	4
PXE	10	10
Total	**100**	**100**
Table 6. Status of the Eyes (n=100)		

Out of the 100 eyes of 50 patients 81% of the eyes were normal which was significantly higher (Z=8.76; p=0.0001). 10.0%, 4.0%, 3.0% and 2.0% of the eyes had PXG, POAG, PACG and NTG respectively.

Surgical Treatment	Number	Percentage
Yes	9	18
No	41	82
Total	**50**	**100**
Table 7. Distribution of Surgical Treatment		

Only 18.0% of the patients underwent surgical treatment. No surgical intervention was required for rest 82.0% of the cases.

Surgical Complication	Number	Percentage
Yes	2	22.22
No	7	77.78
Total	**9**	**100**
Table 8. Distribution of Surgical complication		

Overall surgical complication was observed in 22.22% of the cases.

DISCUSSION

PEX has a greater prevalence in the older population.[4] Hence, data on the clinical profile of PEX is important due to the increasing age of general population in many parts of the world. In the current study, most of our patients with PEX were between 55 years and 74 years, which is comparable to the previously published reports.[9] The incidence of PEX tends to increase with age, and is less common below the age of 60 years.[27] In accordance with all previous studies, most of the patients in our study (84.0%) were >54 years which was significantly higher (Z=5.65; p=0.0001). Only 16.0% of the patients were ≤54 years.

There are conflicting reports of gender differences in PEX.[28] We found a male to female ratio of 1.6:1.

Chi-square test showed that there was no significant association between age and gender (p=0.44). Thus the

disease was evenly distributed over age among both males and females.

Patients with PXG have higher IOP with greater fluctuations and marked spikes that likely cause more severe optic neuropathy compared to patients with POAG. PXG develops in approximately 50% of patients with PEX syndrome over time and is recognized as the most common type of secondary open angle glaucoma. The PEX patients in our study had 10.0%, 4.0%, 3.0% and 2.0% of PXG, POAG, PACG and NTG respectively.

We found 81% eyes were non-glaucomatous. This is comparable to a previous study from South India, in which 92.5% of the study population was non-glaucomatous.[10] In our study, the mean IOP (mean ± s.d.) of the patients was 17.48 ± 2.58 mmHg with range 14- 23 mmHg and the median was 17 mmHg. Of the eyes in which optic disc assessment was possible, features of glaucoma (raised IOP and CDR ≥ 0.7) were seen in 18% eyes.

92.0% of the eyes showed open angle and rest 8.0% had occludable angles (Z=11.87; p=0.00001)and none of the patients presented with closed angles coroborating the higher incidences of occludable angles.[2]

Unilateral PEX occurs in 48-76% of patients and converts to bilateral disease in up to 50% of patients within 5 to 10 years.[12]

In our study, Most of the patients (54.0%) had disease in both eyes followed by right eye (30.0%) which was significant (Z=3.43; p=0.0006). 16.0% of the patients had disease in left eye.

Surgical complication was observed in 22.22% of the cases, but this was not further analysed since very few patients (9 patients) underwent surgical procedures.

The major drawback of our study was that we were unable to document reliable visual fields in many of our patients with visually significant cataract. Hence, visual field could not be used as a reliable indicator of disc damage caused by raised IOP. Therefore, these data were ignored in the analysis.

CONCLUSION

"Pseudoexfoliation syndrome is associated with higher IOP, incidence of cataract, and poor vision. Further population based longitudinal studies are warranted to assess the prevalence of PEX and associated risk factors. In our study we found results similar to other studies with respect to age distribution of pseudo exfoliation patients. Higher incidence was found in age group of patients more than 55 years. So, we can draw a conclusion that one have to be more careful while diagnosing a case of PEX, particularly when we are going to treat the patient surgically for lental changes and for the management of glaucoma in such patient complicated with the PEX. Proper preoperative assessment and peroperative surgical care can only bring out a safe and successful outcome.

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