Case Report

Relapsing Sudden Deafness with Benign Paroxysmal Positional Vertigo

Jiann-Jy Chen,[1-3] Dem-Lion Chen[4]

Department of Otorhinolaryngology, School of Medicine, Fu Jen Catholic University, Taipei, Taiwan

ABSTRACT

We describe a 44-year-old woman who had right tinnitus up to six times in recent seven years. Right reversible sensory hearing loss was identified, but vertigo did not attribute to canulolithesis of semicircular canal. Although it is yet beyond the current image techniques and equipment to solve the enigma in such a case, we suggest an unknown recurrent vasculopathy, rather than Ménière's disease or migrainous vertigo acted on the underlying presumably abnormal microcirculation of the inner ear and left behind the right transient cochleopathy with partial vestibulopathy. (Rawal Med J 2010;35:).

Key words: sudden deafness, cochleopathy, vestibulopathy, benign paroxysmal positional vertigo, Ménière's disease, migrainous vertigo.

INTRODUCTION

Sudden deafness is defined as hearing losses up to 30dB hearing level (dBHL) over three continuing audiogram frequencies within three days. Unilateral sufferers are much more than bilateral ones. It is prevalent in adult Taiwanese between 40 and 60 years old, with the average

of 43.20.[1] The severity was judged by the average of hearing thresholds between 250 Hz and 4,000 Hz.[1] Sudden deafness is yet of unknown etiology, possibly a central lesion, a virus infection, a vasculopathy or an autoimmune disease. Relapsing sudden deafness with vertigo is often attributable to Ménière's disease or migrainous vertigo.[2,3] Most patients with benign paroxysmal positional vertigo (BPPV) are primary or idiopathic, approximately 15% of them follow an acute vestibular neuritis. Furthermore, although 8%~44% of them have a chronic ipsilateral sensorineural hearing loss, they are rarely associated with simultaneous ipsilateral sudden deafness,[4] and herein we report such a rare case from Taiwan.

CASE PRESENTATION

A 44-year-old robust female Taiwanese has been bothered with right tinnitus up to six times after several nights' staying up in recent seven years. The symptomatic duration was between one and two weeks and the symptom-free interval was between six and twelve months. When she was symptomatic, several seconds' vertigo could be induced several times a day by lying down or turning head rightward. There was no headache, blurred vision, paresthesia, nausea, ataxia or other focal neurologic symptoms. The previous four attacks were conservatively treated at other hospitals till August 2008 when she visited us for the 5th attack. Right Dix-Hallpike test was positive (Fig. 1A). The average hearing threshold was 38 dBHL in the

right ear and 5 dBHL in the other. Over the following week, she was conservatively treated with prednisolone 20mg orally daily. The tinnitus and episodic vertigo remitted and average hearing threshold was 12dBHL in the right ear and 9 dBHL in the other.

Fig 1 here

During the 6th attack on March 2008, she visited the author again for a further study. The right Dix-Hallpike test showed positive again and the positioning vertigo could be cured by three times Epley's canulolith repositional procedures (Rollover test: positive) but recurred in simply half an hour. The head-shaking test could provoke spontaneous leftward nystagmus. The average hearing threshold was 34 dBHL in the right ear and 8 dBHL in the other. The short increment sensitivity index over 1,000 Hz, 2,000 Hz and 4,000 Hz was respectively 10%, 20% and 25% in the right ear, and all 0% in the other. The speech discrimination score was both 100%. A Romberg test and tests for tandem gait, diadochokinesia, finger to nose tracing, knee to heel coordination and orthostatic hypotension did not show any abnormality.

