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PSEUDOEXFOLIATION SYNDROME IN ETHIOPIAN GLAUCOMA PATIENTS
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PSEUDOEXFOLIATION SYNDROME IN ETHIOPIAN GLAUCOMA PATIENTS

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ABSTRACT

Background: **Pseudoexfoliation syndrome (PXS) has variable prevalence rates in the glaucoma population depending on geographic location and racial composition of the population studied. There is no data available on this subject in Ethiopia.**
Objective: **To determine the prevalence of PXS among Ethiopian glaucoma clinic population.**
Design: **A prospective study.**
Setting: **The Glaucoma Clinic of Menelik-II- Hospital, which is a tertiary eye care centre in Addis Ababa, Ethiopia.**
Participants: **All new patients with glaucoma or ocular hypertension (ocular HPN)seen during the study period from 1st April to 30th September 1996 were included in the study.**
Results: **Out of the 432 new patients of glaucoma or ocular HPN seen during the study, 108 patients (25%) had PXS. The age of PXS cases ranged from 32 years to 86 years (mean 62.3 years). There were 80 males and 28 females. Of the PXS patients, 104 (96.3%) were cases of chronic open angle glaucoma, while angle closure glaucoma and ocular HPN were rare comprising only two patients (1.85%) each.**
Conclusion: **The relatively high prevalence rate of PXS among Ethiopian glaucoma clinic population found in this study stresses the need for further community-based research in order to prevent blindness due to glaucoma associated with PXS.**

INTRODUCTION

The presence of pseudoexfoliative material as grayish flecks on the pupillary border in patients with chronic glaucoma was first described by Lindberg in 1917(1). Recently, Schlotzer-Schrehardt *et al*(2) produced evidence for fibrillin as an intrinsic component of pseudoexfoliation (PXF) fibers and that enhanced expression of fibrillin or abnormal aggregation of fibrillin-containing micro fibrils may be involved in the pathogenesis of PXS.

PXS is characterised clinically by the formation or deposition of fibrillo-granular material throughout the anterior segment of the eye. It is often accompanied by elevated intraocular pressure with or without glaucomatous optic nerve and visual field damage(3). There are strong indications that PXS is not a purely intraocular disease but represents ocular manifestation of a systemic disorder involving an aberrant connective-tissue metabolism throughout the body(4,5,6).

PXS was originally associated with Scandinavian countries, but it occurs worldwide with variable reported prevalences(1). The prevalence of PXS ranges from none among the Eskimos to as high as 38% in Navaho Indians(7,8).

Glaucoma occurs more commonly in eyes with PXS than without it(1). The causative factors in the development of a special type of secondary open-angle glaucoma in PXS are the mechanical obstruction of outflow channels by pseudoexfoliative material of exotrabecular origin as well as accumulation of locally produced PXF-material in juxta canalicular tissue of trabecular meshwork, followed by dysfunction of endothelial cells and disorganisation of juxtacanalicular tissue and Schlemm's canal(9).

The reported prevalence rates of PXS in the glaucoma population varies too, ranging from practically zero to as high as 93%(1). PXS was thought to be rare in black persons(10,11), but Luntz found the prevalence of PXS to be 1.4% in white and 20% in Bantu glaucoma patients in South Africa(12). One of the main reasons for the variability of reported prevalence rates of PXS in geographical locations worldwide is the racial and ethnic composition of the population studied(1). Therefore, it is important to look into the situation among Ethiopian glaucoma patients for the prevalence of PXS.

To our knowledge, there is no reported data on the prevalence of PXS among Ethiopian glaucoma patients. Such a study is very important since there is

a significant difference in management and prognosis of glaucoma associated with PXS than primary open angle glaucoma (POAG). Therefore, the main purpose of this study was to determine the prevalence of PXS among Ethiopian glaucoma clinic population.

MATERIALS AND METHODS

This was a six-month prospective study (from April 1 to September 1, 1996) of PXS patients seen at the Glaucoma Clinic of Menelik-II Hospital, Addis Ababa, Ethiopia. All patients with glaucoma or ocular HPN seen in the General-OPD were referred to the Glaucoma Clinic. These patients underwent a thorough work-up in the Glaucoma Clinic, which included a brief history, visual acuity (VA) and intraocular pressure (IOP) measurement, slit-lamp microscope (SLM) examination, gonioscopy, funduscopy and visual field (VF) testing.

