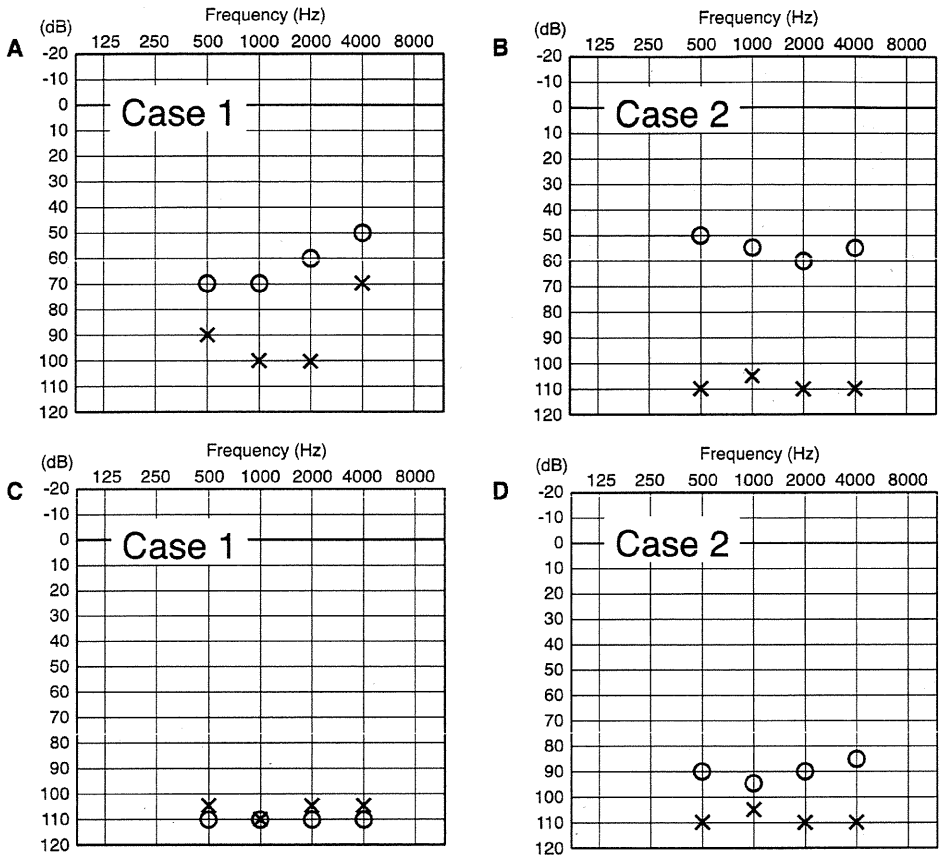

Fig. 1. (A) Case 1; a 5-year-old girl with asymptomatic congenital CMV infection (threshold using ASSR test). (B) Case 2; a 5-year-old girl with asymptomatic congenital CMV infection. These were results of first diagnosed with hearing loss. (C and D) Deterioration in hearing, for one month and for one year, respectively.

significant (Fig. 1D). She was referred to our hospital for further examinations, and her preserved umbilical cord demonstrated a positive result for congenital CMV infection. Late-onset and slowly progressive hearing loss for one year was suggested. There were no inner ear abnormalities seen in the CT findings. She underwent CI surgery in the left ear at the age of 4 years 9 months.

Each child received the same rehabilitation according to auditory oral method by the same speech therapist after implantation.

Table 1
The activated areas of the brain in profoundly deaf individuals during speech-reading.

Case	Gender/age (years)	Activated areas	
		Right hemisphere	Left hemisphere
1	Female/5	Superior temporal gyrus [BA22] Cingulate gyrus [BA31] Middle frontal gyrus [BA9]	Middle temporal gyrus [BA21] Inferior parietal lobe [BA40] Occipital gyrus [BA19] Precueus [BA7]
2	Female/5	Middle temporal gyrus [BA21] Postcentral gyrus [BA3/1/2] Middle occipital gyrus [BA20] Middle frontal gyrus [BA9]	Precentral gyrus [BA4] Precuneus [BA31] Precuneus [BA19] Cingulate gyrus [BA24]

3. Results

3.1. Brain imaging with PET

Table 1 and Fig. 2 show the areas that were activated in each child during a speech-reading task. The following cortical areas showed significantly higher metabolism during speech-reading in the children compared to normal hearing control subjects. In Case 1, the activated areas were the bilateral auditory association area [BA21], the bilateral precuneus, somatosensory cortex [BA7], the left secondary visual area [BA19], and the left inferior parietal lobule [BA40].

The activated areas in Case 2 were similar to those in Case 1, but the activation of the visual association areas in the parietal lobe were lower and smaller than in Case 1.

3.2. Assessment before cochlear implant, and outcome

Table 2 shows the children's scores in the *K*-test before CI, in the word and sentence discrimination tests, and in the Japanese WISC-III at one year after implantation. *K*-test scores that assessed Cognitive-Adaptive development of each child were almost similar. Both cases showed 30–40 dB of aided hearing thresholds at all frequencies with CI. One year after CI, the results of the Japanese WISC-III showed a clearer difference in VIQ than PIQ, in which Case 1 had a better score compared with Case 2. Case 1 did

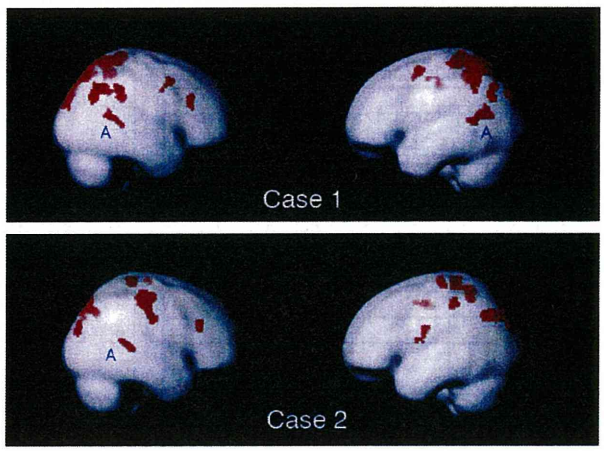

Fig. 2. Cortical activation by language-related visual stimuli in two profoundly deafened with congenital CMV infection cases. Case 1 and 2 showed significant activation in auditory association areas (A) (SPM2, $p < 0.001$, uncorrected).

Table 2
The results of total development before and after cochlear implant, and auditory assessment.

	Before CI	One year after CI	
	K-test (Cognitive-Adaptive)	WISC-III (Japanese version)	Infant word and sentence discrimination
Case 1	99	PIQ 101 VIQ 84	Word 98% Sentence 90%
Case 2	93	PIQ 93 VIQ 56	Word 100% Sentence 53%

CI, cochlear implant; *K*-test, Kyoto scale of psychological development; WISC, Wechsler Intelligence Scale for Children; PIQ, performance IQ; and VIQ, verbal IQ.

better as well in the sentence discrimination component of the auditory perception ability assessment while their results were similar regarding words in the word and sentence discrimination test for children.

4. Discussion

This is the first report on the evaluation of cortical processing of language in hearing loss children with congenital CMV infection. In infants with congenital CMV infections, as many as 20% will suffer from some degree of SNHL, either fluctuating or progressive [14]. This may present a late onset hearing loss, even if the results of newborn hearing screening were normal. The clinical courses of hearing loss in Cases 1 and 2 were typical for asymptomatic congenital CMV infection. Performance and outcome of children with CIs have a strong relation to hearing variables such as onset and course of hearing loss, age of hearing aids fitting, and social background variability, which depends on habilitation and education. According to Fukushima et al. and Kawasaki et al., children with *GJB2* mutations as the etiology for hearing loss have an advantage in their CI outcomes and speech acquisition with normal cognitive development compared with children with unknown etiologies, but this is because the hearing loss is of cochlear origin [15,16]. On the other hand, widely varying conclusions regarding CI outcomes with congenital CMV infection have been reported. Some studies reported the efficacy being not inferior to that of other CI recipients, while others reported it being much poorer [9,17–20]. Accordingly, prediction of CI outcomes for hearing loss with CMV infection is still difficult, unclear, and inconsistent because of various manifestations, progression and the possibility of involvement of higher brain function. Yamazaki et al. suggested that CI with CMV infection outcomes vary widely depending on the psycho-neurological disorders, with their differences in proportion and severity [19].

