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Audiovestibular impairments associated with intracranial hypotension

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ABSTRACT

Objective: To investigate the patterns and mechanisms of audiovestibular impairments associated with intracranial hypotension.
Methods: We had consecutively recruited 16 patients with intracranial hypotension at the Neurology Center of Pusan National University Hospital for two years. Spontaneous, gaze-evoked, and positional nystagmus were recorded using 3D video-oculography in all patients, and the majority of them also had pure tone audiometry and bithermal caloric tests.
Results: Of the 16 patients, five (31.3%) reported neuro-otological symptoms along with the orthostatic headache while laboratory evaluation demonstrated audiovestibular impairments in ten (62.5%). Oculographic analyses documented spontaneous and/or positional nystagmus in six patients (37.5%) including weak spontaneous vertical nystagmus with positional modulation (n = 4) and pure positional nystagmus (n = 2). One patient presented with recurrent spontaneous vertigo and tinnitus mimicking Meniere's disease, and showed unidirectional horizontal and torsional nystagmus with normal head impulse tests during the attacks. Bithermal caloric tests were normal in all nine patients tested. Audiometry showed unilateral (n = 6) or bilateral (n = 1) sensorineural hearing loss in seven (53.8%) of the 13 patients tested.
Conclusions: Intracranial hypotension frequently induces audiovestibular impairments. In addition to endolymphatic hydrops and irritation of the vestibulocochlear nerve, compression or traction of the brainstem or cerebellum due to loss of CSF buoyancy may be considered as a mechanism of frequent spontaneous or positional vertical nystagmus in patients with intracranial hypotension.

1. Introduction

Intracranial hypotension is characterized by orthostatic headache due to low CSF volume caused by either spontaneous or post-traumatic dural laceration [1–5]. The headache is typically provoked by sitting or standing, and relieved by lying flat. Associated symptoms include nausea, vomiting, pain and tight feeling in the neck, diplopia, and blurred vision. A study of 30 consecutive patients with intracranial hypotension has described neurotological symptoms including dizziness (30%), tinnitus (20%), aural fullness (20%), and hearing loss (3%) [1], while another reported auditory symptoms in approximately 70% of the patients [6]. Earlier reports have also documented unilateral or bilateral low-frequency sensorineural hearing loss on audiometry with or without vertigo, likewise in Meniere's disease (MD) [6–17]. However, previous studies have mostly focused on auditory dysfunction, and no study has attempted objective documentation of vestibular dysfunction in intracranial hypotension.

To determine the patterns and mechanisms of audiovestibular impairments associated with intracranial hypotension, we performed neurotological evaluation in 16 consecutive patients.

2. Materials and methods

2.1. Subjects

We had consecutively recruited 16 patients with intracranial hypotension according to the International Classification of Headache Disorders criteria for headaches attributed to spontaneous (or idiopathic) low CSF pressure at the Neurology Center of Pusan National University Hospital for two years from November 2011 to November 2013 [18]. All patients had orthostatic headache, and brain MRIs showed strong contrast enhancements and thickening of the pachymeninges in ten (62.5%), compatible with the typical findings of intracranial

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hypotension. Lumbar CSF studies were also performed in 13 patients. Eleven patients had spontaneous intracranial hypotension while the remaining five developed the symptoms just after lumbar puncture for spinal anesthesia due to operation (n = 3, rectal cancer, hemorrhoids, and penile curvature respectively), or chiropractic manipulation for chronic lower back pain (n = 2). The duration of intracranial hypotension ranged from 1 to 21 days (6.1 ± 5.2 days). The patients included seven men and nine women with the ages ranging from 23 to 64 years (mean ± SD = 41.9 ± 10.1 years).

All the experiments followed the tenets of the Declaration of Helsinki and were approved by the Institutional Review Board of Pusan National University Hospital. Informed contents were obtained after the nature and possible consequence of this study had been explained to the participants.

2.2. Neuro-otological evaluation

All patients had full bedside neurological and neuro-otological evaluation by the authors. Eye movements were recorded binocularly at a sampling rate of 60 Hz using 3D video-oculography (SensoMotoric Instruments, Teltow, Germany). Spontaneous nystagmus (SN) was recorded both with and without fixation. Gaze-evoked nystagmus was induced with horizontal (±30°) and vertical (±20°) target displacements. For positional nystagmus (PN), patients bent down, straightened, and turned their heads to either side while supine. Patients were also subjected to the straight head hanging. The presence of spontaneous and positional nystagmus in the patients was defined only when the mean slow phase velocity (SPV) of the spontaneous nystagmus and maximal SPV of the positional nystagmus exceeded the values (mean + 2SD) observed in normal controls (horizontal SN ≥ 1.1°/s; vertical SN ≥ 1.8°/s; torsional SN > 0°/s; horizontal PN ≥ 2.6°/s; positional upbeat nystagmus ≥ 3.8°/s, positional downbeat nystagmus ≥ 0°/s; torsional PN ≥ 0.9°/s). In patients with spontaneous nystagmus, the SPV of positional nystagmus was determined by subtracting the SPV of spontaneous nystagmus.

