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Article

Silent Vestibulopathy in Asymmetric Hearing Loss Can Be a Sign of a Cerebellopontine Angle Tumor

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Abstract: The presence of an ipsilateral cerebellopontine angle (CPA) tumor should be ruled out in patients with asymmetric sensorineural hearing loss (ASNHL). Although many patients with CPA tumors have ipsilateral vestibular hypofunction, some of them do not experience dizziness even with ipsilateral vestibular hypofunction. We analyzed the incidence of CPA tumors among patients with ASNHL without subjective dizziness based on the presence of vestibulopathy. We hypothesized that a patient with silent unilateral vestibular hypofunction (UVH) is more likely to be diagnosed with a CPA tumor. Among 157 subjects who underwent MRI for ASNHL, those who did not have "subjective dizziness" were selected. All subjects underwent hearing and vestibular function tests. UVH was diagnosed if canal paresis $\geq$ 25%, positive head-shake nystagmus, or gain of head-impulse test < 0.8 were detected. The diameters of the CPA tumors were measured along the petrosal ridge on the axial plane of MRI. Among the enrolled subjects, 44 (28.02%) were diagnosed with a CPA tumor. The 37 patients (84.1%) with a CPA tumor had silent UVH, while only 33 of the 113 patients (29.2%) without a CPA tumor had silent UVH (chi-square test, odds ratio = 12.8, $p < 0.001$). Silent UVH in patients with ASNHL may be a sign of a CPA tumor.

Keywords: asymmetric sensorineural hearing loss; cerebellopontine angle tumors; unilateral vestibular hypofunction

1. Introduction

Asymmetric sensorineural hearing loss (ASNHL) is characterized by a binaural difference at more than one frequency. The etiological diagnosis of ASNHL is extensive, and a precise diagnosis can be elusive. ASNHL can be caused by cochlear disorders and injuries, including noise exposure and a retrocochlear disorder. Patients with vestibular schwannomas (VSs) present with ASNHL 85–97% of the time [1–4]. The possibility of an ipsilateral cerebellopontine angle (CPA) tumor should always be considered in patients with ASNHL. Although CPA tumors are benign, their growth may result in significant morbidity and mortality if left untreated. Earlier detection of these tumors has resulted in a smaller tumor size at the time of diagnosis, which has significantly reduced the morbidity of treating CPA tumors.

Although gadolinium-enhanced magnetic resonance imaging (MRI) is the current gold standard for diagnosing CPA tumors, not all patients with ASNHL can directly undergo MRI as the primary screening procedure because of the significant cost of this examination. Cost effectiveness must be

considered when determining who should undergo MRI evaluation. The use of MRI selectively for patients with a high probability of having a CPA tumor based on clinical suspicion is more cost effective.

Approximately 5–10% of all intracranial tumors develop in the CPA [5], and 60–90% of CPA tumors are VSs [6]. Meningioma of the CPA is the second-most common neoplasm, constituting 10–15% of all masses at this site [7]. CPA tumors are manifested by cranial neuropathies related to their location. Despite that VSs typically originate in the inferior vestibular nerve (62–91%), followed by the superior vestibular nerve (6–38%) [8–10], the rate of vertigo reported in patients with a VS is variable, ranging from 19 to 61% [2,4,11–13]. Clinical manifestations of a CPA meningioma do not differ significantly from those of a VS [14], and many patients do not experience vertigo.

The slow-growing nature of most CPA tumors and relatively large area for tumor expansion lead to slowly progressing impairment in vestibular function. This slowly progressing dysfunction allows for gradual central vestibular compensation. We surmised that this compensation minimizes symptoms, such as acute vertigo or dizziness, in patients with a CPA tumor who may have ipsilateral vestibular hypofunction.

Evaluation of vestibular function is an important step in the evaluation of dizziness to know if the cause of dizziness is vestibular origin. The caloric test and head impulse test (HIT) are valuable methods for determining unilateral or bilateral vestibular hypofunction by measuring the vestibulo-ocular reflex (VOR) in response to horizontal semicircular canal stimulation. Head-shake nystagmus (HSN) is also a valuable screening tool for detecting vestibular asymmetry [15]. Moreover, the importance of examining for vestibular dysfunction extends beyond merely diagnostics in CPA tumors, being also meaningful for identifying in these patients both from a multisensory deficit and balance prognostic factors, and likewise, it provides them with the opportunity to rehabilitate it with therapy.

