Research Article / Araştırma Makalesi

Audiometric and Radiological Evaluation of Patients with Single-Sided Sensorineural Hearing Loss; Single Center Experience

Tek Taraflı Sensorinöral İşitme Kayıplı Hastaların Odyolojik ve Radyolojik Olarak Değerlendirilmesi; Tek Merkez Deneyimi

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Abstract

Background: In this study, It was aimed to make audiological and radiological evaluations of patients in the pediatric age group with unilateral sensorineural hearing loss, to reveal the degree of hearing loss, frequency of inner ear anomalies and the 8th cranial nerve anomaly according to age groups.
Materials and Methods: Forty-four pediatric patients who applied to Harran University Medical Faculty Otorhinolaryngology outpatient clinic between June 2014 and June 2015 and diagnosed with unilateral sensorineural hearing loss were included in the study. Patient's age, gender, lateralization of hearing loss, otoscopic examination findings, tympanometric and audiological findings, and radiological inner ear pathologies were evaluated.
Results: Nineteen patients were female (43.2%) and 25 were male (56.8%). The age range was between 0 to 16 years. The mean age of male patients was 9.88 years, while the mean age of female patients was 9.84 years. Twenty (45.5%) of the patients had sensorineural hearing loss in the right ear and 24 (54.5%) in the left ear. In the patients audiometric examination, 2 (4.5%) patients had unilateral moderate; five patients had (11.4%) unilateral moderate-severe, 11 (25%) patients had unilateral severe, and 26 (59.1%) patients had unilateral profound sensorineural hearing loss. In the radiological examinations, 33 (75%) patients had no pathological findings, while 11 (25%) patients had internal ear pathology. Four patients (9.1%) had bilateral internal inner ear pathology, 4 (9.1%) had a right inner ear, and 3 (6.8%) had left inner ear pathology.
Conclusions: Audiological and radiological imaging should be performed in patients with unilateral sensorineural hearing loss. High resolution computed tomography, and magnetic resonance imaging should be performed for the possible presence of an inner ear anomaly and evaluation of the 8th cranial nerve.

Key Words: Hearing loss, Sensorineural, Inner ear anomalie, Imaging findings

Öz.

Amaç: Bu çalışmada tek taraflı sensorinöral işitme kaybı saptanan pediatrik yaş grubundaki hastaların; odyolojik ve radyolojik değerlendirmelerinin yapılması, elde edilen veriler sonucunda yaş gruplarına göre işitme kaybının derecesi, iç kulak anomali sıklığı ve 8.kranial sinirin değerlendirilmesi amaçlanmıştır.
Materyal ve Metod: Haziran 2014 - Haziran 2015 tarihleri arasında Harran Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Hastalıkları polikliniğinde tek taraflı sensorinöral işitme kaybı tanısı konulan 44 çocuk hasta çalışmaya alındı. Hastaların yaş, cinsiyet, işitme kaybının lateralizasyonu, otoskopik muayene bulguları, timpanometrik inceleme bulguları, odyolojik bulguları ve radyolojik iç kulak patolojileri değerlendirildi.
Bulgular: Hastaların 19'u kız (%43.2), 25'i erkek (%56.8) idi. Yaş aralığı 0-16 yıl aralığındaydı. Erkek hastaların yaş ortalaması 9.88 yıl, kız hastaların yaş ortalaması ise 9.84 yıl olarak olarak saptandı. Hastaların 20'sinde (%45.5) sağ kulakta, 24'ünde (%54.5) ise sol kulağında sensorinöral işitme kaybı mevcuttu. Hastaların odyometrik incelemelerinde 2'sinde (%4.5) tek taraflı orta, 5'inde (%11.4) tek taraflı orta-ileri, 11'inde (%25) tek taraflı ileri, 26'sında (%59.1) tek taraflı çok ileri derecede sensorinöral işitme kaybı tespit edildi. Hastaların görüntülemeleri incelendiğinde; 33 (%75) hasta normal, 11 (%25) hasta iç kulak patolojisine sahipti. 4 hastada (%9.1) bilateral iç kulak patolojisi, 4 hastada (%9.1) sağ iç kulak ve 3 hastada (%6.8) sol iç kulak patolojisi tespit edildi.
Sonuç: Tek taraflı sensorinöral işitme kaybı tanılı hastalarda odyolojik ve radyolojik görüntüleme yapılmalıdır. Yüksek çözünürlüklü bilgisayarlı tomografi ve manyetik rezonans görüntüleme ile iç kulak anomalisinin varlığı ve 8.kranial sinirin değerlendirilmesi gerekmektedir.