This study involved all new cases referred to the Glaucoma Clinic during the study period. All patients with PXS and glaucoma or ocular HPN were included in this study, except those secondary glaucoma cases due to trauma, intraocular inflammation and anterior adherent leucoma.

For the purpose of this study, glaucoma is defined as an IOP≥22 mmHg with glaucomatous cupping of the optic disc and visual field loss, and ocular HPN if IOP ≥ 22mmHg without glaucomatous cupping or visual field loss(13). Pseudoexfoliation syndrome was diagnosed by the presence of typical flakes of exfoliative material on the pupillary margin or presence of peripheral exfoliation band on anterior lens capsule as revealed by diagnostic mydriasis or both(14).

The following data were collected for each patient: identification card number, name, age, sex, address, VA, IOP, SLM - findings of anterior segment, gonioscopic findings, cup/disc ratio and funduscopic findings, VF results and diagnosis.

RESULTS

During the study period a total number of 432 new patients were seen at the glaucoma Clinic of Menelik-II Hospital, Addis Ababa, Ethiopia. Out of these, 108 patients had pseudoexfoliation syndrome. Thus the prevalence rate of PXS cases among glaucoma clinic population in this series was found to be 25%. The age of PXS cases ranged from 32 years to 86 years, with a mean age of 62.3 years (Table 1). There were 80 males and 28 females, giving a male to female ratio of 2.85:1.

Table 1

Age distribution of PXS cases with glaucoma or ocular HPN

Age group in years	No. of patients	Percentage
30-39	1	0.92
40-49	7	6.48
50-59	26	24.1
≥60	74	68.5
Total	108	100

Of the PXS cases, 104 (96.3%) had COAG and two patients (1.85%) had ocular HPN and two others had ACG (Table 2). Of the 104 PXS cases with COAG, 64 patients had bilateral and 40 patients unilateral glaucoma (Table 3).

Table 2

Type of glaucoma among PXS cases

Type of glaucoma	No. of patients	Percentage
COAG	104	96.3
Ocular HPN	2	1.85
ACG	2	185
Total	108	100

Table 3

Presentation of the 108 patients with PXS PXF

Type of glaucoma	No. of Patients	Bilateral	Unilateral
COAG	104		
Bilateral	64	62	2
Unilateral	40	16	24
Ocular HPN	2	-	
Bilateral	2	2	-
Unilateral	-	-	-
CACG	2		
Bilateral	1	1	-
Unilateral	1	1	-

Of the 64 PXS cases with bilateral COAG, 62 had PXF in both eyes, but in the remaining two PXF was unilateral. Of the cases with unilateral COAG and unilateral PXF, the glaucoma involved invariably the eye with PXF. Two patients presented with bilateral ocular HPN and bilateral PXF. One patient each presented with bilateral ACG and unilateral ACG, but both had bilateral PXF.

DISCUSSION

In this study the prevalence of PXS among Ethiopian glaucoma clinic population was found to be 25%. This relatively high prevalence rate is close to the findings of Luntz(12), who reported a prevalence rate of 20% among the Bantu South African glaucoma clinic population.

Age is an important determinant in the incidence of PXS. In all reviews, the incidence of PXS and glaucoma increases with age(1). The fact that PXS is

overall the disease of the elderly is also reflected in the mean age of 62.3 years found in this study. In this study 92.6% of the patients were 50 years of age and above. The youngest patient in this study was 32 years old. However the youngest reported patient was 22 years of age(15).

In agreement with the literature(1), the vast majority of PXS cases (96.3%) in this study presented with COAG. ACG and ocular HPN were very rare each constituting only 1.85% of the patients in this series.

Among the unilateral COAG cases, 60% had unilateral PXF on the side of glaucomatous eye, but 40% of patients had bilateral PXF. The presence of PXS in the fellow eye of patients with unilateral glaucoma constitutes a serious risk factor(16). Therefore in cases of unilateral glaucoma, it is imperative to preserve the fellow eye.

