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Chasing dizzy chimera: Diagnosis of combined peripheral and central vestibulopathy

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ABSTRACT

Diagnosis of combined peripheral and central vestibulopathy remains a challenge since the findings from peripheral vestibular involvements may overshadow those from central vestibular disorders or vice versa. The aim of this study was to enhance detection of these intriguing disorders by characterizing the clinical features and underlying etiologies. We had recruited 55 patients with combined peripheral and central vestibulopathy at the Dizziness Clinic of Seoul National University Bundang Hospital from 2003 to 2013. Peripheral vestibular involvement was determined by decreased caloric responses in either ear, and central vestibulopathy was diagnosed with obvious central vestibular signs or the lesions documented on MRIs to involve the central vestibular structures. Combined peripheral and central vestibulopathy could be classified into four types according to the patterns of vestibular presentation. Infarctions were the most common cause of acute unilateral cases while cerebellopontine angle tumors were mostly found in chronic unilateral ones. Wernicke encephalopathy and degenerative disorders were common in acute and chronic bilateral disorders. Twenty five (45.5%) patients showed only vestibular findings with or without auditory involvements, but association with gaze-evoked nystagmus, impaired smooth pursuit or central types of head shaking nystagmus indicated a central vestibular involvement in most of them (23/25, 92.0%). Given the requirements for urgent treatments and potentially grave prognosis of combined vestibulopathy, central signs should be sought even in patients with clinical or laboratory features of peripheral vestibulopathy. Scrutinized bedside evaluation, however, secured the diagnosis in almost all the patients with combined vestibulopathy.

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1. Introduction

Due to potentially grave prognosis of central vestibular disorders, differentiation of central from peripheral causes has always been the prime goal in the diagnostic approaching of the vestibular symptoms such as dizziness, vertigo, and unsteadiness [1]. Recent progress in clinical neurotology and neuroimaging, however, markedly improved diagnosis of isolated peripheral or central vestibular disorders [2]. Especially, the introduction of head impulse tests (HITs) and HINTS (negative HIT, direction-changing nystagmus, and skew deviation) has greatly enhanced bedside differentiation of central from peripheral vestibular disorders [3,4].

Several disorders may involve both peripheral and central vestibular structures, and failure to identify central signs in these chimeric disorders may lead to disastrous outcome since the prognosis is mostly dependent upon central vestibular involvements. However, combined peripheral and central vestibulopathy frequently poses a diagnostic difficulty since the peripheral vestibular signs may overshadow the central ones or vice versa [5]. For example, the HINTS may not be enough to detect central disorders such as anterior inferior cerebellar artery (AICA) infarction that mostly presents acute prolonged vertigo from lesions involving the brainstem and cerebellum as well as the inner ear [6]. Furthermore, recent studies described positive HITs in various lesions involving the brainstem or cerebellum, making the distinction between the central and peripheral vestibular lesions more difficult [7–9]. The aim of this study was to enhance detection of these intriguing disorders by characterizing the clinical features and underlying etiologies.

2. Methods

At the referral-based Dizziness Clinic of Seoul National University Bundang Hospital, 55 patients (27 men, mean age = 63.0, age range = 31–86) had a diagnosis of combined peripheral and central vestibulopathy from 2003 to 2013. Peripheral vestibular involvements were determined when the patients showed caloric paresis (CP) in either ear.

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Central vestibulopathy was defined by obvious central neurotologic signs including HINTS, head-shaking nystagmus (HSN) with central patterns, or by lesions involving the central vestibular structures, as was documented on MRIs. The ocular motor findings other than HIT and skew deviation were based on the results of oculography. According to the chronologic patterns of neurotologic presentation and the lesion side, the combined vestibulopathy was classified into four types; acute unilateral, acute bilateral, chronic unilateral, and chronic bilateral.

Acute vestibular syndrome was defined as sudden onset of vestibular symptoms and signs with their peak within days and gradual improvements thereafter while the chronic syndrome was defined as persistence or worsening of vestibular symptoms and signs for more than three months. Some patients have been described previously [6,8,10].

The patients had bedside evaluation of the ocular alignment using cover and alternate cover tests, spontaneous and evoked nystagmus, saccades, smooth pursuit (SP), visually enhanced vestibulo-ocular reflex (VVOR), in addition to routine neurological examination [11]. The nystagmus was also evoked by horizontal head-shaking, and positional maneuvers that included head bending, and leaning, lying down, head turning to either side while supine, and straight head-hanging. Spontaneous nystagmus (SN) was observed both with and without fixation, and the evoked nystagmus was evaluated only without fixation using video Frenzel goggles (SLMED, Seoul, Korea). Bedside HITs were performed manually with a rapid rotation of the head of ~20° amplitude in the planes of all semicircular canals (SCCs). The HIT was considered abnormal if a corrective saccade consistently supplemented the inadequate slow phase in the plane of the SCCs stimulated [12].

Nystagmus was recorded binocularly at a sampling rate of 60 Hz using a video-oculography (SensoMotoric Instruments, Teltow, Germany) [13]. Detailed methods have been described previously [13,14]. GEN was defined by the direction-changing nystagmus that beat in the direction of gaze in the both horizontal (±30°) planes. Central patterns of HSN included HSN beating to the lesion side, HSN in the opposite direction of SN, and perverted HSN (mainly downbeat nystagmus developing in response to horizontal head-shaking) [6,15–17].