However, there is a lack of specificity and sensitivity with these screening procedures [16], as small tumors may result in false-negative results, and the diversity of peripheral vestibular disorders may result in false-positive results.

We hypothesized that subjects with silent unilateral vestibular hypofunction (UVH) are more likely to be diagnosed with a CPA tumor. To test this hypothesis, we analyzed the incidence of CPA tumors based on the presence of ipsilateral vestibulopathy among patients with ASNHL without a history of dizziness.

2. Materials and Methods

This study protocol was approved by Seoul National University Bundang Hospital Internal Review Board (No. B-1604-343-111). We adopted a cross-sectional design, and this study is based on a retrospective analysis of the patients' charts. Therefore, this study did not result in any risk to subjects, and patient consent was not required.

Data from patients > 18-years of age who underwent brain MRI from January 2013 to June 2015 were collected from an electronic medical record database using the search words (1) brain MRI, (2) pure tone audiometry, and (3) vestibular function tests (VFTs) including HSN, bithermal caloric test, and video HIT (v-HIT). A total of 439 patients were retrieved by this search procedure. Among them, 91 patients with a history of dizziness were excluded. ASNHL was defined as a $\geq$ 15 dB difference at one or more frequencies with no conductive component or $\geq$ 15% difference in speech discrimination scores (SDS) between ears [17,18]. Of the 348 patients, 157 met the ASNHL criteria and were finally included in our study. The study group comprised 69 males and 88 females (mean age, 56.2 $\pm$ 13.2 years; range, 19–91 years).

2.1. Audiometry and Tumor Size

Pure tone audiograms were analyzed based on the guidelines suggested by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology—Head and Neck Surgery [19]. The results of the audiometric assessment were available at frequencies of 250, 500, 1000, 2000, 3000, 4000, and 8000 Hz. Thresholds of 0.5, 1, 2, and 3 kHz were used to calculate the pure tone average

(PTA). The mean hearing threshold (MHT) was calculated separately for all measured frequencies. Speech audiometry, including measurement of speech discrimination at the suprathreshold, was also recorded.

Radiological assessment of tumor size was performed by MRI using 2–3 mm axial slices. Tumor size was determined by measuring the maximum diameters of the extracanalicular tumors, which were parallel to the petrous ridge. The size of an intracanalicular tumor limited to the internal auditory canal was measured along the long axis of the canal. The MRI results were categorized as intracanalicular, small (<1.5 cm), medium (1.5–3.0 cm), and large (>3.0 cm) tumors.

2.2. Videonystagmography (VNG)

The slow phase velocities of spontaneous nystagmus (SN) and HSN were measured using a VNG system (Chartr ENG; ICS Medical, Schaumburg, IL, USA) with the subject in the sitting position. HSN was assessed after 15 s of passive head shaking with 30° neck flexion at a frequency of 2 Hz. After subtracting the amount of SN, the direction and intensity of the horizontal component of corrected HSN were obtained as parameters for analysis. The criteria of HSN were defined as having been met when the following values were exceeded: horizontal $\geq 3°/s$; vertical $\geq 2°/s$; torsional $\geq 2°/s$), and when the nystagmus lasted ≥ 5 s [20].

2.3. Bithermal Caloric Test

The bithermal caloric test was performed in the supine position with upward head flexion of 30°. Eye movements were recorded using a binocular video oculography system (I-Portal®, Neuro Kinetics Inc., Pittsburgh, PA, USA) to track horizontal eye movements. Caloric irrigation was performed using binaural alternate irrigation for 30 s with 30 s with 300 mL of cold (30 °C) and warm (44 °C) water (ICS NCI-480®, GN Otometrics, Taastrup, Denmark). The interval between individual irrigations was at least 5 min. Percent canal paresis (CP) was calculated using the Jongkees formula. The result was considered abnormal or UVH when the CP score was >25%.

2.4. v-HIT

The examinees were instructed to stare at a stationary target 1 m in front of them, while short-lasting head rotations around an earth-vertical axis were randomly applied from behind the examinees. A video oculography system was used for acquisition and analysis of eyeball and head movements (ICS impulse®, GN Otometrics). The test was repeated at least 10 times on each side in an unpredictable direction of 5–10° and peak acceleration of 750–6000°/s^2. Only head rotations with a defined waveform were accepted within the predefined velocity and acceleration window. Movements of the right eyeball and head were recorded. The areas under the velocity curves of these two movements were obtained from head-impulse onset to the back crossing of zero. The VOR gain was defined as the ratio of the area under the velocity curve of the right eye to that of the head (http://www.icsimpulse.com). The result was considered abnormal or UVH when the VOR gain was <0.8 [21].

