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Bilateral simultaneous sudden sensorineural hearing loss☆

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ABSTRACT

Objective: This study adopted an inner ear test battery and MR imaging in patients with bilateral sudden sensorineural hearing loss (SSNHL) to investigate their causes, disease extent, and evaluate hearing outcome.
Patients and methods: From 1995 to 2014, 16 patients with bilateral SSNHL received audiometry, caloric test and MR imaging. Vestibular-evoked myogenic potential (VEMP) test was added to the test battery after 2000.
Results: Percentages of abnormal mean hearing level (MHL), cervical VEMP test, ocular VEMP test, and caloric test in patients with bilateral SSNHL were 100% (32/32), 100% (12/12), 100% (4/4), and 81% (26/32), respectively, implying that not only the cochlear part but also the vestibular part was severely affected in both ears. Causes of bilateral SSNHL were neoplasm in 5 patients, stroke in 5, meningitis in 1, and unknown in 5. Post-treatment MHL did not significantly differ from pre-treatment MHL indicating poor hearing outcome. Seven patients (44%) had passed away within 5 years after onset, 2 patients were lost, and 7 patients survived.
Conclusion: Via MR imaging, causes of bilateral SSNHL were identified for 69% of cases. Both cochlear and vestibular endorgans/afferents were identified to be severely affected bilaterally by the vestibular test battery and resulted in poor hearing outcome. A high mortality rate (44%) indicates that bilateral SSNHL is an ominous sign for a more sinister underlying disease.

1. Introduction

Sudden sensorineural hearing loss (SSNHL) is defined as a rapid decline (less than three days) of more than 30 dB sensorineural hearing loss in at least three contiguous frequencies [1]. The estimated incidence of SSNHL in the United States ranges from 5 to 20 per 100,000 annually [2,3]. In contrast, a minor feature of SSNHL is bilateral involvement accounting for 0.4–3.4% of all SSNHL patients [4,5]. Most patients with bilateral SSNHL encompass a serious systemic disease with poor prognosis for hearing recovery despite immediate treatment. In the past, most researchers paid much attention on the dominant cochlear features of bilateral SSNHL, but less on the vestibular involvement.

Stimulation *via* loud sound or bone vibration enables recording of vestibular-evoked myogenic potential (VEMP) from cervical muscles (called cervical VEMP, cVEMP) and extraocular muscles (called ocular VEMP, oVEMP). A recent investigation showed that the cVEMP test assesses saccular function while the oVEMP test predominantly evaluates utricular function [6]. Thus, an inner ear test battery comprising audiometry, oVEMP test, cVEMP test, and caloric test help map the affected territory in the cochlea, utricle, saccule, and semicircular canals, and has been widely applied to patients with Meniere's disease [7], cerebellopontine angle tumor [8], unilateral SSNHL [9], etc. This study adopted an inner ear test battery in patients with bilateral SSNHL to investigate their causes, map the affected territory and evaluate hearing outcome.

2. Patients and methods

2.1. Patients

From 1995 to 2014, 1459 patients of SSNHL within 2 weeks were consecutively encountered at the clinic of the university hospital. Of them, 16 patients (1.1%) were diagnosed as bilateral SSNHL. Diagnosis of SSNHL is based on a rapid decline (less than three days) of more than 30 dB sensorineural hearing loss in at least three contiguous frequencies [1]. Bilateral SSNHL is defined as SSNHL simultaneously affected in both ears within 72 h.

Exclusion criteria included concurrent middle or inner ear anomaly, previous ear surgery, autoimmune inner ear disorders, progressive sensorineural hearing loss, recurrent sudden deafness, and bilateral sequential SSNHL (>72 h).

In total, 16 patients with bilateral SSNHL including 10 males and 6 females with mean age of 51 ± 21 years were enrolled in this study (Table 1). All patients were admitted to our ward and received a detailed history taking, local check-up of the ears, and a battery of tests including blood examination, neurological examination, audiometry, caloric test with electronystagmographic (ENG) recordings, and MR imaging examination. After 2000 and 2008, cVEMP and oVEMP tests were added to the test battery, respectively.

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Table 1
Clinical information of 16 patients with bilateral sudden sensorineural hearing loss.

No.	Age	Sex	Underlying disease	Caloric (R/L)	cVEMP (R/L)	Diagnosis	Outcome
1	56	F	Nil	n/n	No test	Neurofibromatosis type 2	Expired
2	17	F	Nil	Absent/absent	No test	Neurofibromatosis type 2	Alive
3	40	M	Nil	Absent/absent	Absent/absent	Neurofibromatosis type 2	Alive
4	42	M	Dyslipidemia	cp/n	Absent/absent	Neurofibromatosis type 2	Alive
5	51	M	Dyslipidemia	cp/absent	Absent/absent	Basilar artery occlusion	Expired
6	64	F	CAD, DM, hypertension	cp/cp	No test	Basilar artery occlusion	Expired
7	59	M	Dyslipidemia	Absent/cp	No test	Stroke	Alive
8	79	M	Nil	Absent/absent	Absent/absent	Stroke	Expired
9	70	F	CAD, DM, dyslipidemia, hypertension	n/n	Absent/absent	Stroke	Lost
10	61	F	Lung cancer	Absent/absent	No test	Brain metastasis	Expired
11	46	M	Dyslipidemia	cp/cp	No test	Cryptococcal meningitis	Alive
12	5	M	Thalassemia	cp/cp	No test	Bilateral SSNHL	Alive
13	72	F	Hypertension	cp/cp	No test	Bilateral SSNHL	Lost
14	60	M	Irradiated NPC	cp/absent	No test	Bilateral SSNHL	Expired
15	52	M	Irradiated NPC	n/absent	Absent/absent	Bilateral SSNHL	Alive
16	39	M	Irradiated NPC	Absent/absent	No test	Bilateral SSNHL	Expired

