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Inner Ear Disorders Associated with Hearing and Vestibular Loss: A Case Series

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[Portions of the manuscript have been redacted to ensure privacy]

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Abstract 1

Introduction: Sudden sensorineural hearing loss (SSNHL), a hearing loss of at least 30 dB at three or more consecutive frequencies within a 72-hour period, is typically idiopathic. Potential causes are thought to include viral, autoimmune, vascular, neurologic, neoplastic, or traumatic origins. Treatment consists of steroids. Poor patient follow up and compliance with rehabilitation negatively affect outcomes. Poor follow-up also results in the inadequate medical documentation of outcomes. Case Presentation: a 20-year old female presented with unilateral SSNHL with high-frequency tinnitus in the right ear. Her audiological history includes a known profound hearing loss in the left ear, as well as, occasional dizziness combined with headaches and tinnitus that are exacerbated by exercise. Discussion: Results of treatment are variable. Partial or complete recovery of audiological thresholds is more likely with timely diagnosis and treatment. Conclusion: Further research is needed to determine the causes of poor follow up and rehabilitation compliance.

Case Presentation 1

Sudden Sensorineural Hearing Loss: A Case Report

Introduction

Sudden sensorineural hearing loss (SSNHL) is characterized by objective and subjective reports of rapid onset hearing loss, often accompanied by tinnitus or dizziness (Kuhn et al., 2011; Sone et al., 2009). Systemic steroids are typically the first treatment approach and hearing amplification devices are recommended for patients with incomplete recovery of hearing acuity. Reverse sloping configurations have been associated with a better prognosis compared to high-frequency hearing loss (Kuhn et al., 2011). The prognosis of SSNHL can be difficult to track due to inadequate follow up from patients.

Case Presentation

A 20-year-old college student presented for sudden tinnitus and decreased hearing in the right ear. The high pitched tinnitus was constant and co-occurred with the hearing loss, which began four days prior. The patient had a congenital profound sensorineural hearing loss in the left ear, and an extensive history of symptoms including headaches, dizziness, slurred speech, loss of balance, and nausea that were exacerbated by exercise or leaning forward. Due to the history, this individual had routine audiological evaluations. It was reported that the most recent audiogram was completed less than a year prior and revealed normal hearing in the right ear. These results were not available for review. An audiogram from 2014 was obtained and verified normal hearing in the right ear. At that time, hearing thresholds were not measured in the left ear due to the extent of the congenital loss. Results from the 2014 audiogram are shown in Figure 1 with pure tone air conduction thresholds for the right ear. Tests for the vestibular symptoms

reportedly included two magnetic resonance imaging (MRI) tests and a computerized tomography (CT) scan; results were not available for review. The patient also stated that a diagnosis of Meniere's disease had been contraindicated. The patient denied a history of pressure equalization tubes or ear surgery.

An audiological evaluation was performed and revealed normal hearing sensitivity from 250-3000 Hz, precipitously sloping to a moderately-severe sensorineural hearing loss at 6000-8000 Hz. Figure 2 shows these results for the right ear with pure tone air and bone conduction thresholds. Tinnitus matching revealed a subjective perception of tinnitus to be at 8000 Hz at 5 dB sensation level (SL).

Management

Referral to an otolaryngologist was made. The patient was seen by an otolaryngologist later that day. She was diagnosed with sudden sensorineural hearing loss in the right ear. Treatment included oral administration of 60 mg of prednisone for 7 days. It was suggested that the patient return in 7 days for a follow-up audiologic evaluation. The results of subsequent audiologic evaluations are unknown due to a lack of follow up at the referring clinic.

Discussion

Sudden sensorineural hearing loss is uncommon, making up about 1% of all sensorineural hearing loss (Jaffe, 1973). Four thousand new cases are reported in the United States each year (Mattox & Simmons, 1977). This disorder may be preceded by an upper respiratory infection. Systemic steroids, administered through an intratympanic injection or oral tablets, are the primary treatment approach. Outcomes of steroid treatment are variable. Patients

who are treated promptly and appropriately typically have some improvement in audiologic symptoms, although complete recovery is rare. The benefits of prompt treatment for most patients qualifies SSNHL as a clinical audiologic emergency. Not all patients require treatment. Spontaneous recovery has been documented in 45-65% of cases (Byl, 1984). Medical professionals such as otolaryngologists have no way of predicting who will experience spontaneous recovery. The roles of audiologists in the treatment process include evaluating hearing thresholds after treatment, and fitting the patient with amplification if warranted. Research has shown poor follow-up and compliance with audiological rehabilitation recommendations in individuals who experienced SSNHL (Xie et al., 2019). For this case, the patient did not follow-up at the referring clinic and it is suspected that follow-up care was conducted at the otolaryngology office where the prednisone was prescribed.

