

Cerebellopontine angle tumor patients with and without sudden hearing loss

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Objective: To assess significant differences of clinical features between cerebellopontine angle tumor (CPAT) patients with and without sudden hearing loss (SHL).

Methods: All 153 patients in this study underwent clinical tests to assess pure tone audiometry, speech audiometry (SDS), auditory brainstem response (ABR), and caloric test.

Results: Of the 153 patients, SHL was revealed in 42 and not in 111 patients (27% and 73%, respectively). The most common audiogram pattern in both groups was that of high-tone loss. The proportion of patients with SHL who had extremely poor discrimination of SDS (2%–30%) was higher than that of those without SHL. The ABR of 41 patients with SHL and 91 patients without SHL were recorded. Moreover, 8 and 5 patients with and without SHL, respectively, had normal responses on the affected side.

Conclusions: High-tone loss was the most common audiogram pattern observed among CPAT patients. ABR of patients presenting with SHL should be assessed. Regarding audiogram patterns, CPAT patients usually present with high-tone loss and reveal extremely poor discrimination in SDS. MRI is helpful for a diagnosis when CPAT volume increases based on ABR.

Key words: cerebellopontine angle tumor, sudden hearing loss, clinical features, audiological findings, neuro-otological findings

Introduction

Otolaryngologists often encounter patients with sudden hearing loss (SHL). This condition can be caused by different factors, such as viral infections, vascular compromise, and cochlear membrane rupture.¹ Considering that cerebellopontine angle tumor (CPAT) may also cause SHL, physicians should consider it as part of their differential diagnosis for SHL. The objective was to discover significant differences in clinical features and audiological and neuro-otological findings between CPAT patients with and those without SHL.

Materials and Methods

Of 153 patients diagnosed with CPAT at Hongo Neurosurgery Clinic between April 2017 and March 2020, 42 (27.5%) presented with SHL and 111 (72.5%) did not. Age, sex, diagnosis, and audiological and neuro-

otological findings were obtained from the clinical records of the patients. The patients underwent clinical tests to assess pure tone audiometry, speech audiometry, auditory brainstem response (ABR), and caloric test. Tumor size was evaluated by measuring the maximum diameter on magnetic resonance imaging (MRI) and classified as small (≤ 1.5 cm), medium (1.6–2.9 cm), and large (≥ 3.0 cm) based on a previous study.² Pure tone average was calculated as the average hearing level at 500, 1,000, and 2,000 Hz. Hearing level and audiogram patterns were categorised based on a previous study.³ The 67-S speech audiometric test list developed by the Japan Audiological Society was used to record speech audiometry. Using air conduction ear phones, the speech-recognition threshold was obtained at the lowest hearing level in which at least 50% of the test words on the list were identified correctly. Speech discrimination scores (SDSs) were evaluated based on a previous study.³ The participants were divided into 4 groups based on the SDS: unable to understand

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any syllables (0%), extremely poor discrimination (2%–30%), poor to moderate discrimination (32%–60%), and good discrimination (62%–100%). To record the ABR, the surface electrodes were placed on the patient's forehead and mastoid, and 0.1 msec clicks (80 dBnHL) were presented at a rate of 10 Hz, with 1,000 responses averaged twice. The signals were amplified and bandpass-filtered (100–3,000 Hz). Caloric responses to ice-water irrigation were evaluated using electronystagmography.

The χ^2 test with the Statistical Package for the Social Sciences Software version 23 (IBM, New York, USA) was used to compare the groups with P values of <0.05 considered statistically significant.