Anahtar kelimeler: İşitme kaybı, Sensorinöral, İç kulak anomalisi, Görüntüleme bulguları

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Introduction

Hearing loss and deafness are conventionall it was defined and measured as a result of the hearing tests (1). Sensorineural hearig loss (SNHL) is a permanent hearing loss. Permanent hearing loss prevalence has been repoted as 1.2 to 1.86 per 1000 live births, and rhis is the most common sensory disorder in childhood (1). Unilateral hearing loss is defined as the presence of 20 dB or more hearing loss in the ear while the hearing is normal on the opposite side. Bilateral hearing loss defines the presence of hearing loss for both ears (2). There are various classifications related to hearing loss made according to etiological causes, type of hearing loss, and side/sides of hearing loss. Perinatal infections, structural anomalies, genetic disorders are listed among the causes of congenital hearing loss. Structural anomalies and internal ear abnormalities can be easily detected in 30-40% of patients with hearing loss by high resolution computed tomography (HRCT) (3). This study aimed to determine the degree of hearing loss, lateralization, and the detection and frequency of inner ear anomalies in pediatric patients with unilateral SNHL.

Materials and Methods

Forty-four pediatric patients who applied to Harran University Medical Faculty Otorhinolaryngology outpatient clinic between June 2014 and June 2015 with unilateral SNHL were included in the study. Study approval was obtained from the ethics committee of Harran University School of Medicine (12.12.2014 dated and 12 numbered session 07 numbered decision). Patients who had a parotid infection, head trauma, sudden hearing loss, neonatal period risk factors, noise exposure, meningitis, systemic disease, and meniere disease were excluded. All cases were evaluated with a detailed neuro-otologic examination, audiometric tests, and HRCT and magnetic resonance imaging (MRI). The patients' hearing loss level was determined by pure tone audiometry and auditory brainstem behavior (ABR) tests. Hearing loss levels were determined by Clark classification with audiological test results. With Clark classification, the level of hearing loss between 16 -25 dB HL was classified as slight, those with 26-40 dB HL were classified as mild, and 41-55 dB HL were moderate, while 56 -70 dB HL were moderately severe, 71- 90 dB HL were severe, and > 90 dB HL were classified as profound hearing loss (4). The inner ear structures are evaluated anatomically and morphologically and the determined inner ear anomalies are grouped by using Sennaroğlu classification (5). Accordingly, cochlear malformations were classified according to the following headings; Michel deformity, cochlear aplasia, common cavity deformity, incomplete partition-type I (IP-I), cochlear hypoplasia and incomplete partition type II (IP-II) (Mondini deformity). Vestibular malformations were classified as Michel deformity, dilated vestibule, vestibule hypoplasia, vestibule aplasia, common cavity deformity. Semisurgical duct anomalies were classified as semicircular canal hypoplasia, aplasia, dilatation. Internal acoustic canal anomalies were classified as narrow, wide and aplasic internal acoustic channels. Vestibular and cochlear aquaducts were defined as wide or normal. The frequency of each detected anomaly was calculated.

Statistical Analysis

Statistical Package for the Social Sciencesm (SPSS 21 Inc. Chicago, IL, USA) was used for statistical analyses. To analyze data, descriptive analysis (frequency and percentage) and mean±standard deviation were chosen.

