

Auditory Neuropathy: A Case of Near Misdiagnosis

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Abstract

Auditory neuropathy is a type of hearing loss. This is a case study that describes an 18 months old child presenting with hearing loss reported by his parents. The parents reported that although he exhibited signs of hearing loss, they were told by multiple experts that tests confirmed that he was not deaf. The child sometimes acted as if he was deaf and sometimes did not, which lead to confusion about additional diagnoses that might be included in the diagnosis. An early diagnosis of auditory neuropathy is time sensitive as early development of learning and language is more beneficial for the child. Listening to parents and understanding the subtleties that segregate the diagnosis of auditory neuropathy from auditory processing disorder, autism or other similar presentations is important in the diagnosis and treatment of auditory neuropathy in children.

Introduction

Auditory neuropathy is a disorder that effects the neural processing of auditory stimuli in the brain and does so either directly or indirectly [1]. Dowley et al. [2] explain that auditory neuropathy is a disorder of the cochlear inner hair-cell and/or eighth-nerve function. Auditory Neuropathy Spectrum Disorder is the full name for this disorder. Other names that are often used interchangeably are Auditory Neuropathy (AN) and Auditory Dyssynchrony (AD). For the discussion in this article AN will be used. AN is the main cause of hearing impairment in about 10% or more of deaf patients [3]. Due to the fact that there are considerable variations in patient presentation of symptoms, one must not only understand the physiological testing that must be used but the subtleties in patient presentation that are often overlooked [1].

Diagnostic testing in the evaluation of AN includes a battery of hearing tests [4]. The tests that are considered the most useful consist of the otoacoustic exam (OAE), auditory brainstem response (ABR), and middle ear reflex (MEMR) [4]. One differentiation in auditory neuropathy versus other ontological diseases is the presence of normal outer hair cell function along with abnormal inner hair cells [4]. Since OAEs may initially be present in the newborn screening, retesting for those children with suspected hearing problems or at risk children is important in order to rule out other suspected diagnoses. The hallmark of testing for AN is a positive OAE and negative or abnormal ABR [1]. An auditory brainstem response is an auditory evoked potential that is measured from the electrical activity in the brain. It is measured using electrodes placed on the head [5]. The test for auditory brainstem response shows a more in depth frequency than does the otoacoustic exam. ABR's are used to estimate severe to profound hearing loss ranges [5]. A Middle Ear Muscle Reflex (MEMR) tests how well the ear responds to loud sounds. This is a test of neural measurements through stapedial (stapedius muscle) reflex thresholds [6].

Clinical presentation of AN is typically a difficulty listening in noisy situations as well as fluctuations in the ability to hear. Other signs of AN are delayed speech and language development. There are also patients that report elevated temperatures with AN. Diagnosis of AN involves an understanding of the physiological responses of this disorder as these are key factors to be considered [1]. The main characteristics of auditory neuropathy are listed in the table below [4]. The difficulty in diagnosing AN is the ambiguity of symptoms that may be confused with other diagnoses such as auditory processing disorder, autism, and mental retardation.

Case Report

The patient is a Caucasian male, presented to CorpusCARE Family

Medicine at the age of 20 months. His parents reported frustration with his previous care and requested a second opinion. They stated that they had noticed that the child sometimes acted as he was deaf and sometimes he was able to hear. At birth, the patient became a full resuscitation and was admitted into the NICU for 7 weeks. During his time in the NICU, he had a history of multiple blood transfusions due to thrombocytopenia as well as hepatomegaly and hyperbilirubinemia. Gentamycin was given following his birth for 2 days for a total of 4 doses because of the concern that he might be septic. Although the patient had multiple complications, his otoacoustic emissions (OAE) was present when tested at 48 hours after admission to the NICU and he was not suspected as having a hearing disorder. It was observed that sometimes the patient would react to sounds and voices but at other times he would not startle and would stare into space even when there were loud sounds.

