Sudden Sensorineural Hearing Loss as the Initial Symptom in Acoustic Neuroma Patients

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Abstract

Previous studies have shown that patients with acoustic neuroma (AN) sometimes present with sudden sensorineural hearing loss (SSNHL) as an initial symptom. The purpose of this research was to investigate the clinical characteristics, diagnosis and treatment of AN in patients diagnosed as SSNHL initially. A total of 10 patients presented with SSNHL as the initial symptom and diagnosed as AN by magnetic resonance imaging (MRI) finally were enrolled in this study. Patient demographics, associated complaints (mostly tinnitus and vertigo), severity of hearing loss, audiogram configurations, auditory brainstem response (ABR) and MRI examination were reviewed and analyzed. Treatment outcome and management protocols were also included in this study. These patients exhibited varying severity of hearing loss and a variety of audiogram configurations. All patients showed an abnormal ABR. According to the Koos grading standard, there were 5 grade I (intracanalicular) tumors, 3 grade II tumors and 2 grade III tumors. The treatment outcome revealed that 2 patients exhibited recovery of the average hearing of impaired frequency by more than 15 dB, and 6 patients showed no recovery. Four patients were referred to undergo surgical treatment after being diagnosed with AN, 1 patient accepted stereotactic radiation therapy, and the remaining 5 patients were taken "waiting and scan" strategy. The findings of this study show that hearing recovery after corticosteroid treatment for SSNHL does not exclude the presence of AN and all patients diagnosed as SSNHL initially should undergo MRI and ABR to prevent misdiagnosis and delays in potential treatment.

1. Introduction

Sudden sensorineural searing loss (SSNHL) is defined as a rapid-onset sensorineural hearing loss of [?]20 decibels (dB), affecting at least 2 consecutive frequencies that occurred within 72h with no identifiable cause [1]. The incidence of SSNHL is reported to be 5–20 per 100,000 individuals, with about 66,000 new cases per year in the United States [2]. An epidemiological survey in Japan revealed that the incidence of SSNHL was 60.9 per 100 000 population [3]. In China, the prevalence of SSNHL has been on the rise in recent years, but large samples of epidemiological data are lacking. For patients suffering SSNHL, more than 90% cases are idiopathic and the remainder are due to causes such as acoustic neuroma, stroke, malignancy, Meniere's disease, trauma, autoimmune disease, syphilis, Lyme disease, and perilymphatic fistula [4, 5].

Acoustic neuroma (AN) is known as a benign tumor that originates from superior or inferior vestibular branch of the cochleovestibular nerve in the internal auditory canal (IAC) [6]. Sensorineural hearing loss is the major presentation of patients with AN and patients with AN could present with SSNHL as an initial symptom occasionally [7-9]. According to previous studies, the reported prevalence of AN in patients presenting with SSNHL ranges from 1.8% to 5.2% [10, 11]. As a result of the increasing widespread use of magnetic resonance imaging (MRI), more patients with AN than expected have been detected among those with SSNHL. MRI is regarded as the gold standard for imaging diagnosis of AN. In the meantime, auditory brainstem response (ABR) test is recommended for the initial assessment of SSNHL patients when appropriate, and it is highly sensitive to AN larger than 10 mm in size [10]. AN is the most frequently observed MRI abnormality in

patients with SSNHL [9]. According to the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines, MRI or ABR should be used for the retrocochlear pathology evaluation of patients with SSNHL [2].

Previous studies have found that hearing can improve with corticosteroid treatment in AN patient initially presenting with SSNHL [12-14] and that drug therapy is usually administered before MRI is performed. As a result, some physicians may assume that patients with SSNHL who respond to steroid therapy could effectively exclude the presence of AN, which leads to delays in the diagnosis. Since a small number of SSNHL patients whose hearing loss is caused by AN, which is also prone to clinical misdiagnosis and missed diagnosis.

In the present study, we conducted a retrospective study of 10 AN patients presenting with SSNHL as an initial symptom, with the aim of clarifying the detailed clinical features and the efficacy of treatment of AN with SSNHL and of guiding the clinic to prevent misdiagnose and missed clinically.

2. Methods

2.1 Ethical considerations

The study was approved by the Committee of Medical Ethics of the Hospital. Written informed consent was obtained from all participants in this study.