2.5. Statistical Analyses

Statistical analyses were performed using SPSS statistical software (SPSS® v19.0, IBM, Armonk, NY, USA). The data are presented as means ± standard deviation. We used Pearson's chi-square test for 2 × 2 tables to compare sex, tumor side, and presence of UVH between the tumor and non-tumor groups. Mean ages, tumor sizes, asymmetries of SNHL, and asymmetries of SDS between the two groups were compared using the unpaired *t*-test. Comparisons of mean asymmetries of SNHL (MASNHL) at each frequency in the CPA tumor group were analyzed using the paired *t*-test. A p-value < 0.05 was considered significant.

3. Results

Of the 157 patients, CPA tumors were identified in 44 patients (18 males and 26 females), and CPA tumors were absent in the remaining 113 patients (51 males and 62 females). The mean age of the patients without CPA tumors (the non-tumor group) was 57.8 ± 13.3 years, which was significantly higher than that of the patients with CPA tumors (the tumor group) (52.3 ± 12.3 years; $p = 0.019$).

3.1. Characteristics of CPA Tumors

A VS was the most common CPA tumor and accounted for the majority of the abnormal MRI findings ($n = 39$, 88.6%), representing 24.8% of all MRI scans performed. The next most common tumor ($n = 3$, 6.8%) was meningioma, which represented 1.9% of all MRI results evaluated. The remaining CPA tumors detected were a facial nerve schwannoma ($n = 1$, 2.3%) and a lower cranial nerve schwannoma ($n = 1$, 2.3%).

The mean tumor size was 2.1 ± 1.2 cm (range, 0.2–5.5 cm). The tumor was intracanalicular in one patient (2.3%), small (<1.5 cm) in 12 patients (27.3%), medium (1.5–3.0 cm) in 23 patients (52.3%), and large (>3.0 cm) in eight patients (18.2%). In the CPA tumor group, 25 were on the right side and 19 on the left side.

3.2. Audiological Results

The patients with CPA tumors had a mean PTA of 49.9 ± 28.0 dB, whereas those without a tumor had a mean PTA of 61.0 ± 28.5 dB. The patients in the tumor group had a better speech discrimination score than did those in the non-tumor group (57.0 ± 38.4% vs. 42.0 ± 39.5% of the mean SDS, respectively). Significant differences were observed in the mean PTA and mean SDS between the tumor and non-tumor groups ($p = 0.029$ and 0.033, respectively).

The MHT for the affected ear and the MASNHL, which is the difference in the hearing threshold by frequency between the two ears, are represented in Table 1. Figure 1 shows the estimated MASNHL by frequency. In the CPA tumor group, MASNHL at mid-to-high frequencies (1000–8000 Hz) were significantly higher than MASNHL at low frequencies (250–500 Hz) ($p < 0.05$). In particular, the tumor group showed the highest asymmetries at frequencies of 3000 and 4000 Hz. On the other hand, there were no significant differences in MASNHL within frequencies in the non-tumor group.

Table 1. Pure tone audiometry results from the affected side according to tumor presence.

Frequencies (Hz)	MHT (dB)		MASNHL (dB)		*p*-Value *
	CPA Tumor (*n* = 44)	**Non-Tumor (*n* = 113)**	**CPA Tumor (*n* = 44)**	**Non-Tumor (*n* = 113)**	
250	34.3 ± 31.3	50.5 ± 31.7	25.9 ± 29.6	42.4 ± 31.6	0.003
500	36.4 ± 32.2	58.1 ± 31.5	27.1 ± 31.1	48.4 ± 32.3	<0.001
1000	48.4 ± 31.9	62.3 ± 31.4	35.8 ± 31.2	50.0 ± 32.4	0.013
2000	54.8 ± 31.0	60.9 ± 29.2	40.11± 31.3	47.4 ± 30.1	0.180
3000	59.9 ± 31.0	62.6 ± 29.0	45.2 ± 32.2	45.6 ± 29.9	0.949
4000	62.2 ± 30.1	68.1 ± 28.7	45.9 ± 31.1	47.5 ± 29.7	0.763
8000	71.9 ± 31.3	82.2 ± 25.4	43.0 ± 28.5	46.9 ± 29.0	0.447

* MASNHL difference between two groups, unpaired *t*-test. Numbers in bold are statistically significant. Abbreviations: MHT, mean hearing threshold; MASNHL, mean asymmetries of sensorineural hearing loss.