Out of 16 patients, 13 received pure tone audiometry in sitting position, and nine had bithermal caloric tests in supine position with the head inclined at 30°. The remaining patients refused to receive the tests due to profound orthostatic headache. The caloric stimuli comprised alternative irrigation for 25 s with about 250 ml of cold and hot water (30 °C and 44 °C). Caloric paresis was defined by a response difference of 25% or more between the ears using the Jongkees' formula.

Medications that could potentially affect the vestibular system were not allowed during the study.

2.3. Treatment strategy and follow-up evaluation

All patients received conservative treatment (n = 6) or epidural blood patch (n = 10). After resolution of orthostatic headache with the treatments, the patients with abnormal eye movements previously had a follow-up recording of eye movements.

3. Results

3.1. Clinical characteristics

Detailed demographic and clinical profiles of the patients are described in Table 1.

Five (5/16, 31.3%) had neuro-otological symptoms along with the orthostatic headache. Two patients (patients 1 and 3) developed both orthostatic dizziness and tinnitus, and the remaining three had orthostatic dizziness (patient 6) or orthostatic tinnitus (patients 9 and 10). Two patients (patients 1 and 3) reported tinnitus in both ears while another two (patients 9 and 10) noticed tinnitus only in one ear. One patient (patient 1) without a previous history of medical or neuro-otological diseases presented with recurrent spontaneous vertigo, earfullness, and tinnitus in either ear along with the orthostatic dizziness, tinnitus, and headache. The attacks lasted for several hours and occurred 1–2 times per day, which mimicked MD.

3.2. Neuro-otological evaluation

Six (6/16, 37.5%) patients showed spontaneous and/or positional nystagmus (Table 2). Three patients (patients 1, 2, and 6) developed weak spontaneous downbeat nystagmus without visual fixation in sitting position. In patient 1, the downbeat nystagmus did not change after taking supine position, but changed into upbeat nystagmus during straight head hanging (Fig. 1A). Patient 2 showed slight attenuation of downbeat nystagmus while supine, but no change during straight head hanging. In patient 6, downbeat nystagmus increased significantly and accompanied counter-clockwise torsional and left-beating components in supine and straight head hanging positions (Fig. 1B and Video 1). Patient 3 disclosed weak spontaneous upbeat nystagmus without visual fixation in sitting position, which was exaggerated in supine and straight head hanging positions (Fig. 1C). Counter-clockwise torsional and left-beating components were also accompanied in the supine position, and left-beating component was associated in the straight head hanging position. The remaining two patients exhibited isolated positional nystagmus including pure upbeat (patient 4) or mainly upbeat nystagmus mixed with right-beating and clockwise torsional components (patient 5) in the straight head hanging position. In all six patients, positional nystagmus lasted more than one minute. On resuming sitting position, reversal of the positional nystagmus occurred in two patients (patients 1 and 5).

During two attacks of vertigo in patient 1, we observed unidirectional horizontal and torsional nystagmus beating to the right or left without visual fixation (Video 2), but bedside horizontal head impulse tests were always normal.

Bithermal caloric tests were normal in all nine patients tested, and seven (54%) of the 13 patients with audiometry showed unilateral low-frequency sensorineural hearing loss (SNHL) (n = 6) or bilateral low- and high-frequencies SNHL (n = 1). The degree of hearing loss was mild in all the patients.

3.3. Follow-up evaluation of the audiovestibular dysfunction

All patients had resolution of orthostatic headache immediately or within several days after epidural blood patch or conservative treatments. Six patients also showed the disappearance of spontaneous and positional nystagmus immediate or within several days after resolution of orthostatic headache. In four patients (patients 1, 2, 5, and 6), spontaneous and positional nystagmus disappeared within several days ranging from 1 to 7 days (mean = 4 days) after epidural blood patch. In patient 1, recurrent attacks of vertigo and auditory symptoms improved after diuretics treatment for three months.

Four (patients 1, 3, 5, and 10) of the seven patients received follow-up evaluation of auditory function at different periods. Three (patients 1, 3, and 10) of them showed resolution of hearing loss at three months, 20 days, and four months respectively after the resolution of orthostatic headache. However, hearing loss did not improve at two weeks after the resolution of orthostatic headache in patient 5.

4. Discussion

The present study demonstrated that approximately 30% of patients with intracranial hypotension report neuro-otological symptoms including dizziness, earfullness, and tinnitus along with orthostatic headache, while laboratory evaluation discloses audiovestibular impairments in 63%. Unilateral or bilateral SNHL was found in about half of the patients, and 38% exhibited spontaneous or positional nystagmus. These results support earlier findings of frequent audiovestibular impairments among the patients with intracranial hypotension [1,6]. Of interest, one

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