2.2 Study design and participants

We reviewed retrospectively medical records of all patients treated as SSNHL initially and diagnosed as AN finally after undergoing MRI, who were admitted to our hospital between 2008 and 2021. These patients met the clinical diagnostic criteria for SSNHL, which is defined as sensorineural hearing loss of 20 dB or more over at least 2 contiguous frequencies that develops within 3 days [1]. Data collection included patient demographics, associated complaints (mostly tinnitus and vertigo), results of pure-tone audiometry (PTA), ABR and MRI examination. Exclusion criteria included known previous or progressive hearing loss, AN has been diagnosed before SSNHL occurred and another inner-ear disease. Patients who did not undergo MRI examination were also excluded.

2.3 Audiological assessment

All patients received the essential audiological examinations, including pure-tone audiometry and ABR. The severity of hearing loss was classified based on criteria of the World Report on Hearing published by World Health Organization (WHO) in 2021 [15]. According to this criterion, severity of hearing loss is categorized into 7 grades based on hearing thresholds measured with PTA at 0.5, 1, 2, and 4 kHz:

1. Normal hearing, mean hearing thresholds less than 20 dB;
2. Mild hearing loss, mean hearing thresholds is 20 to $<$ 35 dB;
3. Moderate hearing loss, mean hearing thresholds is 35 to $<$ 50 dB;
4. Moderately severe hearing loss, mean hearing thresholds is 50 to $<$ 65 dB;
5. Severe hearing loss, mean hearing thresholds is 65 to $<$ 80 dB;
6. Profound hearing loss, mean hearing thresholds is 80 to $<$ 95 dB;
7. Complete or total hearing loss/deafness, mean hearing thresholds is 95 dB or greater.

We also analyzed the patterns of hearing loss and the configuration of audiogram was categorized into 7 forms: low-frequency ascending form, U-shaped form, high-frequency descending form, flat form, profound form, dip form and other form [16] (see Table S1).

Results of the ABR were considered abnormal when they met at least one of the following criteria: (1) absent evoked response upon the compatible auditory threshold; (2) desynchronization of waves other than wave I; (3) interpeak latency (IPL) between waves I and III > 2.5 ms; (4) IPL between waves I and V > 4.4 ms; (5) Wave V interaural latency difference (ILD) > 0.2 ms; (6) interaural difference of IPL between waves I and V > 0.2 ms [17].

2.4 Imaging examination

All enrolled patients underwent a targeted MRI scan of the brain. The scans included high-resolution T2 sequences and contrast-enhanced T1-weighted MRI directed to the IAC and cerebellopontine angle (CPA). The tumor size was measured on MRI images, and the type of tumor within the IAC (intracanalicular) and the maximum diameter of the tumor in CPA were used according to the recommendation of the Summary and consensus in 7th International Conference on acoustic neuroma [6]. According to Koos grading standard, the tumor size is classified as follows: grade I, tumor is confined to the IAC (intracanalicular) and the maximum diameter is [?] 1 cm; grade II, small tumor protrusion into CPA without contact with the brain stem, diameter ranges from 1.1 to 2 cm; grade III, tumor occupying the CPA with no brainstem displacement, diameter ranges from 2.1 to 3 cm; grade IV, large tumor with brainstem and cranial nerve displacement, diameter is more than 3.0 cm [18].

2.5 Treatment evaluation

All patients enrolled in the study were treated in agreement with standardized drug therapeutic protocols in accordance with the principles of treatment for SSNHL. All patients underwent audiogram tests on the first day of treatment and 1 or 2 weeks after treatment. The evaluation of treatment outcome was based on the hearing recovery criteria described in the Guidelines for the diagnosis and treatment of SSNHL (published in China at 2015) [1]:

1. Complete recovery: the hearing thresholds of impaired frequency returned to normal, or reached the level of contralateral ears or the original level.

2. Significant recovery: the hearing thresholds of impaired frequency increased [?]30 dB on average.

3. Slight recovery: the hearing thresholds of impaired frequency increased [?]15 dB and < 30 dB on average.

4. No recovery: the average hearing threshold of impaired frequency increased < 15 dB.

We evaluated the rates of hearing recovery in the study groups and a summed rate of both complete recovery, significant recovery, slight recovery was defined as the “effective” rate.

2.6 Statistical Analysis

Data analysis was performed with GraphPad Prism 8 and Microsoft Office Excel. Continuous variables were summarized using means, standard deviations, and range values when normally distributed;categorical data were summarized as number (percentage) and analyzed by the $\chi 2$ or Fisher tests when normally distributed. Spearman’s rank correlation analysis was used to determine the relation among the investigated factors and the results were presented with a confidence interval (CI) of 95%. $P < 0.05$ was considered statistically significant.