In this study, the auditory association area in the temporal lobe was activated bilaterally in Case 1 and unilaterally in Case 2. Fujiwara et al., in a FDG-PET study using the same methods and tasks as the present study, showed that subjects with better spoken language skills had less temporal lobe activation [7]. These cases exhibited nearly identical cortical activation patterns to those of congenitally deafened children, suggesting that they did not have enough hearing to develop the cortical network for audition. Previous studies have suggested that plastic changes in auditory cortices were strongly determined by the duration of auditory deprivation [21,22]. However, our two cases of children with CMV-related hearing loss were affected with severe bilateral hearing loss over a short period and were able to acquire spoken language with only a little delay for their age group. It is an interesting but unsolved question why they exhibited results that were the same as previous reports of pre-lingually deafened patients who did not receive sufficient auditory signals and therefore depended on visual cues. One possibility was that high speech-reading activation in the temporal auditory area might be linked to the condition of lacking auditory speech skills at that point, rather than reflecting a consequence of replacement by visual cross-modal plasticity due to a hearing loss of long duration. Besides, visual language activation in the auditory area may change even if affected by hearing loss of a short duration, or it might be influenced by the age-related metabolic changes during the critical period for spoken language acquisition. Another possibility was that these results might indicate that both cases had not received sufficient hearing stimulation as a foundation of language during their early years, which may be attributed to the central nervous system impairment of CMV infection.

Regarding the results of assessment after CI, there was a difference of cognitive ability with VIQ and hearing ability of sentence discrimination, with Case 1 having better CI performance than Case 2 (Table 2). In the assessment of auditory performance, Case 2 especially had difficulty in sentence discrimination despite having the same score in word discrimination as Case 1, who had better CI performance. Sentence discrimination tests require not only audible sound coded by CI, but also recognition of semantics and syntax that would be developed and established with hearing experiences during growth. Indeed, because of the differences between our two cases of the brain imaging, especially in the auditory cortex, we were uncertain whether it might be affected by CMV infection or the onset of their hearing loss itself. However, it raised the possibility that involvement of central nerve and high brain function relevant to CMV infection may lead to retardation of sentence discrimination and speech acquisition in Case 2. On the other hand, there was a difference of activation patterns in the parietal visual association areas. Case 2 showed lower and smaller than in Case 1. Fujiwara et al. predicted that the children with deafness were likely to depend more on vision than normal hearing children do. In Case 1, when someone talked to her, she might have been able to pay much more attention to their facial expression, gestures and visual cues for understanding better than Case 2. Lee et al. reported the comparison of brain metabolic activity between good and poor CI outcomes [23]. The activity patterns in the parietal regions of those with good CI outcomes in their study were similar to our result in Case 1.

We considered that these results might indicate that the differences of cortical activities according to hearing abilities could have been influenced by CMV infection that involves higher function of the brain directly and/or affects the cochlea peripherally. Additionally, if CMV infection might have affected only the

cochlea, these cortical activation patterns were influenced secondary by the time course of hearing loss characterized by CMV infection, which had varied manifestations.

Accurate diagnosis of hearing loss and early cochlear implantation are important for successful speech development. The approach using PET could help those involved in the habilitation and education of pre-lingually deafened children to decide upon the appropriate mode of communication for each individual. Brain imaging technologies to evaluate the neural basis for auditory speech skills have been developed and much evidence has been reported; however, correlation with hearing loss etiology, pathology and cross-modal plasticity of auditory cortex remains contentious. Further evaluations of the cortical metabolism before and after implantation are necessary for establishing appropriate personalized audiologic rehabilitation programs for individuals based on their etiology and brain function.

Conflicts of interest statement

We, the authors, declare that there were no conflicts of interest in conjunction with this paper.

Acknowledgements

We thank A.C. Apple-Mathews and Nora Prachar for help in preparing the manuscript. We also thank Masanori Sakaguchi, M.D. and radiologic technologists, Kouichi Anraku and Hiroyuki Fujimoto, for excellent technical assistance.

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Acta Oto-Laryngologica. 2014; 134: 58–65

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ORIGINAL ARTICLE

Clinical features of rapidly progressive bilateral sensorineural hearing loss

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Abstract

Conclusion: Rapidly progressive bilateral sensorineural hearing loss (SNHL) often develops as a symptom of intracranial diseases or systemic vasculitis. For early diagnosis and treatment of these potentially fatal diseases, a history of hearing deterioration within 2 months and associated symptoms may be important. *Objectives:* To reveal clinical features and causative diseases for rapidly progressive bilateral SNHL. *Methods:* The inclusion criterion was patients with bilateral progressive SNHL, who had experienced difficulty in daily conversation within 4 days to 1 year after the onset of hearing loss awareness. This study was a retrospective evaluation of 12 patients with rapidly progressive bilateral SNHL who visited our hospital between 2007 and 2011. *Results:* The causative disease for hearing loss was identified in 11 of 12 patients; intracranial lesions including nonbacterial meningitis, meningeal metastasis of lymphoma, and superficial siderosis in 4 patients, systemic vasculitis in 2, auditory neuropathy spectrum disorder in 1, and an isolated inner ear disorder in 4. Relatively rapid hearing deterioration within 2 months showed a significant association in six patients with an intracranial lesion or systemic vasculitis. Moreover, all these six patients complained of dizziness and/or non-cochleovestibular symptoms such as fever, headache, and/or altered mental state in addition to hearing loss.

Keywords: *Auditory perception, intracranial disease, systemic vasculitis, magnetic resonance imaging, hearing threshold*

Introduction

Sensorineural hearing loss (SNHL) is caused by various disorders, including sudden deafness, presbycusis, hereditary hearing loss, drug-induced hearing loss, and Meniere's disease. Various clinical data are used to diagnose the cause of SNHL, of which the time course of hearing deterioration may be particularly important for estimating the nature of the disorder. For example, sudden deafness has an onset period of < 72 h [1], while presbycusis deteriorates by 1–2.5 dB per year over a long period of time. We also encounter patients with bilateral SNHL whose hearing deteriorates more slowly than that in sudden deafness but more quickly than that in presbycusis. Such patients often have serious complicating diseases, although only a few studies have examined this type of hearing loss. In this study, we report 12 cases of rapidly progressive bilateral SNHL and analyze the clinical features and causative diseases for hearing loss.

Material and methods

The study was a retrospective review of medical records. Of the 908 patients diagnosed with bilateral SNHL who visited the Department of Otolaryngology at Kobe City Medical Center General Hospital from

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(Received 17 June 2013; accepted 30 July 2013)
ISSN 0001-6489 print/ISSN 1651-2251 online
DOI: 10.3109/00016489.2013.831993

Table I. Characteristics of 12 patients with rapidly progressive bilateral sensorineural hearing loss.

Case no.	Onset (age in years)	Time from onset to difficulty in daily life (days)	Gender	Causative disorder	Category of causative disorder	Worst hearing (dB)		Hearing after treatment (dB)		Clinical symptoms
						R	L	R	L	
1	33	4	M	Cryptococcal meningitis	Intracranial lesion	115	115	68.3	25	Fever, headache, altered mentation, dizziness
2	45	60	M	Chronic herpes meningitis + labyrinthitis		115	108.3	No improvement		Fever, tinnitus
3	60	6	M	Meningial metastasis of lymphoma		75	50	45	48.3	Fever, dizziness
4	79	30	F	Superficial siderosis		65	61.7	No improvement		Dizziness, tinnitus
5	73	45	F	Cogan's syndrome	Systemic vasculitis	115	115	No improvement		Fever, headache, dizziness
6	44	4	F	Vasculitis syndrome		93.3	81.7	51.7	38.3	Fever, headache, altered mentation
7	26	7	F	Auditory neuropathy	ANSD	115	113.3	No improvement		Tinnitus
8	63	120	F	Isolated inner ear disorders	Isolated inner ear disorder	65	56.7	No improvement		Tinnitus
9	67	90	M	Isolated inner ear disorders		103.3	103.3	No improvement		Tinnitus
10	69	360	M	Isolated inner ear disorders		95	115	No improvement		Tinnitus
11	69	360	F	Isolated inner ear disorders		80	73.3	No improvement		Tinnitus
12	61	14	F	Undefined disorder	Undefined	53.3	55	41.7	41.7	Fever, backache

January 2007 to December 2011, 12 (1.3%, 5 males and 7 females; Table I) who met the following criteria for rapidly progressive bilateral SNHL were selected: (1) pure-tone audiometry data showing bilateral SNHL and average hearing thresholds at 500, 1000, and 2000 Hz of ≥ 50 dB; (2) difficulty in daily conversation without lip-reading or sign language within 4 days to 1 year after the onset of hearing loss awareness; and (3) exclusion of cases with bilateral Meniere's disease or functional hearing loss. Wegener's granulomatosis [2], Churg-Strauss syndrome [3], and eosinophilic otitis media [4], are also known to induce progressive hearing loss, but were excluded from this study because these diseases lead to mixed hearing loss rather than SNHL. The median age at onset of hearing loss was 62 years (range 26–79 years). The precise deterioration speed of the patients' pure-tone audiometric thresholds could not be calculated because most of them came to our hospital after having moderate or severe SNHL and their initial pure-tone audiometry thresholds before the onset of hearing loss had not been tested. Therefore, we defined progressive bilateral SNHL on the basis of subjective time course of deterioration in auditory perception.

