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Case Report of Non-Organic Hearing Loss: Literature Review

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ABSTRACT

Although more advanced audiological tests including auditory brain response (ABR), transient-evoked otoacoustic emissions (TEOAEs), and distortion product otoacoustic emissions (DPOAEs) have been used to diagnose non-organic hearing loss (NOHL). NOHL may still be missed or misdiagnosed in clinical practice. We present 5 cases of NOHL in our department over the past 2 years. Two cases (Case 1 and Case 4) were initially misdiagnosed and prescribed inappropriate treatment, with poor outcomes. The other 3 patients were diagnosed correctly after ABR testing. In all cases, the audiogram was not reliable. Then ABR and OAEs were used to measure hearing thresholds. As suspected, discrepancies between PTA and ABR were found in all 5 cases. Although the presence of distinctive clinical features (age, gender, past history, and occupation) may be sufficient for a clinical suspicion of NOHL, objective audiological tests such as ABR and OAEs are essential for diagnosis. We suggest that otolaryngologists/audiologists should consider the diagnosis of NOHL when the patient shows poor test retest reliability on pure tone audiometry.

Keywords: Non-Organic Hearing Loss (NOHL); Pure tone audiometry; Conversion disorder; ABR; OAE

INTRODUCTION

Non-Organic Hearing Loss (NOHL), unconscious deafness, is defined as hearing loss that cannot be explained by any organic abnormality in the anatomy or physiology of the hearing system [1,2]. In the scientific literature, terms that are interchangeable for NOHL include: psychogenic hearing loss, conversion deafness, functional hearing loss, pseudohypacusis, and malingering [2]. When information related to the etiology of the patient's condition is lacking, the most likely diagnoses are NOHL, functional hearing loss, psychogenic hearing loss, and conversion deafness. The diagnoses of pseudohypacusis and malingering usually refer to patients who intentionally exhibit impaired hearing for self-gain (e.g., financial compensation, drugs), to win a lawsuit, or to avoid military service [3]. In most cases encountered in clinical practice, experienced clinicians may easily confirm NOHL using conventional audiological procedures [4]. More advanced audiological tests including auditory brain response (ABR) [5], Otoacoustic Emissions (OAEs) [6], Transient-Evoked Otoacoustic Emissions (TEOAEs) [7], and distortion product otoacoustic emissions (DPOAEs) [8] are often used as tools for diagnosis. However, the accurate diagnosis of NOHL remains challenge, especially in cases with high risk for actual hearing loss. For example, a diagnosis of NOHL may be overlooked in a patient with chronic hearing loss or hearing loss caused by head trauma. The prevalence of NOHL is less than 2% of the general population [9]. In children, NOHL with new onset hearing loss is frequently misdiagnosed as sudden hearing loss. The incidence of childhood NOHL is reported to range from 2%-7%, but these figures may have been underestimated [2]. Patients who have been misdiagnosed may be prescribed inappropriate treatment, such as steroids, exploratory surgery, and rehabilitation.

The etiology and risk factors of NOHL have been investigated in

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numerous studies over the past 10 years. The etiology of this condition differs between pediatric and adult patient populations. Familial, social, or emotional conflict is the most common precipitating factor among pediatric patients [10]. Factors considered to contribute to the development of NOHL include depression, personality disorder, anxiety, and history of a conversion disorder [11]. Conversion disorder is defined in the Diagnostic and Statistical Manual of Mental Disorders (5th ed.) As the presence of symptoms or deficits in voluntary motor or sensory functions, accompanied by clinical findings that cannot be explained by a neurological or medical condition [12,13]. Although much is known regarding the physical characteristics, comorbid symptoms, psychological makeup, and neuropsychological performance of patients with conversion disorder, the mechanism behind it is still poorly understood. Neuroimaging studies of conversion disorder have shown different activity patterns in different parts of the brain [14-16]. Reduced activity in the thalamus may be responsible for reduced sensory perception, which may contribute to the elevated hearing thresholds observed in many patients with NOHL at the time of their initial presentation [17].