Results

Forty-four pediatric patients with SNHL were included in the study. Table-1 shows the demographic characteristics, physical examination, and tympanometry test findings of the patients. Of forty-four patients, 19 were female (43.2%), and 25 were male (56.8%). The age range was between 0 to 16 years. The mean age of male patients was 9.88 years, while it was 9.84 years for female patients. Twenty (45.5%) of the patients had SNHL in the right ear and 24 (54.5%) in the left ear. In the otoscopic examination of the right ear of patients, 37 (84.1%) patients were evaluated as natural, 4 (9.1%) patients had opaque right tympanic membrane, and 3 (6.8%) patient's right tympanic membrane was retracted. In the otoscopic examination of the left ear of patients, 33 (75%) patients were evaluated as natural, 8 (18.2%) patients had opaque left tympanic membrane and 3 (6.8%) patient's left tympanic membrane was retracted. In the 226 Hz tympanogram test, 37 (84.1%) patients were type-A, 4 (9.1%) patients were type-B, and 3 (6.8%) patients were type-C for the right ear. 33 (75%) patients were type-A, 8 (18.2%) were type-B, and 3 (6.8%) were type-C for the left ear. The degree of hearing loss in patients audiological evaluation results were as follows; 2 (4.5%) patients had unilateral moderate SNHL, 5 (11.4%) patients had unilateral moderate-severe SNHL, 11 (25%) patients had unilateral severe SNHL and 26 (59.1%) patients had unilateral profound SNHL (Table 2). When HRCT and MRI examinations of the patients were examined, 33 (75%) patients had normal findings, while 11 (25%) patients had internal ear pathology (Table 3). Four patients (9.1%) had bilateral internal inner ear pathology, 4 (9.1%) had a right inner ear, and 3 (6.8%) had left inner ear pathology. In the evaluation of inner ear pathology type, 1 (2.3%) patient had cochlear hypoplasia. The patient with cochlear hypoplasia had a small, rudimentary cochlea (Figure-1). The vestibule and semicircular canals were normal. Four patients (9.1%) had mondini malformation (Figure-2), 2 (4.5%) patients had an IP-I anomaly in the cochlea (Figure-3). It was seen that patients with a Mondini malformation had a cochlea with an incomplete 1.5-time turn.

Table 1. Demographic, physical examination, tympanometry test findings of the patients

			n(%)
Age (Mean±Sd)	Male		9.88±5.60
	Female		9.84±5.81
Gender	Male		25 (56.8%)
	Female		19 (43.2%)
Otoscopic examination findings	Right Ear	Natural	37 (84.1%)
		Retracted	3 (6.8%)
		Opaque	4 (9.1%)
	Left Ear	Natural	33 (75%)
		Retracted	3 (6.8%)
		Opaque	8 (18.2%)
Tympanometry test findings	Right Ear	Type- A	37 (84.1%)
		Type- B	4 (9.1%)
		Type- C	3 (6.8%)
	Left Ear	Type- A	33 (75%)
		Type-B	8 (18.2%)
		Type- C	3 (6.8%)

Table 2. The degree of hearing loss in patients audiological evaluation results

	The degree of hearing loss
Unilateral moderate sensorineural hearing loss	2 (4.5%)
Unilateral moderate-severe sensorineural hearing loss	5 (11.4%)
Unilateral severe sensorineural hearing loss	11 (25%)
Unilateral profound sensorineural hearing loss	26 (59.1%)
Total	44 (100%)

Table 3. Radiological imaging findings of patients

	Computed Tomography and Magnetic Resonance Imaging findings
Natural	33 (75%)
Cochlear hypoplasia	1 (2.3%)
Mondini malformation	4 (9.1%)
İncomplete partition type-I	2 (4.5%)
Left internal acoustic canal dilatation	1(2.3%)
Bilateral wide vestibular aquaduct	2 (4.5%)
Right internal acoustic duct narrowing and the 8th cranial nerve aplasia	1 (2.3%)
Total	44 (100%)