Results

Initial symptoms

In total, 42 CPAT patients with SHL had the following symptoms: SHL (n = 42), tinnitus (n = 23), vertigo (n = 7), dizziness (n = 5), ear fullness (n = 1), and ear pain (n = 1). The following symptoms were observed in 111 CPAT patients without SHL: HL (n = 79), tinnitus (n = 74), vertigo (n = 25), dizziness (n = 35), ear fullness (n = 2), ear pain (n = 1), headache (n = 5), numbness in the face (n = 4), facial palsy (n = 2), and decrease in taste (n = 1) and facial tactile sensation (n = 1). However, 5 patients had no symptoms. The incidence of hearing loss was higher in patients with SHL than in those without SHL, and that of dizziness was higher in patients without SHL

Table 1. Initial symptom of CPAT patients with and without SHL

	With SHL	Without SHL
Hearing loss	42*	79
Tinnitus	23	74
Vertigo	7	25
Dizziness	5	35*
Ear fullness	1	2
Ear pain	1	1
Headache	0	5
Numbness in the face	0	4
Facial palsy	0	2
Decrease in taste	0	1
Decrease in facial tactile sensation	0	1
No symptoms	0	5

Note that 5 patients without SHL were asymptomatic.

*P < 0.05

Table 2. Characteristics of CPAT patients with and without SHL

Age	With SHL			Without SHL		
	Men	Women	Total	Men	Women	Total
≤10	0	0	0	0	0	0
11–20	0	0	0	1	0	1
21–30	4	0	4	5	2	7
31–40	4	4	8	7	7	14
41–50	4	1	5	11	12	23
51–60	6	8	14	12	22	34
61–70	4	6	10	8	16	24
71–80	1	0	1	1	7	8
≥81	0	0	0	0	0	0
Total	23	19	42	45	66	111

In both groups, the majority of patients were in their 50s.

than in those with SHL ($P < 0.05$) (Table 1).

Diagnosis

Of the 42 CPAT patients with SHL, 41 presented with schwannomas, and 1 patient presented with a lipoma. Of 111 CPAT patients without SHL, 93 presented with schwannomas, 16 with meningiomas, and 2 with epidermoid cysts. There were more schwannomas than any other tumors in both groups.

Patient characteristics

The data of the 42 patients with SHL (23 men and 19 women) and 111 patients without SHL (45 men, 66 women) are summarised in Table 2. The average age of

the SHL group was 50.3 (range, 25–74) years. That of the non-SHL group was 52.1 (range, 20–79) years. In both groups, the majority of patients were in their 50s (SHL, 33.3%; non-SHL, 30.6%). In the SHL group, the right side was affected in 24 patients and the left side in 18 patients. In the non-SHL group, the right side was affected in 52 patients and the left side in 59 patients.

Tumor size

Of the 42 CPAT patients with SHL, 20 (47.6%), 15 (35.7%), and 7 (16.7%) presented with small (≤ 1.5 cm), medium (1.6–2.9 cm), and large (≥ 3.0 cm) tumors, respectively. Of 91 CPAT patients without SHL, 21 (23.1%), 37 (40.7%), and 33 (36.3%) presented with