Upon presentation, the patient appeared well nourished and was observant of his mother and father. He was alert, active and was smiling. Vital signs were unremarkable. He weighed 22 lbs. and was 30.5 inches long, which placed him in the 5 to 10th percentile for height and weight. His ear exam was unremarkable with cone of light present; tympanic membranes were grey and crisp. His ear canals were non-erythemic without drainage and external ear structures anatomically correct. Developmental screening included fine motor skills, gross motor skills, personal and social milestones, and language milestones; he was appropriate for all developmental screening with the exception of speech and language skills. His vocabulary included around 10 words. According to Kliegman et al. [7], the normal vocabulary for a 20 month old child is from 20 to 50 words.

Previous labs from the pediatric ear, nose and throat (ENT) specialist were unremarkable. Studies included testing by the pediatric ENT in a soundproof booth. The patient passed this hearing screen. The pediatric ENT diagnosed otitis media with effusion and recommended equalization tube placement at 12 months old. He therefore concluded that a hearing disorder was unlikely.

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The patient was referred to UT Dallas Callier Center for Communication Disorders by this office after his initial visit. Audiological testing was ordered including an otoacoustic exam (OAE), auditory brainstem response (ABR) and a middle ear muscle reflex (MEMR). The OAE was now absent, the ABR was abnormal and the MEMR was abnormal. An MRI was ordered of his head in order to inspect the internal structures of his ears. His MRI was within normal limits and showed the presence of all normal structures bilaterally.

The following chart shows the primary tests for auditory neuropathy and the possible outcomes. The highlighted words are the actual results for this patient (Table 1) [8].

Discussion

Based on the results of his tests, the child was diagnosed with Auditory Neuropathy. The diagnosis of Auditory Neuropathy, as demonstrated by this case, can be very elusive and take multiple evaluations to discover. The patient presented in this case study had gone through multiple tests for hearing including the newborn screening in which he had OAE's present at that time and later were absent with testing at the Callier Center. Certain factors might have negatively affected the result of the patient's first OAE test. One was his hyperbilirubinemia that began 24 hours after his birth and remained until his time of discharge. Another was the gentamycin antibiotic that he had received during his stay in the NICU. Dowley et al. [2] reports that the loss of the inner hair cells of the cochlea have been found in patients with a history of hyperbilirubinemia as well as ototoxic medications such as gentamycin. Hyperbilirubin and the use of gentamycin during this time frame can both affect the results of the OAE test in newborns.

According to Smolkin et al. [9], 92% of the first OAE tests are done within the first 48 hours of age. This child had hyperbilirubinemia as well as multiple rounds of gentamycin. The pediatric ENT looked for other etiologies in order to explain the hearing deficit. The specialist wanted to rule out fluid behind the tympanic membrane as a possible cause of a decrease in hearing. He was also referred to a child psychologist to be evaluated for a possible diagnosis of autism based on the parent's descriptions of his behaviors.

The importance of future clinical assessments includes the dissemination of information that should be aimed at all primary

care providers as they are often the first line of interaction with these patients [1]. For those patients that have been given gentamycin or have had hyperbilirubinemia as an infant, testing for hearing should be recommended at a later date and parents should be informed of signs and symptoms in which to be aware. Advances in the diagnosis of AN require the ability to more accurately distinguish the variations that we observe in order to better understand the patient with AN [1]. The ultimate advantage in understanding AN will be a more informed approach in the management of those patients with this disorder [1]. A pre-audiometric triage including tympanometry and MEMR is the recommendation from Dr. Berlin et al. [3] for all the children seen in consultation.

Auditory Neuropathy is often difficult to diagnose so that timely attention leads to sooner intervention [1]. Making a differential diagnosis is sometimes cumbersome for the practitioner. In the diagnosis of AN, a thorough knowledge of the pathophysiology and usual clinical picture of the disease is needed by the practitioner in order to rule out other disease processes.

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Tests	Auditory Neuropathy
Tympanogram	✓ <i>Normal</i>
MEMR (Middle Ear Muscle Reflex)	✓ <i>Abnormal</i> or absent
OAE	✓ Present or <i>absent</i> (over time) (was present when tested in NICU and now is absent)
ABR	✓ <i>Abnormal</i> or absent
Pure-tone thresholds/audiometry	✓ <i>Mild</i> to severe loss
Word recognition (quiet)	✓ <i>Excellent</i> to poor
Word recognition (noisy)	✓ <i>Poor</i>

Table 1: Results of his tests.