The diagnoses of causative diseases of rapidly progressive bilateral SNHL were based on medical interviews, physical findings, and examinations by otologists, internal medicine specialists, and radiologists. The examinations included blood autoantibody tests, microbiological culture tests, radiographic examinations (CT and MRI), and cerebrospinal fluid (CSF) tests, as well as conventional otological examinations including pure-tone audiometry, distortion product otoacoustic emissions (DPOAEs), and auditory brainstem response (ABR). The causative diseases were categorized into five groups: (1) an intracranial lesion for which CT, MRI, and/or CSF tests revealed an abnormality in the central nervous system; (2) systemic vasculitis, diagnosed by positive blood tests for autoantibodies and systemic inflammation and vasculitis-specific skin lesion, retinal vasculitis, or non-syphilitic interstitial keratitis; (3) auditory neuropathy spectrum disorder (ANSD), diagnosed on the basis of good responses in DPOAE and a lack of obvious responses in ABR; (4) isolated inner ear disorder, with no abnormality on CT or MRI scans and no symptoms other than cochleovestibular symptoms; and (5) an undefined disorder with symptoms other than cochleovestibular symptoms.

The time course of hearing deterioration was evaluated using subjective manifestations. The time course was defined as the time period from the onset of hearing loss awareness to the onset of difficulty in understanding speech in daily life, and it was classified as follows: (1) 4 days to 1 week, (2) 1 week to 1 month, (3) 1–6 months, and (4) 6 months to 1 year. We also focused on clinical manifestations other than hearing loss, which were divided into cochleovestibular symptoms including tinnitus and dizziness and noncochleovestibular symptoms including fever, headache, and altered mental state.

Results

Clinical manifestations

The time course of hearing deterioration was from 4 days to 1 week in four patients, from 1 week to 1 month in two patients, from 1 to 6 months in four patients, and from 6 months to 1 year in two patients. The median hearing level (i.e. the worst value for each patient) of the 12 patients was 94 dB for the right ear and 93 dB for the left ear (Table I). With respect to manifestations related to noncochleovestibular disorders, fever was the leading symptom and was observed in six patients (50%). Among these patients with fever, three also complained of severe headache and two of these further suffered from altered mental state. Tinnitus was observed in seven patients including all six patients without noncochleovestibular symptoms. Dizziness was reported in four patients and three of these were also associated with a noncochleovestibular symptom, but the other complained of only tinnitus and dizziness (Table I).

MRI findings

Brain MRI was performed in nine patients including all six with a noncochleovestibular symptom, one with both tinnitus and dizziness, and two with tinnitus. Association of noncochleovestibular symptoms and dizziness with bilateral SNHL suggests the presence of systemic or intracranial lesions in the former and a retrocochlear or unusual inner ear disease in the latter. In fact, the diagnosis of an intracranial lesion or systemic vasculitis was confirmed or supported by MRI in five of seven patients with a noncochleovestibular symptom or dizziness (Figure 1). In case 4, T2-weighted MRI revealed superficial hypointensity on the surface of the brainstem and cerebellum, which was diagnosed as superficial siderosis. In the other four patients, gadolinium-enhanced T1-weighted MRI showed abnormal enhancement in the inner ear or internal auditory canal. In five cases complaining solely of tinnitus in addition to hearing loss, only two underwent brain MRI. In the other three cases, results of neurological examinations implied that the lesion was restricted in the cochleae and, therefore, careful follow-up of pure-tone audiometry, ABR,

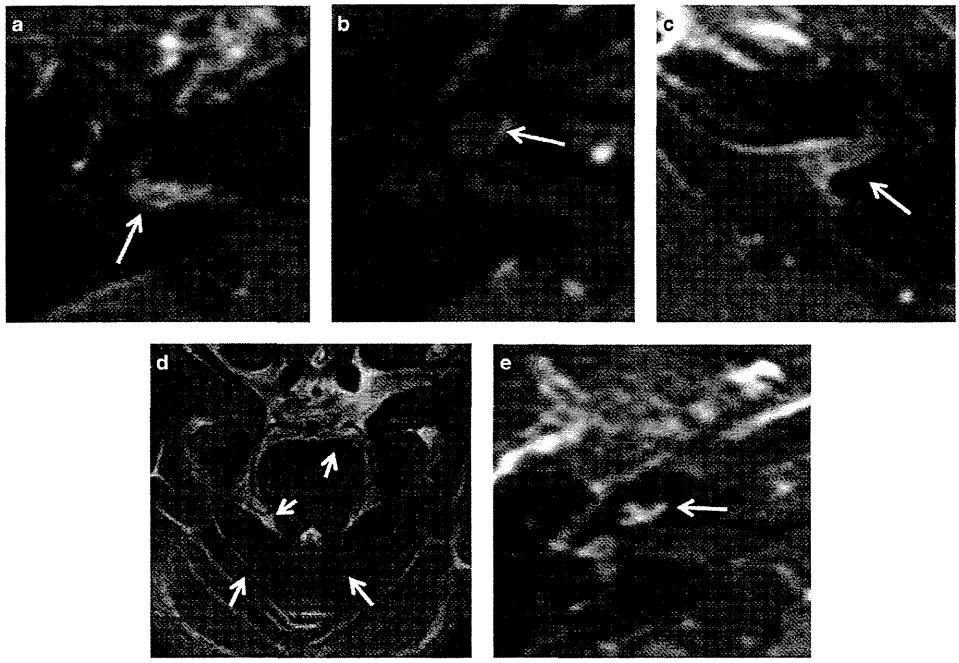

Figure 1. (a) Case no. 1. Cryptococcus meningitis with enhancement of bilateral internal auditory canal (IAC) on gadolinium-enhanced MRI. The enhanced right IAC is shown. (b) Case no. 2. Chronic viral meningitis plus labyrinthitis with enhancement of bilateral cochlea on gadolinium-enhanced MRI. The enhanced basal turn of the right cochlea is shown. (c) Case no. 3. Meningeal metastasis of lymphoma with enhancement of bilateral IAC on gadolinium-enhanced MRI. Enhanced left IAC is shown. (d) Case no. 4. Superficial siderosis with hypointensity along the brainstem and cerebellum on T2-weighted MRI. (e) Case no. 5. Cogan's syndrome with enhancement of bilateral cochlea on gadolinium-enhanced MRI. The right whole cochlea is enhanced.

DPOAE, and/or blood tests for autoimmune antibodies rather than brain MRI were conducted to evaluate cochlear disorders.

Categories of causative diseases

The causative diseases for hearing loss are shown in Table I. Systemic evaluation showed abnormalities restricted to the inner ear in four patients (isolated inner ear disorder). Intracranial lesions were detected in four patients and systemic vasculitis in two, with these disorders diagnosed as the causes of bilateral SNHL. The intracranial lesions included Cryptococcus meningitis, chronic meningitis due to herpes simplex virus, meningeal metastasis of lymphoma, and superficial siderosis. The two patients with systemic vasculitis were diagnosed with Cogan's syndrome and Sjögren syndrome with aseptic meningitis, retinal vasculitis, and skin lesions.

Relationship between category of causative diseases and clinical manifestations

The time course for deterioration in auditory perception was ≤ 60 days in the six patients with an

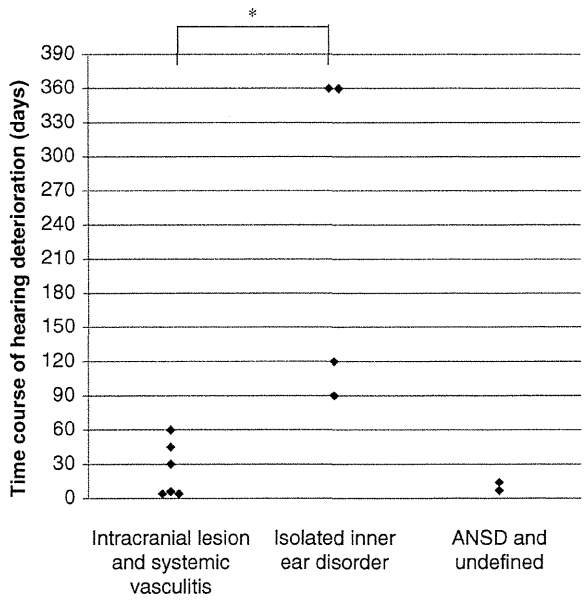

Figure 2. Time course of hearing deterioration in different categories of causative disorders. There was a significant difference between patients with intracranial lesion and systemic vasculitis, and those with an isolated inner ear disorder.
*: $p < 0.05$

Table II. Characteristics of six patients with an intracranial lesion or systemic vasculitis.