Although some clinical characteristics of NOHL have previously been reported, some patients with this condition may still be misdiagnosed, leading to unnecessary costs and lost time. Therefore, an appropriate protocol for diagnosis must be established. In this study, we present five cases of NOHL and review the related literature to increase awareness of this condition among otolaryngologists and/or audiologists.

CASE PRESENTATION

Case 1

A 40 year old female presented at our department with history of bilateral hearing loss over the last 4 years. She also complained of tinnitus. The initial examination revealed no remarkable findings in the eardrums. Although pure tone audiometry (PTA) was performed 3 times, no satisfactory threshold was identified. The patient was suspected to have NOHL. The results obtained on OAE and ABR testing confirmed normal hearing in both ears, confirming the suspected diagnosis **(Figures 1)**. The patient was diagnosed with NOHL and referred to a psychologist. She recovered after completing a course of psychological therapy.

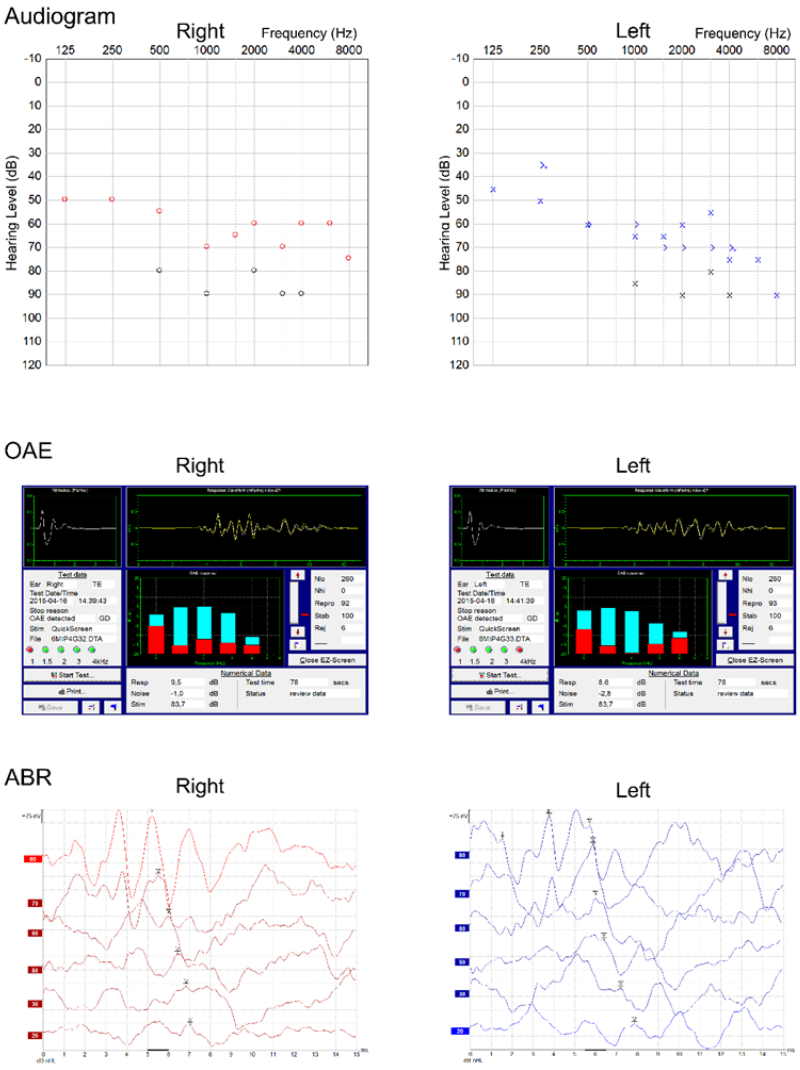

Figure 1: Outcomes of the repeated PTA assessment as performed on the day of the patient's presentation at our clinic. Because of uncertainty related to the patient's responses, it was impossible to reliably calculate hearing thresholds. Subsequently, the patient was sent for OAE and ABR. The results of these tests indicated that the patient had normal hearing.