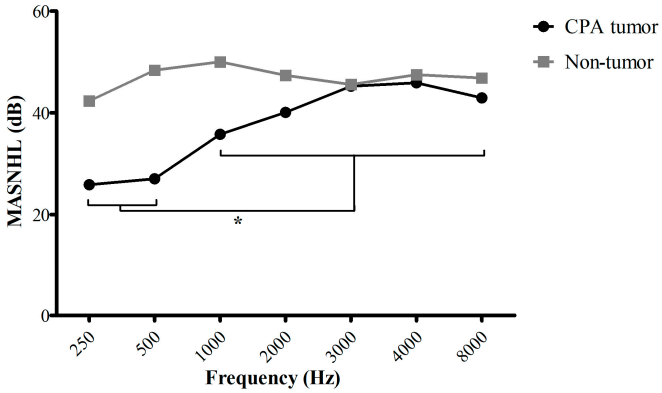

Figure 1. MASNHL by frequency in the CPA tumor and non-tumor groups. * Significant difference between two frequencies in the CPA tumor group, $p < 0.05$. MASNHL, mean asymmetries of sensorineural hearing loss; CPA, cerebellopontine angle. Abbreviations: MASNHL, mean asymmetries of sensorineural hearing loss; CPA, cerebellopontine.

3.3. VFTs

Of the 157 patients, 82 underwent the caloric test, 107 underwent VNG, and 52 underwent v-HIT. Seventy patients who showed at least one abnormal test result were diagnosed with UVH. All vestibular deficits were on the same side as the ear with hearing loss.

The percentage of UVH in the CPA tumor group was 84.1%, compared with 29.2% in the patients without a CPA tumor (Table 2). Silent UVH was a significantly reliable marker of a CPA tumor in patients with ASNHL (odds ratio = 12.8, $p < 0.001$).

Table 2. Association between CPA tumors and vestibular function.

UVH	No. Patients (%)		*p*-Value *	OR (95% CI)
	CPA Tumor	Non-Tumor		
Positive	37 (84.1%)	33 (29.2%)		
Negative	7 (15.9%)	80 (70.8%)	<0.001	12.8 (5.2–31.6)
Total	44	113		

* Pearson's chi-square test. Abbreviations: UVH, unilateral vestibular hypofunction; CPA, cerebellar pontine angle; OR, odds ratio; CI, confidence interval.

The size of the tumor was significantly associated with UVH in the CPA group ($p = 0.027$), whereas it was not associated with the degree of hearing loss or SDS ($p = 0.743$ and 0.406, respectively). The mean tumor size was 2.2 ± 1.2 cm in the 37 patients with UVH and 1.5 ± 0.5 cm in the 7 patients without UVH. However, other variables, including age, sex, PTA, and SDS, were not associated with UVH.

4. Discussion

In this study, we investigated whether vestibular dysfunction based on VNG, the caloric test, and the v-HIT were predictive of CPA tumors in patients with ASNHL not experiencing dizziness. We found that a significant increase in the rate of positive MRI results was associated with silent UVH. In this specific study group, the sensitivity and specificity of the VFT battery were 84.1% (37/44) and 70.8% (80/113), which were comparable with those of the auditory brainstem response (71% sensitivity and 74% specificity) for evaluating ASNHL [22].

According to the International Classification of Diseases-10, as the standard diagnostic tool, unilateral SNHL can be used to indicate a diagnosis of SNHL in only one ear, with unrestricted hearing on the contralateral side. Patients with hearing problems present more commonly with asymmetric

hearing loss rather than unilateral hearing loss. However, there are no standard criteria for defining ASNHL. While CPA tumors need to be ruled in adults presenting with asymmetric SNHL, this is not common; nor is the case in pediatric asymmetric hearing loss, in which cochlear nerve aplasia, congenital CMV and cochleovestibular anomalies predominate as the etiology in this type of hearing loss. Therefore, the incidence of CPA tumors in patients with ASNHL varies widely from 2.1% to 7.7%, depending on the definition of ASNHL [22,23]. In this study, to take into account all of the different definitions used in previous studies [17,22–25], ASNHL was defined as a 15 dB difference at any single frequency or a 15% difference in SDS between ears. As our current study was retrospective, the smaller asymmetries for defining ASNHL do not imply less strict criteria for undergoing MRI.