Case no.	Diagnosis	Treatment	Time before treatment (days)	Hearing improvement
1	Cryptococcal meningitis	Antifungal drug	3	Improved
2	Chronic herpes meningitis + labyrinthitis	Steroid and anti-HSV agents	Unknown	Not improved
3	Meningial metastasis of lymphoma	Steroid and anticancer drug	6	Improved
4	Superficial siderosis	No treatment		Not improved
5	Cogan's syndrome	Steroid	90	Not improved
6	Sjögren syndrome	Steroid	4	Improved

intracranial lesion or systemic vasculitis and ≥ 90 days in the four patients with an isolated inner ear disorder. The Mann–Whitney U test showed a significant difference ($p < 0.05$) between these groups (Figure 2). As shown in Table I, all patients with an intracranial lesion or systemic vasculitis complained of dizziness and/or noncochleovestibular symptoms in addition to hearing loss. Four of these six patients had dizziness and five of them had fever, headache, or altered mental state. These symptoms were not observed in patients with ANSD or an isolated inner ear disorder, who had only tinnitus as an associated symptom.

Hearing improvement after treatment for the causative diseases

The causative disease was treated in five patients with an intracranial lesion or systemic vasculitis, except in case 4 who had superficial siderosis (Table II). Hearing improved in three patients, who did not require hearing aids in daily life. The delay from the onset of hearing loss awareness to the beginning of treatment was within 1 week in cases 1, 3, and 6, who showed an improvement in hearing. However, it took as long as 90 days in case 5, who showed no change in hearing threshold after treatments. In case 4, the origin of bleeding that caused hemosiderosis was not determined despite radiographic evaluations, including brain and spinal MRI, and the patient showed no improvement in hearing at follow-up. Improvement in hearing loss did not occur in any of the patients with ANSD or an isolated inner ear disorder, despite systemic administration of steroids and/or circulation activators.

Discussion

This study was performed as a retrospective review of 12 cases with progressive bilateral SNHL who complained of difficulty in daily conversation within 4 days to 1 year after the onset of hearing loss awareness. The patients with bilateral SNHL presenting this time course of deterioration were relatively rare and accounted for only 1.3% of those with bilateral SNHL in this study. However, retrospectively, distinguishing this type of SNHL from others was meaningful because 6 of these 12 patients (50%) developed SNHL from an intracranial lesion or systemic vasculitis, which can be fatal without appropriate treatment. It is also noteworthy that all three patients with an intracranial lesion or systemic vasculitis, who showed improvement in hearing, underwent early treatment of the causative diseases, suggesting that accurate diagnosis and appropriate treatments for the causative disease at its early stage may be important to restore hearing as well as to lower the mortality. In the present study, the rapidly progressive SNHL was also caused by ANSD or an isolated inner ear disorder, but clinical manifestations of intracranial lesions and systemic vasculitis were different from those observed in other categories of causative diseases. Our study showed that in patients with intracranial lesions and systemic vasculitis, the time from onset of hearing loss to difficulty in daily life was within 2 months and significantly shorter than that in patients with an isolated inner ear disorder. In addition to the rapidly progressing hearing loss, noncochleovestibular symptoms and/or dizziness were always associated with intracranial lesions and systemic vasculitis, while all five patients with an isolated inner ear disorder or ANSD complained of only tinnitus. Among noncochleovestibular symptoms, fever was the leading symptom (6 of 12 patients), followed by headache and an altered mental state. In all cases with fever, the origin of fever was difficult to identify at first and systemic inflammation or intracranial infection was identified later based on systemic evaluations by otologists, internal medicine specialists, and radiologists. The presence of headache and an altered mental state also suggests that lesions may

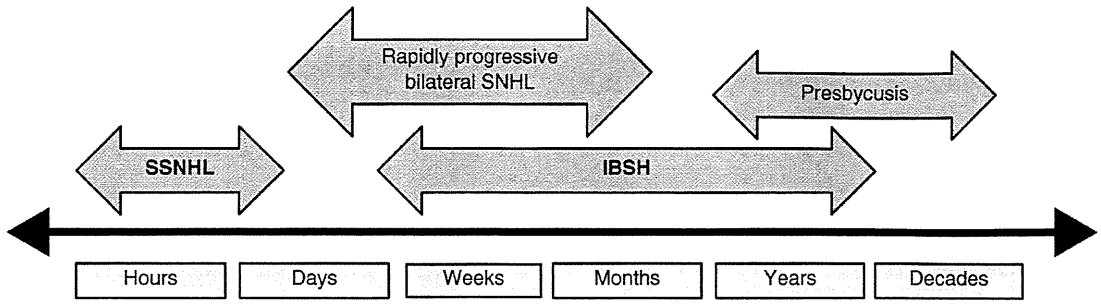

Figure 3. The time course in various types of bilateral sensorineural hearing loss (SNHL). IBSH, idiopathic bilateral SNHL; SSNHL, sudden SNHL.

involve other areas of the central nervous system in addition to the auditory neural pathway. Interestingly, obvious vestibular dysfunction was not observed in patients with an isolated inner ear disease, although four of the six patients with an intracranial lesion or systemic vasculitis had dizziness. The inner ear lesions in the present series may have been limited to the cochlea, with central compensation possibly making the vestibular symptoms less prominent despite the presence of some vestibular involvement.

We performed brain MRI in nine patients including all seven with a noncochleovestibular symptom or dizziness. Headache, altered mental state or other abnormal neurological findings in addition to the eighth cranial nerve dysfunction suggests the presence of an intracranial lesion. In this situation, brain MRI is necessary to evaluate intracranial diseases. Even though the neurological disorders were limited to the eighth cranial nerve, association of dizziness with SNHL might be caused by labyrinthitis or lesions in internal auditory canals and brain MRI may be recommended. Prolonged unknown origin of fever associated with bilateral SNHL is also an indication for brain MRI to evaluate labyrinthitis and nonbacterial meningitis.

In the present study, pure-tone hearing thresholds were improved in case 1 with Cryptococcus meningitis and case 3 with meningeal metastasis of lymphoma after the intracranial administration of antifungal and anticancer drugs, respectively. Hearing recovery is usually difficult in patients with Cryptococcus meningitis [5], although a patient with this disease was reported to show partial recovery of hearing after treatment [5]. Hearing improvement after treatment has also been reported in patients with bacterial and viral meningitis [6,7]. Vasculitis causes SNHL in patients with connective tissue diseases such as systemic lupus erythematosus and polyarteritis nodosa [8], with this type of hearing loss reported to improve following plasmapheresis or immunosuppressive therapy using steroids or cyclophosphamide [2,9]. In our study, case 6, who had Sjögren syndrome, showed hearing improvement after steroid treatment. In contrast, hearing loss in case 5, who had Cogan's syndrome, was not improved by steroids. Although hearing improvement has been described in a patient with Cogan's syndrome [10], it is often difficult to improve hearing loss in such patients.

Previous case reports indicate that the etiology of bilateral SNHL, which deteriorates more slowly than sudden deafness and more quickly than presbycusis, also includes meningeal carcinomatosis [11], metastasis of carcinoma in the bilateral internal auditory canal [12], mitochondrial neurogastrointestinal encephalopathy (MINGIE) [13], and polyarteritis nodosa [14]. These diseases were not found in the present study due to the small size of the study. The rapidly progressive bilateral SNHL can be induced by various types of diseases with different etiologies described above and, moreover, within each type of a disease, severity of symptoms may vary widely between patients. Therefore, further study investigating more patients with rapidly progressive bilateral SNHL is needed to lead to definite conclusions about the importance of clinical manifestations and indications for MRI for diagnosis of the causative diseases.