The auditory brainstem response (ABR) revealed the latency of waves I, III and V was respectively 2.12 msec, 3.90 msec and 6.29 msec in the right ear, and 1.71 msec, 3.96 msec and 6.04 msec in the other (Fig. 1B). The posturagraphy showed that the body weight-center located rightward and afterward (Fig. 1C). The air-conduction vibrated cervical vestibular evoked myogenic potential (ACV-cVEMP) revealed the latency (amplitude) of p13 and n23 was respectively 14.50 msec (-45.35 μV) and 23.35 msec (59.19 μV) in the right side, and 16.38 msec (-78.16 μV) and 24.18 msec (94.89 μV) in the other (Fig. 1D). The electronystagmogram for pursuit, saccade, optokinetic-nystagmus and optokinetic-after-nystagmus did not show any

abnormality. The caloric nystagmus (20°C tap water) of the right ear did not show any rhythmic eyeball velocity waves and that of the left ear showed rightward rhythmic waves with a positive visual suppression (Fig. 1E).

T1, T2, fluid-attenuated inversion recovery, diffusion weighted magnetic resonance image with 3D time-of-flight angiogram did not show any abnormality. All blood examinations were within normal ranges, including C-reactive protein, erythrocyte sedimentation rate, anti-nuclear antibody, rheumatoid factor and anti-SSA/SSB. Over the following week, she was conservatively treated again with oral steroids. The tinnitus and episodic vertigo remitted and the average hearing threshold was 12 dBHL in the right ear and 6 dBHL in the other. The short increment sensitivity index was both 0% and the speech discrimination score was both 100%. The following ten months were uneventful.

DISCUSSION

Otoacoustic emission, electrocochleogram or ABR respectively investigate the hair cell, cochlea or auditory pathway;[5] however, simply ABR is available at our hospital, so we additionally applied pure tone audiometry, short increment sensitivity index and speech discrimination score to her. When she was symptomatic, the right hearing impairment with the positive short increment sensitivity index, the negative speech discrimination score test and the

normal ABR (Fig. 1B) indicated that the right cochlea was sick and the right cochlear nerve was healthy. [6] She did not have any headache, so migraine was unlikely cause. The duration of frequent vertigo was simply several seconds, so her disease did not meet the diagnostic criteria of Ménière's disease, in which vertigo should last for at least twenty minutes. [7] Although the right positive Dix-Hallpike test (Fig. 1A) and the positive Rollover test explicated BPPV, the positioning vertigo could recur in half an hour, indicating canulolithesis of the right posterior semicircular canal was unlikely causative.

The posturagraphy showed the body weight-center deviated in the right and posterior direction (Fig. 1C), indicating the predominance of her left and anterior vestibular function. An ACV-cVEMP study resorts to a loud sound for a sacculo-collic reflex in spite of deafness or normal hearing. The pathway passes through the lower brainstem, including the saccule, the inferior vestibular nerve, the cochlear nucleus and the spinal accessory nucleus. In healthy adult Taiwanese, the latencies of p13 and n23 are respectively 16.3±3.2 msec and 24.4±5.0 msec. [8] When she was symptomatic, the ACV-cVEMP's wave latencies were all at reference range (Fig. 1D), so the right sacculo-collic reflex pathway was healthy, including the right inferior vestibular nerve.

Fig 2 hare

A caloric test investigates the vestibular ocular reflex pathway, passing through the upper brainstem; besides, its visual suppression investigates the flocculus.[9] The cold-water caloric

test for her right ear did not provoke a significant rhythmic nystagmus (Fig. 1E), but that for her left ear provoked the rightward rhythmic nystagmus with a positive visual suppression, indicating the right vestibulopathy and the left health vestibule. Because the image study did not show any abnormality, a space-occupying lesion in the cerebellar-pontine angle was ruled out.

After the all above objective studies were concluded, the cochleopathy with partial vestibulopathy was impressed in the right ear. The function of the cochlear nerve, the saccule and the inferior vestibular nerve was well. However, the partial vestibulopathy is attributable to the superior vestibular neuropathy, or/and the lateral and anterior semicircular canulopathy. Because of the BPPV, the function of the right posterior semicircular canal was confirmed well. A vasculopathy, a vestibular neuropathy and a local viral reactivation are possible etiologies of acute unilateral vestibulopathy with sudden deafness. [4] In our patient, it might be a vasculopathy contributing to the cochleopathy with partial vestibulopathy because of recurrence. We suggest an unknown recurrent vasculopathy occurred after her exhaustion and acted on the underlying presumably abnormal microcirculation of the inner ear.

