Case Report

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Audiological findings from an adult with thin cochlear nerves

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Summary **Reported here are audiological findings from an adult with thin cochlear nerves. Magnetic resonance imaging (MRI) revealed that he had a thinner cochlear nerve in the left ear than in the right ear. He had a higher degree of hearing loss in the left ear and poor speech recognition scores for both ears. He had normal middle ear and cochlear functioning. The auditory brainstem response and acoustic reflexes were absent, indicating a retrocochlear pathology. Long latency responses (LLR) revealed normal cortical functioning. Hence, implantation of an auditory brainstem implant might be an option, but the patient would need to be aware of its limitations. This case highlights the importance of MRI in evaluating congenital malformations of the cochlear nerve when audiological findings indicate a retrocochlear pathology.**

Keywords: Magnetic resonance imaging, cochlear nerve, auditory brainstem response, long latency responses

1. Introduction

The assessment of structural malformations of the inner ear and vestibulocochlear nerve plays an important role in assessment and management of individuals with hearing loss. A structural abnormality in the auditory nerve could be the possible reason for the limited benefit of hearing aids and cochlear implants. Magnetic resonance imaging (MRI) plays an important role identifying such structural abnormalities (*1,2*). Audiological findings from individuals with auditory nerve malformations are key to providing appropriate counseling to the patient. In addition, audiological findings are also key to deciding rehabilitation options. Miyanohara *et al*. (*3*) reported a case of a 6-year-old child who had congenital absence of the cochlear nerve and moderate high-frequency sensorineural hearing loss. Otoacoustic emissions were present and an auditory brainstem response was absent. Furuta *et al*. (*4*) reported a 12-year-old child with a thin cochlear nerve in one ear. The child had a structurally normal cochlea but MRI revealed a thin cochlear nerve. Furuta *et al*. posited that the thin cochlear nerve could have been due to mumps or a developmental malformation that caused sensorineural hearing loss in the affected ear. However, there is a dearth of information on audiological findings from adults with thin cochlear nerves. Thus, reported here are findings from an audiological evaluation of an adult with thin cochlear nerves. This report also attempts to describe the possible pathophysiology that resulted in those findings.

2. Case Report

A 30-year-old adult male was seen by the audiology clinic with reduced hearing sensitivity in both ears since childhood. Hearing sensitivity in the right ear was described as better than that in the left ear. He described tinnitus in both ears, with more intense tinnitus in the left ear. He also complained of difficulty understanding speech, especially in situations involving a large amount noise. He mentioned no vestibular problems. There was no family history of hearing loss. The symptoms had manifested in early infancy and had progressed over time. A body level hearing aid was recommended 10 years prior. He used the hearing aid for 6 months and discontinued its use because of its limited benefit. He underwent a detailed audiological evaluation that included pure tone audiometry, speech audiometry, immittance testing, a transient evoked otoacoustic

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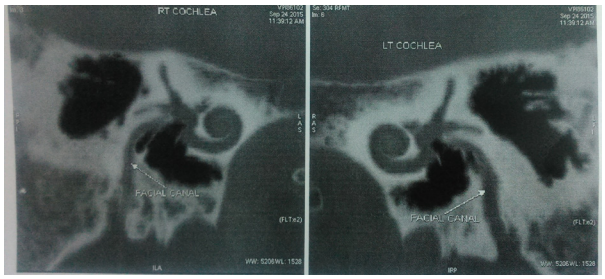

Figure 1. MRI of the left and right cochlea showing abnormal cochlear nerves with respect to the facial canal. MRI, magnetic resonance imaging.

