Bilateral Rapidly Progressive Hearing Loss Followed by Loss of Vestibular Function — Case Report

Duen-Lii Hsieh, Yi-Ho Young

Department of Otolaryngology, National Taiwan University Hospital, Taipei, Taiwan

ABSTRACT

Rapidly progressive sensorineural hearing loss is a specific type of sudden deafness characterized by a progressive stepwise process over a period of a few hours to months with the mechanism attributable to autoimmune reactivity. The hearing organs are always involved and usually the vestibular organs as well. Recently, we have encountered a 33-year-old woman with rapidly progressive sensorineural hearing loss bilaterally. A battery of audiovestibular function tests e.g. audiometry, distortion product otoacoustic emission (DPOAE), auditory brainstem response (ABR), caloric test, and vestibular evoked myogenic potential (VEMP) test were conducted throughout the course. The sequence of audiovestibular dysfunction was absent DPOAE initially, followed by total deafness and absent ABR, and finally absent VEMPs and caloric areflexia, indicating that the cochlea is vulnerable earlier than the vestibule. Hence, this case demonstrates the sequence of degeneration in the inner ear, that is, rapid progressive hearing loss followed by vestibular loss, similar to that seen in the aging process (degeneration) in the inner ear. (Tzu Chi Med J 2006; 18:141-144)

Key words: autoimmune sensorineural hearing loss, idiopathic bilateral sensorineural hearing loss, rapid progressive hearing loss, sudden deafness

INTRODUCTION

There are reports of a specific type of sudden deafness characterized by a bilateral progressive stepwise process over a period of a few hours to months [1-4]. It has many synonyms such as rapidly progressive sensorineural hearing loss (rapid deafness), sudden bilateral sensorineural hearing loss, and idiopathic bilateral sensorineural hearing loss. The condition is progressive, idiopathic, and bilateral. The mechanism of rapidly progressive hearing loss to eventual deafness has been attributed to autoimmune reactivity. In 1979, McCabe [5] reported the first series of patients with progressive bilateral sensorineural hearing loss who were responsive to immunosuppressive therapy. The hearing organs are always involved and usually the vestibular organs as well. However, because vestibular function is affected symmetrically, the frequency of vertigo associated with rapidly progressive hearing loss is low [3]. Therefore, since insufficient vestibular information, it is not known whether the process of vestibular loss parallels that of hearing loss. Recently, we have encountered a patient with rapidly progressive bilateral sensorineural hearing loss and vestibular loss. A battery of audiovestibular function testing was conducted to investigate the sequence of degeneration in the inner ear. Herein, we present this case.

CASE REPORT

A 33-year-old woman visited our clinic due to vertigo accompanied by hearing loss, tinnitus, and fullness...
sensation of the left ear. No one in her family had a history of hearing impairment. At admission on April 16, 2003, local examination of the ear, nose, and throat fields revealed nothing particular. Audiometry disclosed high-tone sensorineural hearing loss with a mean hearing level of 23 dB in the right ear, and moderate sensorineural hearing loss with a mean hearing level of 64 dB in the left ear (Fig. 1A). However, the distortion product otoacoustic emission (DPOAE) test failed to elicit responses, bilaterally. Auditory brainstem response (ABR) demonstrated normal response in the right ear and absent response in the left ear. Electronystagmography (ENG) disclosed spontaneous nystagmus beating toward the right side, while caloric test revealed normal responses bilaterally on April 19, 2003.

Subsequently, vestibular evoked myogenic potential (VEMP) test depicted delayed response on the right side with a latency of p13 of 20.38 ms (norm: <19.1 ms), and normal response on the left side with a latency of p13 of 14.00 ms (Fig. 2A). Under the impression of Meniere's disease in the left ear, she was treated with isosorbide, and discharged three days later.

Unfortunately, bilateral hearing deteriorated four days after discharge. Because of an outbreak of severe acute respiratory syndrome (SARS) at our hospital [6], the patient visited another hospital, where audiometry revealed flat-type sensorineural hearing loss with a mean hearing level of 40 dB in the right ear, but total deafness in the left ear six weeks after initial presentation.

Fig. 1. A serial audiogram of a 33 year-old women demonstrates deterioration of bilateral hearing. (A) The mean hearing levels in the right and left ears were 23 dB and 64 dB, respectively. (B) Hearing on the left side becomes scale out within one week. (C) Deterioration of the hearing in the right ear 2 weeks later. (D) Hearing stabilizes at 78 dB in the right ear and total deafness in the left ear six weeks after initial presentation.
in the left ear (Fig. 1B). She was treated with prednisolone 40 mg daily. One week later (May 1), audiometry demonstrated progressive deterioration in the right ear, with a mean hearing level of 63 dB (Fig. 1C), but the ABR response remained normal in the right ear. She was told as having bilateral sudden deafness, and was treated with intratympanic dexamethasone injections. Altogether she received two injections in the right ear and once in the left ear, with a dosage of 0.5 mg each time. Nevertheless, bilateral profound sensorineural hearing loss persisted.

Four weeks later (May 26), she returned to our clinic, where audiometry revealed profound hearing loss with a mean hearing level of 79 dB in the right ear, and total deafness in the left ear (Fig. 1D). Both ice water caloric test and VEMP test (Fig. 2B) elicited no responses bilaterally. High resolution computed tomography of the temporal bone failed to demonstrate any organic lesion.

