Hearing Loss Associated with Ocular Pseudoexfoliation Syndrome

Thabit Odat MD, Adnan Husban MD**, Hussein Al-Qasim PhD**, Issam Bataineh MD*, Eman Hammouri MD^, Nawwaf Abo Jamoos MD***

ABSTRACT

Objectives: To investigate the type and degree of hearing loss in a group of patients with ocular pseudoexfoliation.

Methods: Two groups of patients were included prospectively in the present study. The study group consisted of 60 patients of both genders, aged between 55 and 80 years (mean age 67.8 years) who were identified to have pseudoexfoliation in one or both eyes on routine ophthalmic clinical examination at Princess Haya Bent Al-Hussein Military Hospital in Aqaba. The control group consisted of 30 healthy individuals matched for age and gender (mean age 56.9 years). All participants underwent full ophthalmological and audiological assessments.

Results: Normal hearing threshold levels ranging between 10-25 decibels were found in 12 (20%) patients. Moderate sensori-neural hearing loss ranging between 50-70 decibels was found in 15 (25%) patients, high frequency sensori-neural hearing loss in the frequency range above 3kilohertz was found in 10 (16.7%) patients, and severe sensori-neural hearing loss ranging between 70-95 decibels hearing level was found in 23 (38.3%) patients.

Conclusion: There is evidence of significant association between ocular pseudoexfoliation and sensori-neural hearing loss.

Key words: Hearing loss, Sensori-neural, Pseudoexfoliation

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Introduction

Pseudoexfoliation (PEX) syndrome has been described as a specific type of stress-induced elastosis, elastic microfibrillopathy, associated with the excessive production and abnormal aggregation of elastic microfibril components which may be promoted by the deficiency of clusterin as a highly efficient extracellular chaperone.[1]

Ocular PEX syndrome is characterized by deposits of fibrillar extracellular material on anterior segment structures of the eye including the zonules, anterior capsule and trabecular meshwork, and may be associated with raised intraocular pressure in up to 50% of affected eyes.[2]

Originally, PEX syndrome was thought to be limited to the anterior segment of the eye, however, recent studies have demonstrated that pseudoexfoliation material is a common age related fibrillopathy of unknown cause and is widely distributed throughout the body, including blood vessels, heart, and lung. These pseudoexfoliation

From the Departments of:
*Ophthalmology, King Hussein Medical Centre, (KHMC), Amman-Jordan
**ENT, (KHMC)
^ Dentistry, (KHMC)
Correspondence should be addressed to Dr. T. Odat, P. O. Box 2740 Irbid 21110 Jordan, E-mail: thabitodat@hotmail.com, thabitodat@doctors.net.uk.
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fibers are consistently associated with fibroblasts, collagen and elastic fibers.[3]

Patients with PEX syndrome and glaucoma are at increased risk of carotid artery stiffness and decrease of baroreflex sensitivity.[4]

Ocular pseudoexfoliation-like deposits have been also found in various other organs and tissues such as liver, kidney, meninges and skin. The correlation between glaucoma and hearing loss has been previously studied but with the exception of normal tension glaucoma and some congenital syndromes, no strong evidence is available for such an association. Clinical impression described by investigators considering the structural characteristics of hearing organs and the possible adverse effect of abnormal deposits and/or vascular abnormalities on these organs would not be surprising.[5]

The inner ear is a complex organ where sound waves are translated into bioelectrical energy and afferent electrical nerve impulses. Embryologically, the anterior segment structure of the eye as well as the tectorial and basilar membranes in the inner ear are all derived from neural ectoderm. Fibrils of PEX material that accumulate on lens, pupillary margin, and other anterior segment structures can also accumulate on the tectorial and basilar membranes as well as stria vascularis in the inner ear.[6]

Hearing loss associated with PEX has been shown in several studies at some tested frequencies, but no studies have been carried out to determine the type and degree of hearing loss associated with PEX at all audiometric tested frequencies. Cahil *et al.* assessed the relationship between PEX and sensori-neural hearing loss in 69 patients with pseudoexfoliation at tested frequencies: 1, 2, and 3 kHz. They found that a large proportion of patients with pseudoexfoliation have sensori-neural hearing loss.[2] Similarly, Shaban and Asfour investigated the relationship between ocular PEX and sensori-neural hearing loss in 41 patients with PEX at three pure tone tested frequencies; they found that the majority of patients with ocular PEX had sensori-neural hearing loss.[6]

Yazdin *et al.* compared hearing threshold levels of 83 subjects with ocular PEX and compared with 83 age and gender matched controls without PEX at sound frequencies important for speech comprehension (1,2, and 3 kHz). They concluded that hearing threshold levels at frequencies that are important for speech comprehension are significantly worse in individuals with ocular PEX than in matched controls.[5]

Turacli *et al.* investigated the possible relationship between PEX and sensori-neural hearing loss in a group of 51 patients with PEX and 22 controls without PEX. They found that sensori-neural hearing loss was detected in the majority of PEX patients.[7] Aydogan *et al*. tested 75 subjects with PEX with a well matched control group, performing pure tone audiometry at 250, 500, 1000, 2000, 4000, and 8000 Hz. They found that mean thresholds at speech frequencies and mean absolute hearing thresholds at 2, 4, and 8 kHz were higher in PEX patients than controls; however, absolute hearing thresholds at 250, 500, and 1000 Hz individually did not differ significantly between cases and controls.[8]

The purpose of the present study is to determine the type and the degree of hearing loss at all tested audiometric frequencies and to determine which part of basilar membrane is affected.