The definition of rapidly progressive SNHL in previous reports varies, including SNHL deteriorating in days [15] or in weeks to months [14,16–18]. However, the disease entity described in these reports is almost identical, which is the SNHL that progresses more slowly than sudden deafness and more rapidly than presbycusis. Thus, in line with those previous reports, we defined rapidly progressive SNHL as the one that deteriorates in days to months. The time course of rapidly progressive bilateral SNHL compared with that of other types of common bilateral SNHL is illustrated in Figure 3. Idiopathic bilateral SNHL (IBSH) is a progressive bilateral SNHL of unknown etiology and

was proposed as a clinical entity in 1976. In IBSH, hearing loss usually progresses over several years; therefore, deterioration in hearing loss is slower than that observed in the current patients [19], suggesting different etiologies. In the current study, the four patients with isolated inner ear disorders showed a significantly slower deterioration in hearing loss compared with the other patients. IBSH sometimes shows rapid progression of hearing loss within several days or weeks; therefore, patients with similar pathology to that observed in IBSH could meet our criteria for rapidly progressive bilateral SNHL if they visit a hospital in the rapid phase of the disease.

A noteworthy aspect of the patients reported in this study was that early treatment of intracranial lesions and systemic vasculitis improved hearing loss, suggesting the importance of early diagnosis of the causative disease, although further investigation of large numbers of patients is necessary to prove the effectiveness of early treatment. Early diagnosis is also important because the causative diseases for rapidly progressive bilateral SNHL include fatal conditions such as meningitis or malignant diseases, or diseases that may result in irreversible neurological deficits such as superficial siderosis. In patients with superficial siderosis, decreasing the risk for a poor outcome requires early diagnosis of the disease and identification and ablation of the bleeding source [20].

Conclusion

Rapidly progressive bilateral SNHL is rare, but it often develops as a symptom of intracranial disease or systemic vasculitis, both of which are potentially fatal. Hearing may recover in patients who undergo treatment at an early stage of the causative disease. This indicates that early diagnosis followed by appropriate treatment of the causative disease is critical for the management of these patients.

Acknowledgments

We would like to thank Dr Michi Kawamoto and Dr Nobuo Kohara in our institute for advice about diagnosis and treatment of patients. This study was supported by a Grant-in-Aid for Scientific Research (C) (22591894) and a Grant-in-Aid for Young Scientists (B) (22791642) from the Japanese Ministry of Education, Culture, Sports, Science, and Technology.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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Notice of correction

The Early Online version of this article published online ahead of print on 21 Nov 2013 was missing information about the authors.
The corrected version is shown here.

HRCT-BASED PREDICTION FOR COCHLEAR IMPLANT OUTCOMES OF CASES WITH INNER EAR AND INTERNAL AUDITORY CANAL MALFORMATIONS

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Introduction

Inner ear and internal auditory canal (IAC) malformations account for approximately 20-35% of congenital sensorineural hearing loss[1,2] and an increasing number of children with inner ear and/or IAC malformations underwent cochlear implantation. According to Sennaroglu's classification of inner ear malformations, which is the most widely accepted, the inner ear malformations are divided into labyrinth aplasia, cochlear aplasia, common cavity (CC), incomplete partition type I (IP-I), type II (IP-II), and type III (IP-III), cochlear hypoplasia type I (CH-I), type II (CH-II), and type III (CH-III), and large vestibular aqueduct syndrome (LVAS).[1,3] This classification is essential to investigate the etiology of the inner ear malformations, but with respect to predicting cochlear implant (CI) outcomes, it might not be enough, because it does not include IAC malformations such as narrow IAC (NIAC) and hypoplasia of the bony cochlear nerve canal (HBCNC). These IAC malformations are highly associated with cochlear nerve deficiency (CND), which has a negative impact to CI outcomes.[4,5]

The purpose of this study was to establish a new CT-based categorization which is simple and includes both inner ear and IAC malformations for predicting CI outcomes.

Materials and methods

Between 2004 and 2010, 98 subjects who were under 20 years old underwent cochlear implantation at Kobe City Medical Center General Hospital. Among them, CT revealed that 24 subjects had inner ear and/or IAC malformations at the implanted side.

We evaluated inner ear and IAC malformations at the implanted side based on CT findings. Sennaroglu's classification was used to classify inner ear malformations and the IAC malformations were classified into NIAC and HBCNC. NIAC was diagnosed when the maximum diameter of the IAC was less than 2 mm.[2] The width of the bony cochlear nerve canal (BCNC) was evaluated at the mid-portion between the base of the modiolus of a cochlea and the fundus of the IAC on axial images. When the diameter of the BCNC is less than 1.5 mm, it is diagnosed as HBCNC.[5] CND was diagnosed when a cochlear nerve (CN) appeared smaller than the facial nerve on the parasagittal MR imaging.

We categorized inner ear and IAC malformations into four groups by two criteria: (1) the presence or absence of a bony modiolus in the cochlea; and (2) the diameters of IAC and BCNC. In this categorization, both Group 1 and Group 3 have a bony modiolus in the cochlea, while Group 2 and Group 4 lack this component. Both IAC and BCNC are normal in Group 1 and Group 2, but NIAC or HBCNC was observed in Group 3 and Group 4. Sennaroglu's classification of inner ear malformations clearly discriminates between

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Cholesteatoma and Ear Surgery – An Update, pp. 371-373
Edited by Haruo Takahashi

the presence and absence of a bony modiolus in the cochlea. According to his classification, a bony modiolus is present in IP-II, CH-III, LVAS, and a normal inner ear, while CC, IP-I, IP-III, CH-I, and CH-II have a cystic cavity without a bony modiolus.[3]

We evaluated CI outcomes by category of auditory performance (CAP) scores,[6] hearing thresholds of pure-tone sounds, infant word speech discrimination scores, and monosyllabic word speech discrimination scores at one to three years after implantation. A subject with 0-4 CAP scores could not even understand common phrases without visual language and, therefore, we defined 5-7 CAP scores as a good CI outcome and 0-4 CAP scores as a poor one.

Results

We categorized our patients based on the two criteria as described above. In this study, there was no case categorized in Group 4. Group 1, Group 2, and Group 3 consisted of 11, 7, and 6 cases, respectively. MR imaging revealed CND in all cases of Group 3.

The post-operative CAP score was equal or over five in all cases of Group 1, but did not exceed four in all of Group 3. In Group 2, the post-operative CAP score was still four in two cases even after three years of CI usage, but reached to five or six in the remaining five cases. As shown in Figure 1, using our new categorization instead of the existing classifications, we can better discriminate between a good and poor outcome.

We examined speech discrimination scores of 22 cases except for two cases of Group 3 whose response to voice was poor. The correct percentage of the closed-set infant word discrimination test was ≥ 80 in all cases of Group 1, while the score ranged from 40 to 60 in tested cases of Group 3. The correct percentage of Group 2 widely varied between cases, ranging from 55 to 100. The open-set monosyllabic word discrimination test is much more difficult than the closed-set infant word discrimination test and, therefore, only 17 of 24 patients, who were over five years old and used their CI for more than two years, underwent this examination. All tested cases of Group 1 and 3 cases of Group 2 could answer correctly in equal or over 80% of accuracy. The correct percentage of the remaining cases, including all tested cases of Group 3, was ≤ 30.

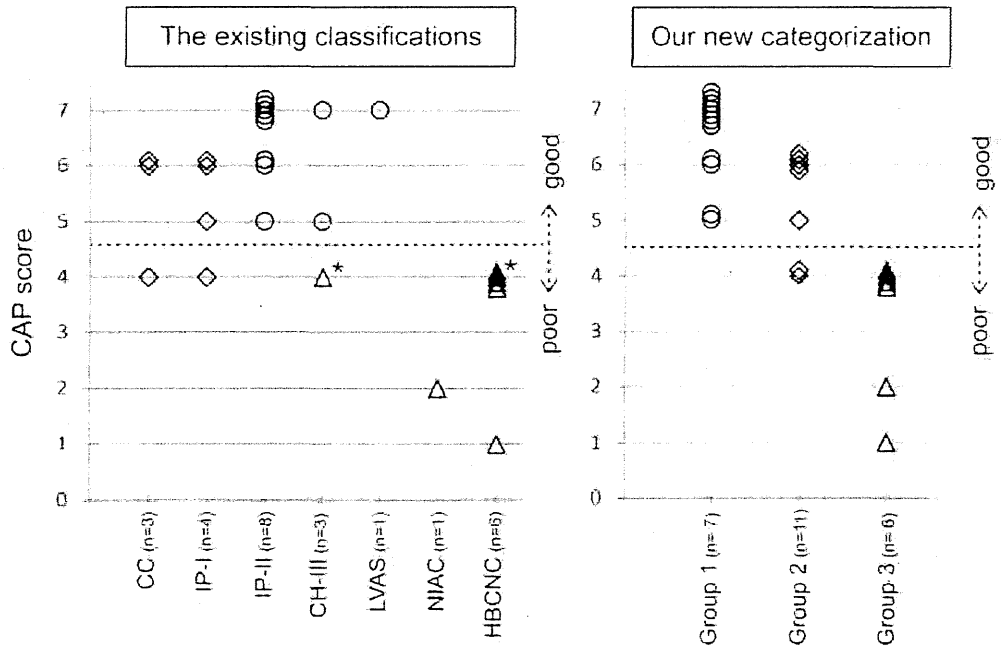

Fig. 1. A. The post-operative CAP score of each type of malformations based on the existing classifications. One case with both CH-III and HBCNC is plotted twice (*). B. The post-operative CAP score of each group of our new categorization. In both graphs, the members of Group 1, Group 2, and Group 3 are represented by a circle, diamond, and triangle, respectively.