According to the definition of ASNHL proposed above, the current study showed that 44 (28%) of 157 cases with ASNHL had CPA tumors. This value is almost four-fold the highest rate reported in previous studies. Such a high prevalence of CPA tumors in our current study may be because patients experiencing dizziness were excluded from the study. In fact, CPA tumors are exceedingly rare in the general population. Considering VS, which accounts for 90% of CPA tumors, the realistic annual incidence was recently suggested to be 0.0013% of the general population [26]. Therefore, the decision to perform MRI to diagnose a CPA tumor should be based on clinical symptoms and historical features. The most common symptom of VS is asymmetric hearing loss. At the time of diagnosis, 94% of patients present with hearing loss [27]. Conversely, the 1-year incidence of CPA tumors also increases to 8.3% in subjects with ASNHL [22].

In contrast, the incidence of vertigo decreases in these subjects. Cueva (2004) prospectively evaluated 312 patients with ASNHL, including 24 cases of VS and 2 cases of glomus jugulare tumors; none of these patients presented with both a CPA tumor and vertigo [22]. An epidemiological study reported that the 1-year prevalence rates of vertigo and dizziness at the population level are 48.3% and 35.6% [28]. Vestibular hypofunction is exceedingly common in individuals with hearing loss period (prevalence ranges from 20–70% in the literature), and rarely are these individuals dizzy, with the exception only of those that experience a rapid change in vestibular function (i.e., due to cochleovestibular loss). Because dizziness and vertigo are so prevalent in the general population, the absence of vertigo in subjects with ASNHL is strongly associated with VS [17,22]. Saliba et al. reported that the risk of developing VS reaches 86% in patients with ASNHL who do not experience vertigo.

The current study showed that the mean patient age was significantly higher in the non-tumor group than in the CPA tumor group, although the difference was small (5.5 years). In the previous large-scale prospective study by Cueva, the mean age of patients with negative MRI findings was 53.6 years, similar to but slightly younger than that in our study (57.8 $\pm$ 13.3 years) [22]. In addition, Kentala et al. reported that the mean age of the tumor group at the onset of symptoms was 47 years [27]. One of the factors contributing to the older age distribution of the non-tumor group may be the reason for undergoing brain MRI. Older patients may have received MRI for reasons other than an ASNHL work-up, such as a routine health check-up. This is supported by a recent report that asymmetric hearing loss is common in the older population and not indicative of retrocochlear pathology [29].

Asymmetries in the tumor group were greater at mid-to-high frequencies (1000–8000 Hz) than at low frequencies (250–500 Hz). Greater asymmetries were also seen at frequencies of 3000 and 4000 Hz than 1000 Hz. Similarly, Saliba et al. suggested that the asymmetry at 3000 Hz is the most representative value of all frequencies between 250 and 4000 Hz [17]. Table 1 also shows that the pattern of hearing loss differed between the two groups. The non-tumor group had a relatively flat type of hearing loss, and higher frequencies were more affected than lower frequencies in the tumor group. Several studies have also shown that patients with VS have high-tone hearing loss [30–32]. The middle- and high-frequency cochlear nerve fibers lie on the outer surface [33]. In addition, the cisternal segment of the cochlear nerve has tonotopic organization, so high frequencies are closer to the internal acoustic meatus [34]. These anatomical findings support that tumor-related compression neuropathy can cause mid-to-high frequency ASNHL.

Our results showed that the presence of UVH was significantly correlated with tumor size (p = 0.027). Similarly, Kentala et al. reported that tumor size affects performance on the VFT [27]. Although VS originates from the inferior vestibular nerve (IVN) in more than 60% of cases [8–10], both studies used VFT techniques only to assess the horizontal semicircular canals that are innervated by the superior vestibular nucleus (SVN), so small tumors arising from the IVN do not compress the SVN sufficiently, and may present normal function. Thus, the additional use of other techniques to assess IVN lesions, such as vestibular-evoked myogenic potential (VEMP), would aid the early diagnosis of CPA tumors. Ushio et al. examined the diagnostic value of the VEMP in patients with VS and suggested that VEMP is more sensitive for detecting small tumors than is the caloric test [35]. He et al. also reported that tumors originating from the IVN are smaller than those originating from the SVN [10]. v-HIT, as well as scleral search coils, are presently accepted as the gold standard for detecting vertical canal dysfunction, but at the time of the study, we did not have a verified normal VOR gain value for vertical canals. In a future study, we will further evaluate the efficacy of detecting small tumors and determining the nerve of VS tumor origin using v-HITs.