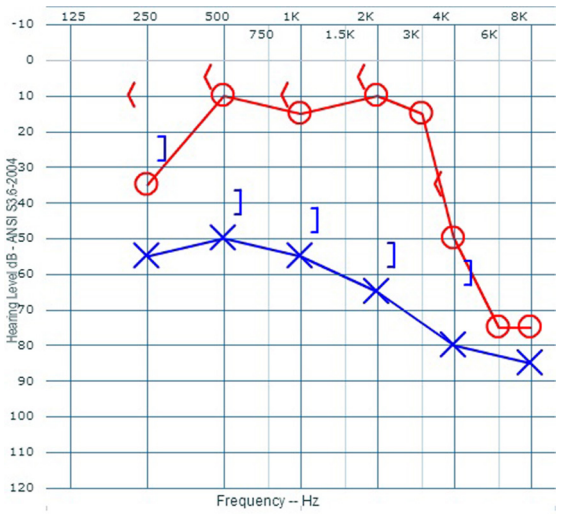

Figure 2. A pure tone audiogram showing air- and bone-conduction thresholds for the left and right ear. Thresholds for the right ear are plotted in red and those for the left ear are plotted in blue.

emission (TEOAE) test, a distortion product otoacoustic emission (DPOAE) test, an auditory brainstem response (ABR) test, and a long latency response (LLR) test using standard protocols. Neurological evaluation was performed and included a clinical neurological examination and a computed tomography (CT) scan of the brain with MRI of the temporal bone to identify any space-occupying lesion in the auditory nerve. Informed consent to participation in this research and publication of its findings was obtained.

Clinical neurological evaluation suggested that the man had a retrocochlear pathology and X-rays were obtained. X-rays revealed that the man had thin cochlear nerves. The left cochlear nerve was thinner than the right cochlear nerve. Figure 1 shown the MRI image of both auditory nerves with respect to the facial canal. The internal auditory canals were normal in size. The vestibular branch of the vestibulocochlear nerve was normal in size. MRI revealed no signs of an abnormal mass in the canal or the nerve. No abnormalities were detected in the structures of the middle and inner ear. No fracturing of the temporal bone was noted. X-rays verified the thinness of the cochlear nerves, and no other abnormalities were detected in the structures of the middle or inner ear.

An audiological evaluation revealed that the man had minimal sensorineural hearing loss in the right ear and moderately severe sensorineural hearing loss in the left ear based on Clark's classification of degree of hearing loss (*5*). The pure tone average of 500 Hz, 1,000 Hz, 2,000 Hz, and 4,000 Hz in the right ear was 21.25 dB HL and that in the left ear was 62.5 dB HL. An audiogram for both ears is shown in Figure 2. Speech recognition scores were determined using the word list developed by Yathiraj and Vijayalakshmi (*6*). The speech recognition scores were 28% in the right ear and 12% in the left ear. He had a type 'A' tympanogram with acoustic reflexes absent for pure tones at 500 Hz, 1,000 Hz, 2,000 Hz, and 4,000 Hz in response to both ipsilateral and contralateral stimulation. TEOAE and DPOAE were present in both ears with a signal-to-noise ratio greater than 6 dB, indicative of normal outer hair cell functioning. ABR testing was performed (90-dB click stimuli at a repetition rate of 11.1/s), and the ABR was absent in both ears. LLR testing was performed (click stimuli at a repetition rate of 1.1/s), and LLR was present in the right ear at normal latencies and amplitude but absent in the left ear. Thus, the audiological evaluation revealed a higher degree of hearing loss in the left ear with poor speech recognition in both ears. He had normal cochleae and auditory cortices with abnormal auditory nerve functioning.

Ethical Considerations: All testing was done using non-invasive techniques approved by the ethics committee of the Institute and in accordance with the Declaration of Helsinki. All test procedures were explained to the individual and his family before testing and informed consent was obtained.

3. Discussion

MRI revealed thin cochlear nerves. MRI results also revealed that all of the structures of the middle and inner ear were normal, suggesting a cochlear nerve abnormality alone. Studies have reported that MRI is an important and accurate means of identifying structural abnormalities of the cochlea and auditory nerve (*1,2*). Adunka *et al*. (*7*) found that most individuals with a cochlear nerve deficiency had a normal internal auditory canal (IAC) morphology. Thus, Adunka *et al*. suggested that high-resolution MRI is a better way to identify cochlear nerve deficiencies than CT. In the current case, MRI was used and it revealed that the patient had normal IACs but thin cochlear nerves in both ears. Hence, MRI is a key imaging study when a retrocochlear pathology is indicated during audiological evaluation.

