Curable Sensorineural Hearing Loss and Critical Hearing Loss

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Abstract: Sensorineural hearing loss has generally been believed to be incurable, but it has become known in recent years that some cases of sensorineural hearing loss are curable. A typical example of such cases is sudden deafness. However, the diagnosis of sudden deafness should not easily be used; when a case was finally evaluated to be of unknown cause on subdivision diagnosis, it should be diagnosed as sudden deafness, because there seem to be variable causes of sudden deafness. Therefore, the diagnosis of sudden deafness made on initial examination may often be corrected thereafter. The cases, which should be differentiated from sudden deafness, include perilymphatic fistula, steroid responsive sensorineural hearing loss, and so on, and they are curable. On the other hand, the cases, which are diagnosed as sudden deafness from the past history on initial examination as critical hearing loss, include acoustic neuroma and brainstem infarction. Only the case, in which these diseases are ruled out, can be regarded as being of unknown cause.

Key words: Curable hearing loss; Critical hearing loss; Sudden deafness; Perilymphatic fistula; Steroid responsive hearing loss

Introduction

Sensorineural hearing loss is a general term of hearing loss cases due to the disturbance of the auditory pathway involving the area ranging from the cochlea to the auditory cortex of the cerebrum. The incidence of cochlear hearing loss attributed to the disturbance of the cochlea, which is one of such sensorineural hearing loss cases. Sensorineural hearing loss hardly becomes curable long after the occurrence of the disturbance, because sensory cells of the cochlea are hardly regenerated once they have been impaired. A typical example of the sensorineural hearing loss cases is deafness due to aging (senile hearing loss).

Curable Cases of Sensorineural Hearing Loss

Since about the middle of the 20th century, it has become known that some acute cases of

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sensorineural hearing loss are curable.[1,2] These diseases were named as sudden deafness, which is currently a common name, by Rasmussen (1949).[3] In Japan, the term, sudden deafness, was used by Tsuiki (1955).[4] It has also become known that the disease must be dealt with as medical emergency because it shows a good prognosis by early treatment.

It has subsequently become known that some sensorineural hearing loss cases other than sudden deafness are also curable. One of them is hearing loss with which immunological abnormalities are involved, and McCabe[5] used the term, autoimmune sensorineural hearing loss, for calling such a case of hearing loss in 1979.

In Japan, the authors[6] reported in 1975 that sensorineural hearing loss accompanying Takayasu disease or aortitis syndrome is steroid responsive sensorineural hearing loss. In 1981, the authors[7] reported that some cases of this type of sensorineural hearing loss show high levels of immune complex. With regard to these cases, no reliable diagnostic method has been found. Although it is one diagnostic method to clinically demonstrate the response to steroids, there are no methods of demonstrating in any case without response to steroids. Even if hearing is improved by administration of steroids, it cannot be immediately concluded that these cases show hearing loss related to immunological abnormalities, because it is difficult to differentiate the improvement in response to steroid administration from spontaneous recovery. It has therefore been estimated from animal experiments that immunological abnormalities are involved with the occurrence of some deafness cases, but there has been no diagnostic methods in the actual setting of clinical examinations under the present situation. In the possibility of such a deafness case (that will be described later), however, it is a realistic method to administer steroids from the early stage of the treatment.

Among sensorineural hearing loss cases, perilymphatic fistula has been clarified as being curable. There is a bottleneck in the diagnosis of the disease; i.e., the definite diagnosis of this disease cannot be made, unless perilymphatic leakage from either the round window or oval window or both of them is confirmed by exploratory tympanotomy. In some cases of perilymphatic fistula, hearing loss has been improved by covering the two windows using a piece of perichondrium. Therefore, there has been much argument about the diagnosis and timing of surgery. Deafness in perilymphatic fistula, which occurs suddenly, is required to be differentiated from sudden deafness.

Thus, some diseases have become excluded from the category of sudden deafness in recent years. At present, the diagnosis of sudden deafness is restricted to the cases of unknown cause. The diseases, which have become excluded from the category of sudden deafness, include the above-described deafness cases due to immunological abnormalities, perilymphatic fistula, mumps hearing loss without swelling of the parotid gland, acoustic neuroma (AN), other cerebellopontine angle tumors, brainstem infarction or cerebral hemorrhage, Ménière disease, and so on.