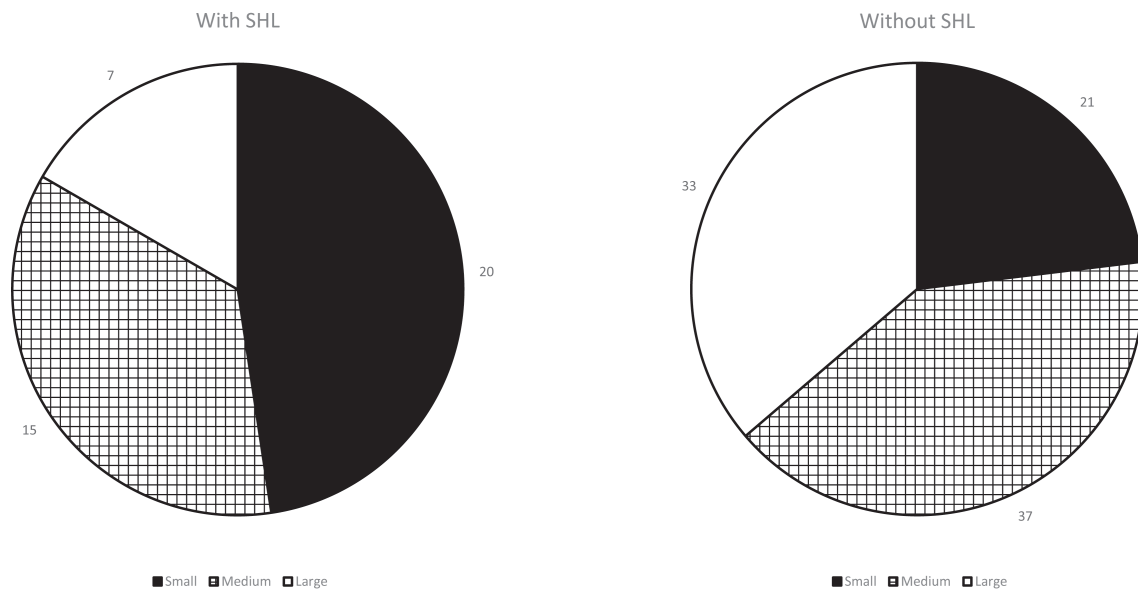


Figure 1. Tumor size

CPAT patients with SHL were more likely to have small tumors, and CPAT patients without SHL were more likely to have large tumors.

Table 3. Hearing level and audiogram pattern

PTA	With SHL					Without SHL				
	High-tone loss	Flat loss	Low-tone loss	Trough-shape loss	Total	High-tone loss	Flat loss	Low-tone loss	Trough-shape loss	Total
0–20 dB	2	4	0	0	6	10	20	1	2	33
21–50 dB	13	1	0	5	19	29	1	3	8	41
51–70 dB	3	3	1	3	10	8	2	1	5	16
≥ 71 dB	3	4	0	0	7	4	10	2	1	17
Total	21	12	1	8	42	51	33	7	16	107

The majority of patients with and those without SHL had relatively mild hearing loss. The most common audiogram pattern in both groups was high-tone loss.

small, medium, and large tumors, respectively. There were more patients with SHL who had small tumors than those without SHL, and more patients without SHL had large tumors than those with SHL ($P < 0.05$) (Figure 1).

Audiological findings

The hearing level and audiogram patterns during the initial examination were evaluated (Table 3, Figure 2). The majority of patients with and those without SHL had relatively mild hearing loss. The most common audiogram pattern in both groups was high-tone loss (50.0% and

47.7%, respectively). In total, 8 (19.0%) of 42 patients with SHL and 16 (15.0%) of 107 patients without SHL had trough-shape audiograms. The hearing level and audiogram patterns did not significantly differ in either group.

The findings on the maximum SDS are depicted in Figure 3. In total, 38 (55.3%) patients with SHL and 86 (61.6%) patients without SHL had good discrimination scores. The proportion of patients with SHL who presented with extremely poor discrimination was higher than that of those without SHL ($P < 0.05$).

The ABRs of 41 patients with SHL and 91 patients

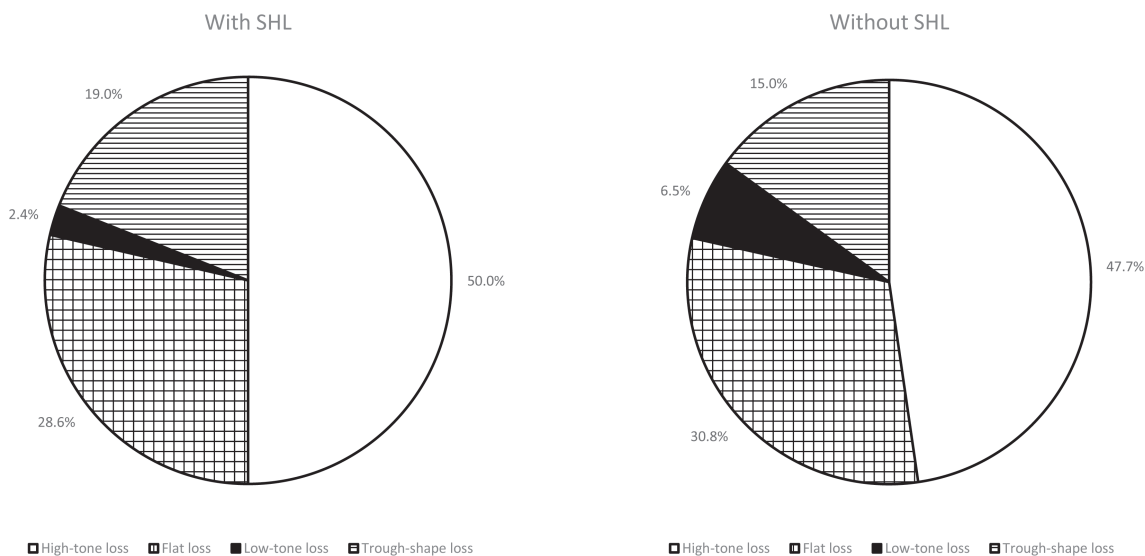


Figure 2. Audiogram pattern

The most common audiogram pattern in both groups was high-tone loss.