Discussion

In this study, we established a new CT-based categorization including both the inner ear and IAC malformations. This categorization is defined by two criteria; (1) the presence or absence of a bony modiolus in the cochlea; and (2) the diameters of IAC and BCNC. We focused on these structures because the bony modiolus contains spiral ganglion cells, the major target of CI-mediated electrical stimulation, and their axons go through BCNC and IAC.

Group 1, which is defined by the presence of a bony modiolus of the cochlea with a normal IAC and BCNC, showed the best CI-aided hearing performance among three groups. The high proportion of post- or peri-lingually deaf cases might also contribute to the high CI outcomes of this group.[7] Group 2 is defined by the absence of a bony modiolus with a normal diameter of IAC. The CAP score and speech discrimination score varied widely between cases in this group, but five out of seven cases could understand common phrases without visual languages. Group 3 is defined by the presence of a bony modiolus in the cochlea with NIAC or HBCNC and their post-operative improvement of hearing performance was limited. Visual languages were necessary for them to understand common phrases even after long usage of their CI. MR imaging revealed CND in all cases of Group 3, which might be responsible for their poor outcomes.

Conclusion

Our new CT-based categorization, which was based on the presence or absence of a bony modiolus in the cochlea and the diameters of IAC and BCNC, was effective in predicting CI outcomes of children with inner ear and/or IAC malformations. The CI outcomes were the best in Group 1, followed by Group 2 and Group 3. All cases of Group 1 showed good CI outcomes and could communicate orally. On the other hand, all cases of Group 3 showed poor CI outcomes and used lip-reading or sign language to understand common phrases. The CI outcomes of Group 2 varied between cases, but many of them showed good CI-aided hearing performance.

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小さな common cavity 例の人工内耳手術

内藤 泰

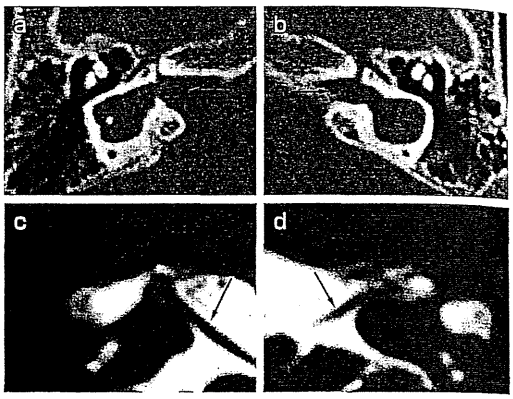
小さな common cavity 例の手術は難しい

人工内耳手術は内耳奇形例でも可能であるが，蝸牛と前庭が分離せずに1つの腔になっている common cavity 奇形では内耳形態にさまざまなバリエーションがあり，個々の症例に応じた工夫が必要である[1]．本項では，内耳奇形の人工内耳のなかでもとくに難度が高い，小さな common cavity 例の手術について述べる．

症例は女児で，言語発達の遅れにより耳鼻科を受診し両側高度難聴の診断が確定した．その後の補聴器装用で効果が得られず，当科紹介となった．2歳5か月時の所見で，聴性定常反応（ASSR）で両側無反応．乳幼児有意味聴覚統合スケール（IT-MAIS）は2点（40点満点），新版K式発達検査では，認知適応領域の発達指数（DQ）104に対して，言語社会領域の DQ が48と低い成績であった．

画像検査所見

側頭骨 CT では両側とも common cavity 奇形があり，内耳道から内耳まで軟部組織陰影が連続している（❶a, b→）．MRI では，両側で第8脳神経が明瞭に観察される（❶c, d→）が，蝸牛神経と前庭神経の分離は確認できない．内耳道と内耳腔のあいだの隔壁は MRI でも不明瞭で，内耳開窓で gusher（脳脊髄液の噴出）をきたす可能性がある．cavity は右のほうが若干大きいので，右側の手術を行う方針とした．



❶ 側頭骨の画像検査所見（a, c：右，b, d：左）

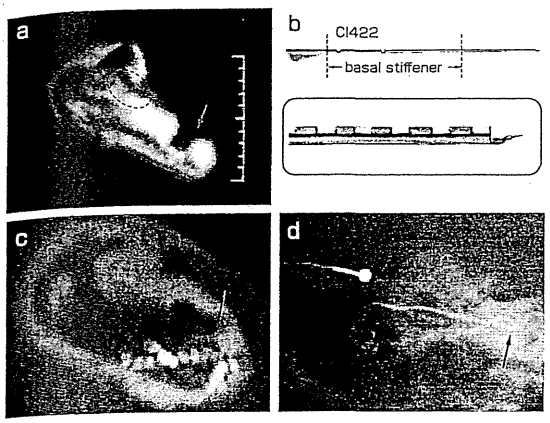
cavity の大きさを計測して電極を選択する

右内耳の三次元再構築 MR 像（❷a）をみると，cavity 前方の蝸牛相当部分の直径は3 mm 程度であり，その後方に顔面神経迷路部に該当する内腔の切れ込み（❷a→）がある．このように小さな空間に電極を敷設するには，できるだけ細い電極が有利と考え，コクレア社の CI422 電極（❷b，拡大図は電極アレイ先端部分の形状）を選択した．この電極は先端付近の直径が0.3 mm，根元が0.6 mm と細く，アレイの片側だけに電極がある half band 構造になっている．内耳奇形例で通常用いられる同社のストレート電極の先端付近の直径は0.4 mm と若干太く，また電極が全周にある full band 構造になっているので，狭い空間内では電極同士の接触によるショートの可能性もある．このため，本例では CI422 電極を選択し，通常とは逆に電極面を外にして弯曲させ，cavity 内腔壁に密着するように敷設する計画とした．

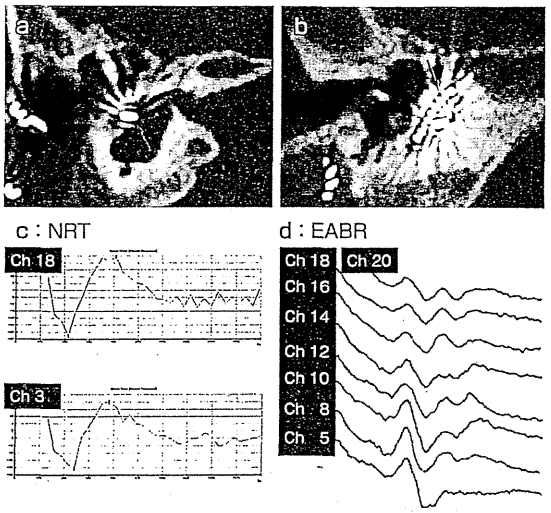
手術時の留意点

小さな common cavity では，できるだけ広い手術視野を確保し，cavity 内を明視して電極を入れないと思いどおりの位置に電極が入らない．本例では外側半規管隆起の前端付近に径3～4 mm の大きな開窓を行って内腔を観察した．gusher をきたしている所が内耳道底と考えられ，その前方がおおむね cavity の蝸牛該当部分であると推測した．

実際には cavity のどの部分に蝸牛神経が分布しているかわからないので，内腔の前端を中心にできるだけ広い範囲をカバーできるように，電極を中央で曲げて挿入し，先端の小さな空間だけでなく，その後方の cavity にも電極が触れるように工夫した．この電極の根元寄りには電極を若干固くする basal stiffener という構造がある（❷b）



