Pseudoexfoliation syndrome in various ethnic population of Nepal

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ABSTRACT

Pseudoexfoliation syndrome is an ocular condition characterized by a distinctive deposition of fibrillar material in the anterior segment of the eye. Pseudoexfoliation syndrome is frequently associated with glaucoma. Though it is seen in all the population, prevalence varies considerably in different races. A hospital based study done in Nepal by our group showed that Gurung population is frequently affected by this disease than any others. To determine the fact a larger population based study was necessary. Thus a research was designed to survey the various populations in the community. Different population from Kathmandu valley, Kavrepalanchowk, Pokhara, and Ghandruk were evaluated in two different phases to include major ethnic population living in Nepal. Household surveys done to bring all the targeted population to base hospital and detail examination including anterior segment evaluation after dilatation of pupil, optic disc evaluation, intraocular pressure measurement, gonioscopy, visual field recording and photographic documentation was done. Total of 4430 population surveyed while only 2135 fulfilled the criteria for inclusion. Gurungs irrespective of their place of residence showed higher prevalence of Pseudoexfoliation syndrome accounting for 7.8% in Kathmandu valley while 12.0% in Ghandruk. *Tamangs* were affected very rarely by this disease accounting only for 0.3%. Gurungs are at higher risk of developing Pseudoexfoliation syndrome than any others in Nepal.

Keywords: Pseudoexfoliation syndrome, glaucoma, *Gurungs*.

INTRODUCTION

Pseudoexfoliation syndrome (PXS) was first described by Linberg in 1917 among Finnish population.[1] The clinical diagnosis is made by the presence of typical pseudoexfoliation material (PXM) on the anterior capsule surface. In addition to PXM, other features include corneal endothelial pigmentation, loss of pupillary ruff, iris transillumination, Sampaolesi's line, and pigment deposition in the trabecular meshwork. Reported prevalence rates of PXS vary widely in different geographic locations.[2-4] Reasons for such variations have been thought to be racial and ethnic composition of the population studied, patient selection and clinical criteria for the diagnosis. The prevalence of PXS reported are England (4.0%), Germany (4.7%), Norway (6.3%), Eskimos (0%), Russia (12.0%), Finland (22.0%), Iceland (29.0%), Greece (16.1%), Australia (0.98%), and Iran (9.6%).[5-8] *Gurungs* living in Nepal has been reported to be predominantly affected by this disease entity by one of the hospital based studies originating from Kathmandu Valley.[9]

PXS is associated with various ocular complications. Elevated intraocular pressure and glaucomatous nerve damage had been demonstrated in patients with PXS.[10-14] Cataracts were reported to be more common in patients with PXS.[15,16] The prime interest in PXS is due to its association with glaucoma, which is the foremost cause of irreversible blindness all over the world. The primary aim of this study was to assess the prevalence of PXS in various ethnic populations in Nepal.

MATERIALS AND METHODS

A community based cross sectional descriptive study was conducted to include major ethnic population living in different parts of Nepal. Study area was selected purposefully as per the settlement pattern of different ethnic groups targeted for the study. Gurungs surveyed from Ghandruk, Pokhara and Kathmandu Valley, *Newars*, *Chetri* and *Brahmin* were surveyed from Kavre, *Sherpas* from Boudha Kathmandu and *Tamangs* from Ichangu village in Kathmandu. Gurung population was taken from Ghandruk as well as Pokhara and Kathmandu valley so that the effect of high altitude on the disease can be evaluated. Individuals who are 30 years and above residing in the study area and cooperated for the detail examination were included in the study. Household survey was performed by the local enumerators hired for the study to bring the entire individuals fulfilling the criteria for inclusion to the base clinic.

Complete examination of eye was performed including anterior segment evaluation after dilating the pupil, fundus examination, intraocular pressure (IOP) measurement, gonioscopy, anterior segment photography and Humphrey visual field analysis (for

Table-1: Demographic features of population studied (n= 2135)