3. Results

3.1 Patients

In total, 10 patients were enrolled in our study. The demographic characteristics of the patients were summarized in Table 1. There were 4 men and 6 women, ranging in age at diagnosis of SSNHL from 26 to 70 years with a mean age of 46.2 ± 13.16 years. Of the 10 patients enrolled in this study, 9 showed a tumor on the same side of SSNHL, with 4 on the left side and 5 on the right. There was one patient with SSNHL in bilateral ear who had an incidental finding of AN in the right ear (case 8). Among the 10 patients,

9 (90%) complained of tinnitus and 2 patients (20%) complained of dizziness as accompanying symptoms. Three patients underwent vestibular function examination with 1 patient showed reduced function of the left horizontal semicircular canal, 1 patient showed reduced function of the right horizontal semicircular canal and 1 patient with positive positional test.

3.2 Audiology

3.2.1 The severity of hearing loss

Among the 10 patients, the severity of hearing loss of the affected ear was characterized as mild in 2 ears (20%), moderate in 1 ear (10%), severe in 4 ears (40%), profound in 1 ear (10%) and deafness in 1 ear (10%). In addition, one patient present normal hearing threshold at frequencies of 0.25, 0.5, 1 and 2 kHz, 45 dB hearing loss in 4 kHz. The PTA ranged from 18.75 dB to 120 dB, with a mean range of 60.75 ± 30.39 dB.

3.2.2 Audiogram configuration

The audiograms of the affected ears of all 10 patients when diagnosed with SSNHL were depicted in Figure 1a. For the 7 forms of audiogram configurations, 2 ears (20%) were classified as U-shape, 3 (30%) as high-frequency descending, 2 (20%) as flat, 1 (10%) as dip, 1 (10%) as profound and 1 (10%) as other.

3.2.3 ABR

As for ABR examinations, all the 10 patients were evaluated as having an abnormal ABR according to the diagnostic criteria mentioned above. The sensitivity of ABR for the detection of AN was 100% in our study. Table 2 demonstrated the latency of ABR waves of the 10 enrolled patients. In these patients who showed an abnormal ABR waveform pattern, no response was detected in 2 patients (20%) at 100 dB SPL, 2 patients (20%) had only wave V present, 3 patients (30%) showed the absence of wave I, 2 patients (20%) with IPL between waves I and III $>$ 2.5 ms, 4 patients (40%) with IPL between waves I and V $>$ 4.4 ms. Significantly, except for 2 patients with no response, the ILD of wave V were all $>$ 0.2 ms in the remaining 8 patients, with a mean value of 1.01 ± 0.43 ms.

3.3 Imaging findings

MRI was performed in all 10 patients revealing the tumors ranging from 0.7 cm to 2.7 cm, with a mean size of 1.44 ± 0.63 cm. Figure 1b demonstrated the axial cranial MRI of these patients. According to the Koos grading standard, there were 5 grade I (intracanalicular) tumors, 3 grade II tumors and 2 grade III tumors. The audiograms of patients with grades I–III tumor are shown in Figure 2. There was no obvious relation between tumor size and hearing loss with regard to the audiometric pattern. We also conducted correlation analyses between tumor size and grade of hearing loss and configuration of audiograms. There was no significant correlation between tumor size and grade of hearing loss (r = 0.1136, P = 0.7533; Spearman's rank correlation test). Tumor size and configuration of audiograms were also unrelated (r =0.0528, P =0.8831; Spearman's rank correlation test).

3.4 Management Results

Among the 10 patients in our study, 8 recorded detailed PTA results before and after drug treatment (see Figure 3), while 2 had either subjective report only or incomplete audiometric data available. The treatment outcome revealed that 2 (25%) of the 8 patients exhibited recovery of the average hearing of impaired frequency by more than 15 dB, and 6 (75%) patients showed no recovery. Figures 4 illustrated pure tone thresholds of the 8 patients who recorded detailed PTA results at diagnosis of SSNHL and after drug treatment. Overall, there was no obvious recovery before and after drug treatment. In addition, we also followed up the speech recognition score (SRS) of 4 patients after drug treatment, and 2 of them showed significant improvement in SRS compared with that before treatment: case 6 showed an increase in SRS from 68% to 100%, and case 7 showed a significant increase from 8% to 96%. For the final management of the 10 patients, 4 patients were referred to undergo surgical treatment after being diagnosed with AN, 1 patient accepted stereotactic radiation therapy, and the remaining 5 patients were taken "waiting and scan" strategy (observation and follow-up of MRI). Among the 4 patients with surgical treatment, 3 were

performed with a translabyrinthine approach and 1 with a retrosigmoid approach, and the postoperative hearing of the affected ear of the 4 patients were all totally deafness (see Figure S1).