Case 2

A 17 year old male presented at an ear, nose, and throat clinic complaining of hearing loss that had started immediately after his head was hit with a field hockey stick. His previous medical history was unremarkable. At the time of injury, there was no loss of consciousness, headache, nausea, or photophobia. The patient was able to walk normally immediately after the injury. He had neither tinnitus nor vertigo at the time of injury or afterwards. Physical examination revealed a normal external ear canal and tympanic membrane bilaterally. No hemotympanum was seen. The results of nasal and oral exams were also normal. There was no evidence of Cerebrospinal Fluid (CSF) oto-or-rhinorrhea, or any external sign of basal skull fracture. CT and physical examination showed that patient's cranial nerves and cerebellum were normal. However, the patient's results on the Weber test were lateralized to the left, and results on the Rinne test were equivocal.

An audiogram demonstrated normal left sided hearing with profound right sided sensorineural hearing loss. The physician diagnosed the patient with right side hearing loss. A hearing aid was provided at the patient's request. Use of hearing aids did not improve the patient's right sided hearing loss. He came to our hospital for further treatment 4 year later.

Pure tone audiometry was performed 3 times because the threshold could not be identified with accuracy. We therefore suspected NOHL. The results on ABR, OAE, computed tomography (CT) and blood tests were normal. A diagnosis of NOHL was made. The patient's parents were informed that the patient had normal hearing and that he did not need the hearing aid. A telephone interview performed during follow up confirmed that the child's hearing had normalized.

Case 3

An 18 year old female in her first year of high school presented to our department with a complaint of sudden bilateral hearing loss that had persisted for 2 weeks. The patient denied tinnitus and vertigo. Physical examination revealed normal tympanic membranes. PTA revealed flat sensorineural hearing loss bilaterally. The results of OAEs and ABR indicated normal hearing. A diagnosis of NOHL was made. The patient's hearing subsequently improved.

Case 4

A 53 year old female presented at our clinic complaining of chronic left side hearing loss that had persisted for 3 years. The onset of the patient's hearing loss had occurred when she was hit on the left side of her head with a beer bottle 3 years earlier. Bilateral tinnitus and transient vertigo appeared soon afterward. When the patient presented for treatment in the emergency department, her ears were examined and found to be normal. The hearing loss first appeared after she had returned home. Throughout this period, the patient's right side hearing was normal. She visited an otolaryngologist and was prescribed use of hearing aids. However, use of hearing aids did not improve the patient's hearing loss.

