CASE SERIES

Etiological and epidemiological factors in hearing-impaired students in Van, East of Turkey: A case series

Ahmet Faruk Kiroglu,[1] Muzaffer Ari,[2] Nazim Bozan,[3] Mehmet Parlak,[4] Hakan Cankaya[5]

Abstract

The aim of the present case series was to investigate the causes of hearing loss in the children studying at a primary School for the deaf in Van Province, Turkey. The present series included 95 children with sensorineural hearing loss. Otoscopic and ophthalmological examinations, pure-tone audiometry, and tympanometry were performed for each patient. Measles IgG, rubella IgG, cytomegalovirus (CMV) IgG, toxoplasma IgG, herpes simplex virus (HSV) Type I (HSV-I) IgG, HSV Type II (HSV-II) IgG, and Mycoplasma pneumonia IgG were evaluated in serum samples. Temporal bone anomalies were analyzed on temporal bone computed tomography (CT). A total of 7 radiologically distinct inner ear malformations were detected in 19 children. Twelve children were detected with various fundus pathologies. The most common cause of hearing loss was family-related factors (52%). In conclusion, the etiological and epidemiological factors were consistent with those reported in the literature.

Keywords: Hearing loss, Sensorineural, Etiology, Epidemiology.

Introduction

Hearing loss causes communication problems, thereby leading to language learning problems.[1] These problems vary according to the degree of hearing loss, frequency-based hearing pattern, whether the hearing loss is sudden or progressive, and the onset of hearing loss. Sensorineural hearing loss (SNHL) is a major cause of hearing loss in children which leads to serious challenges not only for the child and the family but also for the physician since the diagnosis of the disease is difficult and there are limited treatment and rehabilitation options.[2]

In this study, we aimed to investigate the causes of hearing loss in the children studying at a primary school for the deaf in Van Province, which is located in the Eastern Region of Turkey, in order to evaluate the children in terms of the use of hearing aid or cochlear implant and, more importantly, to report on the causes of hearing loss for the development of preventive measures to be taken for the individuals with hearing loss.

[1,3,5]Department of Otorhinolaryngology, Head and Neck Surgery, [4]Department of Medical Microbiology, Yuzuncu Yil University, Faculty of Medicine, Van, [2]Department of Otorhinolaryngology, Head and Neck Surgery, Mardin Park Hospital, Mardin, Turkey.

Correspondence: Ahmet Faruk Kiroglu. E-mail: fkiroglu@email.com

Case Series

This study was approved by Yuzuncu Yil University Medical School Human Research Ethics Committee (Approval No. 07.05.2008/01) and conducted between 2008 and 2009. A written consent was obtained from the parents of each patient. The consent of the patient/guardian was taken prior to the writing of the manuscript for publication. The parents were also administered a survey form which queried the prenatal, perinatal, and postnatal diseases associated with SNHL and the use of ototoxic drugs by the mother during pregnancy. Pure-tone audiometry and tympanometry were performed. The defects of vision were detected by using visual acuity test and fundus examination. The temporal bone anomalies were analyzed on coronal images from 30 consecutive temporal bone computed tomography (CT) studies.

The 95 children in the study group included 70 males and 25 females with a mean age of 11.5±2.4 years. Otoscopic examination was performed in 93 children, pure-tone audiometry in 93, and tympanometry was performed in 91 children, that were compatible for the examination (Table-1). Temporal bone CT was carried out in 85 (89%) children. A total of 7 radiologically distinct inner ear malformations were detected in 19 (23%) children including sclerotic cochlea as a sequela of meningitis (n=6), Mondini deformity (n=3), cochlea dysplasia (n=3), large vestibular aqueduct syndrome (LVAS) (n=3), large internal auditory canal (n=2), semicircular canal aplasia (n=1), and semicircular canal dysplasia (n=1). Ophthalmological examination revealed that twelve (13%) children were detected with various pathologies in the fundus, including retinitis pigmentosa (RP) (n=8), maculopathy (n=4), and torch scar (n=1). One child had both RP and maculopathy. The most common cause of SNHL was family-related factors (n=49). These were classified as syndromic (n=8) and non-syndromic factors (n=41). The second most common cause of SNHL was acquired factors (n=43), which included prenatal (n=3)

Table-1: Results of otoscopic examination.

	Right ear			Left ear		
	Female n (%)	**Male n (%)**	**Total**	**Female n (%)**	**Male n (%)**	**Total**
Results of otoscopic examination						
Natural	21 (22.6)	61 (65.6)	82 (88.2)	20 (22)	59 (63)	79 (85)
Perforated	1 (1.1)	3 (3.2)	4 (4.3)	1 (1.1)	4 (4.3)	5 (5.5)
Serous	0 (0)	0 (0)	0 (0)	2 (2.2)	0 (0)	2 (2.2)
Calcareous plaque	3 (3.2)	1 (1.1)	4 (4.3)	1 (1.1)	0 (0)	1 (1.1)
Pseudomembranous	0 (0)	0 (0)	0 (0)	1 (1.1)	2 (2.2)	3 (3.2)
Retracted	0 (0)	3 (3.2)	3 (3.2)	0 (0)	3 (3.2)	3 (3.2)
Degree of hearing loss						
56-70 dB	3 (3.2)	3 (3.2)	6 (6.5)	1 (1.1)	3 (3.2)	4 (4.3)
71-90 dB	4 (4.3)	10 (10.8)	14 (15)	6 (6.5)	10 (10.8)	16 (17.2)
91 dB or greater	18 (19.4)	55 (59.1)	73 (78.5)	18 (19.3)	55 (59.1)	73 (78.5)
Tympanometric Results						
Type A	24 (26.4)	60 (65.9)	84 (92.3)	22 (24.2)	61 (67)	83 (91.2)
Type B	0 (0)	2 (2.2)	2 (2.2)	1 (1.1)	1 (1.1)	2 (2.2)
Type C	0 (0)	5 (5.5)	5 (5.5)	1 (1.1)	4 (4.4)	5 (5.5)

Table-2: Status of owning, using, and utilizing a hearing aid in the children with hearing loss.