The internal auditory canal, cochlea, vestibule, and semicircular canals were all intact. Laboratory examination including complete blood cell counts, blood chemistry, rheumatoid factor, antinuclear antibody, c-reactive protein, and LE cell preparation were all within normal limits. However, serum protein electrophoresis showed increased alpha 1 and alpha 2 globulins and polyclonal hypergammaglobulinemia. Complement analysis also revealed a decreased C4 level (8.9 mg/dL, norm: 27.45 ± 10.72 mg/dL), hence autoimmune sensorineural hearing loss was determined.

One year after initial presentation, she was declared to be totally disabled because of bilateral profound sensorineural hearing loss and vestibular loss. The table summarizes the progression of the audiovestibular dysfunction in this patient.

### DISCUSSION

Rapidly progressive hearing loss with or without vertigo is a debilitating and frightening condition, and autoimmune reactivity is thought to be its etiopathogenic factor [4]. It may cause a constellation of cochleovestibular symptoms that have either sudden or progressive onset, unilateral or bilateral involvement. In this patient, the initial symptoms mimicked Meniere's disease, with fluctuating hearing loss and fullness sensation in the ear, accompanied by acute severe vertigo. Unfortunately, the left ear deteriorated to total deafness within one week (Figs. 1A, B). Furthermore, hearing in the opposite (right) ear also deteriorated at all frequency levels, leading to bilateral profound sensorineural hearing loss within 1 month. Thus, rapid progressive hearing loss was impressed based on a series of audiograms, which helped to differentiate this condition from Meniere's disease and sudden deafness. In addition, deterioration of vestibular function followed the hearing loss, as demonstrated by absent caloric responses and absent VEMP responses.

### Table 1. Progression of Audiovestibular Dysfunction

<table>
<thead>
<tr>
<th></th>
<th>April 16, 2003 (R/L)</th>
<th>May 26, 2003 (R/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean hearing level</td>
<td>23 dB / 64 dB</td>
<td>78 dB/ scale out</td>
</tr>
<tr>
<td>DPOAE</td>
<td>- / -</td>
<td>- / -</td>
</tr>
<tr>
<td>ABR</td>
<td>normal / -</td>
<td>- / -</td>
</tr>
<tr>
<td>Caloric test</td>
<td>normal / normal</td>
<td>- / -</td>
</tr>
<tr>
<td>VEMP test</td>
<td>delayed / normal</td>
<td>- / -</td>
</tr>
</tbody>
</table>

ABR: auditory brainstem response; DPOAE: distortion product otoacoustic emission; VEMP: vestibular evoked myogenic potential; -: absent
VEMP responses 40 days after presentation (Table 1), implying that the vestibular end-organs were also affected.

Schuknecht [7] reported a temporal bone histopathological study in a case of rapidly progressive sensorineural hearing loss, showing the organs of Corti were in an advanced stage of degeneration. The outer hair cells were missing; however, some inner hair cells remained. This pathological feature may explain why our patient had normal hearing (23 dB) in the right ear but an absent DPOAE response initially. Meanwhile, the saccular walls collapsed onto the macula with degeneration of the otolithic membrane and epithelium, correlating with absent VEMPs, since the VEMP test has been validated to evaluate the sacculo-collic reflex [8,9]. Furthermore, histopathological study also showed that all semicircular ducts and ampulla collapsed and the crista were severely degenerated with no remaining sensory epithelium, compatible with bilateral caloric areflexia, 40 days after presentation.

Thus, the sequence of audiovestibular dysfunction in this patient can be traced as follows: an absent DPOAE initially, then total deafness and an absent ABR, and finally absent VEMPs and caloric areflexia. From a phylogenetic viewpoint, vestibular deterioration appears earlier than cochlear deterioration, indicating that cochlea vulnerability appears earlier than vestibular vulnerability [10]. Meanwhile, abnormal VEMP occurs before the development of caloric areflexia, implying that sacular degeneration occurs prior to degeneration of the semicircular canals. Likewise, embryonic development of the semicircular canals occurs earlier than that of the saccula, leading to increased vulnerability of the saccule compared to the semicircular canals. Hence, this case demonstrates that rapidly progressive vestibular loss follows hearing loss, similar to that seen in the aging process (degeneration) in the inner ear.

Clinically, suspicion should be aroused when progressive hearing loss with/without vertigo has no obvious cause. In addition to the clinical manifestations, positive immune laboratory tests and a beneficial treatment response provide other clues for diagnosing this disease. In this patient, polyclonal hypergammaglobulinemia with a decreased C4 level suggested autoimmune reactivity as an etiopathic factor. However, beneficial treatment seems to be no measure to completely stop the disease process, even when intratympanic steroid injection is given [11].

Antigenic epitopes are indeed able through repeated antigenic challenges to induce a cochleovestibular dysfunction. Testing whether passive transfer of specific antibodies against these proteins induces either rapid progressive hearing or vestibular loss would be beneficial [4]. Although there are no tests that provide unequivocal proof of this disease, adults with rapidly progressive sensorineural hearing loss and anti-P0 antibodies have been shown to present a more accentuated hearing loss than anti-P0 antibody-negative patients [12].

REFERENCES