Single-sided deafness related to neural vascular conflict: A case report from Saudi Arabia

Mona S Al Rakaf

ABSTRACT

The impact of single-sided deafness (SSD) is well known, particularly on children. These include difficulties in speech comprehension with background noise, localization, and spatial appreciation. The social and emotional status of the child may be compromised. In addition, parents are keen to understand the etiology and consequences of SSD. Different etiologies are correlated with SSD, and etiology identification is a crucial step toward better management and parent counseling. SSD related to neural vascular conflict (NVC) of the cerebellopontine angle is uncommon. This is the first case, up to my knowledge so far, to be reported in Saudi Arabia. A 13-year-old Saudi girl presented with a history of difficulty in hearing in the left ear, and audiological evaluation demonstrated left-sided profound sensorineural hearing loss. Upon further investigation, magnetic resonance imaging (MRI) showed a left-sided neural vascular conflict of the anterior inferior cerebellar artery (AICA) loop and the left cochlear nerve. In this case, SSD was related to NVC since clinical history, and other investigations did not justify another cause. This report will embrace the role of audiological and radiological investigation, which in turn will help with management recommendations, and help clinicians in dealing with similar challenges.

Keywords: Bone conduction hearing devices, Cochlear implantation, Neural vascular conflict, Single-sided deafness

How to cite this article


doi: 10.5348/101150Z01MR2020CR

INTRODUCTION

Single-sided deafness (SSD) in children is a condition in which they have a non-functional hearing in one ear and normal hearing in the contralateral ear. Individuals with bilaterally normal hearing have a tendency to comprehend speech in noise and have the ability to localize sound precisely [1]. The impact of SSD is well known, particularly in children. Consequences include: difficulties in comprehension speech with background noise, localization, and spatial appreciation. Furthermore, compromised competency of verbal communication and cognitive capacity affected academic performance, and incapacitated social and emotional status [2, 3]. In addition, patients with SSD will experience one-sided auditory deprivation, which will initiate neural plasticity within the auditory cortex and other sensory and cognitive areas [4].

Causes of SSD are congenital cytomegalovirus, meningitis, related syndromes, absence of the cochlear nerve, inner ear abnormality, acoustic neuroma, autoimmune diseases, chronic ear infection, and head trauma [5]. Neural vascular conflict (NVC) of the cerebellopontine angle is considered a rare cause of SSD among children, which can affect the V, VII, VIII, and IX cranial nerves. The commonest presenting symptom of NVC is trigeminal neuralgia, and next to that is hemifacial spasm [6]. Neural vascular conflict of the VIII nerve may provoke symptoms of tinnitus, hearing loss, and vertigo as neurotological symptoms. It was reported that after five centuries of investigations, the exact mechanisms...
beyond the pathogenesis of NVC are still undetermined [7]. Neurological signs related to the pressure effect of vascular loops within the cerebellopontine angle (CPA) on cranial nerves have been documented [8]. Etiological investigations for bilateral sensorineural hearing loss (SNHL) is more toward genetic causation. On the other hand, the temporal bone structural defect is more related to SSD [9]. According to a meta-analysis which composed of five case controls, NVC was significantly associated with SSD [10]. Moreover, 91 patients who were studied retrospectively for unexplained unilateral hearing loss or tinnitus had a negative clinical workup, NVC with MRI confirmation could be the culprit [11]. In this case, MRI showed a left-sided NVC of the AICA loop and the left cochlear nerve. Single-sided deafness was related to NVC in view of the fact that clinical history and other investigations did not justify another cause. In addition, the right side revealed normal hearing and negative MRI findings.

CASE REPORT

A 13-year-old female patient presented to Otorhinolaryngology Head & Neck Surgery (ORL H&NS) clinic at Prince Sultan Military Medical City with a history of difficulty in hearing in the left ear with unknown exact onset. Her father reported that they had been informed by the school teacher that the child did not pass the school’s hearing test on the left ear. The child denied other associated symptoms such as tinnitus and vertigo. No history of previous ear diseases or serious childhood infections. Pre-, peri-, and postnatal history was unremarkable. Past medical, surgical history, and family history were irrelevant. Ear-nose-throat (ENT) examination was without abnormalities. Otoscopic inspection revealed normal eardrums. Cranial nerves evaluation revealed no neurological deficit apart from difficulties in hearing in the left side. Weber’s test localized to the right ear. No observed spontaneous or induced nystagmus. Audiological evaluation disclosed the following: tympanogram showed bilateral type A graph, suggestive of normal middle ear function (Figure 1). Pure-tone audiometry (PTA) demonstrated normal hearing in the right side and left-sided profound sensorineural hearing loss (Figure 2). Auditory brainstem responses (ABR) and auditory steady-state response (ASSR) confirmed PTA results (Figures 3 and 4). Caloric testing revealed left canal paresis of 34% (Figure 5). The results of the caloric test, in provisions of the responses to four air stimuli, were used to attain measures of horizontal canal status. Comprehensive laboratory investigations that included a hormonal panel, biochemistry, and viral serology were unremarkable for underlying pathology. Routine non-enhanced MR sequences were obtained (T1, T2, DWI, FIESTA, and T1 post-contrast and with fat suppression). The MRI revealed that there is a left-sided neural vascular conflict of the AICA loop (Figure 6), which is abutting the cisternal segment of the cochlear nerve proximal to the entrance of the internal auditory canal. The conflict corresponds to faint abnormal enhancement perceived in both axial and coronal acquisitions. Both inner ear structures were within normal limits in terms
of formation and signal intensity. There was no diffusion restriction. The brain showed normal gray-white matter differentiation, normal ventricles, and CSF spaces. Magnetic resonance imaging findings were suggestive of neural-vascular conflict between the AICA loop and the left cochlear nerve. On that basis, single-sided deafness was diagnosed.