In a common inner ear (Fig. 2), the anterior vestibular artery (AVA) supplies the anterior semicircular canal (ASC), the horizontal semicircular canal (HSC), and their ampulla (Am) and

the posterior vestibular artery (PVA) supplies the posterior semicircular canal (PSC) and its ampulla. The common cochlear artery (CCoA) is branched from the anterior inferior cerebellar artery (AICA) and then branches into the AVA and the vestibular-cochlear artery (VCoA). The VCoA branches into the cochlear artery (CoA) and the PVA. In this patient, the CCoA is presumed to branch into the PVA and the VCoA, and the VCoA branches into the CoA and the AVA. A vasculopathy (Fig. 2: the filled pincer) over the VCoA is presumed contributable to the insufficiency of the CoA and the AVA, and thus transiently injured the cochlea, ASC and HSC, but spares the PSC. It is yet beyond the current image technique and equipment to evaluate the microcirculation of a live inner ear and the enigma in such a case should be addressed in the future. Furthermore, the patient responded to steroid treatment in every attack, so the role of autoimmune or inflammatory disease might also be a cause of her symptoms.

[1] Department of Otorhinolaryngology, Tao-Yuan Hospital, Department of Health, Executive Yuan, Taoyuan, Taiwan

[2] Faculty of Medicine, School of Medicine, Fu Jen Catholic University, Taipei, Taiwan

[3] Department of Radiology, Taipei Medical University-Shuang Ho Hospital, Taipei, Taiwan

[4] G-Home Otorhinolaryngologic Clinic, Kaohsiung, Taiwan

Correspondence: Dr. Dem-Lion Chen, G-Home Clinic, 143, Jiannan Road, Nanzi District, Kaohsiung, Taiwan. Tel: +886-7-3515056 Fax: +886-7-3522209

E-mail: jiannjy@yahoo.com.tw

Received: July 6, 2010 Accepted: July 17, 2010

REFERENCES

1. Ku WC, Chang HS, Su MC, Yang E. Prognosis of idiopathic sudden sensorineural hearing loss- a clinical analysis. Chung Shan Med J 2004;15:205-12.

2. Junicho M, Fushiki H, Aso S, Watanabe Y. Prognostic value of initial electronystagmography findings in idiopathic sudden sensorineural hearing loss without vertigo. Otol Neurotol 2008;29:905-9.

3. Eavns RW, Ishiyama G. Migraine with transient unilateral hearing loss and tinnitus. Headache 2009;49:756-8.

4. Karlberg M, Halmagyi GM, Büttner U, Yavor RA. Sudden unilateral hearing loss with simultaneous ipsilateral posterior semicircular canal benign paroxysmal positional vertigo: a variant of vestibulo-cochlear neurolabyrinthitis? Arch Otolaryngol Head Neck Surg 2000;126:1024-9.

5. Okuno H, Watanabe I. Audiological findings of prolonged Menière's disease. Auris Nasus Larynx 1990;17:157-63.

6. Buus S, Florentine M, Redden RB. The SISI test: a review. Part II. Audiology 1982;21:365-85.

7. American Academy of Otolaryngology, Committee on Hearing and Equilibrium. Guidelines for the diagnosis and evaluation of therapy in Menière's disease. Otolaryngol Head Neck Surg 1995;113:181-5.

8. Yang TL, Wu CH, Young YH. Normal value of vestibular evoked myogenic potential. J Taiwan Otolaryngol Head Neck Surg 2001;36:160-4.