Conclusion

This case demonstrates a typical presentation of sudden sensorineural hearing loss with prior vestibular symptoms and unknown recovery due to lack of follow-up with the referring clinic. The importance of compliance with follow-up recommendations should be emphasized to promote positive outcomes.

Abstract 2

Introduction: Labyrinthitis is an impairment of the hearing and vestibular areas of the inner ear. Symptoms of this deficiency include dizziness, tinnitus, and hearing loss that typically presents unilaterally. There are many potential causes of labyrinthitis including bacteria, viruses, parasites, fungi, or toxic substances. Case Presentation: A 78-year-old male sought a diagnosis and treatment for ongoing and progressive vertiginous episodes. His medical history was significant for a sudden sensorineural hearing loss, benign paroxysmal positional vertigo (BPPV), and cancer. The patient's previous treatment included physical therapy without success. Discussion: Several vestibular tests and serial audiograms over the course of multiple appointments were used to diagnose the patient. Diagnosis and treatment of vestibular disorders are essential for the prevention of falls in the elderly population. Conclusion: Continued research in the areas of diagnosis and treatment for vestibular disorders such as labyrinthitis may be helpful in improving the efficiency of diagnostic procedures and the effectiveness of treatment.

Case Presentation 2

Labyrinthitis: A Case Report

Introduction

Labyrinthitis is an infectious or inflammatory process of the inner ear (Kaya et al., 2017). The inner ear, situated in the temporal bone, is a complex maze of bone and membranes. The inner ear includes structures for hearing and balance. The location and composition of the inner ear make it difficult to examine without imaging studies. Medical professionals may use either magnetic resonance imaging (MRI) or computerized tomography (CT) to analyze the inner ear, depending on what structure they want to examine. Labyrinthitis is thought to be caused by bacteria, viruses, parasites, fungi, or toxic substances (Kaya et al., 2017). Inflammation from areas other than the inner ear, such as the middle ear and the meninges can lead to labyrinthitis (Sone et al., 2009). There are several routes for the infective agent to reach the inner ear. Pathways include the oval or round window, a pathologic fistula of the labyrinth, the cochlear aqueduct, and the internal auditory canal (Merchant & Nadol, 2010; Budenz et al., 2014). Loss of auditory and vestibular function are results of a bacterial infection in the inner ear (Merchant & Gopen, 1996). Symptoms can progress for months after the onset of the disease (Sone et al., 2009).

Case Presentation

The patient, a 78 year old male, originally presented 2019 for audiometric and videonystagmography (VNG) testing with complaints of brief and sporadic vertiginous episodes that began five months prior. These episodes were reported to last for a couple of seconds, followed by feeling “off” for a couple of minutes. There was no warning before an episode, and

they occurred when sitting or standing. The patient reported that stationary objects occasionally moved in his field of vision. Tinnitus, decreased hearing, significant head trauma, history of head and neck surgeries, and the use of alcohol or tobacco products prior to the appointment were denied. The patient reported consuming four cups of coffee within 24 hours of the appointment.

The patient's history was significant for a sudden sensorineural hearing loss in 2015, according to patient report. Audiometric thresholds prior to the sudden hearing loss were not available. It is thought that the high frequency sensorineural hearing loss in the left ear as shown in Figure 3 is the permanent result of the sudden hearing loss. He was seen by a neurologist in 2017, at which time a neurological problem was contraindicated. In 2018, the patient reported an episode of vertigo and disequilibrium. Following that, an otolaryngologist was consulted who reportedly diagnosed the patient with right-sided vestibular problems. The patient was treated with physical therapy without improvement. His history also includes a recent diagnosis of BPPV. Documentation revealing the date of this diagnosis and treatments used were not available for review. Reports indicate the BPPV was resolved in March 2019.

Audiometric testing completed in 2019 was asymmetric and revealed normal sloping to severe sensorineural hearing loss in the left ear with a severe to profound sensorineural hearing loss in the right ear. Results of the VNG revealed a complete right-sided caloric weakness. Figure 4 displays the bilateral bi-thermal air caloric results with a typical response in the left ear, and no response in the right ear.