As “sensorineural hearing loss cases that heal up”, sudden deafness, deafness related to immunological abnormalities, and perilymphatic fistula are described below. However, “cases that heal up” should be “curable cases”, correcly describing. All these deafness cases do not necessarily “heal” or “improve”.

1. Sudden deafness

(1) Diagnostic criteria

The diagnostic criteria established by the research group for the disease sponsored by the Japanese Ministry of Health and Welfare are commonly used. They define three key points for the disease: ① It occurs suddenly; ② it shows moderate or severe sensorineural hearing loss; and ③ the cause is unknown. Dizziness or vertigo may be associated with the

Table 1 Diagnostic Criteria for Steroid Responsive Hearing Loss (Established by the author in 1984)

1) Hearing loss is improved by administration of steroids, and hearing loss worsens again by discontinuation of the administration. When hearing level also worsens, hearing level is improved by re-administration of steroids.
2) While a steroid is administered at a reduced dose or at the maintenance dose, hearing level worsens again. Hearing level is improved by the increase in dose.

The cases, which meet either of the above-described conditions 1) and 2) or both of them, are diagnosed as steroid responsive hearing loss.

disease at the time of occurrence of hearing loss, and sudden deafness should also exclude central nervous diseases (brain tumor, cerebral infarction, etc.). Mild cases of deafness including the low-tone disturbance type should be separately dealt with from the usual cases of sudden deafness, because such a case is not necessarily perceived subjectively as hearing loss.

(2) Etiology

The theory of viruses (including the viral reactivation theory), the theory of disturbed microcirculation, etc. have been considered. In recent years, a report has shown that mumps hearing loss, which manifests sensorineural hearing loss alone without parotid swelling as a symptom, is observed by the determination of mumps IgM in 5–6% of sudden deafness cases. These mumps hearing loss cases will be excluded from sudden deafness cases, if the diagnosis is made. Similarly, the deafness cases, which will be clarified to have been caused by viruses owing to the advances in testing methods, are to be excluded from sudden deafness cases in the future. Unfortunately, no certain diagnostic method for cochlear circulatory disturbance has been developed.

(3) Treatment

It is difficult to evaluate the efficacy of remedies, because sudden deafness may show spontaneous recovery. Thus, it can be said that there have been no specific remedies for sudden deafness, which are based on adequate evidence. In the present situation, some drugs are used at rest on the basis of theoretical and empirical evidence. In general, steroids are frequently used, unless the cases are regarded as contraindications for steroids. In addition to steroids, drugs for improving circulation are used, but it remains unclear whether the combined use is more effective than the single use. As a consequence, about a third each of the cases appear to heal, to be improved (to markedly recover and recover), and to be unchanged, respectively.

However, many factors are related to the prognosis: ① hearing level, ② the period (day) from the occurrence to the start of treatment, ③ the presence/absence of dizziness, ④ the rate of improvement 1 week after the start of treatment, ⑤ age, etc. When dizziness or vertigo is severe, hearing impairment is also severe, indicating that the relationship between dizziness and hearing impairment is the same as that between hearing impairment and hearing level; when the rate of improvement in hearing is high ($>50\%$) after the treatment or within 1 week after the occurrence, the prognosis is favorable as a matter of course, whereas the prognosis is poor when the rate of improvement is low ($<50\%$) within 2 weeks after the occurrence.

2. Hearing loss related to immunological abnormalities

(1) Diagnostic criteria

A research group for hearing loss due to immunological abnormalities sponsored by the Japanese Ministry of Health and Welfare established the diagnostic criteria for the research to collect and assess cases of the con-

Table 2 Diagnostic Criteria for Perilymphatic Fistula (Established by the research group by the Japanese Ministry of Health and Welfare in 1990)

Certain cases
Perilymphatic transudation or liquorrhea from either of the vestibular or cochlear window or both of them is confirmed or a fistula is confirmed by surgery (exploratory tympanotomy), endoscopy, etc.
Suspected cases
Some factors predisposing to rapid changes in cerebrospinal fluid pressure and tympanic pressure are followed by the occurrence of a sense of ear fullness, hearing loss, dizziness, and disequilibrium.
Notes 1 through 8
Note 1. The factors predisposing to the changes include the strain, lifting of a heavy thing, nose blowing, anger, diving, traveling by plane, etc.
Note 2. All the symptoms are not necessarily present; either one of them may be present.
Note 3. A sound, pop, may be associated with the condition.
Note 4. It may recur.
Note 5. Sensorineural hearing loss occurs for several hours or days. It may occasionally vary.
Note 6. Hearing loss acutely develops, with which the following condition is associated: "ringing of the ears like water-run" or "a sense of water-run".
Note 7. Dizziness is complained of with compression and decompression to the external and middle ears.
Note 8. A sense of trembling persists, and positional nystagmus is observed on the affected side.