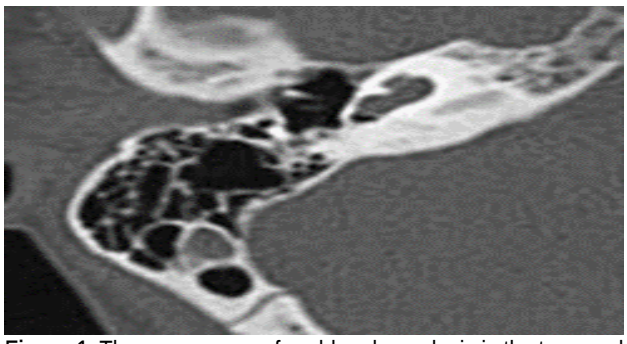

Figure 1. The appearance of cochlear hypoplasia in the temporal HRCT

There was a cystic apex associated with middle and apical folds. Vestibular dilatation and vestibular aquaduct expansion were observed. Two patients with bilateral Mondini malformation also had bilateral internal acoustic canal dilatation. In patients with unilateral Mondini malformation, wide internal acoustic canal pathology was not observed. Two patients had IP-I anomalies. In these patients, the cochlea was cystic, modiolus, and cribriform area were not present. Vestibul was observed in a wide and cystic structure. Four (9.1%) patients had either the vestibular, semicircular canal, internal acoustic canal or vestibular aquaduct anomalies.

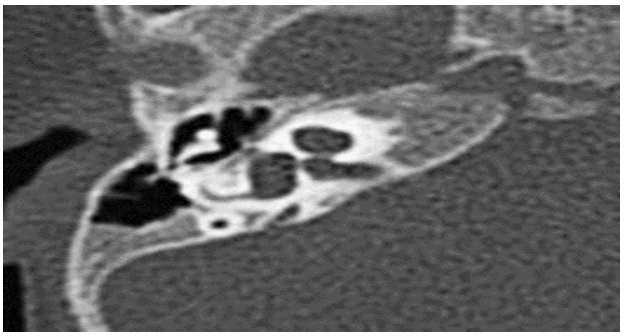

Figure 2. The appearance of mondini malformation in Temporal HRCT

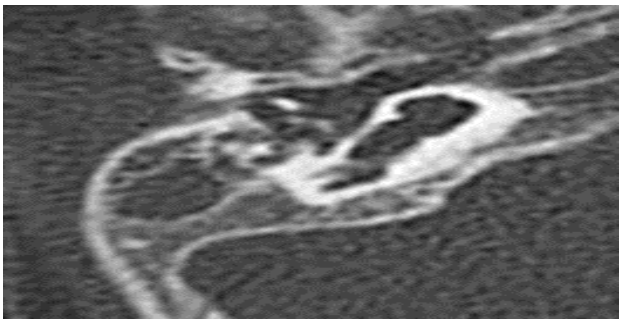

Figure 3. The appearance of incomplete partition Type-I in Temporal HRCT

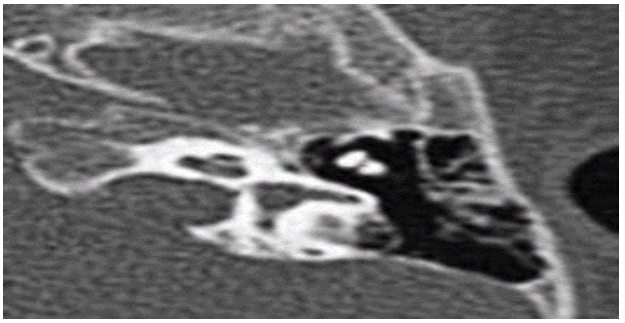

Figure 4. Internal acoustic dilatation in the temporal HRCT

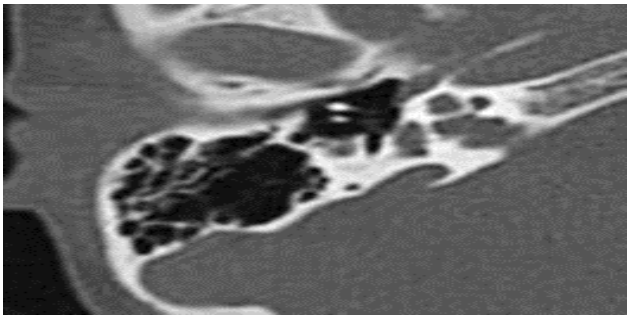

Figure 5. Wide vestibular aqueduct appearance in Temporal HRCT

Left internal acoustic canal dilatation was present in 1 (2.3%) (Figure-4) of these 4 (9.1%) patients, bilateral wide vestibular aquaduct (GVA) was present in 2 (4.5%) patients (Figure-5), right internal acoustic duct narrowing and the 8th cranial nerve aplasia were present in 1 (% 2.3) patient.

