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Development of conductive hearing loss due to posterior semicircular canal dehiscence

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ABSTRACT

We herein report a case of posterior semicircular canal dehiscence (SCD) syndrome who had been audiologically followed up for eight years. The patient originally had sensorineural hearing loss. The audiogram had gradually transformed to pure conductive hearing loss. The posterior SCD was identified in CT scan. The reported case showed the possibility to distinguish the mechanism at play underlying the typical conductive hearing loss in SCD patients by tracing the transition of the hearing loss pattern. This information is of much help to predict the hearing outcomes if surgical intervention were chosen for the treatment.

1. Introduction

In 1986, Wadin et al. radiologically linked inner ear bony dehiscence with the clinical features of hearing loss, vertigo and tinnitus. Among their cases, posterior semicircular canal dehiscence (SCD) induced by high jugular bulb was suggested to be a possible cause of hearing loss [1,2]. After superior SCD case series were first described in 1998 [3], the comprehensive understanding of canal dehiscence including its etiology, topology, mechanisms of the symptoms, and the therapeutic strategy has been promoted.

Posterior SCD has been reported to be very similar to superior SCD in its clinical features [4,5]. SCD patients characteristically present as either sound- or pressure-induced oscillopsia, disequilibrium, vertigo and hearing loss. Some of them suffer from hearing loss with various level of air-bone gap. There are at least two independent mechanisms that explain the conductive hearing loss in SCD patients. One involves the increased air-conducted hearing threshold, and the other is associated with the decreased bone-conducted hearing threshold [6,7].

Although the surgical closure of the dehiscence improved the air-conducted hearing to variable degrees [8–11], some patients showed an increased bone-conducted hearing threshold postoperatively [11]. These results also indicated that there are multiple mechanisms underlying the hearing loss in SCD patients. If we can distinguish the mechanisms at play in individual patients, it may be possible to select candidates who are likely to be successfully treated by surgical intervention. One way to distinguish the reasons for the conductive hearing loss is to evaluate how the hearing changes in SCD patients during the course of the development of SCD.

We herein report a patient with posterior SCD who initially showed sensorineural hearing loss. During a long-term follow-up over eight years, the patient's hearing gradually transformed to conductive hearing loss. A computed tomography (CT) scan of the affected side of the temporal bone showed dehiscence of the posterior semicircular canal to the jugular bulb.

This may be the first case report describing how the typical conductive hearing loss developed during the course of disease progression.

2. Case report

A 14-year-old female came to our institute complaining of left-sided hearing loss, which was first found during a routine medical examination at her elementary school when she was six years old. She was initially diagnosed to have left sensorineural hearing loss at another institute when she was 13 years old (Fig. 1 Oct, 2000 and Mar, 2001). MRI of her brain showed no particular abnormalities. She had no significant past medical or family history. She had never experienced dizziness.

On examination, both eardrums were found to be normal. Neither spontaneous nor positional nystagmus was observed. The

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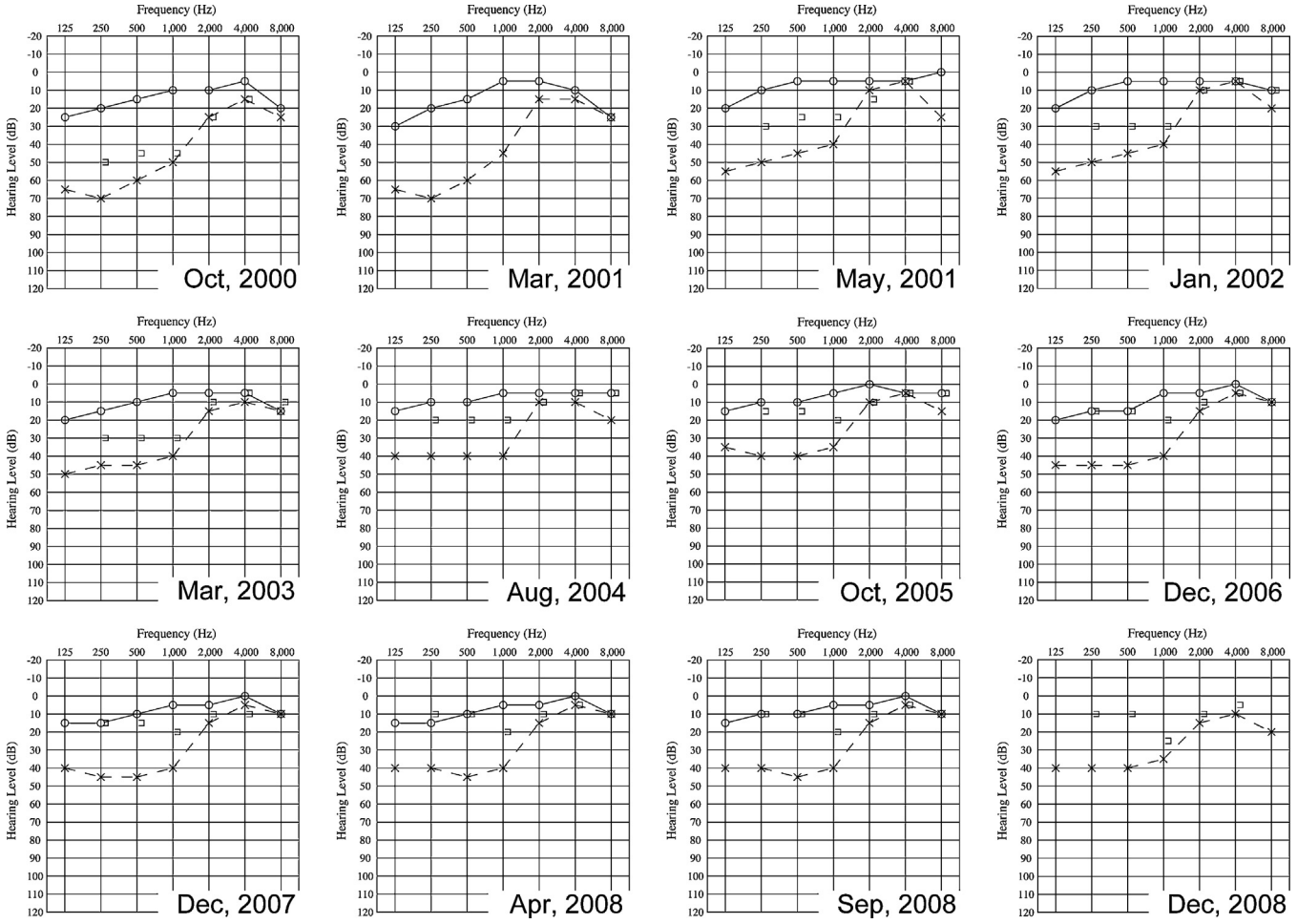