Computerized dynamic posturography (CDP) was conducted in 2019 to evaluate for abnormalities in the patient's use of the three sensory systems that contribute to postural control: somatosensory, visual, and vestibular; as well as his brain's ability to integrate these inputs. All results from this procedure were in the normal range.

The patient returned to the clinic in 2019 for audiometric and cervical vestibular evoked myogenic potential (cVEMP) testing. At that time, changes in hearing and tinnitus were denied. Changes in medical history included a diagnosis of lung cancer that was treated with radiation, a "tiny spot" on his cerebellum and the right frontal lobe for which treatment is unknown, recently experienced drop attacks, and a change in vertigo. Audiometric thresholds are displayed in Figure 3 with bilateral air and bone conduction thresholds. These were consistent with results from April 2019. The cVEMP results were within normal limits, but the morphology of the right waves were difficult to replicate. The evaluating clinician suggested interpreting these results with caution. The VNG, audiogram, and cVEMP responses were indicative of right-sided labyrinthitis.

Management

Regular audiologic evaluations were recommended to monitor for changes in hearing acuity. The recommendation of a BiCROS hearing system trial was made to address his asymmetric hearing loss. The patient declined amplification. He was encouraged to continue with physical therapy to minimize the effects of the vertiginous episodes. Follow up with medical professionals as directed was suggested due to his extensive medical history.

Discussion

Labyrinthitis is a progressive inflammation that can present with various clinical symptoms. Several tests, including comprehensive audiometry, VNG, CDP, and cVEMPs were required to obtain all the relevant information for a proper diagnosis. Each test evaluated a different aspect of the inner ear with each result providing more information that eventually

differentiated the diagnosis. There were several components to the VNG, each one analyzing different structures and functions. Ocular motor testing was used to identify potential ocular motor disorders or a central pathology that was contributing to the patient's symptoms. Positional testing examined the stationary position holding and the gravitational orientation of the vestibular system. The last subtest of the VNG, caloric irrigation, separately stimulated both inner ears to compare each ear's response to the contralateral ear. Ice water caloric irrigation in the right ear is another evaluation that would be of interest when required equipment is available. Additionally, ice water calorics may elicit some response from the horizontal semicircular canal when a response cannot be obtained with warm or cool water stimulation. The ability to use the three systems that contribute to balance was assessed with CDP. The patient's saccule and inferior portion of the vestibular nerve were assessed during the cVEMP. Although the battery was timely, each evaluation contributed meaningful information.

Accurate diagnosis of labyrinthitis is important for establishing an effective treatment plan. Dizziness occurs more commonly in the elderly population. Management of vestibular disorders can prevent falls which is especially important in people with advanced age. Accidents during old age can be devastating and result in severe injuries that require timely and expensive rehabilitation. For these reasons, diagnosis and treatment of vestibular disorders are priorities for audiologists that ultimately aim to promote a high quality of life.

Conclusion

This case demonstrates a complex and long-developing case of right-sided labyrinthitis. An extensive test battery was required to determine the proper diagnosis. Management included a recommended trial of amplification and continued vestibular rehabilitation physical therapy.

Further research may be beneficial for improving diagnostic and treatment techniques in patients with labyrinthitis.