In this study, patients in the non-tumor group had higher hearing thresholds and lower speech discrimination scores than did those in the tumor group. Since the 1960s, audiometric data have suggested that poor speech discrimination is associated with retrocochlear disorders such as CPA tumors [36,37]; thus, our findings seem to be in contrast to those of previous reports. However, our audiometric results in the tumor group were similar to those of recent studies [22,31,32]. Harner et al. collected audiometric data from a large group of patients (n = 619) with VS with a mean age and mean tumor size comparable to those in our patients and showed a similar level of sloping hearing loss (MHT varied from 35.7 $\pm$ 25.6 dB at 500 Hz to 65.8 $\pm$ 33.7 dB at 4000 Hz) with serviceable word recognition (51.8 $\pm$ 38.2%) [32]. Van dijk et al. (2000) suggested that since the advent of MRI, more VSs have been detected prior to severe hearing deterioration [31]. Therefore, this discrepancy between our data and others may be explained by the worse-than-expected results in the non-tumor groups of earlier studies. Only 11% of our cases in the non-tumor group had bilateral asymmetric hearing loss, whereas 89% had purely unilateral SNHL. On the other hand, 58% of patients without a tumor had bilateral/asymmetric hearing loss in the prospective study by Cueva. Perhaps it is because in clinical practice, MRIs are ordered for patients with greater asymmetry than the inclusion criteria applied in this study of 15 dB/15%. Such a population bias due to the retrospective study design might have contributed to the relatively significant hearing deterioration and poor speech discrimination in the non-tumor group.

This study presented a relatively large population study in order to analyze the audiometric, imaging, and vestibular results in patients with ASNHL. However, the limitation of this study deserve discussion. The cause could only be ascertained in the CPA tumor group (28%). For most patients with ASNHL in the non-tumor group, the cause remained unclear. To confirm other etiologies related to ASNHL in the non-tumor group, additional investigations, including genetic examinations and immunological tests, might be performed. Atturo et al. reported that 66.7% of the patients with unilateral SNHL were recognized as immune-mediated inner ear disease [38]. However, genetic examination might not be valuable in determining the cause of ASNHL. To date, there are no genetic mutations reported to be specifically associated with unilateral hearing loss [39].

5. Conclusions

In patients showing asymmetric hearing loss without history of vertigo, the odds ratio of CPA tumor was 12.8 if the subjects have ipsilateral vestibular hypofunction. Hidden vestibular hypofunction may be a clue to suspect cerebellopontine angle tumor in unilateral hearing loss patients. This study suggested an important role for vestibular evaluations in predicting CPA tumors in patients with unilateral sensorineural hearing loss without a history of vertigo.

Author Contributions: W.S., Y.J.J. and J.-W.K. conceived and designed the study concepts; W.S., Y.J.J., H.G.P. and Y.S. acquired the data; W.S. and Y.J.J. analyzed the data; B.Y.C. and J.-J.S. contributed the critical revision of the manuscript for important intellectual content; W.S. and Y.J.J. wrote the paper. J.-W.K. supervised the study. All authors reviewed the manuscript.

Conflicts of Interest: The authors declare no conflict of interest.