Fig. 1. The full course of the pure tone audiograms. Oct, 2000–Mar, 2001, audiograms taken at another institute. May, 2001–Dec, 2008, audiograms taken at the Kyushu University Hospital. Apr, 2008, the last audiogram taken before exploratory tympanotomy.

pure-tone audiometry (PTA) showed left mixed hearing loss with 15–20 dB air-bone gap over the frequency of 125 to 1000 Hz (Fig. 1 May, 2001). The tympanometry findings were normal on both sides. The distortion-product otoacoustic emission (DPOAE), tested at 1.5, 2.0, 3.0, 4.0, 5.0, and 6.0 kHz, was normal on the right side, but no distortion product was detected except for 3 kHz in the left ear, which is consistent with the PTA result. Her auditory brainstem response (ABR) confirmed left-sided hearing loss compatible with a 1 kHz threshold in PTA, which showed 50 dB, 20 dB, and 10 dB thresholds on the left ear, and 10 dB, 0 dB, and 10 dB thresholds on the right ear for 1 kHz, 2 kHz, and 4 kHz respectively; thus, functional and psychogenic hearing loss were ruled out.

A CT scan of the temporal bones showed no signs of infection in the middle ear or the mastoid cavity. No anomaly was noted within the scan. However, there was a high-riding jugular bulb on the left side (Fig. 2A).

We followed up the patient's PTA every six months for eight years (Fig. 1 May, 2001–Apr, 2008). The bone conduction threshold had gradually decreased and reached a near-normal level in 2008, without much change in the air conduction hearing; thus, her hearing had transformed to a pure conductive hearing loss (Fig. 1 Apr, 2008). Otosclerosis was suspected based on the hearing tests in 2007–2008, although we were aware of the atypical clinical features of this patient: that she had unusual progression of the air-bone gap and the fact that her stapedial reflex was positive on both sides.

The patient underwent exploratory tympanotomy in 2008. The incudo-stapedial joint and the stapes footplate were judged to be mobile. No abnormalities that could explain her conductive hearing loss were detected. The patient's middle ear was retained unchanged. The postoperative audiogram did not change (Fig. 1 Sep, 2008 and Dec, 2008).

A temporal bone CT was performed again in 2011 for the middle ear re-evaluation, in which dehiscence was detected between the high-riding jugular bulb and the posterior semicircular canal of the left ear (Fig. 2B and C). Retrospective re-evaluation of her first CT in our hospital showed a thin bony wall between posterior semicircular canal and jugular bulb (Fig. 2A).

The vestibular evoked myogenic potential (VEMP) showed an elevated amplitude on the affected side, which was more than twice as high as the right side at the same stimulation level. The findings indicated an abnormally low VEMP threshold on the affected side, which was typical of Tullio's phenomenon often seen in SCD patients.

3. Discussion

Conductive hearing loss is usually the result of either external or middle ear sound transmission problems. Therefore, patients with conductive hearing loss with intact external and middle ear structures are usually offered an option of exploratory tympanotomy with possible replacement of ossicles. However, conductive hearing loss can also occur when dehiscence of the bony