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Figure 1

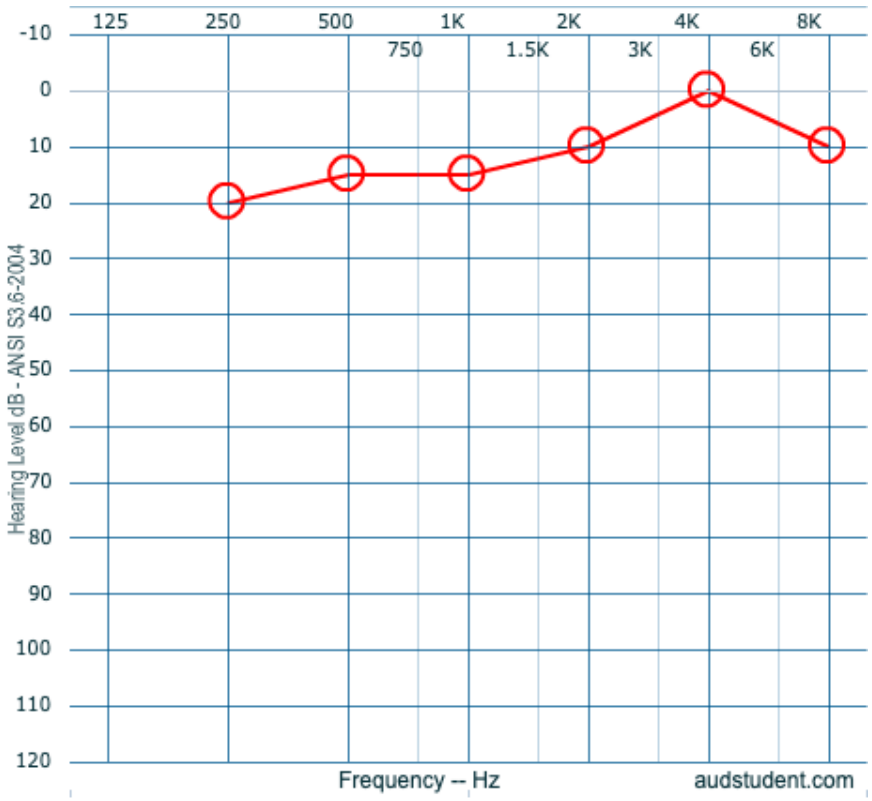

Figure 1. Audiometric thresholds from 2014.

Figure 2

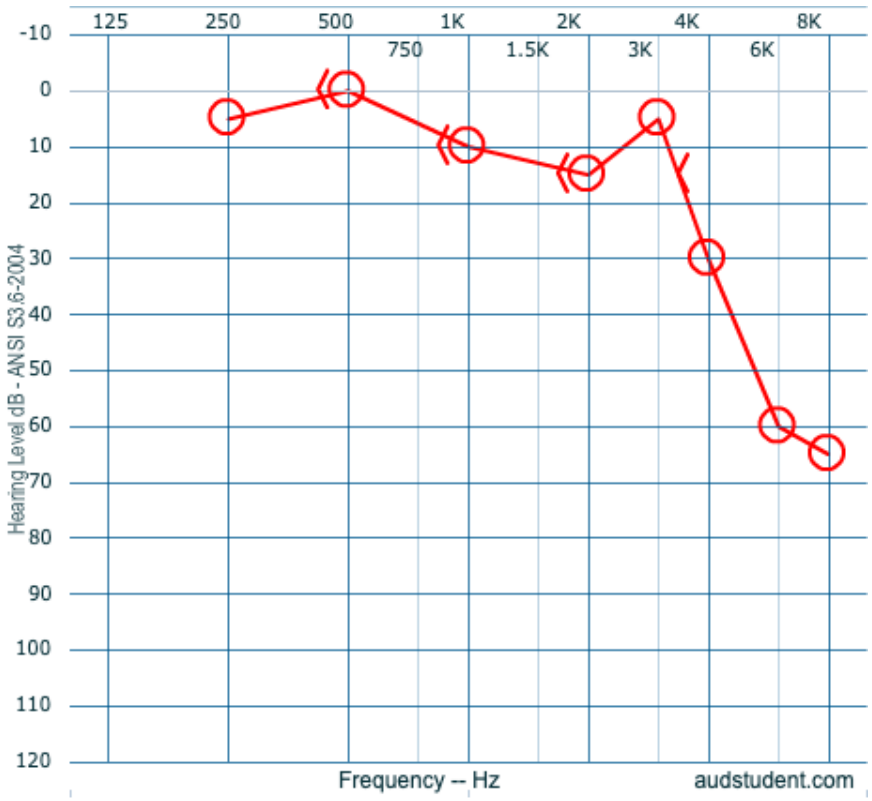

Figure 2. Audiometric thresholds after a sudden sensorineural hearing loss.