A pure tone audiogram revealed a higher degree of hearing loss in the left ear than in the right ear. Thus, more severe hearing loss in the left ear suggested that

the cochlear nerve in the left ear was less intact. These findings coincided with MRI results, which revealed a thinner cochlear nerve on the left. Similar results were obtained by Furuta *et al*. (*4*), who reported a child with a thin cochlear nerve in one ear who had a higher degree of hearing loss in the affected ear. The current patient had worse speech recognition scores for the left ear (12%) compared to those for the right ear (28%). In individuals with a poor understanding of speech, low speech recognition scores are more indicative of a retrocochlear pathology than a cochlear pathology (*8*). The auditory nerve in the left ear was thinner and could have led to poorer speech recognition scores in the left ear compared to that in the right ear. However, speech recognition scores for both ears differed little. This suggests that even a slight thinning of the auditory nerve can significantly affect speech perception scores and may not be correlated with auditory nerve thickness. The current patient had normal middle ear functioning with absent acoustic reflex, which correlates with clinical findings of a retrocochlear pathology (*8,9*). The absence of appropriate conduction of sound from an abnormal auditory nerve could have led to the absence of acoustic reflexes. The current patient had normal otoacoustic emissions, indicating normal cochlear functioning. MRI revealed no abnormalities in the cochleae, which clearly indicates that the disorder is retrocochlear.

In the current case, an auditory brainstem response was absent in both ears. This also indicates abnormal functioning of the auditory nerve. ABR can be an indication of a retrocochlear pathology, suggesting the need for further investigation using X-rays. ABR is usually reported to be abnormal in individuals with structural anomalies of the cochlear nerve (*3,4*) and auditory neuropathy spectrum disorder (*8,9*). LLR testing assesses the auditory cortex and was performed in the current case. LLR testing revealed normal latencies and amplitudes in the right ear, suggesting normal cortical functioning. The LLR to click stimuli was absent in the left ear because of a higher degree of high-frequency hearing loss in that ear. Normal auditory cortical function was noted, suggesting that auditory problems were mainly due to abnormal cochlear nerves. Other peripheral and central structures appeared to be normal. Thus, hearing aids and cochlear implants may provide a limited benefit. Auditory brainstem implants (ABIs) are recommended for individuals with cochlear nerve deficiencies since they bypass the cochlear nerve and directly stimulate the cochlear nucleus (*2,10*). Thus, an ABI might be an option to improve the current patient's ability to understand speech. However, studies of ABIs recommend that an ABI be implanted early on for more benefit and those studies indicate that an ABI mainly improves awareness of sound (*11,12*). There are also risks associated with implantation of an ABI and an ABI provides a limited benefit in terms of understanding speech (*11,12*). Given these limitations, implantation of an ABI may be an option, but the patient would need to have realistic expectations of what benefit the ABI would have. In the current case, detailed immunological testing of the inner ear and genetic testing were not performed, and such testing could provide further insight into the pathophysiology of the patient's condition.

4. Conclusion

Reported here are audiological findings from an adult with thin cochlear nerves. The individual in question had minimal hearing loss in the right ear and moderately severe sensorineural hearing loss in the left ear. The degree of hearing loss was greater in the ear with the thinner cochlear nerve. He had normal otoacoustic emissions, suggesting normal cochlear functioning. ABR was absent in both ears, indicating a retrocochlear pathology. LLRs were present in the right ear, suggesting normal auditory cortex functioning. Thus, implantation of an ABI may be an option, albeit with limited benefit. Larger samples of individuals with thin cochlear nerves need to be studied further.

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