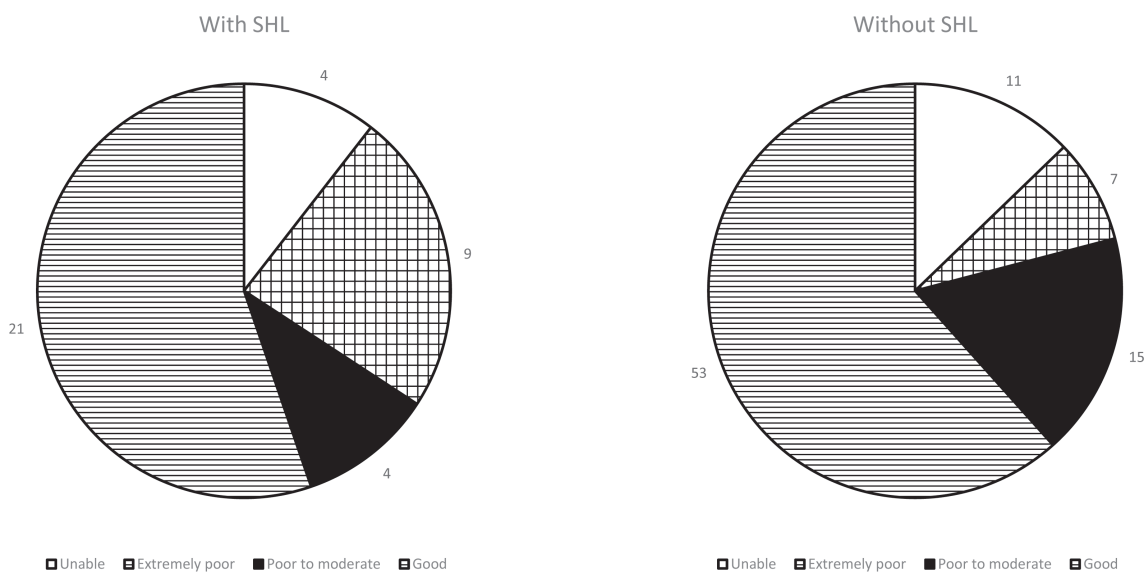


Figure 3. Speech audiometry

Most had good discrimination scores. Patients with SHL had extremely poor discrimination (2%–30%) but higher than those of patients without SHL.

without SHL revealed 8 (19.5%) patients with and 5 (5.5%) without SHL had normal responses on the affected side. The proportion of patients with SHL who had a normal response was higher than that of those without SHL ($P < 0.05$) (Figure 4). In 8 patients with SHL, the clinical diagnosis was schwannoma; however, in those without SHL, the diagnosis was schwannoma in 4 and 1 patient had a meningioma.

Neuro-otological findings

Caloric responses were not observed in 10 of 42 patients

with SHL but decreased (canal paresis $> 20\%$) in 19 patients. Thirteen patients had normal responses. Among them, 12 were diagnosed with schwannomas and 1 with a lipoma. Caloric responses were not observed in 40 of 100 patients without SHL but decreased in 36 patients. Twenty-four patients had normal responses. Among them, 19 were diagnosed with schwannomas, 4 with meningiomas, and 1 with an epidermoid cyst. The caloric responses did not significantly differ in either group (Figure 5).

