

A CLINICAL STUDY OF PSEUDOEXFOLIATION IN A TERTIARY EYE CARE CENTRE

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ABSTRACT

BACKGROUND

Pseudoexfoliation is characterised by the deposition of homogeneous grey white material, which is produced by the abnormal basement membrane of epithelial cells of trabecular meshwork, lens capsule or iris gets deposited in various ocular structures. The material is composed of protein core surrounded by glycosaminoglycans. Glaucoma is classically of the open angle and however angle closure can occur due to pupil block. Pupil block in pseudoexfoliation patients occur either due to zonular instability and subsequent anterior displacement of lens-iris diaphragm or due to posterior synechiae associated with a rigid non-dilating pupil.

It is a prospective study to find out the associated features in patients with pseudoexfoliation.

MATERIALS AND METHODS

The study was conducted in Medical College Hospital over a period of one year from October 2016 to September 2017; 68 patients with pseudoexfoliation were examined.

RESULTS

In our study, we found the incidence of pseudoexfoliation was higher in patients between 51 - 70 years of age. It was unilateral in 59% of cases. Intraocular pressure was raised in 22% of cases. Only 14% of cases had closed angle. Glaucomatous optic neuropathy was seen in 19% of cases.

CONCLUSION

The prevalence of pseudoexfoliation is higher in elderly age group. The incidence of glaucoma is higher than in normal population. The other associated features like trabecular pigmentation and nuclear cataract are also more common in patients who had pseudoexfoliation.

KEYWORDS

Pseudoexfoliation, Glaucoma, Bull's Eye Configuration, Trabecular Hyperpigmentation.

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BACKGROUND

Pseudoexfoliation (PEX) material gets deposited as whitish grey protein on the lens, iris, ciliary epithelium, corneal endothelium and trabecular meshwork.¹ Pseudoexfoliation glaucoma is the most common form of open angle glaucoma worldwide with an identifiable aetiology.² Since pseudoexfoliation material can be present throughout the body, it is also called pseudoexfoliation syndrome. Recent studies interpret PXE as an inherited microfibrilopathy. LOXL1 gene on chromosome 15q24 has been found to be associated with the disease. Genetic studies suggest that inheritance is by maternal transmission (mitochondrial X-

linked inheritance). Additional factors such as oxidative stress, ischaemia, low-grade inflammation and impaired cellular protective mechanism play a role. This leads to production and accumulation of fibrillar extracellular matrix in various tissues. This is composed of protein core surrounded by glycosaminoglycans. Pseudoexfoliation is classically diagnosed by slit-lamp visualisation of white powdery deposit in a bull's eye configuration on the lens capsule. This gets deposited on the anterior lens capsule as a central zone of visible material measuring 1 - 3 mm in diameter combined with middle clear zone and a peripheral cloudy ring. The middle clear zone is created by the posterior surface of iris rubbing off the pseudoexfoliation material from the lens. This results in a loss of iris pigment, which leads to transillumination defects on the iris.³ The pupil dilates poorly due to dilator muscle atrophy and can complicate cataract surgery. Such eyes also have weak zonular attachment, which may result in subluxation of lens. Pseudoexfoliation glaucoma commonly presents unilaterally, but may affect the other in more than 40% of patients.⁴ The higher intraocular pressure observed in pseudoexfoliation,

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glaucoma can lead to more rapid optic nerve damage and visual field loss.

The prevalence rate of pseudoexfoliation varies from 10% - 20% in the general population over the age of 60 years.^{5,6} but 2.3% in Blue Mountains Eye Study in Australia.⁷ The prevalence of PEX as reported by population-based survey from South India varies between 7.5 - 13.5%.⁸ The percentage of exfoliation syndrome presenting with glaucoma is different for every population. Overall studies indicate about 40% of exfoliation syndrome patients develop glaucoma.⁹ Pseudoexfoliation glaucoma accounts for about 25% of all open angle glaucomas. It has a more aggressive clinical course and worse prognosis than primary open angle glaucoma. It is typically associated with higher mean IOP levels, greater diurnal pressure variations, higher frequency and more severe optic nerve damage. They also undergo more rapid visual field loss, poorer response to medications and a greater necessity for surgical intervention. The chamber angle is often narrow presumably due to anterior movement of lens-iris diaphragm, which is related to zonular weakness. Increased awareness of this condition and its associated clinical signs are important in the detection and management of glaucoma and preoperative determination of these patients at increased risk of surgical complication. The narrow anterior chamber angle due to anterior movement of lens-iris diaphragm resulting from zonular weakness also adds to the surgical complication. The pupil often dilates poorly. Phacodonesis and Iridodonesis may occur due to zonular weakness, which leads to vitreous loss and lens dislocation during cataract surgery.