PXF is known to be a definite risk factor for the development of elevated IOP and glaucoma. Henry *et al*(17), found the five and 10 year cumulative probabilities of initially non-glaucomatous eyes with PXF developing glaucoma to be 5.3% and 15.4% respectively, a significantly higher rate than would be expected in a similar group of patients without PXS.

This study has demonstrated a relatively high prevalence rate of 25% for PXS among Ethiopian glaucoma clinic population. Therefore, further community-based research is needed to determine the prevalence of PXS among the population at large in order to prevent loss of vision due to glaucoma associated with PXS.

REFERENCES

1. Layden, W.E. Exfoliation Syndrome: In: The Glaucomas, Vol.2, (Eds), Shields MB and Krupin, T. C.V. Mosby Co., 1989, 997-1015.
2. Schlotzer-Schrehardt, U., von-der Mark, K. Sakai, L.Y. and Naumann, G.O:. Increased extracellular deposition of fibrillin-containing fibrils in pseudoexfoliation syndrome. *Invest,-Ophthalmol-Vis-Sci.* 1997; **38:** 970-84.
3. Prince, A.M. and Ritch, R. Clinical signs of pseudo-exfoliation syndrome. *Ophthalmology.* 1986; **93:** 807-7.
4. Schlotzer-Schrehardt, U., Kuchle, M. and Naumann, G.O. Electron-microscopic identification of pseudoexfoliation material in extrabulbar tissue. *Arch. Ophthalmol,* 1991; **109:**565-70.
5. Streeten, B.W., Li, Z.Y., Wallace, R.N., Eagle, R.C. Jr. and Keshgegian, A.A. Pseudoexfoliation fibrillopathy in visceral organs of a patient with pseudoexfoliation syndrome. *Arch. Ophthalmol.* 1992; **110:**1757-62.
6. Schlotzer-Schrehardt, U.M., Koca, M.R., Naumann, G.O. and Volkholz. H. Pseudoexfoliation syndrome. Ocular manifestation of a systemic disorder? *Arch. Ophthalmol.* 1992;**110:**1752-6.
7. Faulkner, H.W. Pseudoexfollation of the lens among Navaho Indians. *Amer. J. Ophthalmol.* 1971; **72:**206-7.
8. Forsius, H. Prevalence of pseudoexfoliation of the lens in Finns, Lapps, Icelanders, Eskimos, and Russians. *Trans. Ophthalmol. Soc.* UK 1979; **99:** 296-8.
9. Schlotzer-Schrehardt, U.M. and Naumann, G.O. Trabecular meshwork in pseudoexfoliation syndrome with and without openangle glaucoma. A morphometric, ultrastructural study. *Invest. Ophthamol- Vis-Sci.* 1995; **36:**1750-64.
10. Aasved, H. The geographical distribution of fibrillopathia epitheliocapsularis. *Acta. Ophthalmol.* 1969;**47:**792-8 10.
11. Gradle, H.S. and Sugar, S. Glaucoma capsulare. *Amer. J. Ophthalmol* 1947; **30:**12-19.
12. Luntz, M.H. Prevalence of pseudoexfoliation syndrome in an urban South African clinic population. *Amer. J. Ophthalmol.* 1972; **74:**581-7.
13. Kozart, D.M. and Q. Yanoff, M. Intraocular pressure status in 100 consecutive patients with exfoliation syndrome. *Ophthalmology.* 1982; **89:**214-8.
14. Horven, I. Exfoliation syndrome, *Arch. Ophthalmol.* 1966; **76:**505-11.
15. Sugar, H.S. Pigmentary glaucoma and the glaucoma associated with the exfoliation -pseudoexfoliation syndrome. *Ophthalmology.* 1984; **91:**307-10.
16. Brooks, A.M.V. and Gillies, W.E. The presentation and Prognosis of Glaucoma in pseudoexfoliation of the lens capsule. *Ophthalmology.* 1988; **95:**271-6.
17. Henry, J.C. Long-term follow-ups of pseudoexfoliation and the development of elevated intraocular pressure. *Ophthalmology* 1987; **94:**545-52.