4. Discussion

AN is the most common posterior cranial fossa tumor in adults, accounting for 6%–8% of all intracranial tumors and more than 80% of tumors in the CPA [19]. The early symptoms in patients with AN are mainly unilateral sensorineural hearing loss, tinnitus and vertigo [20]. SSNHL occurs in 10% to 20% of AN patient at some point in their medical history, but the incidence of AN is somewhat lower in patients with SSNHL [4]. Sensorineural hearing loss is the major presentation of AN patients and is often accompanied with tinnitus. In the present study, 90 percent (9/10) of AN patient who present SSNHL as an initial symptom complained of tinnitus, which is consistent with the incidence of tinnitus in patients with AN (51%˜ 92%) that reported in the previous studies [21-23]. Even though tinnitus is a nonspecific symptom, clinicians should still be alert to patients with unilateral SSNHL and tinnitus to avoid missing the diagnosis. Meanwhile, the absence of accompanying symptoms does not eliminate the possibility of a tumor.

Previous studies have shown that the audiogram configuration may be trough shaped or U-shape in AN patient who presenting with SSNHL as a primary symptom [7, 21]. Furthermore, a recent study has found that the trough shaped or U-shape audiogram was significantly more prevalent in patients with AN than in ordinary idiopathic SSNHL patients, and the incidence of AN in SSNHL patients with trough shaped or U-shape audiogram was significantly higher in SSNHL patients with other audiogram configurations [24, 25]. This study suggests that a trough shaped or U-shape audiogram in patients with SSNHL may indicate the presence of AN. In our study, the incidence of U-shape audiograms (20%, 2/10) in SSNHL patients with AN was not significantly specific compared to other configurations, which may be due to the small sample size.

As the gold standard for AN diagnosis, MRI is the preferred examination and can provide exquisite tumor characterization, surgical planning, and post-therapeutic assessment [19]. High-resolution MRI can detect tumors smaller than 1cm located in the IAC and differentiate AN from other masses such as facial nerve schwannoma, meningioma, epidermoid cyst, arachnoid cyst, aneurysm, and metastasis [26, 27]. According to previous studies, AN can be successfully diagnosed and largely differentiated from other lesions with 96% to 100% sensitivity and 88% to 93% specificity with the combination of T1 and T2- weighted MRI [28]. In the present study, all the enrolled SSNHL patients were undergone contrast-enhanced MRI and 10 were diagnosed as AN. In addition to MRI, ABRs have been used widely as a screening procedure for the diagnosis of AN, particularly when MRI is not available. In our study, abnormal ABR results were obtained in all patients, and the overall ABR sensitivity in diagnosing AN in SSNHL was 100%. Due to the small sample size, our results do not indicate the sensitivity of the ABR. We have known that ABR testing has limits. The reported sensitivity of ABR for the diagnosis of AN varies between 63 and 97%, however, for small AN, its sensitivity decreases significantly to 8%-42% and ABR is not possible when the hearing loss exceeds 80 dB in the 2000 to 4000Hz frequency [4, 29]. A recent modelling study has found that the cost-saving with ABR prior to MRI does not seem to outweigh the number of missed patients with AN and other important pathologies that would have been detected when using standalone MRI [30]. Therefore, for SSNHL patients, we recommend that ABR and MRI should be combined to improve the accuracy of detection and prevent misdiagnosis and missed diagnoses, especially for small AN.

Previous studies have pointed out that the pathogenesis of SSNHL in AN patient involves mechanical compression of the adjacent cochlear nerve, based upon the conjecture that the nerve fibers responsible for middle-frequency hearing are in a position more susceptible to tumor compression [14]. Although hearing loss due to nerve compression is theoretically progressive, a sudden enlargement of the tumor (e.g., hemorrhage or cystic degeneration) could compress the cochlear nerve enough to cause sudden hearing loss [31]. Nevertheless, it has been reported that tumor size is not to be correlated with the grade of hearing loss, and the correlation between tumor size and the incidence of SSNHL is also controversial [9, 16]. In this study, we found no significant correlation between tumor size and grade of hearing loss. In addition, the tumor size and configuration of audiograms were also unrelated. These results were consistent with other previous studies.