MATERIALS AND METHODS

The study was conducted in a Medical College Hospital over a period of 1 year from October 2016 to September 2017. Sixty eight patients with pseudoexfoliation were examined. All cases were examined before and after dilatation of pupil with the slit-lamp biomicroscope. All patients were dilated with 1% tropicamide and 5% phenylephrine eye drops. Anterior lens capsule, pupillary margin and angle structures were examined for evidence of pseudoexfoliation material. The established stage of pseudoexfoliation (PEX) was defined as the presence of white, flaky dandruff-like material at the pupillary margin and/or on the anterior lens capsule.¹⁰ Early pseudoexfoliation was defined as small specks of radial, brown pigmented lines on the anterior lens capsule. Fundus examination was done with direct ophthalmoscope and with +90D lens to note optic disc changes. Intraocular pressure was determined by Goldmann’s applanation tonometer and Gonioscopy was done in all cases. Gonioscopic grading of the angle was performed based on the structures visualised. An occludable angle was defined as non-visibility of the pigmented posterior trabecular meshwork >75% of angle circumference in the primary position without manipulation.¹¹

Pupillary dilatation was assessed in all patients as poor pupillary dilatation has been well documented in PEX.^{12,13} Pupillary dilatation was assessed using a slit-lamp in a dark room. The illumination system of a slit lamp was placed

horizontally and a slit beam was adjusted according to pupillary dilatation and the measurement was made. A pupillary dilatation equal or greater than 7 mm was considered good. Visual fields were done with Humphrey’s automated perimeter whenever it was possible. All cases were investigated to find out the incidence of glaucoma and its type.

Inclusion Criteria

All patients aged above 40 years who have presented with pseudoexfoliation were taken up for the study.

Exclusion Criteria

1. Patients having history of any previous ocular surgery.
2. Patients having glaucoma without pseudoexfoliation.

RESULTS

	Sex	No. of Cases	Percentage
1.	Male	51	75
2.	Female	17	25

Table 1. Sex Incidence

The ratio of occurrence in males-to-females is 3: 1. The gender differences varies in different studies.¹⁴

Age Group	No. of Cases	Percentage
41-50	1	1.5
51-60	28	41.2
61-70	29	42.6
71-80	8	11.8
>80	2	2.9

Table 2. Age Incidence

The pseudoexfoliation was higher in the age group of 51 - 70 amounting to almost 84%.

The mean pupillary diameter was 5.6 mm in eyes with established pseudoexfoliation. In eyes with early pseudoexfoliation, the mean pupillary diameter was 7.15 mm. The difference in pupillary dilatation between patients established in early PEX was characteristically significant.

IOP in mmHg	No. of Cases	Percentage
Below 20	53	78
21-24	07	10
>24	08	12

Table 3. IOP in Pseudoexfoliation

Overall incidence of increased intraocular pressure in our series was found to be 22%, whereas the incidence in random population in same age group was 3%.

Width of Angle	No. of Cases	Percentage
Occludable angle	09	13
Open angle	59	87

Table 4. Width of Angle in Pseudoexfoliation

Only 13% of cases had closed angle. Rest had open angle.

Cup-Disc Ratio	No. of Cases	Percentage
0.3	16	24
0.4	17	25
0.5	15	22
0.6	7	10
0.7 or more	13	19
Total	68	100

Table 5. Cup-Disc Ratio

The cup-disc ratio was higher in 19% of cases; 63% (43 out of 68) of patients had trabecular pigmentation.

Type of Cataract	No. of Cases	Percentage
Nuclear	11	65
Cortical	6	35

Table 6. Association with Type of Cataract

Incidence of nuclear or brown cataract was higher than cortical cataract.