dition. The key points of the criteria are as follows: ① The disease is dependent on steroids (Table 1); ② existing autoimmune diseases are associated with the disease; and ③ abnormalities are revealed by immunological examination. The authors include ①+②+③ in the category of the systemic type (hearing loss associated with Takayasu disease), and ①+③ in the local type. It is necessary to assess whether the local type will be transferred to the systemic type if the local type is left untreated. The authors have experienced no cases of such transfer from the local type to the systemic type, probably because the local type has been treated.

(2) Etiology

The site of the inner ear, which is involved in the occurrence of hearing loss, or the immunological mechanism underlying the occurrence of hearing loss is quite unclear. As the site of impairment, the stria vascularis and the endolymphatic sac have been estimated from animal experiments.

(3) Clinical symptoms

Cases of steroid responsive sensorineural hearing loss are divided into the type of deafness accompanying Takayasu disease and the local type without known autoimmune disease or any disturbance other than deafness.[8] Both types are frequently observed in women in their 30s and 40s. Most cases are bilateral. In many cases, the unilateral side is revealed on the initial examination to have already had severe deafness or total deafness. Some cases show mixed deafness, suggesting that some morbid conditions of the eustachian tube are suspected. However, exudative otitis media is not observed. No specific findings have been revealed by any immunological examination conducted in clinical routine tests. Many cases show increased sedimentation rate and high total gamma-globulin and IgM levels in serum. Sedimentation rate is increased at the time of the exacerbation of hearing in some cases.

(4) Treatment

Prednisolone (PSL) as the steroid and Sairei-to (Chai-Ling-Tang) are used together.[8] The combined use of Sairei-to has facilitated to reduce the dose of PSL, as compared to the single use of PSL, probably because the herb medicine can enhance the effect of steroid. Severe deafness cases do not respond to PSL. The criterion for reduction in dose of PSL is that the dose ($\leq$10 mg) is decreased by 1 mg over the 4-month period. If hearing worsens during the reduction, the dose may be increased by 50% of the maintenance dose,

then be gradually decreased again.

3. Perilymphatic fistula

(1) Diagnostic criteria

The key points were described earlier. When perilymphatic fistula is suspected on interview, the item as shown in Table 2 are used for reference.

(2) Etiology

According to the hypothesis of Goodhill,[9] there are an explosive route and an implosive route for the cause of the condition. With the former route, either one of the cochlear windows or both are broken in the direction from the cochlear side to the middle ear side in response to increased intracranial pressure. With the latter route, the cochlear window is broken in the direction from the middle ear side to the cochlear side in response to increased middle ear pressure. Changes in intracranial pressure and middle ear pressure due to barotrauma, head injury, and middle ear injury are responsible for perilymphatic fistula. There are also some cases of unknown cause.

(3) Clinical symptoms

Hearing loss shows various symptoms as follows: some cases occur suddenly; some cases show rapid progression and exacerbation within several days after the onset; deafness itself is mild, and dizziness and tinnitus (ringing of the ears) are the main symptoms in some cases; and disequilibrium is remarkable in some cases. Anamnesis is important for the diagnosis, while fistula test is important for examination. The fistula test is a method of observing nystagmus under compression and decompression by closing up tightly the external auditory meatus. Observation of nystagmus is facilitated by the use of infrared photography. It is secure to record findings of nystagmus on videotape or with a nystagmograph (ENG).

(4) Treatment

Before long (within 2 weeks) after the onset of hearing loss, spontaneous closure of fistula is expected by resting with drug therapy, as in the case of sudden deafness. Exploratory tympanotomy is conducted on the following cases: vertigo or deafness do not improve even 2 weeks after the onset; and vertigo is severe or hearing deteriorates. In surgical cases, the cochlear window is closed with the tragal perichondrium and fibrin sealant is used for the attachment, regardless of the presence or absence of perilymphatic transudation.