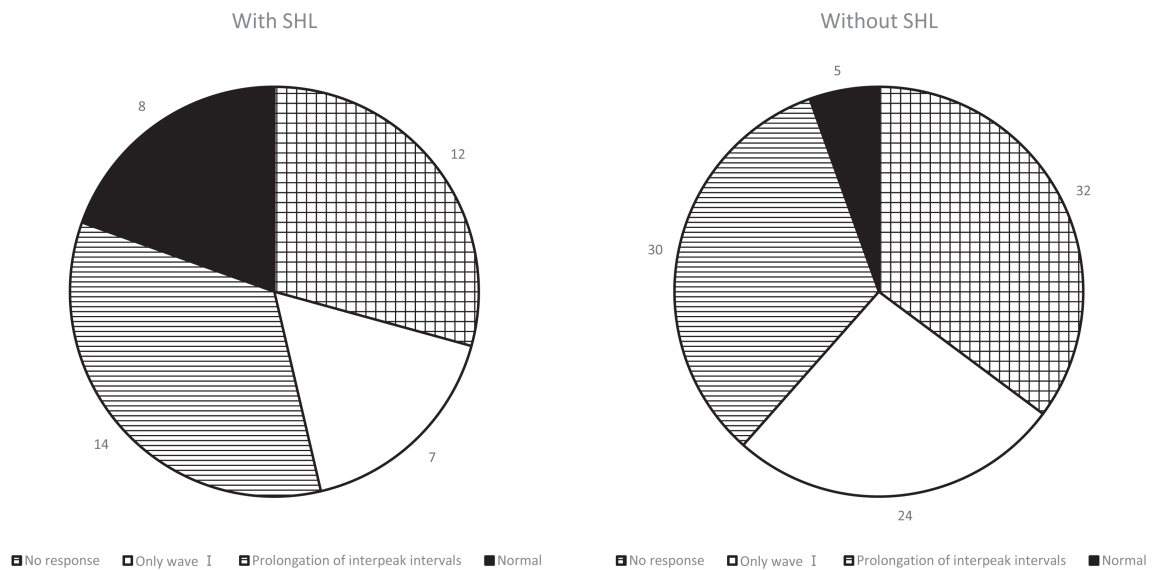


Figure 4. ABR

Eight patients with SHL and 5 patients without SHL had normal ABRs, which were higher in patients with SHL.

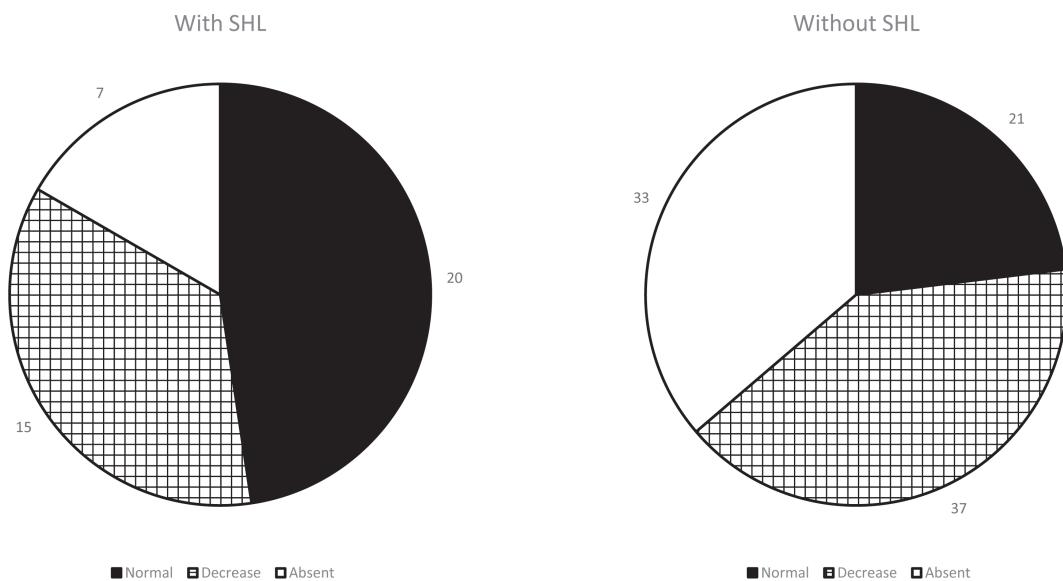


Figure 5. Caloric test

Thirteen patients with SHL and 24 without SHL had normal responses.