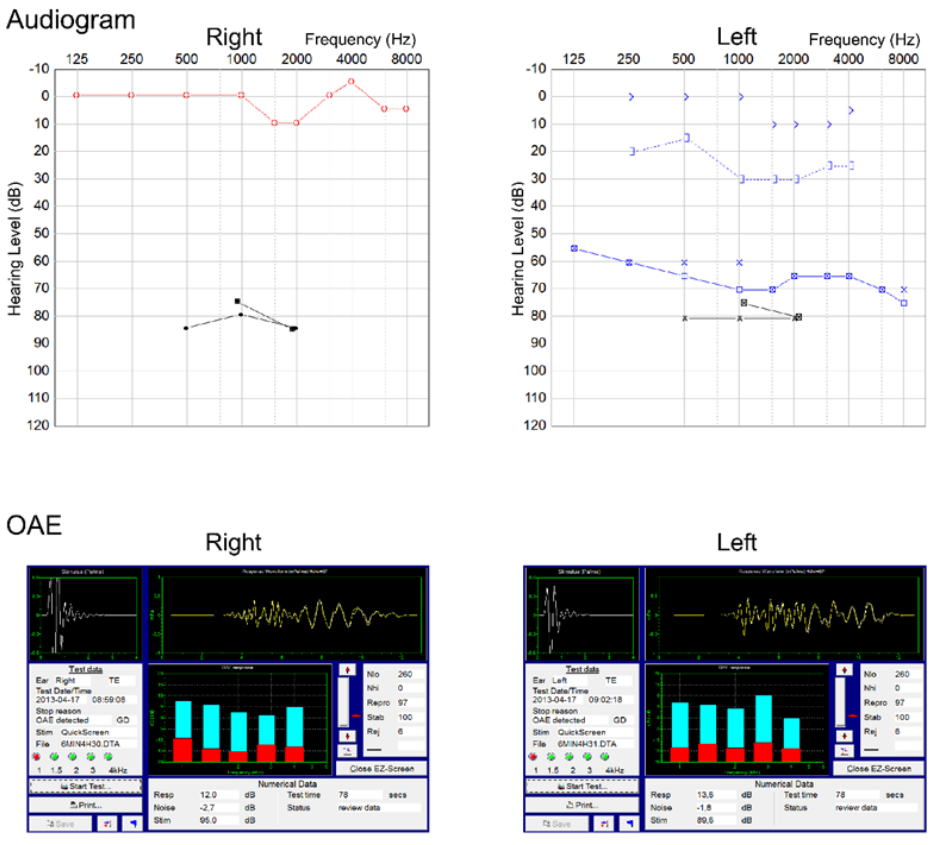

Figure 2: Outcomes of the repeated PTA evaluation performed on the day the patient presented to our department. The results showed left-side hearing loss; the right side indicated normal hearing. The patient was then sent for OAE. The results of this examination indicated normal hearing.

The patient first presented to our department 1 year ago. Physical examinations of the patient's ear and nervous system were normal. The results of PTA showed hearing loss on the left side **(Figure 2)**. OAE was subsequently performed. The results were normal. We explained the results to the patients and told her that use of the hearing aids was unnecessary. She was referred to a psychologist. The symptoms reported by the patient to the psychologist included poor sleep, inattention, and a negative outlook on life. The psychologist diagnosed the patient with post-traumatic stress disorder by the psychologist. She participated in a course of psychological therapy.

Case 5

A 47 year old female visited us complaining of hearing loss, aural fullness, hyperacusis, and tinnitus, with symptoms stronger on the right side. The patient reported issues with balance and frustration related to her tinnitus. The results of her physical examination were normal. The patient had undergone PTA testing 3 years earlier, and was found to be completely deaf. The patient's hearing subsequently improved. Last winter, she presented again at our department. PTA was performed. The results indicated that her left side hearing had improved since her last visit to our clinic. Two months later, she visited us again because her hearing had worsened. The results of PTA showed that the patient's hearing was impaired in comparison to the quality observed at the time of her second visit to the clinic, but nonetheless remained superior to the results observed at the time of the patient's first visit to our clinic. The results of OAE and ABR testing indicated a diagnosis of NOHL **(Figure 3)**. The results of OAE indicated normal hearing bilaterally. The thresholds obtained by ABR testing were 25 dB on the left side and 40 dB on the right side.