Many studies have observed that hearing recovery occurred in some AN patients presented with SSNHL after corticosteroid therapy and reported the recovery rate ranging from 16.7% to 44.4% [10, 31, 32]. It is well-known that SSNHL patients with different types of audiogram configurations have obvious differences in the hearing recovery [1]. Several studies have found that the recovery rate of SSNHL in AN patient was also significantly related to audiogram pattern [24, 25]. In 2021, Wasano et al. [16] revealed that the recovery of hearing in patients with U-shaped audiograms was significantly greater than in patients having the other audiogram forms, and the recovery rate decreased as the SSNHL episodes in patients increased. In 2017, Cho et al. [27] reported that non-tumorous lesions (intra-labyrinthine hemorrhage and labyrinthitis) showed a poorer treatment response than that of AN in patients with SSNHL. Hearing recovery may be due to the regression of tumor edema caused by corticosteroid treatment and/or the absorption of hemorrhage from the tumor itself or in the vicinity of the tumor [33]. In the present study, 25% (2/8) SSNHL patients diagnosed as AN showed hearing recovery after drug treatment. This rate was consistent with previous studies. These findings suggested that a therapeutic response to corticosteroid treatment for SSNHL does not exclude the presence of AN and all patients with SSNHL should undergo MRI to prevent misdiagnosis and delays in potential treatment.

5. Conclusion

In conclusion, we have reported on a series of 10 AN patients presented SSNHL as a primary symptom and treated as SSNHL initially. MRI is the most effective examination for the diagnosis of small AN. This study demonstrated that the hearing loss of these patients may improve with corticosteroid treatment. Therefore, we recommend that all patients presenting with SSNHL, regardless of whether the hearing loss responds to drug treatment, should undergo MRI to rule out AN and avoid delayed treatment due to missed diagnosis. In addition, as an effective screening procedure, ABR is also important for the diagnosis of AN.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

Conflicts of Interest: The authors disclose no conflicts of interest.

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Authors' Contributions:WHY designed the work; WDY provided data resources; LJ, CGH and ZXL acquired and analyzed data; SMT and WHY drafted, revised and approved the manuscript; WQJ agree to be accountable for all aspects of the work. All authors have reviewed, discussed and approved the manuscript.

Supplementary Materials

Figure S1: Postoperative hearing results of the 4 patients who were treated with surgical operation. Case 3, 5, 6 were taken with translabyrinthine approach and case 7 with retrosigmoid approach. A red triangle with an arrow was used for the right ear and a blue square with an arrow was used for the left ear to indicate that there was no response at maximum air conduction with masking.

Table S1: Criteria for different configurations of audiograms.

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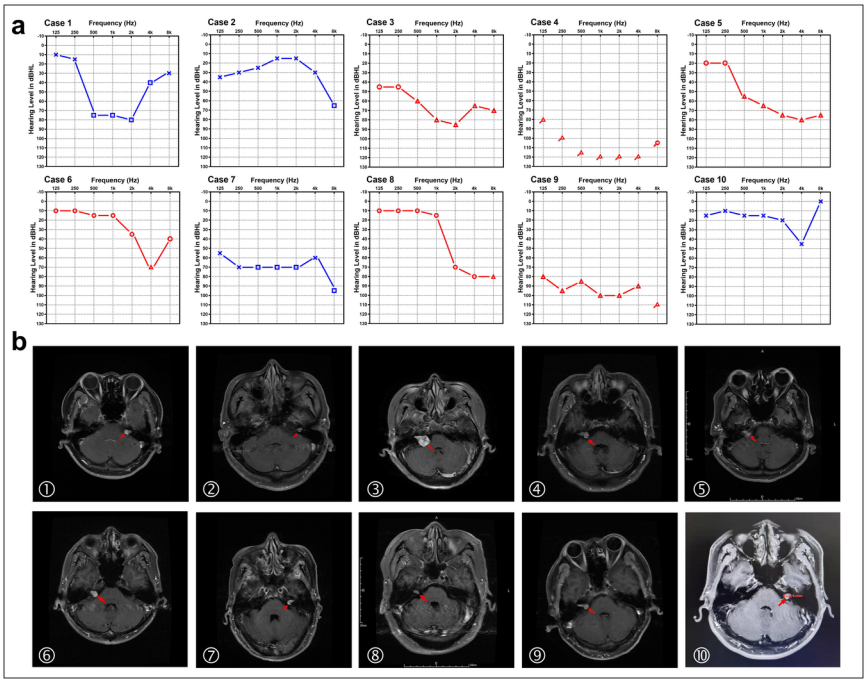