Discussion

There was a study comparing CPAT patients with SHL and sudden deafness (SD),⁴ however, no detailed comparison between CPAT patients with and those without SHL was made. Therefore, in the present study, I aimed to determine whether or not there were any significant differences in the clinical features and audiological and neuro-otological findings between this particular cohort of patients.

In 1951, Edwards and Paterson⁵ first reported about acoustic tumors (ATs) in patients who present with SHL. Since then, several investigators have assessed the incidence of SHL in CPAT (range, 3.2% – 18.9%) (Table 4)^{2,5-9} without consideration of the results of the present study in which 42 (27.5%) of 153 patients presented with SHL. Recently, the incidence of SHL among CPAT patients has been increasing due to a greater number of physicians who believe that patients with CPAT may present with SHL and because of the development of diagnostic tools including MRI.

Several patients with and without SHL experience HL and tinnitus. However, not all patients with HL experience a sudden onset of symptoms. In the present study, the initial symptoms experienced by CPAT patients without SHL included numbness in the face, facial palsy, and decrease in taste and facial tactile sensations. These symptoms are correlated with damage in cranial nerves other than the eighth cranial nerve. However, they were not observed in CPAT patients with SHL.

A majority of patients were diagnosed with schwannomas in both groups with no noted relationship between SHL and CPAT. In another study, schwannomas accounted for 80% of CPAT cases and meningiomas for less than 10%.¹⁰ Other less common lesions in the cerebellopontine angle include epidermoid tumors, those that metastasised from other sites, sarcoidosis, paraganglioma, cholesterol granulomas, and

hemangiomas.¹¹

Regarding age, participants in their 50s and 60s comprised 57.1% of patients with SHL and 52.3% of patients without SHL. Previous reports^{12,13} had similar results. Therefore, age is evidently not a significant difference.

In the present study, CPAT patients with SHL were more likely to present with small tumors, whereas CPAT patients without SHL were more likely to have large tumors. However, the tumor sizes in some previous reports varied.^{2,3,8,9} Therefore, whether or not a specific tumor size causes SHL cannot be confirmed.

Regarding audiogram patterns, 50.0% of 42 patients with SHL and 47.7 % of 107 patients without SHL had high-tone loss, and through-shape loss was observed in 19.0% and 15.0% of patients with and without SHL, respectively. Moreover, Takahashi et al.¹⁴ reported that high-tone loss was observed in 38% of the patients in their study. The anatomical features are such that, the neurofibers of middle and high tones go along the dorsum of the cochlear nerve,¹⁵ and any direct pressure on that nerve, caused by tumor growth volume, may result in high-tone loss.

The ABR test has proven effective in diagnosing CPAT. In Pensak et al.,² abnormal responses were found in all the AT patients in their study. Kanzaki⁸ obtained a similar finding that 96% of AT patients had abnormal results. In the present study, 33 of 41 patients with SHL and 86 of 91 patients without SHL (80.5% and 94.5%, respectively) had abnormal ABR test results. Even though this rate is lower than that in previous reports, it is still high. This slight discrepancy might be attributed to the fact that 8 patients (19.5%) with SHL and 5 patients without SHL had normal responses, of whom 5 and 3 patients, respectively, had small tumors. In the present study, 20 of 42 patients (47.6%) with SHL had small tumors. Therefore, this study involved a higher number of small tumors associated with SHL than did any of the

Table 4. Incidence of SHL in CPAT

Authors	Year	Total patients	Patients with SHL	Incidence of SHL (%)
Edwards, et al. ⁵	1951	157	5	3.2
Higgs, et al. ⁶	1973	44	4	9.1
Pensak, et al. ²	1985	498	69	13.9
Berg, et al. ⁷	1986	133	17	12.8
Kanzaki ⁸	1986	90	12	13.3
Yanagihara, et al. ⁹	1993	111	21	18.9
Ochiai	2020	153	42	27.5

The incidence rate of SHL in CPAT ranged from 3.2% to 27.5%, and it has been increasing recently.

previous studies.