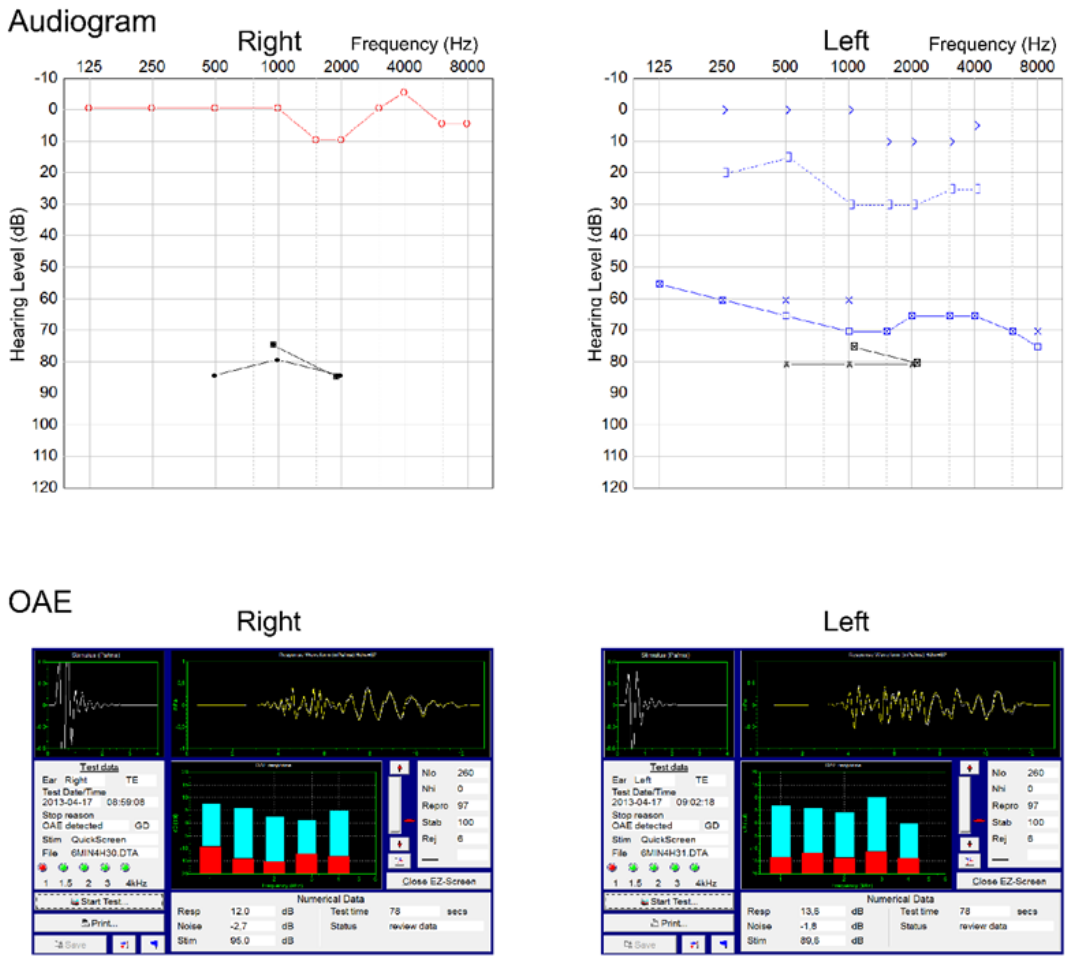

Figure 3: Outcomes for the repeated PTA evaluation performed on the day that the patient arrived at our department. The results obtained for the left and right sides indicated severe hearing loss. However, the audiometry diagram did not clearly identify a reliable hearing threshold. OAE and ABR were also performed. The results of OAE indicated normal hearing. The results of ABR indicated a hearing threshold of25dB on the left side and 40dB on the right side.

DISCUSSION

The mainstay of the NOHL diagnosis is differentiate actual and measured hearing. However, objective and subjective audiometric tests may not be performed at the same time for each patient who presents to us with hearing loss. Failure to recognize the possibility of NOHL may result inconsiderable expense related to unnecessary or even harmful hearing aid fittings [18]. Therefore, it is important to document the features of this condition. PTA is the conventional hearing test used most commonly to identify hearing thresholds. Investigators previously reported several indicators on standard audiometry that