Discussion

Unilateral hearing loss is defined as the presence of 20 dB or more hearing loss in the ear while the opposite ear has normal hearing. Unilateral hearing loss has various descriptions in different studies in the literature; therefore, the data about its frequency is variable. However, the incidence of unilateral hearing loss was between 0.19-2/1000 in the newborn period and 3-5/1000 in the school period (6-8). In the study of Yelverton et al. (7) the incidence of unilateral hearing loss was reported as 0.5 in 1000 individuals. Unilateral hearing loss cannot be expressed by the child in early childhood and often do not cause a complaint because families cannot recognize this problem. Therefore, the possibility of treatment in the early period is eliminated. When the families of these children are questioned, parents generally state that the child's school success is low and mention problems such as balance pathologies, difficulty in distinguishing between speech in noisy environments.

In the study of Song et al. (3) conducted on 322 patients, ten patients (3.1%) had mild hearing loss, 39 patients (12.1%) revealed moderate hearing loss, 29 patients (9%) had severe hearing loss ve 244 patients (75.8%) had profound hearing loss. In the study of Stadio et al. (9) conducted on 154 children, unilateral profound SNHL was reported in 4 patient and unilateral moderate SNHL was repoted in 3 patients. In the study of Niu et al. (10) conducted on 75 patients, 1 patiente had mild unilateral hearing loss, 35 patients had moderate unilateral hearing loss, 32 patients had severe unilateral hearing loss, 6 patientes had profound unilateral hearing loss, 1 patient undetermined unilateral hearing loss.

In our study, 26 (59.1%) of 44 cases had unilateral profound, 11 (25%) patients had unilateral severe, 5 (11.4%) patients had unilateral moderate-severe, and 2 (4.5%) patients had unilateral moderate SNHL. Children with unilateral hearing loss should be investigated in various aspects. Unilateral hearing loss may be congenital or acquired due to many different causes. It is not always possible to reach the etiology. However, in addition to genetic evaluation, it is recommended to investigate the viral infections, to evaluate the other systems regarding various syndromes that might cause hearing loss and to perform a radiological evaluation for the inner ear and other anomalies. Any level of lesions can cause unilateral SNHL in the auditory pathway from the cochlea to the central nervous system. The lesion is most often in the cochlea, especially in the sensory epithelium and hairy cells. Less often seen localizations are spiral ganglion or higher levels. Identifying the localization of the lesion is very important in terms of prognosis and treatment (11). Most of the inner ear anomalies occur as a result of the cessation of development in the first trimester of pregnancy. This condition may be caused due to a genetic error or exposure to a teratogenic factor during embryogenesis of the inner ear. Early diagnosis and appropriate rehabilitation are very important for normal speech and language development (12). The inner ear is a complex organ composed of two intertwined structures called bone labyrinths and membranous labyrinths. In approximately 20% of cases with SNHL, rarely seen inner ear bone labyrinth anomalies can be observed (13). However, 80% of patients have hearing loss secondary to pathologies in the membranous labyrinth of pathologies at the cellular level, while the inner ear bone labyrinth structure is normal. No radiological findings can be detected in patients with hearing loss secondary to cellular pathology. As with all hearing losses, it is controversial which method should be chosen as the first choice in the radiological evaluation of unilateral hearing loss. In particular, in recent years, imaging modalities such as HRCT and MRI with increasing sensitivity can provide data to the clinician that cannot be reached in any other examinations. HRCT provides superior information about bone defects in the temporal bone and is actually the first recommended imaging modality and MRI provides superior information in demonstrating especially the 8th cranial nerve anomalies. An early detectable anomaly in the inner ear and cerebellopontine corner tumors can be seen together with unilateral hearing loss. Unilateral SNHL should be considered as an entity different from bilateral hearing loss, and imaging methods should be used to eliminate the suspicion of a mass lesion (3).