References

1. Moffat, D.A.; Baguley, D.M.; von Blumenthal, H.; Irving, R.M.; Hardy, D.G. Sudden deafness in vestibular schwannoma. *J. Laryngol. Otol.* **1994**, *108*, 116–119. [CrossRef] [PubMed]
2. Selesnick, S.H.; Jackler, R.K.; Pitts, L.W. The changing clinical presentation of acoustic tumors in the mri era. *Laryngoscope* **1993**, *103*, 431–436. [CrossRef] [PubMed]
3. Mathew, G.D.; Facer, G.W.; Suh, K.W.; Houser, O.W.; O'Brien, P.C. Symptoms, findings, and methods of diagnosis in patients with acoustic neuroma. *Laryngoscope* **1978**, *88*, 1893–1903. [CrossRef] [PubMed]
4. Matthies, C.; Samii, M. Management of 1000 vestibular schwannomas (acoustic neuromas): Clinical presentation. *Neurosurgery* **1997**, *40*, 1–9. [PubMed]
5. Smirniotopoulos, J.G.; Yue, N.C.; Rushing, E.J. Cerebellopontine angle masses: Radiologic-pathologic correlation. *Radiographics* **1993**, *13*, 1131–1147. [CrossRef] [PubMed]
6. Heier, L.A.; Comunale, J.P., Jr.; Lavyne, M.H. Sensorineural hearing loss and cerebellopontine angle lesions. Not always an acoustic neuroma—A pictorial essay. *Clin. Imaging* **1997**, *21*, 213–223. [CrossRef]
7. Roser, F.; Nakamura, M.; Dormiani, M.; Matthies, C.; Vorkapic, P.; Samii, M. Meningiomas of the cerebellopontine angle with extension into the internal auditory canal. *J. Neurosurg.* **2005**, *102*, 17–23. [CrossRef] [PubMed]
8. Komatsuzaki, A.; Tsunoda, A. Nerve origin of the acoustic neuroma. *J. Laryngol. Otol.* **2001**, *115*, 376–379. [CrossRef] [PubMed]
9. Khrais, T.; Romano, G.; Sanna, M. Nerve origin of vestibular schwannoma: A prospective study. *J. Laryngol. Otol.* **2008**, *122*, 128–131. [CrossRef] [PubMed]
10. He, Y.B.; Yu, C.J.; Ji, H.M.; Qu, Y.M.; Chen, N. Significance of vestibular testing on distinguishing the nerve of origin for vestibular schwannoma and predicting the preservation of hearing. *Chin. Med. J.* **2016**, *129*, 799–803. [PubMed]
11. Edwards, C.H.; Paterson, J.H. A review of the symptoms and signs of acoustic neurofibromata. *Brain* **1951**, *74*, 144–190. [CrossRef] [PubMed]
12. Erickson, L.S.; Sorenson, G.D.; McGavran, M.H. A review of 140 acoustic neurinomas (neurilemmoma). *Laryngoscope* **1965**, *75*, 601–627. [CrossRef] [PubMed]
13. Morrison, G.A.; Sterkers, J.M. Unusual presentations of acoustic tumours. *Clin. Otolaryngol. Allied Sci.* **1996**, *21*, 80–83. [CrossRef] [PubMed]
14. Su, C.H.; Chen, C.M.; Young, Y.H. Differentiating cerebellopontine angle meningioma from schwannoma using caloric testing and vestibular-evoked myogenic potentials. *J. Neurol. Sci.* **2013**, *335*, 155–159. [CrossRef] [PubMed]
15. Morawiec-Bajda, A.; Brzydlo-Golinska, G. head shaking test (HSN) using to diagnose vestibule damage. *Otolaryngol. Pol.* **2006**, *60*, 407–413. [PubMed]
16. Gianoli, G.J.; Soileau, J.S. Acoustic neuroma neurophysiologic correlates: Vestibular-preoperative, intraoperative, and postoperative. *Otolaryngol. Clin. N. Am.* **2012**, *45*, 307–314. [CrossRef] [PubMed]
17. Saliba, I.; Martineau, G.; Chagnon, M. Asymmetric hearing loss: Rule 3,000 for screening vestibular schwannoma. *Otol. Neurotol.* **2009**, *30*, 515–521. [CrossRef] [PubMed]
18. Ahsan, S.F.; Standring, R.; Osborn, D.A.; Peterson, E.; Seidman, M.; Jain, R. Clinical predictors of abnormal magnetic resonance imaging findings in patients with asymmetric sensorineural hearing loss. *JAMA Otolaryngol. Head Neck Surg.* **2015**, *141*, 451–456. [CrossRef] [PubMed]
19. Monsell, E.M. New and revised reporting guidelines from the committee on hearing and equilibrium. American academy of otolaryngology-head and neck surgery foundation, inc. *Otolaryngol. Head Neck Surg.* **1995**, *113*, 176–178. [CrossRef]
20. Huh, Y.E.; Koo, J.W.; Lee, H.; Kim, J.S. Head-shaking aids in the diagnosis of acute audiovestibular loss due to anterior inferior cerebellar artery infarction. *Audiol. Neurootol.* **2013**, *18*, 114–124. [CrossRef] [PubMed]