Figure 1: Figure 1: (a) Pure-tone audiograms of the affected ear of the 10 acoustic neuroma patients at diagnosis of sudden sensorineural hearing loss (air conduction). The red line represents the right ear and the blue line represents the left ear. (b) The axial cranial MRI of the 10 patients diagnosed with acoustic neuroma in our study. Red arrows indicate the tumor location.

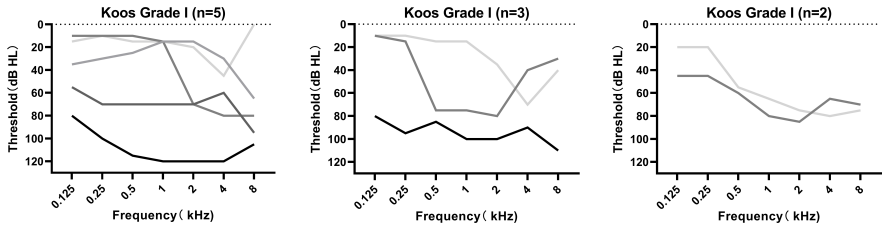

Figure 2: Figure 2: Audiograms of the 10 patients with acoustic neuroma diagnosed as sudden sensorineural hearing loss initially classified according to the Koos grading system. Audiometry found no consistent trend with regard to Koos grade.

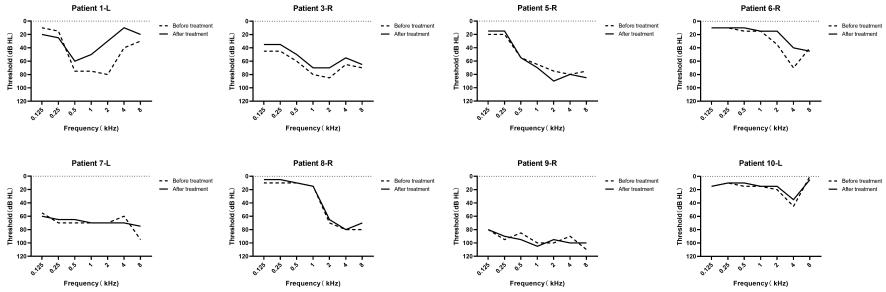

Figure 3: Figure 3: Audiograms of the eight acoustic neuroma patients initially presenting with sudden sensorineural hearing loss who recorded completed pure tone audiometry results before and after drug treatment. The "L" and "R" means the tumor side. The dashed line represents the hearing threshold before treatment, and the solid line represents the hearing threshold after treatment.

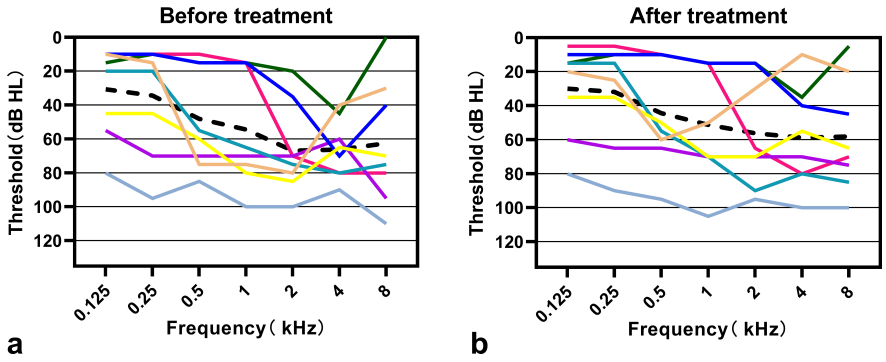

Figure 4: Figure 4: (a) Pure-tone thresholds at the time of diagnosis of sudden sensorineural hearing loss in all 10 patients. (b) Pure-tone thresholds after drug treatment of sudden sensorineural hearing loss in 8 patients who recorded completed pure tone audiometric results. The black dashed line represents the average hearing thresholds at each frequency.

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