In the studies of Haffey et al. (6) and Simons et al. (14) it was reported that 33% to 35% of cases with unilateral SNHL had positive findings in HRCT. In our study, the rate of positive findings in HRCT was 25% in 44 cases. In the study of Kaya et al. (15) conducted on 1516 ears; reported that 89.29% of the ears had cochlovestibular, 2.5% vestibular, 0.14% semicicular canal, 0.77% internal acoustic canal, 1.1% cranial nevre malformation. In their study, Aldhafeeri and alsonasi (16) detected an iner ear anomaly in 24 patients. They reported GVA in 8 (33.3%) patients, mondini malformation in 7 (29.1%), semicircular canal dysplasia in 8 (33.3%) patients, and cochlear hypoplasia in 1 (4.1%) patient. In a study by Song et al. (3) which included 322 patients with unilateral SNHL, 93 patients were found to have inner ear anomaly after an HRCT scan (28.9%). When 93 inner ear anomalies are examined, 49 (52.7%) cases showed a cochleovestibular anomaly, 27 (29%) patients were found to have isolated vestibular anomaly, 17

(18.3%) patients had an anomaly of vestibular or cochlear aquaducts. Of the 49 cochleovestibular anomaly patients, IP-II was the most common type (28 of 49), followed by IP-I (10 of 49). Fifteen of 19 unilateral IP-II and 5 of 9 bilateral IP-II cases were combined with EVAS. In the study of Tuzcu et al. (17) among 54 patients who were diagnosed with SNHL, 15 (27.8%) cases were found to have internal ear anomalies after MRI and HRCT imagings. Seven of the cases (46%) had Mondini deformity, 5 (33%) had isolated GVA, 1 (7%) had Michel aplasia, 1 (7%) had cochlear aplasia, and 1 (7%) had joint cavity anomaly.

In our study, cochlear hypoplasia was present in one patient (2.3%), Mondini malformation was present in 4 patients (incomplete partition type-II) (9.1%), IP-I anomaly was present (4.5%) in two patients and other internal ear anomalies accompanied with normal cochlea (9.1%) were present in 4 patients. Mondini malformation was bilateral in 2 patients and was seen only in the right ear in 2 patients. IP-I anomaly in 2 patients was present only in their right ears. Cochlear hypoplasia was also present in the right ear. Other inner ear anomalies accompanying normal cochlea were present in 4 (9.1%) patients. Left internal acoustic canal dilatation was present in 1 (2.3%) of these 4 (9.1%) patients, while bilateral GVA was present in 2 (4.5%) patients, right internal acoustic duct narrowing and the 8th cranial nerve aplasia was present in 1 (2.3%) patient. According to Bamiou et al. (18) HRCT in children with unilateral SNHL has been shown to be useful in demonstrating bilateral ear pathology. According to Uweara et al. (19) it was observed that new-onset hearing loss in the functioning ear was 10.6% in a 12-week follow-up. Marcus et al. (20) reported that 25 patients with unilateral hearing loss had also developed a hearing loss in the intact ear detected with HRCT findings.

In our study, including 44 patients with unilateral SNHL, 11 patients (25%) had an inner ear pathology on the HRCT scans. In 4 (9.1%) of 11 (25%) pediatric patients with internal ear pathology, internal ear anomalies were also detected in the intact ear.

Conclusion

Audiometric and radiological imaging should be performed in pediatric patients who are diagnosed with unilateral SNHL. Intrauterine anomalies and nerve pathologies should be investigated by HRCT and MRI. Also, it should be kept in mind that in children with unilateral SNHL, there may be internal ear anomalies in the intact ear, and SNHL may also develop in the intact ear later in life.

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Conflict of Interest

No conflict of interest was declared by the authors.

Financial Disclosure

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Ethical Approval:

Ethics committee approval was received for this study from the ethics committee of Harran University School of Medicine (12.12.2014 dated and 12 numbered session 07 numbered decision).

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