❷ 術後検査所見
a：三次元再構築 MR 像，b：使用した電極，c：術後の傍冠状断平均 CT 画像，d：術後の単純 X 線像



❸ 術中・後の検査所見
a：cavity への入口部（矢印）の CT 像，b：cavity 内の電極，c：NRT 波形，d：EABR 波形

が，この適度の硬さが狭い空間内で電極を操作するのに役立った．なお，gusher は筋肉や筋膜片を cavity 内に充填することで制御できた．

手術結果

術後の CT（❷c，傍冠状断平均 CT 画像）では，計画どおり，cavity 前端付近を中心に内腔壁に密着して電極アレイを敷設できていることが確認された（❷c→）．アレイの先端と根元は前端の小さな空間から後ろにはみ出て伸びている．単純 X 線像では，アレイの固い部分の前端が狭い内腔の前端に位置しているように見える（❷d→）．軸位断 CT 像では，電極アレイが cavity の外側中央付近から挿入され（❸a→），前半部分の内腔に密着して敷設できているのが観察できる（❸b→）．

術中の電気生理学的検査（反応波形を❸c，d に示す）では，NRT で 2 番から 22 番，EABR で 5 番から 21 番電極において反応が確認された．術前に蝸牛相当部分と予想していた空間内にはおおむね 7 番から 17 番電極が収まっているが，実際にはその後方にも蝸牛神経が分布していたことがわかる．とくに cavity 内腔の下面ではほぼ電極先端まで反応があり，蝸牛神経支配がかなり尾側後方まで及ぶと推測され，今後，同様症例の手術を行ううえで参考になる．

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1）内藤　泰．内耳奇形例における人工内耳手術手技の留意点．神崎　仁編．1. 耳疾患．耳鼻咽喉科・頭頸部外科診療のコツと落とし穴．東京：中山書店；2006. p. 167-9.

プレークスルーのポイント

- 小さな common cavity 例では術前の CT，MRI 検査で内腔の大きさを計測し，現在臨床使用できる電極のうちどれが最もフィットするか十分検討することが大切である．
- 手術では cavity をできるだけ大きく開窓し，内腔を直視しながら，あらかじめ曲げた電極を内腔に密着するように敷設する．
- 術中に単純 X 線撮影と NRT や EABR 検査を行うと，電極アレイが適切に敷設できているか否か，cavity のどこに蝸牛神経が分布しているか確認できる．

ランチョンセミナー

治療の観点から見た耳疾患の画像診断

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Imaging of ear disorders seen from a viewpoint of treatment.

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Textbooks on medical imaging usually show key images for diagnosing a certain disease, but seldom describe its treatment and prognosis leaving them to clinical or surgical textbooks. We encounter many patients and diseases during our clinical practice. Each disease has its unique etiology, diagnostic process, treatment and its result, which cannot be understood well without seeing the stages after the diagnosis. A viewpoint of treatment is, thus, essential for clinically appropriate imaging diagnosis. This article reports cases of infectious and inflammatory ear diseases, temporal bone traumas and inner ear anomalies, in which not only their diagnostic key images but also their treatment findings and prognoses are described and discussed.

はじめに

一般の画像診断解説では画像による疾患診断までは示されるが，それに対する実際の治療法や治療結果には言及されることは稀であり，他の教科書や手術書などに委ねられるのが通常である．我々は臨床を続けるうちに多彩な疾患・患者に遭遇するが，個々の疾患には，それぞれの病因，診断，治療，予後という一連の流れがあり，各々についてその診断までを見ただけでは，全体像を把握することはできない．臨床的に適切な画像診断を行うためには，治療の観点が不可欠である．本稿では，耳疾患のうち，感染・炎症性疾患，側頭骨外傷，内耳奇形を取り上げ，各々の画像診断上の要点を治療も含めて概説する．なお，本稿では，筆者の著書「画像でみる耳の診断と治療－小児編」[1]で取り上げた症例をいくつか呈示している．読者には，あらかじめご承知いただくとともに，さらに詳細な所見等については同書をご参照いただきたい．

1．感染・炎症性疾患

中耳炎は日常臨床で最も頻繁に遭遇する耳疾患である．通常の急性中耳炎は臨床症状と鼓膜所見で十分な診断ができ，また短期に治癒するので，側頭骨CTなどの高度の画像検査は不要であるが，合併症を伴う急性中耳炎や慢性中耳炎では，

治癒を阻害している要因の探求や耳小骨連鎖の病態観察，手術の要否などの治療方針決定のために画像診断が必要になる．また，手術治療を行った症例では，術後の経過観察にも画像診断が大きな役割を果たす．

症例1：5歳　男児

主　訴：左耳痛，左側頭部の頭痛

現病歴：左耳痛で近医耳鼻科受診．急性中耳炎の診断で左鼓膜切開と抗菌薬投与を受けたが，翌日になっても耳痛が改善しなかった．発症2日後に，最寄りの市中病院を経て当科紹介となった．依然として左耳痛と頭痛を強く訴えている．

局所所見：左外耳道に淡血性耳漏あり．耳後部に発赤，腫脹なし．

検査所見：白血球10100/μl，CRP21.1mg/dl，意識清明．

CT所見：側頭骨ターゲットCT（Fig. 1：a）では，発育良好な乳突蜂巣全体に軟部組織陰影が充満しているが，骨破壊像は見られず，S状静脈洞周辺は均一な軟部組織陰影にしか見えない（Fig. 1:a→）．軟組織ウィンドウの頭部CT像（Fig. 1: b）では左S状静脈洞が腫脹していることが観察でき，血栓による静脈洞の閉塞腫脹と推測される．

MRI所見：腫脹した左S状静脈洞はT2強調像では低信号（Fig. 2：c→）であるが，T1強調像では中等度の信号強度（Fig. 2：d→）で血流による無信号域（flow void）はなく，凝血塊として矛盾のない所見である．MR静脈撮影像（MR venography）（Fig. 2）では上矢状静脈洞から右の横静脈洞，S状静脈洞への流れはきれいに描出されているが，左側の横静脈洞からS状静脈洞は全く描出されていない（Fig. 2：a ↓）．

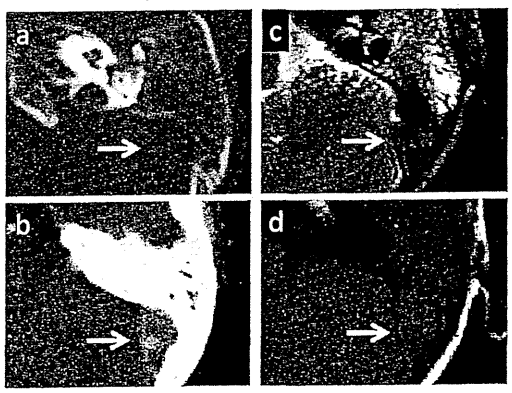

Fig. 1　CT and MRI of sigmoid sinus thrombosis

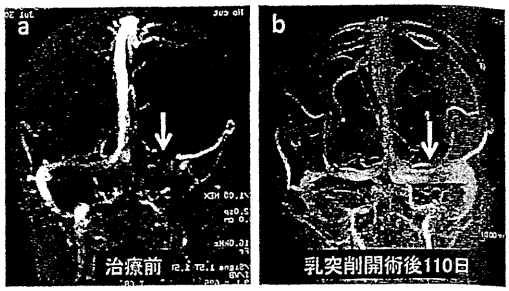

Fig. 2　MR venography before and after treatment of sigmoid sinus thrombosis

治療とその経過：患児到着4時間後に全身麻酔下に左乳突削開術を行った．乳突蜂巣は炎症性肉芽で充満しており，削開中には肉芽から通常より遥かに強い出血があったが削開が完了すると出血もほぼ停止した．S状静脈洞内にはゼリー状の凝血塊が充満していた．静脈開窓部を閉鎖し，上鼓室から鼓室への交通路も清掃，確保して手術を終了した．手術により左中耳炎は完治し，術後110日では左S状静脈洞の再疎通が確認された（Fig. 2: b ↓）．

解　説：S状静脈洞血栓症は急性および慢性中耳炎や頭部外傷の重篤な合併症の一つであり[2]，対応が遅れると死亡に至る場合もある．本症の診断は臨床症状とCT，MRI検査による．CTではS状静脈洞の造影効果のない拡大が診断の鍵になり，S状静脈洞そのものは内部の血栓によってやや高濃度となる．同じCT検査でも，側頭骨ターゲット撮影ではウィンドウ幅が広く設定され，血栓と周囲の脳脊髄液や小脳などとの濃度コントラストがつかないので注意を要する．血栓症の画像ではMRIの有用性が高いが，発生から時間が経つと共にT1およびT2強彫像での信号強度パターンが変化してゆくので時間に応じて読影しな