Methods

Two groups of patients were included prospectively in the present study. A group of 60 patients of both genders, aged between 55 -80 years (mean age 67.8 years) who were identified as having PEX in one or both eyes in routine ophthalmic clinical examination at Princess Haya Bent Al-Hussein Military Hospital at Aqaba, were included in the study group. The control group consisted of 30 healthy age and gender matched individuals without PEX (mean age 56.9 years).

Full medical history was taken from all patients, including current or previous medical and surgical treatment for eyes and any ear, nose and throat diseases. Exclusion criteria were history of previous ear surgery, history of tympanic membrane perforation, history of head trauma, history of noise exposure, or working in a noisy environment, or repeated exposure to gun fire, use of ototoxic drugs, and concurrent upper respiratory tract infection at time of examination. Each patient underwent a full ocular examination including Snellen visual acuity test, slit lamp biomicroscopy, gonioscopy, applanation tonometry, and dilated fundoscopy. After that, all patients were referred to the Ear-Nose-Throat (ENT) unit for audiological assessment. Assessment was as follows:

1. Otoscopic examination: otoscopic investigation of the external and middle ear was carried out on each ear of the participants

Table I. The distribution of PEX among the study group (60 patients)

	Single eye	Both eyes	Total
Male	12	21	33
Female	9	18	27

Table II. Means and Standard Deviations of hearing threshold levels of control group (30 patients)

Frequency (Hz)	Mean of hearing threshold levels (dB)	Standard Deviation SD of Hearing threshold levels (dB)
250	17.25	6.29
500	13.5	5.08
1000	12.4	4.8
2000	10.5	4.0
3000	10.75	4.16
4000	13.75	5.25
6000	18.25	3.5

Table III. The distribution of hearing loss among 60 patients with PEX

Number of Patients	Degree of hearing loss	Type of hearing loss	Frequency range tested(Hz)
12	Normal hearing (10-25 dB HL)	No hearing loss	250-6000
15	Moderate (50-70 dB HL)	SNHL	250-6000
10	HFHL	SNHL	250-6000
23	Severe HL (70-95 dB HL)	SNHL	250-6000

SNHL: Sensori-neural hearing loss
HFHL: High frequency hearing loss > 3 kHz

of both groups by ENT doctors to evaluate any disorders of the external and middle ear.

2. Pure tone audiometry: hearing threshold levels measurement was done at 0.25, 0.5, 1, 2, 3, 4, and 6 kHz using air and bone conduction audiometry to determine the type and the degree of hearing loss. All measurements were carried out on each ear of the participants using descending of 10 dB step size and ascending of 5 dB step size to determine the lowest hearing threshold levels at all tested frequencies for each ear. A qualified audiologist in an isolated test using the same clinical diagnostic audiometer for all subjects carried out all measurements.
3. Tympanometry: test of the middle ear function was carried out on each ear of the participants of both groups using middle ear analyzer. The findings revealed no abnormalities that may interfere with the accuracy of pure tone audiometry.

Results

Ophthalmological investigation:

Sixty patients with unilateral or bilateral PEX who were attending the ophthalmic clinic at Princess Haya Military Hospital at Aqaba were selected randomly and enrolled in the study group during the period January 2007 and June 2007.

Table I shows the distribution of PEX in the study group and it shows that PEX is more frequent in males (male **:** female, 1.2 **:** 1) and both eyes (unilateral **:** bilateral, 1 **:** 1.86) are more frequently affected in both genders.

Audiological investigation:

Otoscopic examination revealed no abnormalities in the study group as well as in the control group.

Pure tone audiometery showed that hearing threshold levels were within normal levels at all tested frequencies (ranged between 5-20 dB HL).

Table II shows the mean and standard deviation of hearing threshold levels of the control group. Table III shows the degree and type of hearing loss of 60 patients (study group).

Discussion

In the present study, there is a significantly high prevalence of sensori-neural hearing loss in patients with PEX as compared with the age and gender matched group. This high prevalence was observed in the majority of patients at all tested frequencies ranging between 250-6000 Hz, which reflects the effect of PEX syndrome on all parts of the basilar membrane.

Our results are consistent with the results obtained with Cahill *et al.*[2] Yazdin *et al.*[5] Shaban and Asfour,[6] Turacli *et al.*[7] and Aydogan *et al.*[8] that ocular PEX syndrome has an association with sensori-neural hearing loss. No research study had shown contrast to these findings.

Sensori-neural hearing loss may be attributed to

various etiologies especially in old ages such as toxic agents, acoustic neuroma, or the aging process, however, the exact mechanism is unknown.[3]

In the present study, all patients with ocular PEX had various degrees of sensori-neural hearing threshold change compared with age matched control group, so we believe that there is an association between PEX and sensori-neural hearing loss. Structural alteration of the tectorial and basilar membranes and stria vascularis in the inner ear by deposition of PEX fibrils may change the way in which vibratory energy is conducted to the sensory hair cells and alter their surrounding chemical environment resulting in sensori-neural hearing loss.

Conclusion

It can be concluded that the majority of patients with ocular PEX had various degree of sensori-neural hearing loss. This indicates that there is an association between hearing loss and PEX.

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