CAD: coronary artery disease; cp: canal paresis; cVEMP: cervical vestibular-evoked myogenic potential; DM: diabetes mellitus; n: normal; NPC: nasopharyngeal carcinoma.

Blood examination consisted of complete blood cell counts, biochemistry profile, serological tests for syphilis, c-reactive protein, antinuclear antibody, rheumatoid factor, lupus erythematosis preparation, and serum protein electrophoresis. Viral titers against herpes family e.g. herpes simplex, varicella zoster, EB virus, or cytomegalovirus were also included.

This study was approved by the institutional review board, and each subject signed the informed consent to participate.

2.2. Audiometry

Mean hearing level (MHL) was defined as averaged hearing thresholds at four frequencies of 500, 1000, 2000 and 4000 Hz. Mean hearing gain indicates the difference between the pre-treatment MHL and the post-treatment one. Outcome of hearing recovery was evaluated according to the literature. “Cured” indicates a patient in whom the MHL recovers to the threshold of 20 dB or better. “Improved” indicates those in whom the mean hearing gain is more than 10 dB, whereas “Unchanged” means that the mean hearing gain is equal or less than 10 dB [9].

2.3. Caloric test

Bithermal caloric test was conducted with an ENG recorder. The norm of slow phase velocity (SPV) of caloric nystagmus at our laboratory is 31 ± 7 (mean ± SD)°/s. Canal paresis is defined when the mean SPV of caloric nystagmus in the lesion ear is <17°/s, or as a greater than 25% difference between maximum SPV measurements for each ear, when compared with the sum of SPVs from each ear. If caloric response was not elicited, the subject underwent ice water (4 °C, 10 mL) caloric testing to further confirm the caloric areflexia.

2.4. oVEMP test

The subject was in a sitting position. Two active electrodes were placed around 1 cm below the lower eyelids. The other two reference electrodes were positioned 1–2 cm below the active ones, and one ground electrode was on the sternum. During recording (Smart EP 3.90, Intelligent Hearing Systems, Miami, USA), the subject was instructed to look upward at a small fixed target >2 m from the eyes. The stimulation rate was 5/s. The duration of analysis of each response was 50 ms, and 30 responses were averaged for each run. The operator held the vibrator and delivered a repeatable tap on the forehead. If oVEMP was not elicited, alternatively, tapping at the ipsilateral mastoid site (2 cm behind the opening of external ear canal) was performed [10]. The input signal was 500 Hz sine wave, driven by a custom amplifier. The drive voltage was adjusted to produce a peak force equivalent to 144 dB force level.

The initial negative–positive biphasic waveform comprised peaks nI and pI. Consecutive runs were performed to confirm the reproducibility of peaks nI and pI, and oVEMPs were deemed to be present.

2.5. cVEMP test

Each subject was in a supine position. Two active electrodes were placed on the upper half of the sternocleidomastoid (SCM) muscles; one reference electrode was positioned on the suprasternal notch, and a ground electrode was situated on the forehead. The other settings were the same as in the oVEMP test, except that the vibrator delivered a repeatable tap on the subject's head at inion [11]. Subjects were given feedback of the level of EMG activity in their SCM muscles and were required to keep a background muscle activity of at least >50 μV. The subjects elevated their heads during testing. A total of 50 responses were averaged and recorded bilaterally.

The first positive and second negative polarities of biphasic waveform were termed waves p13 and n23, respectively. Consecutive runs were performed to confirm the reproducibility of peaks p13 and n23, and cVEMPs were deemed to be present.

2.6. Statistical methods

The MHL between the right and left ears, before and after treatment were compared by paired t test. A significant difference indicates $p < 0.05$.

3. Results

3.1. Clinical manifestation

Clinical manifestation in 16 bilateral SSNHL patients comprised tinnitus (63%), vertigo/dizziness (50%), headache (38%), ataxia (31%), nausea/vomiting (13%), and fullness sensation (6%). Six patients (38%) had underlying systemic diseases *i.e.* dyslipidemia in 5, hypertension in 3, diabetes mellitus in 2, and coronary artery disease in 2.

3.2. Audiometry

Types of audiogram consisted of total loss in 10 ears, flat type in 8 ears, down-slope type in 7 ears, high-tone loss in 4 ears, and saucer type in 3 ears (Table 2). Of them, similar type on both ears was observed in 10 patients (62%).

Average hearing thresholds for the 16 bilateral SSNHL patients at frequencies of 500, 1000, 2000 and 4000 Hz were 68 ± 33 (mean ± SD),