Ethnic Population	Parameters	Numbers (%)
Gurungs		
Ghandruk	Total Population	275
	Males	130 (47.3%)
	Females	145 (52.7%)
	Mean Age (SD)	60 (9.446)
Pokhara	Total Population	350
	Males	121 (34.6%)
	Females	229 (65.4%)
	Mean Age (SD)	58 (9.740)
Kathmandu	Total Population	270
	Males	112 (41.5%)
	Females	158 (58.5%)
	Mean Age (SD)	59 (8.262)
Sherpa	Total population	365
	Males	152 (41.6%)
	Females	213 (58.4%)
	Mean Age (SD)	59 (10.110)
Tamangs	Total population	340
	Males	167 (49.1%)
	Females	173 (50.9%)
	Mean Age (SD)	62 (10.262)
Newar	Total population	211
	Males	101 (47.9%)
	Females	110 (52.1%)
	Mean Age (SD)	57 (10.245)
Brahmin	Total population	206
	Males	99 (48.0%)
	Females	107 (52.0%)
	Mean Age (SD)	61 (9.667)
Chetri	Total population	118
	Males	50 (42.4%)
	Females	68 (57.6%)
	Mean Age (SD)	56 (8.467)

those needed). Presence of exfoliative material on the pupillary border was the first tool to suspect PXS. After dilating the pupils, the presence of exfoliation changes over the anterior surface of the lens capsule was the

Table-2: Prevalence of pseudoexfoliation (n=2135)

Ethnic population	Total Number	Total PXS	%
High altitude Gurung	275	33	12.0%
Pokhara valley Gurung	350	30	8.6%
Kathmandu valley Gurung	270	21	7.8%
Sherpa	365	0	0
Tamang	340	1	0.3%
Newar	211	0	0
Brahmin	206	0	0
Chetri	118	0	0

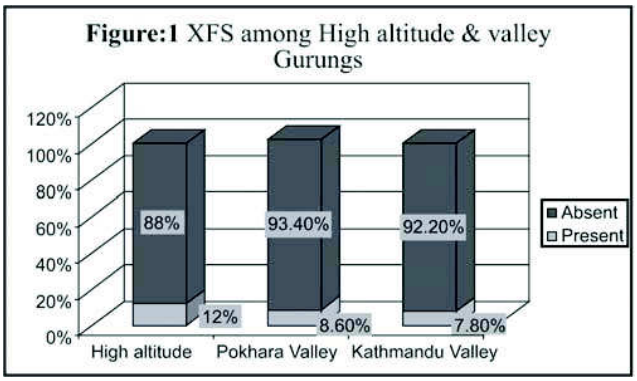


Figure:1 XFS among High altitude & valley Gurungs

determining sign for the diagnosis. Other signs like presence of corneal pigment deposition, transillumination defect at pupillary margin, exfoliation material over the angles of the anterior chamber were noted. Glaucoma was diagnosed only when there is presence of optic nerve head changes with visual field defect with or without rise in IOP.

RESULTS

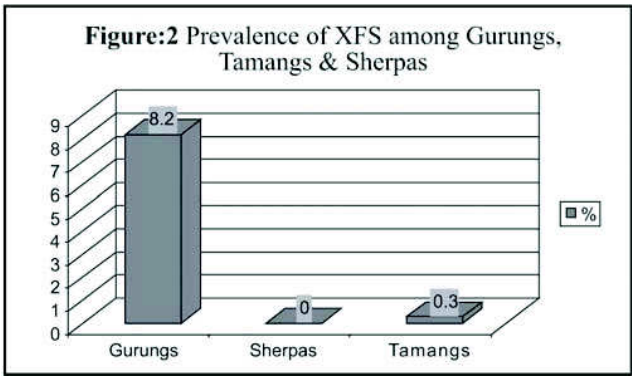
Total of 4430 population surveyed while only 2135 fulfilled the criteria for inclusion. Among total population examined, majority were females accounting for more than 50.0% in all ethnic groups. Mean age groups ranged from 56 to 62 in all groups of population enrolled in the study (Table-1).

Gurung population showed higher number of Pseudoexfoliation cases in contrast to any other, irrespective of their place of residence ranging from 7.8% in Kathmandu Valley to 12.0% in Ghandruk high land (Table-2). While comparing the prevalence among high altitude *Gurungs* and valley *Gurungs*, it was found that PXS was seen among both these population (Fig. 1). The odds ratio is 1.45 (95.0% confidence interval being 0.84-2.53) while high altitude *Gurungs* from Ghandruk were compared with Pokhara Valley *Gurungs*. The X^2 test showed no significant difference with P value > 0.1 at one degree of freedom. Comparison between High altitude *Gurung* and Kathmandu Valley *Gurung* also showed no significant difference. The odds ratio is 1.62 (95.0% confidence interval being 0.88-2.99) and p value 0.99.