Critical Hearing Loss

Critical hearing loss indicates the deafness case with an underlying disease that will become life-threatening if it is left untreated. There is another case of bilateral progression of deafness, although it does not accompany any fatal disease. According to the severity of disturbance, some cases may be regarded as semi-critical deafness, because they become disturbance of communication in social life. The disease described as critical hearing loss herein is restricted to sensorineural hearing loss.

Typical examples of the causative diseases underlying critical hearing loss are AN [including bilateral AN (NF 2)], other cerebellopontine angle tumors (meningioma, epidermoid, etc.), brainstem infarction, cerebral hemorrhage, multiple sclerosis, and autoimmune diseases (Takayasu disease, systemic lupus erythematosus, recurrent polychondritis, etc.).

Typical examples of the diseases causing a disturbance of communication as a consequence of bilateral progression of sensorineural deafness are NF 2, perilymphatic fistula, dilated aqueduct of the vestibule, Takayasu disease, labyrinth syphilis, and Cogan syndrome. NF 2, Takayasu disease, and Cogan syndrome are regarded as “critical hearing loss” as well.

AN and brainstem infarction are described herein.

1. AN

(1) Etiology

Bilateral AN, which is called neurofibromatosis 2 (NF 2), is believed to be a hereditary abnormality. The causative gene for unilateral AN still remains unknown. Unilateral AN is described herein.

(2) Clinical symptoms

The main symptoms of the disease are hearing loss and tinnitus in almost 90% of the patients when they visited hospitals for examination. To be important, AN is hardly differentiated from sudden deafness in 10–20% of AN cases, because hearing loss suddenly occurs. It also escapes an examiner's attention, because hearing level is improved in some AN cases. If AN is left untreated without assessment because it is regarded as an example of incurable sudden deafness, "late diagnosis" or "erroneous diagnosis" may occur. Therefore, all cases, which are considered to be sudden deafness, should be suspected to show the condition as the initial symptom of AN. Such an attitude is needed.

(3) Diagnosis

It has conventionally been believed that there is no audiometric configuration characteristic of AN, but it became increasingly known that some cases of small tumors, which manifest as sudden deafness, show the trough type of audiometric configuration. The trough type was indicated first by the authors.[10] Then, transorbital technique and Stenvers projection are conducted as a method for plain roentgenography of the internal auditory meatus. However, no abnormalities are observed in approximately 20% of small tumor cases. With regard to auditory brainstem response (ABR) test, the indication for the test is restricted to the cases in which the mean level for frequencies 4 kHz and 8 kHz is 70 dB or lower. Approximately 15% of small tumor cases (<10 mm in size) show no abnormalities. On the basis of the situation, ABR and magnetic resonance imaging (MRI) are conducted on suspected cases, especially suspected young patients with good hearing, while MRI is conducted on the cases with 70 dB or higher of hearing level, because they are excluded from the indication for ABR. From the aspect of cost, several new methods for screening are also being assessed; with these methods, 3-dimensional Fourier transform fast recovery fast spin echo are used or the amount of contrast medium are reduced to half, and the number of the images taken is also limited.

(4) Treatment

Unless there are problems with general condition in patients aged 60 years or under, surgery may be recommended as the first choice. If the tumor tends to increase in the elderly patients, patients with risk factors, and patients refusing surgery, stereotaxic radiotherapy may be selected. In cases of small tumors localized in the internal auditory meatus, which accompany the favorable preoperative hearing level, hearing level is postoperatively to be sustained to 70–80% by the middle cranial fossa method. The operation is highly safe, and the mortality with the operation is 1% or lower. The rate of preservation of the facial nerves, which becomes the most issue among the sequelae, is being increased in recent years. The incidence of postoperative sequelae is high among the recurrent cases after stereotaxic radiotherapy, and the postoperative function of the facial nerves is extremely poor.

2. Brainstem infarction

Some cases of small brainstem infarction show sudden deafness and dizziness or vertigo alone, which accompany no other cerebral nervous symptoms. Since dizziness or vertigo may be associated with sudden deafness of unknown cause, it is necessary for brainstem infarction to be ruled out, in addition to differentiation of brainstem infarction from sudden deafness and Ménière disease. In the elderly patients and patients with a past history of hypertension particularly, MRI is an essential means for test.

The treatment of deafness in patients with brainstem infarction follows that for sudden deafness. In the cases with past histories of hypertension, diabetes mellitus, and hypercholesterolemia, as well as those with risk factors, the control of these conditions is required.

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