The cause of SHL in CPAT remains unclear. Neeley¹⁶ hypothesized that SHL is caused by: pressure on the eighth cranial nerve, vascular compromise in the inner ear, biochemical changes within the inner ear, and diminished fiber mass. Higgs et al.⁶ and Yanagihara et al.⁹ showed that factors, such as the site of origin, histopathology, growth rate, and vascular variation, may be associated with development of SHL. Moreover, the tumor likely compresses the labyrinthine artery, or hemorrhage in an intracanalicular tumor occurs. Because the labyrinthine artery is an end artery, acute ischemia of the cochlea could result from the compression of this artery due to tumor volume expansion. A reduction in the degree of edema of the tumor mass with steroids may improve the blood supply to the cochlea and result in hearing recovery.

High-tone loss was the most common audiogram pattern observed in CPAT patients. There were significant differences in hearing loss as an initial symptom, and extremely poor discrimination based on the SDS between CPAT patients with and those without SHL. Therefore, when patients present with SHL, pure tone audiometry should be performed as well as SDS when pure tone audiometry reveals a high-tone loss, and an ABR test when SDS shows extremely poor discrimination. MRI is helpful to obtain a diagnosis when the CPAT volume increases as evidenced by the ABR.

Conflicts of Interest: None

References

1. Zadeh MH, Storper IS, Spitzer JB. Diagnosis and treatment of sudden-onset sensorineural hearing loss: A study of 51 patients. *Otolaryngol Head Neck Surg* 2003; 128: 92-8.
2. Pensak ML, Glasscock ME 3rd, Josey AF, et al. Sudden hearing loss and cerebellopontine angle tumors. *Laryngoscope* 1985; 95: 1188-93.
3. Johnson EW. Auditory test results in 500 cases of acoustic neuroma. *Arch Otolaryngol* 1977; 103: 152-8.
4. Nageris BI, Popovtzer A. Acoustic neuroma in patients with completely resolved sudden hearing loss. *Ann Otol Rhinol Laryngol* 2003; 112: 395-7.
5. Edward CH, Paterson JH. A review of the symptoms and signs of acoustic neurofibromata. *Brain* 1951; 74: 144-90.
6. Higgs WA. Sudden deafness as the presenting symptom of acoustic neurinoma. *Arch Otolaryngol* 1973; 98: 73-6.
7. Berg HM, Cohen NL, Hammerschlag PE, et al. Acoustic neuroma presenting as sudden hearing loss with recovery. *Otolaryngol Head Neck Surg* 1986; 94: 15-22.
8. Kanzaki J. Present state of early neurotological diagnosis of acoustic neuroma. *ORL* 1986; 48: 193-8.
9. Yanagihara N, Asai M. Sudden hearing loss induced by acoustic neuroma: significance of small tumors. *Laryngoscope* 1993; 103: 308-11.
10. Sekhar LN, Jannetta PJ. Cerebellopontine angle meningiomas. Microsurgical excision and follow-up results. *J Neurosurg* 1984; 60: 500-5.
11. Brackmann DE, Bartels LJ. Rare tumors of the cerebellopontine angle. *Otolaryngology Head Neck Surg* 1980; 88: 555-9.
12. Lee SH, Choi SK, Lim YJ, et al. Otologic manifestations of acoustic neuroma. *Acta Otolaryngol* 2015; 135: 140-6.
13. Kim SH, Lee SH, Choi SK, et al. Audiologic evaluation of vestibular schwannoma and other cerebellopontine angle tumors. *Acta Otolaryngol* 2016; 136: 149-53.
14. Takahashi K, Yamamoto Y, Ohshima S, et al. Audiometric patterns in acoustic neuromas: an analysis of 105 cases. *Otol Jpn* 2011; 21: 23-8 (in Japanese).
15. Sando I. The anatomical interrelationships of the cochlear nerve fibers. *Acta Otolaryngol* 1965; 59: 417-36.
16. Neely JG. Gross and microscopic anatomy of the eighth cranial nerve in relationship to the solitary schwannoma. *Laryngoscope* 1981; 91: 1512-31.