HITs were measured in 14 patients. To quantify HITs, the head and eye movements were recorded using a magnetic search coil technique in a 70 cm cubic search coil frame (Skalar, Delft, The Netherlands) [10]. Detailed description on the methods and normative data is available elsewhere [10]. Patients also had evaluation of bithermal caloric tests, ocular torsion using fundus photography, and tilt of the subjective visual vertical [13]. CP was determined when the summated peak slow phase velocities of the induced nystagmus in response to cold and warm water did not exceed 10°/s in either ear.

MRIs were obtained with a 3.0 T or 1.5 T unit (Intera, Philips Medical Systems, Best, The Netherlands) with a section thickness of 3 or 5 mm [18]. The arterial territories were determined according to the previously validated anatomical templates [19].

3. Results

3.1. Etiology

Overall, the etiologies of combined peripheral and central vestibular disorders included infarctions ($n = 23$, 41.8%), tumors ($n = 17$, 30.9%), degenerative disorders ($n = 7$, 12.7%), Wernicke encephalopathy (WE, $n = 5$, 9.1%), cerebral superficial siderosis (n = 2, 3.6%), and infection (n = 1, 1.8%) (Table 1). All the infarctions involved the brainstem or cerebellum. The tumors were invariably located in or around the cerebellopontine angle (CPA). The radiological diagnosis of extra-axial tumors ($n = 12$) was uniformly vestibular schwannoma including the one with neurofibromatosis type 2. Radiologic or pathologic diagnosis of the intra-axial tumors included anaplastic astrocytoma ($n = 2$) and lymphoma ($n = 3$). Patients with degenerative disorders had the diagnosis of cerebellar ataxia and bilateral vestibulopathy (CABV, $n = 5$) [20] or cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome (CANVAS, $n = 2$) [21]. Five patients had WE due to chronic alcoholism (n = 2) or long-term total parenteral nutrition ($n = 3$). The superficial siderosis was idiopathic in one and due to repeated hemorrhages from spinal cord chordoma in the other.

Table 1
Etiologies of combined peripheral and central vestibulopathy.

Etiologies	No.	Percentage (%)
Infarction	**23**	**41.8**
AICA	*13*	*23.6*
AICA + PICA	*8*	*14.5*
PICA	*1*	*1.8*
Venous	*1*	*1.8*
CPA tumor	**17**	**30.9**
Extra-axial	*12*	*21.8*
Intra-axial	*5*	*9.1*
Degenerative disorder	**7**	**12.7**
CABA	*5*	*9.1*
CANVAS	*2*	*3.6*
Wernicke encephalopathy	**5**	**9.1**
Superficial siderosis	**2**	**3.6**
Infection	**1**	**1.8**
Total	**55**	**100.0**

AICA, anterior inferior cerebellar infarction; CABV, cerebellar ataxia and bilateral vestibulopathy; CANVAS, cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome; CPA, cerebellopontine angle; PICA, posterior inferior cerebellar artery.

3.2. Clinical characteristics

All patients experienced dizziness ($n = 9$, 16.4%), vertigo ($n = 27$, 49.1%), or unsteadiness ($n = 19$, 34.5%). Other common findings included hearing loss ($n = 25$, 45.5%), limb ataxia, ($n = 17$, 30.9%), headache ($n = 14$, 25.5%), facial palsy ($n = 13$, 23.6%), and diplopia ($n = 10$, 18.2%). Twenty five (45.5%) patients showed only vestibular findings with or without auditory involvements. In these patients, associated GEN, impaired SP, and the central types of HSN allowed determination of central vestibular involvements in almost all the patients (23/25, 92%) (Table 2).

The combined vestibulopathy was classified into four types; acute unilateral ($n = 21$, 38.2%), acute bilateral ($n = 8$, 14.5%), chronic unilateral ($n = 15$, 29.1%), and chronic bilateral ($n = 11$, 20.0%).

3.3. Acute unilateral combined vestibulopathy

Except one with Ramsay Hunt syndrome, all patients (21/22, 95.5%) in this group had acute cerebellar or brainstem infarction (Fig. 1). The infarctions involved the territories of AICA or posterior inferior cerebellar artery. One patient had a venous infarction involving the middle cerebellar peduncle.

All had acute spontaneous vertigo along with nausea/vomiting (15/22, 68.2%), hearing loss (12/22, 54.5%), limb ataxia (11/22, 50.0%), facial palsy (5/22, 22.7%), or diplopia (4/22, 18.2%). Vertigo or imbalance was the only clinical finding in five patients (22.7%).

GEN occurred in 16 (16/22, 72.7%) and skew deviation in four patients (5/22, 22.7%). Bedside HITs were positive during head turning to the side of CP in 20 (90.9%) patients. Central types of HSN were observed in nine (4.1%) patients. Six (27.3%) patients exhibited CPN, downbeat ($n = 3$), upbeat ($n = 1$) or apogeotropic ($n = 2$). Impaired SP (15/22, 68.2%) was also common. The patient with Ramsay Hunt syndrome showed GEN although brain MRIs were normal. Overall, the HINTS was negative in five patients (5/22, 22.7%), and three of them showed central types of HSN, and another had CPN and impaired SP. The remaining patient (patient 8) had no central signs, but the ocular motor evaluation was incomplete (Table 2).

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