On comparing the prevalence of PXS among Gurungs versus rest of the ethnic groups in valley, it was found that *Gurungs* collectively from Pokhara and Kathmandu

Table-3: Gender distribution of *Gurungs* having PXS

Gender	Total Population (n)	No. of PXS (n)= 84	Percentage Out of total (n)
Male	363	62 (73.8 %)	17.1%
Female	532	22 (26.2 %)	4.1%
			P< 0.001



Valley showed the prevalence rate of 8.2% while the only other ethnic groups showing PXS is *Tamang* and that showed 0.3% of prevalence, the difference of which is highly significant with p value <0.001(Fig. 2).

Gender distribution among cases having PXS in *Gurung* population was studied. It was found that 73.8% out of total cases showing PXS were males and rest were females. The prevalence of PXS therefore among males was 17.1% in contrast only 4.1% of females had PXS. Chi square test showed p value to be < 0.001 (Table-3). Detail evaluation of PXS cases was conducted to find out the laterality of involvement and its association with glaucoma. Laterality did not show much difference on whether being unilateral or bilateral as 46.2% having unilateral involvement (Table-4). Almost one third of the cases of PXS had associated glaucoma (Table-5). PXS was seen more frequently in elderly people in compare to younger ones. The prevalence of PXS was 5.0% among the age group 30-50 whereas it increased to 20.3% among the age groups 70 or more (Table-6).

DISCUSSION

Among total population examined, majority were females accounting for more than 50.0% in all ethnic groups. As per the 2001 census male population was at least 1.4 times greater than their counterparts in majority of these places surveyed for the study. Higher reporting by the females could also be due to the fact that most of the male members of the house holds were out side the village or out of home for the work as the survey time selected was during the day time. This was evidenced by the information gathered during the household survey.

Gurung population showed higher number of pseudoexfoliation cases in contrast to rest of the groups. This was similar to the previous report, which has shown higher prevalence among *Gurungs* than non-*Gurungs*.[10] However there was a slight difference in the prevalence rate between Ghandruk (12.0%), Pokhara (8.6%) and Kathmandu Valley *Gurungs* (7.8%), the prevalence of PXS was significantly seen among *Gurung* population irrespective of the place where they reside. Difference in prevalence rate between Gurungs from various study sites is not significant enough to make a note on it as P value is > 0.1 at one degree of freedom. In contrast rest of the ethnic groups did not show any PXS except the *Tamangs* who showed PXS in only 0.3% of total cases. This difference is highly significant with p value <0.001, which means *Gurungs*, are at higher risk to develop PXS than other ethnic groups. The highest prevalence of PXS had been reported till date from Iceland (29.0%) followed by Finland (22.0%)[6] whereas Russia (12.0%)[6] and Iran (9.6%)[9] had reported a similar prevalence rate as ours. A report from China had shown that PXS is rare in the country accounting for just 0.4% which is similar to that of *Tamangs* in this series.[17]

In this study, males seemed to be predominantly affected by this disease accounting for 73.8% out of total PXS cases despite the fact that more females reported at the time of examination. While trying to find out prevalence rate among the males, it was found to be 17.1% in contrast to just 4.1% among females (p < 0.001). This finding is similar to some of other studies[18] which had shown the higher prevalence in males. A study from China stated that PXS is rare in the country only cases they found to have PXS in their series were both males.[17]

Bilateral PXS was seen in 53.6% of cases in this study which is similar to a study from Greece which reported bilateral PXS to be more frequently seen than unilateral ones.[19] Association of glaucoma with PXS was studied and was found that 33.3% of the PXS had glaucoma where as 64.3% did not have associated glaucoma. This needs to be followed up for longer period as there have been some reports showing increasing prevalence of glaucoma among PXS cases with time. A study from Greece reported that 22.1% of PXS cases had glaucoma.[19] PXS was more frequently seen in elderly people in compare to younger ones showing significantly higher number of PXS in the age group 70 and above

Table-4: Laterality of PXS

Laterality	Number	Percentage
Unilateral	39	46.4 %
Bilateral	45	53.6 %
Total	**84**	**100.0 %**

Table-5: Association of glaucoma in cases of PXS

PXS	Number	Percentage
With glaucoma	28	33.3 %
With no glaucoma	54	64.3 %
With glaucoma suspect	2	2.4 %
Total	**84**	**100 %**

Table-6: Prevalence of PXS in relation to age among *Gurungs*

Age	PXS	Total Population	Percentage
30 - 50	15	298	5.0%
51 - 70	29	400	7.2%
>70	40	197	20.3%

accounting for 20.3% out of total of that age.

Gurungs are at higher risk of developing PXS irrespective of their place of residence. They probably are genetically predisposed to this entity. However, to prove this, a genetic study should be carried out in near future.

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