are often associated with NOHL: poor test retest reliability, a saucer shaped audiogram, a flat sensorineural hearing threshold configuration [19], absence of a shadow curve in the case of total unilateral hearing loss, and a discrepancy between speech audiometry and PTA threshold (based on the audiometric speech frequency average) [20]. Jerger and Herer indicated that patients with NOHL sometimes show a V-typepattern; i.e., the tracings for interrupted tones showed poor hearing, compared to those for continuous tones [21]. Holenweg and Kompis [18] reported that the appearance of an air bone gap on PTA is pathognomonic for NOHL. In our clinical experience, poor test retest reliability is also strongly specific for NOHL. However, audiograms cannot provide a definitive diagnosis. In Cases 1 and 4 presented above, the diagnosis of NOHL was initially missed because they had high test retest variability on PTA. Increased awareness of this condition should decrease the likelihood of a missed diagnosis. NOHL should be included in the differential diagnosis, even in the case of a strong clinical history supporting hearing loss, such as physical trauma or exposure to ototoxic agents [22,23].

As electrophysiology has developed, OAE and ABR techniques have been used to identify objective hearing thresholds. OAE tests, including TEOAEs and DPOAEs, may play a role in the management of NOHL because they are noninvasive and easy and quick to perform. However, OAE testing is present when hearing loss is more than 40-50 dB. For our Case 5, the inconsistency of the results obtained with OAE and ABR testing indicated the NOHL. ABR has the advantage of not depending on a patient's state of consciousness, but it takes some time to perform and requires cooperation from a patient who is awake. As the "gold standard" for the detection and diagnosis of hearing loss, ABR is an irreplaceable tool for diagnosing NOHL. ABR was performed for all cases described above; the results indicated a diagnosis of NOHL in all cases. Because of our ability to precisely diagnose NOHL in an expedient fashion, hearing was completely restored in Cases 2 and 3, without the need for extra medicine or other treatment. Thus, an appropriate audiometric examination is extremely important in the evaluation of any patient with hearing loss.

NOHL has been categorized as a psychological disorder. Post-traumatic stress disorder is a mental disorder that can develop after exposure to stressful or exceptional events, such as assault, warfare, traffic collisions, or other threats on a person's life [13]. The diagnosis of post-traumatic stress disorder (PTSD) is based on the presence of specific symptoms after a traumatic event [24]. The symptoms may include persistent intrusive recollections, avoidance of trauma related cues, negative changes in cognition and emotions, and increased arousal. In Cases 1 and 4, the observed hearing loss was caused by (PTSD). The risk for PTSD is high among immigrants and refugees, particularly those who have immigrated because of armed conflict or political repression. One previous study indicated that 52.0% of Central American refugees from war or political persecution meet the criteria for PTSD [25]. Regarding Case 5, she had undergone several depressing experiences, including domestic violence, caring for a disabled child, and loss of her parents during wartime.

Approximately 95% of NOHL cases recover spontaneously, usually within 2 wk. However, some patients show no improvement or fluctuating hearing levels [26]. The factors that influence prognosis in patients with NOHL are assessment of the predisposing stress, treatment for the symptoms, and lack of any comorbid psychiatric or medical disease. Psychiatric consultation is a helpful strategy for pediatric as well as adult patients, but confrontation and pejorative statements such as, "There is nothing wrong with your hearing." must be avoided [27]. Case 2 resisted transfer to the psychology department, so a more tailored treatment program had to be developed. Shigehito et al. suggested that it is imperative for otolaryngologists to have some psychiatric knowledge and to be able to treat such patients with a primary approach [28]. Numerous studies published over the last 10 years have described functional neurological disorders, often with imaging. Such neuroimaging techniques will also be helpful in the management of NOHL. However, few studies published have focused on NOHL. Additional studies are needed.

CONCLUSION

NOHL is easily diagnosed in cases with a discrepancy between real and measured hearing thresholds. However, the condition is often misdiagnosed. NOHL should be suspected in patients with discrepancies in their results on subjective hearing test; objective tests such as OAE and ABR play important roles in narrowing the differential diagnosis.

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CONFLICT OF INTEREST

Authors declare no conflict of interest

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