21. MacDougall, H.G.; Weber, K.P.; McGarvie, L.A.; Halmagyi, G.M.; Curthoys, I.S. The video head impulse test: Diagnostic accuracy in peripheral vestibulopathy. *Neurology* **2009**, *73*, 1134–1141. [CrossRef] [PubMed]
22. Cueva, R.A. Auditory brainstem response versus magnetic resonance imaging for the evaluation of asymmetric sensorineural hearing loss. *Laryngoscope* **2004**, *114*, 1686–1692. [CrossRef] [PubMed]
23. Urben, S.L.; Benninger, M.S.; Gibbens, N.D. Asymmetric sensorineural hearing loss in a community-based population. *Otolaryngol. Head Neck Surg.* **1999**, *120*, 809–814. [CrossRef]
24. Kesser, B.W. Clinical thresholds for when to test for retrocochlear lesions: Con. *Arch. Otolaryngol. Head Neck Surg.* **2010**, *136*, 727–729. [CrossRef] [PubMed]
25. Jiang, Z.Y.; Mhoon, E.; Saadia-Redleaf, M. Medicolegal concerns among neurotologists in ordering mris for idiopathic sensorineural hearing loss and asymmetric sensorineural hearing loss. *Otol. Neurotol.* **2011**, *32*, 403–405. [CrossRef] [PubMed]
26. Tos, M.; Stangerup, S.E.; Caye-Thomasen, P.; Tos, T.; Thomsen, J. What is the real incidence of vestibular schwannoma? *Arch. Otolaryngol. Head Neck Surg.* **2004**, *130*, 216–220. [CrossRef] [PubMed]
27. Kentala, E.; Pyykko, I. Clinical picture of vestibular schwannoma. *Auris Nasus Larynx* **2001**, *28*, 15–22. [CrossRef]
28. Bisdorff, A.; Bosser, G.; Gueguen, R.; Perrin, P. The epidemiology of vertigo, dizziness, and unsteadiness and its links to co-morbidities. *Front. Neurol.* **2013**, *4*, 29. [CrossRef] [PubMed]
29. Leskowitz, M.J.; Caruana, F.F.; Siedlecki, B.; Qian, Z.J.; Spitzer, J.B.; Lalwani, A.K. Asymmetric hearing loss is common and benign in patients aged 95 years and older. *Laryngoscope* **2016**, *126*, 1230–1232. [CrossRef] [PubMed]
30. Stucken, E.Z.; Brown, K.; Selesnick, S.H. Clinical and diagnostic evaluation of acoustic neuromas. *Otolaryngol. Clin. N. Am.* **2012**, *45*, 269–284. [CrossRef] [PubMed]
31. Van Dijk, J.E.; Duijndam, J.; Graamans, K. Acoustic neuroma: Deterioration of speech discrimination related to thresholds in pure-tone audiometry. *Acta Otolaryngol.* **2000**, *120*, 627–632. [CrossRef] [PubMed]
32. Harner, S.G.; Fabry, D.A.; Beatty, C.W. Audiometric findings in patients with acoustic neuroma. *Am. J. Otol.* **2000**, *21*, 405–411. [CrossRef]
33. Donnelly, M.J.; Daly, C.A.; Briggs, R.J. Mr imaging features of an intracochlear acoustic schwannoma. *J. Laryngol. Otol.* **1994**, *108*, 1111–1114. [CrossRef] [PubMed]
34. De Ridder, D.; Ryu, H.; Moller, A.R.; Nowe, V.; Van de Heyning, P.; Verlooy, J. Functional anatomy of the human cochlear nerve and its role in microvascular decompressions for tinnitus. *Neurosurgery* **2004**, *54*, 381–388. [CrossRef] [PubMed]
35. Ushio, M.; Iwasaki, S.; Murofushi, T.; Sugasawa, K.; Chihara, Y.; Fujimoto, C.; Nakamura, M.; Yamaguchi, T.; Yamasoba, T. The diagnostic value of vestibular-evoked myogenic potential in patients with vestibular schwannoma. *Clin. Neurophysiol.* **2009**, *120*, 1149–1153. [CrossRef] [PubMed]
36. Johnson, E.W. Auditory findings in 200 cases of acoustic neuromas. *Arch. Otolaryngol.* **1968**, *88*, 598–604. [CrossRef] [PubMed]
37. Johnson, E.W. Auditory test results in 500 cases of acoustic neuroma. *Arch. Otolaryngol.* **1977**, *103*, 152–158. [CrossRef] [PubMed]
38. Atturo, F.; Colangeli, R.; Bandiera, G.; Barbara, M.; Monini, S. Can unilateral, progressive or sudden hearing loss be immune-mediated in origin? *Acta Otolaryngol.* **2017**, *137*, 823–828. [CrossRef] [PubMed]
39. Vila, P.M.; Lieu, J.E. Asymmetric and unilateral hearing loss in children. *Cell Tissue Res.* **2015**, *361*, 271–278. [CrossRef] [PubMed]

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