9. Su MC, Young YH. Neurotological aging change of the cerebellum. J Taiwan Otolaryngol Head Neck Surg 1990;30:305-9.

Fig. 1. A: the Dix-Hallpike test. B: the ABR. C: the posturagraphy. D: the ACV-cVEMP. E: the caloric test (20°C tap water).

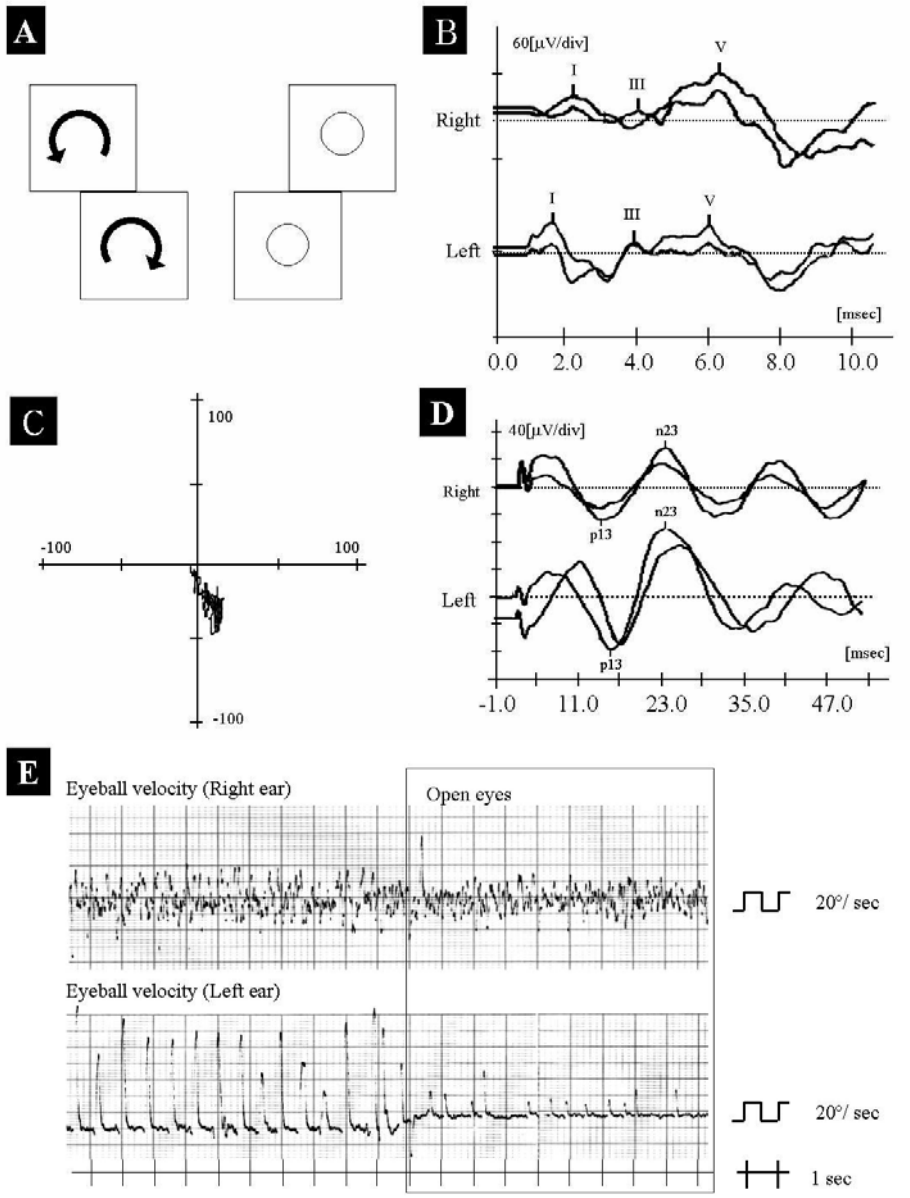

Fig. 2. A common inner-ear microcirculation and our presumption in the patient.