ければならない．また，S状静脈洞血栓症では特にMR venographyによる静脈の状態観察が有用である[3,4]．治療では，早期の乳突削開による中耳炎の外科的制御が有効であり，本章の治療の基本は乳突削開と考えてよい．本例でも，乳突削開と上鼓室の肉芽清掃のみで治癒し，S状静脈洞の再疎通が得られた．

症例2：23歳男性

主　訴：左顔面神経麻痺と難聴，耳鳴

現病歴：左錐体尖真珠腫による顔面神経麻痺，難聴，耳鳴で，7年前に側頭開頭と経乳突法によって真珠腫を摘出し，画像により経過を観察していた．

側頭骨ターゲットCT所見：左錐体の中央部分に手術による骨欠損が見られるが（Fig. 3：a ↓），軟部組織の内部構造は見分けられない．

造影MRI所見：錐体の骨欠損内側部分に，やや低信号で周囲が膜状に造影される領域があり，真珠腫（遺残性再発）とその母膜と診断した（Fig. 3：b ↓）．

non-EP拡散強調MRI：左錐体尖部に塊状の亢進号部分があり，真珠腫と診断できる．また，その外側に小さな高信号腫瘤が2個同定される．

手術所見：左側頭開頭で錐体尖部に到達した．大きな再発真珠腫塊の外側に，硬膜に接して小さな真珠腫塊も確認され，内側の大きな真珠腫とともに剥離，全摘出した．

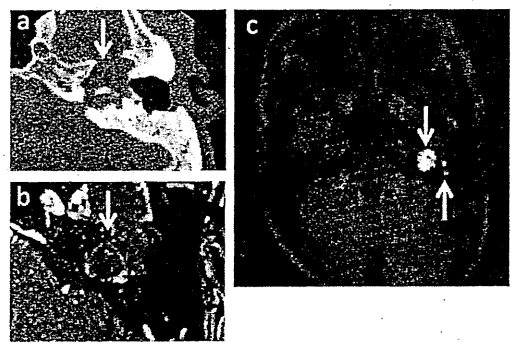

Fig. 3　Follow-up CT and MRI of petrous apex cholesteatoma

解　説：真珠腫のMRI診断には，従来，単純のT1強調像とガドリニウム造影T1強調像の比較，あるいはecho-planar法による拡散強調画像が用いられていたが，近年，同法を用いない拡散強調画像：non-EP-diffusion-weighted MRIの高い診断能力が報告されるようになった[5]．症例2のFig. 3：cでも分かるように，non-EP法はecho-planar法の画像に比べて歪がほとんどなく，小さな真珠腫も検出することができる．今後は，この方法と側頭骨CTの組み合わせが真珠腫の画像診断の中心となってゆくと考えられる．

2．側頭骨外傷

側頭骨外傷は交通外傷や転落事故，転倒，暴力，銃創などで生じる．初期診療では，まず気道の確保，呼吸，循環，中枢神経機能の評価，体温管理を行い（primary survey），次いで身体全域の受傷状態を検索する（secondary survey）．側頭骨骨折の診断は，このsecondary surveyに位置づけられ，それに基づいて根本治療（definitive therapy）に進む[6]．

本症の画像検査では側頭骨高分解能CTが第1選択で，骨折線が錐体の長軸に対して平行であれば縦骨折，直交していれば横骨折と分類されるが，縦骨折の頻度が高く全体の70から90%を占める．機能的観点から側頭骨骨折を迷路骨折の有無で大別する方法も唱えられている[7]．迷路骨折があると基本的に同側の聴覚，末梢前庭機能が完全に失われる．一方，脳組織の損傷についてはsoft-tissue window撮影の頭部CTで急性期の判断を行い，必要に応じて軟部組織コントラストに優れたMRI検査を行う．

症例3：60才　女性

乗用車と接触し，5m引きずられてそのまま車の下敷きになった．当院からドクターカーが出動し，45分後に負傷者とともに帰還．初期診療で状態をひとまず安定させた上で，画像検査が行われた．

頭部CT所見：右中頭蓋窩に厚さ約1cmの急性硬膜

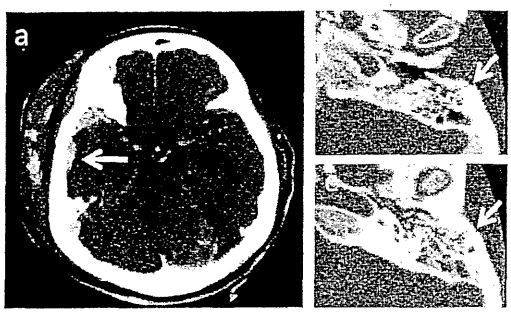

Fig. 4　Brain and temporal bone CT of severe head trauma

外血腫（Fig. 4：a ←），左側頭骨から眼窩上縁の骨折を認めた．硬膜外血腫は脳の圧迫が乏しかったので脳神経外科で保存的に経過観察となった．胸部CT，腹部CTでは異常所見なし．両側下腿X線像で，骨折なし．

全身状態が回復，安定し，顔面の腫脹が軽快した受傷後11日の時点で，左顔面神経麻痺，左難聴が明らかとなり，耳鼻咽喉科受診となった．

側頭骨ターゲットCT所見：骨折線が左乳突部から骨部外耳道，顎関節窩を経て内側前方に走行しており（Fig. 4：b，c ←），乳突部から鼓室まで，中耳腔には軟部組織陰影が充満している．外耳道後壁の骨片が割れて前方の外耳道内に突出しており，耳小骨連鎖ではキヌタ・アブミ関節が離断してキヌタ骨長脚が前方に変位していた．骨折線は骨迷路から離れており，顔面神経管の破壊や断裂は見られない．

治療経過：臨床症状から，内耳障害はなく，難聴は伝音系の問題であり対処は急がないが，顔面神経麻痺が高度であったので，まず顔面神経減荷術を行い，同時に外耳道，鼓膜，耳小骨連鎖の状況を観察する手術計画とした．

受傷18日後に左顔面神経減荷術・鼓室試験解放術を行った．外耳道後壁骨が破壊され，鼓膜も一部穿孔があり，耳小骨連鎖はキヌタ・アブミ関節で離断し，周囲に肉芽形成が見られた．乳突部にも血腫と肉芽が充満しており，これらを除去，清掃した．顔面神経管を膝神経節から鼓室部，乳突部全域にわたって露出，減荷した．顔面神経管の破損，骨片等による神経の圧迫などは確認されなかったが，術中の顔面神経電気刺激で顔面筋の反応は見られなかった．この手術で外耳道と鼓膜，中耳の創傷が治癒，回復を待ち，6か月後に第2段階手術で伝音再建を行った

解　説：側頭骨外傷では，創部の感染には局所処置と抗菌薬の投与，出血には局所止血処置や止血薬の投与，髄液漏には頭部高位での安静などで，まず保存的に対処し，各々効果がなければ手術で対処する．機能検査では全ての脳神経をチェックし，症状に応じて個々の精査を行う．聴覚については，純音聴力検査で感音難聴か伝音難聴かを鑑別する．伝音難聴には後でも対処できるが，急性の感音難聴は時間とともに治療効果が低下するので，できるだけ早く音響外傷に準じた治療を行う．眼振の観察も重要で，耳鳴とともに内耳障害の指標となる．瘻孔症状や変動する難聴など，外リンパ瘻を示す所見があって保存的に治らなければ内耳窓閉鎖術の適応となる．顔面神経麻痺に対しては問診が大切で，即発性のものは重症例が多く，遅発性のものは保存的治療で治る例が多いとされるが，重度の外傷では受傷当初の状況が明確でなく判断に困る場合も少なくない．

3．内耳奇形

先天性難聴に占める内耳奇形の割合は20％程度で[8]，主要な原因の一つである．内耳奇形による難聴でも，他の病因によるもの同様に早期診断，早期介入が重要である．特に，人工内耳が必要になる高度から重度の難聴例では，単なる診断や分類だけでなく，手術計画の観点からも奇形の正確な評価が必要である．我々の最近6年間の集計でも，当科の難聴外来を受診した重度以上の先天性難聴小児91例，182耳中，画像診断で内耳奇形が確認されたのは20例，39耳，21.4％であり，Jacklerらの報告とほぼ同様の結果である．また，この20例中，人工内耳手術に至ったものは内耳や蝸牛の無形成，内耳道狭窄で手術非希望例の3例を除いた17例，85.0％と高率であるが，これ