DISCUSSION

In 1917 Lindberg described cases of chronic glaucoma, in which flakes of whitish material adhered to the pupillary border of iris.¹⁵ This was termed as pseudoexfoliation. Pseudoexfoliation syndrome is a common ocular manifestation of a systemic disease known to cause disease primarily in the eye. Defects in elastin metabolism has been postulated to result in synthesis of pseudoexfoliation material. PEX material is produced by abnormal basement membrane of ageing epithelial cells in the trabeculum, equatorial lens capsule, iris and the ciliary body. This material has also been described in conjunctiva, iris, orbit, orbital vessels, retinal vessels, skin and connective tissues of heart, lung, liver, gall bladder, kidney and cerebral meninges. Hence, it is termed as pseudoexfoliation syndrome. Deposits of white material on the anterior lens surface are the most consistent and important diagnostic feature of pseudoexfoliation. The classic pattern consists of three zones; central disc equal to the diameter of the pupil; a granular often layered peripheral zone and a clear zone separating the two. Phacodonesis may be noted because of damage to the ciliary zonules. The incidence of cataract formation is increased in patients with PXE. Next to the lens, exfoliation material is most prominent at the pupillary border. Pigment loss from the iris sphincter region and its deposition on the anterior chamber structure is hallmark of PXE. The pseudoexfoliative material is found abundantly on the ciliary processes and zonules. This is believed to be the cause for zonular fragility. The zonular weakness leads to phacodonesis, the evaluation of which is particularly important before cataract surgery. Flakes of pseudoexfoliative material are often visible in the anterior chamber angle, most commonly inferiorly. Pigmentation in the angle is also increased, but irregular. Trabecular hyperpigmentation is usually most marked inferiorly. This pigment lies on the surface of the trabeculum and has a patchy distribution. A scalloped band of pigment running on to or anteriorly to Schwalbe’s line, which is termed as Sampolesi’s line is also frequently seen. In patients with PXE,

a distinct type of corneal endotheliopathy may occur which can lead to an early corneal endothelial decompensation.¹⁶ PEX endotheliopathy leads to an increased incidence of bullous keratopathy. Endothelial cell density is significantly decreased. Pleomorphism and Polymegathism occurring in PEX eyes also results from increased intraocular pressure. Corneal endothelial cells are also vulnerable to damage during cataract surgery. True exfoliation of lens capsule or capsular delamination can occur due to trauma or exposure to intense heat as in glass blowers.

Pseudoexfoliation is a major risk factor for development of open angle glaucoma and in some cases of angle closure glaucoma.¹⁷ Unilateral pseudoexfoliation occurs in 48% - 76% of patients and gets converted to bilateral disease in about 50% of patients within 5 - 10 years.¹⁸ In our study, PEX was unilateral in 60% of cases. Most of our patients with PEX were between 51 - 70 years of age, which is comparable to previous published reports. Gender differences varies in different studies. In our study, male-to-female ratio was 3:1. Intraocular pressure was raised in 22% of cases. This was 6-10 times higher than in normal population. Angle of the anterior chamber was occludable in 13% of cases. This agrees with other reports. Elevated IOP in open angle glaucoma in pseudoexfoliation is due to trabecular cell dysfunction, blockage of the meshwork by exogenous and endogenous exfoliation material by liberated iris pigments. Angle closure glaucoma in pseudoexfoliation syndrome can be due to posterior synechiae formation, zonular weakness and associated forward lens movement, rigidity of iris, pupillary block and a smaller pupil.

In our study, we found a significant difference in pupillary dilatation with early and established PEX. 90% of eyes with early PEX had a pupillary dilatation of 7 mm or more, whereas 95% of eyes with established PEX had pupillary dilatation lesser than 6 mm. This observation indicates not all cases of pseudoexfoliation have poor pupillary dilatation.

PEX is associated with greater prevalence of trabecular pigmentation and nuclear cataract. In our study, nuclear cataract was 65% which is in accordance with other studies. Cup: Disc ratio was more than 0.7 in 19% of cases. We were unable to document reliable visual fields in many of our patients with cataractous changes. Hence, visual field could not be used as a reliable indicator of disc damage. We interpreted pseudoexfoliation glaucoma as IOP more than 21 mmHg with disc changes having Cup: Disc ratio > 0.7.

Glaucoma in PEX is challenging to control. Target pressure should be set low, because there is greater diurnal variation of IOP. It can spike in short period. Altered blood-aqueous barrier in PEX leads to increased post-operative inflammation and posterior synechiae formation. All these factors lead to increased complications following cataract surgery. Patients may require aggressive treatment and frequent followup.

CONCLUSION

Pseudoexfoliation was higher in the age group of 51 - 70 years. Glaucoma occurred in 22% of cases. Of these 87%

had open angle and only 13% had occludable angle. We found that increased intraocular pressure, closed angles, increased Cup: Disc ratio, trabecular pigmentation and nuclear cataract were higher in patients with pseudoexfoliation.

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