Owning a hearing aid	**Degree of hearing loss**	**Owning**	**Not owning**
	Moderate (56-70 dB)	2	2
	Severe (71-90 dB)	13	5
	Profound (91 dB or greater)	51	20
	Total (n=93)	66	27
Using a hearing aid **regularly**	**Degree of hearing loss**	**Using regularly**	**Not using**
	Moderate (56-70 dB)	1	1
	Severe (71-90 dB)	8	5
	Profound (91 dB or greater)	20	31
	Total (n=66)	29	37
Utilizing a hearing aid	**Degree of hearing loss**	**Utilizing**	**Not utilizing**
	Moderate (56-70 dB)	1	0
	Severe (71-90 dB)	8	0
	Profound (91 dB or greater)	6	14
	Total (n=29)	15	14

and postnatal factors (n=40). Prenatal factors included Rh incompatibility (n=1), maternal inflammatory disease in pregnancy (n=1), and maternal jaundice in pregnancy (n=1). Postnatal factors included meningitis (n=16), febrile convulsion (n=9), measles and/or parotitis (n=5), and other inflammatory diseases (n=10). The third most common cause of SNHL was idiopathic (n=3). Table-2 presents the status of owning, using and utilizing a hearing aid in the children with hearing loss. The parents of 51 (55%) children were first-degree and the parents of 11 (12%) children were third-degree relatives. In most of the children (61%), hearing loss was diagnosed at the age of 1 or 2, with a mean age of 18 months. Of the 25 female children of the group, 24 (96%) had a with profound hearing loss, which established a significant correlation between gender and the degree of hearing loss (x2= 8.257; standard deviation (SD) = 3; $p<0.05$).

Discussion

Silan et al. reviewed 550 children studying at 5 primary schools for the deaf in the provinces of Istanbul and Zonguldak and detected 33 distinct syndromes in 99 (18%) children.[3] The most common syndromes included Waardenburg syndrome, Usher syndrome, and oculo-otodental syndrome.[3] Similarly, in our study, we also found that Waardenburg syndrome, Usher syndrome, and oculo-otodental syndrome were the most common syndromes in the children with hearing loss. In another study, Turan et al. investigated children with syndromic hearing loss and found that the most common syndrome diagnosed in the parents was Waardenburg syndrome.[4] Similarly, Usher syndrome was found in 15% of the cases

in the study conducted by Yücel et al.[5] In our study, genetic hearing loss was found in 49 (51.6%) of the children with hearing loss.

Turan et al. investigated 2,300 children with genetic SNHL and reported the mean age of diagnosis as 2 years and the mean age at hospital admission as 4 years.[5] In contrast, 52% of the cases in our study had been diagnosed before the age of 1 year.

In a study that investigated 114 children in the USA, hearing loss associated with meningitis was found in 3.5%.[6] Osma et al. reported that 52% of the cases were detected with nongenetic hearing loss, of which 26.5% were caused by meningitis.[7] A study performed in Ankara reported that 2.1% of the cases were caused by prenatal factors and 11.97% of them by perinatal factors, whereas a study performed in Istanbul found that 1.2% of the cases were caused by prenatal and 7.1% of them by perinatal factors.[8,9] In our study, 43 (45.26%) cases were detected with acquired hearing loss. Of these, 3 (3.15%) cases were caused by prenatal and 40 (42.11%) were caused by postnatal factors. These findings of our study were also consistent with the literature.

Coticchia et al. investigated 69 patients with SNHL in the USA and detected inner ear malformations in 17 (25%) patients.[10] Of these, 8 (47%) patients were detected with large vestibular aqueduct syndrome (LVAS). In our study, a total of 7 radiologically distinct inner ear malformations were detected in 19 (22%) out of 85 children. These malformations included sclerotic cochlea as a sequela of meningitis (n=6), Mondini deformity (n=3), cochlea dysplasia (n=3), LVAS (n=3), large internal auditory canal (n=2), semicircular canal aplasia (n=1), and semicircular canal dysplasia (n=1).

In the studies performed in Turkey, the prevalence of SNHL of unknown etiology is reported to range between 15-43%.[7,8] In our study, only 3 (3%) children had SNHL of unknown etiology. The effect of consanguineous marriage on SNHL has been extensively studied in Turkey. The prevalence varies between 31% to 56%.[7,8] Osma et al. reported this prevalence as 39.87% in all cases of neurosensory hearing loss and as 91.11% in cases with hereditary hearing loss.[7] In our study, consanguineous marriage was observed in 62 (67.39%) out of the 92 families that participated in the survey. This is the highest rate reported in the studies conducted in Turkey and shows the risks affiliated with it.

Conclusion

In SNHL was found to be more common in children of consangenous marriages in Turkey.

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