Microvascular decompression (MVD) of the vestibulocochlear nerve is possibly helpful in special cases that reveal pulsatile tinnitus or unbearable positional vertigo. Unfortunately, there is no supporting evidence until this time for the efficacy of MVD for SNHL.

Parents conveyed their distress of their child being rejected or bullied because of the stigma of SSD or wearing hearing aids. This distress leads parents to decide against the usage of hearing aids. Parents and patient were counseled that single-sided deafness cannot be cured. Nevertheless, there is more than one option to lessen the difficulties in hearing from that side. These options are: cochlear implantation (CI) and varying types of hearing aids such as contralateral routing of sound systems (CROSS) hearing aids or bone conduction devices (BCD). This is achieved by transmitting sound from the deaf side to the hearing ear. In addition, the patient was educated on listening strategies and coping through considering the best setting and listening location besides reducing background noise whenever possible, all of this should be with reasonable expectations.

**DISCUSSION**

This case report demonstrated an uncommon pathology for SSD, which was found in a 13-year-old girl, and as mentioned in the previous studies, NVC related to NVC between the AICA loop and the left cochlear nerve was diagnosed.

Figure 3: Auditory brainstem responses with reproducibility for wave V at 20 dB nHL for right ear and absent at 100 dB nHL on left ear.

Figure 4: Auditory steady state response: the thresholds found were 15, 20, 20, and 20 dB, respectively for 500, 1000, 2000, and 4000 Hz, on the right ear, and 95, 90, 100, and 90 dB, respectively for 500, 1000, 2000, and 4000 Hz, on the left ear.

Figure 5: Caloric testing revealed left canal paresis of 34%.

Figure 6: MRI internal acoustic meatus (IAM): Neural-vascular conflict between AICA loop and left cochlear nerve.
is a recognized cause for SSD. There is considerable controversy in the literature regarding the effect of AICA on the VIIIth cranial nerve with a hearing loss sequelae [12]. Contrarily, individuals may have no symptoms in spite of positive MRI findings [13].

This young girl and her family were facing personal challenges in regard to her hearing difficulty. The psychosocial consequences of dealing with the burden of the illness itself and the disfigurement of using a hearing aid, which rooted from the fear of social stigma, were other hurdles to be considered. The role of audiological assessment and follow-up is crucial, not only for clinical diagnosis, but for a rather comprehensive approach that includes the patient and the family’s biopsychosocial needs.

The first step in the audiological evaluation was following the usual model of history taking and audiological evaluation with PTA, ABR, and ASSR. These tests confirmed the profound sensory hearing loss of the left ear and assessed the function of the right ear to be normal without any hearing loss. Hence, ordering an MRI was essential. Moreover, the MRI that was requested to look for an underlying cause found that the NVC was also on the left side only, and this correlates to the area of the patient’s pathology.

Evaluation of patients with SSD is an important measure for appropriate management. Magnetic resonance imaging is an important diagnostic tool that also helps in informed therapy planning, and it is the responsibility of the physician to evaluate the outcome of investigations, the need for further action, and a befitting referral where necessary. In this case, SSD was linked to NVC on the basis of MRI finding and in view of the fact that clinical history and other investigations did not justify another cause.

Etiological evaluation is a crucial step toward better management with an effective counseling process for patients and parents. Magnetic resonance imaging is an important diagnostic tool that also helps in informed therapy planning, and it is the responsibility of the physician to evaluate the outcome of investigations, the need for further action, and a befitting referral where necessary. In this case, SSD was linked to NVC on the basis of MRI finding and in view of the fact that clinical history and other investigations did not justify another cause.

In conclusion, this report described an uncommon pathology for SSD. The causes of SSD may be overlooked when investigations apart from MRI are normal. However, this should not exhaust physicians to look for an underlying cause whenever MRI is available, as in this case, where NVC was the most likely reason for SSD. Having a diagnosis can help the parents and the patient in finding closure for the reason for their illness and move on with the management. Provided with the evidence, this will aid in patient education, counseling, and tailoring the management plan according to the individual needs of the patients and their families. Additional cost-effective studies on the best management for SSD and exploring psychological and social consequences must be carried out, to assure a high standard of delivered care. Evidence-based decision-making empowers professionals to face challenges in convincing the child and parents to offer interventions that tremendously improve the child’s outcomes and outweigh any stigma.

REFERENCES


**********

Author Contributions
Mona S Al Rakaf – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Author declares no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright
© 2020 Mona S Al Rakaf. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.