Figure 3

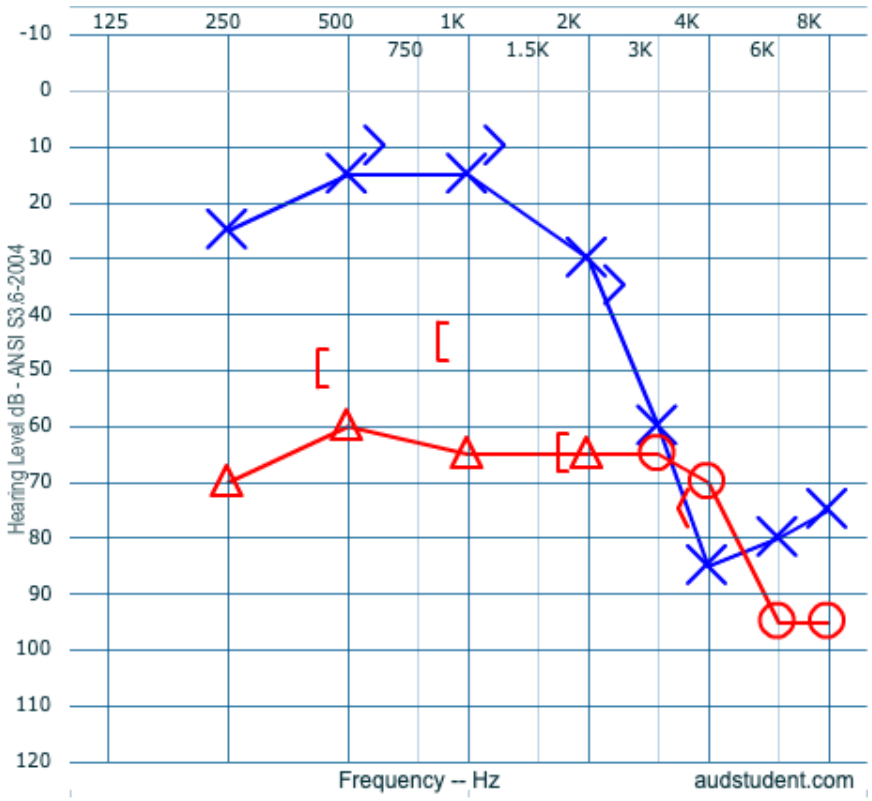

Figure 3. Pure tone air and bone conduction thresholds from July 2019.

Figure 4

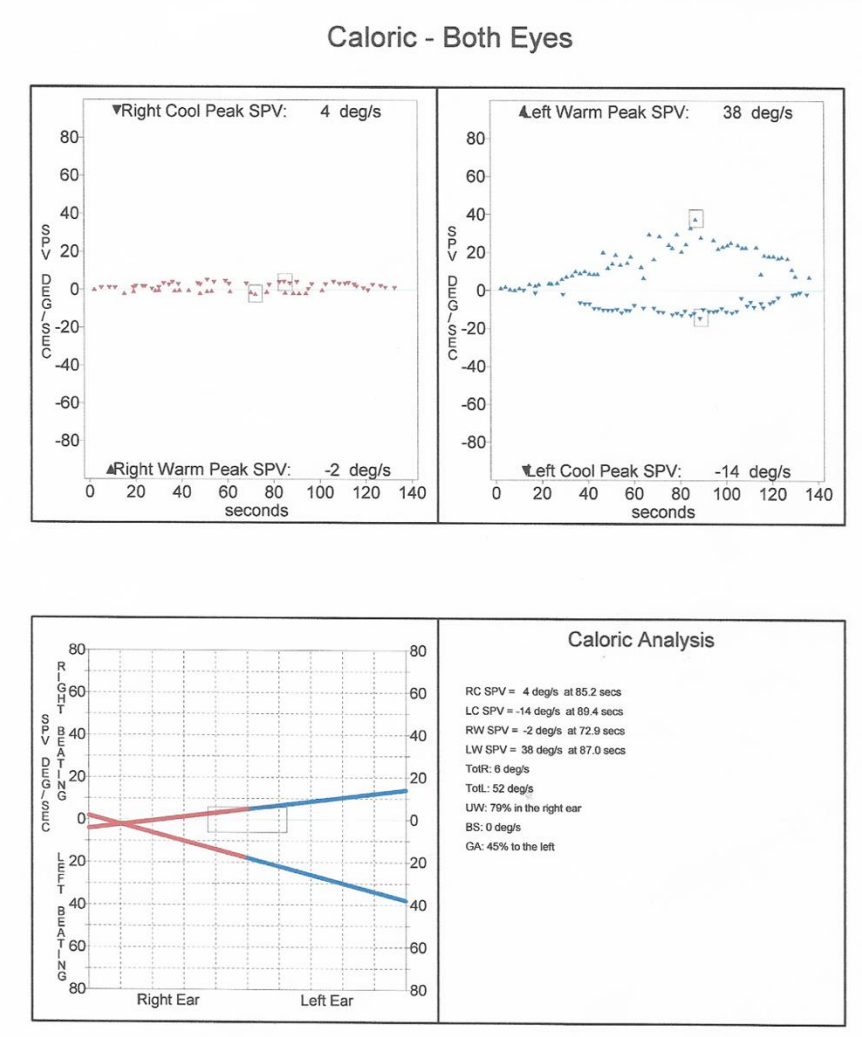

Figure 4. Air caloric irrigation results from April 2019.