Journal of Vestibular Research 24 (2014) 443–451
DOI 10.3233/VES-140524
IOS Press

Combined peripheral and central vestibulopathy

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Received 4 February 2014
Accepted 12 May 2014

Abstract. Diagnosis of central vestibulopathy remains a challenge when it is associated with peripheral vestibular dysfunction because neurotological findings from peripheral vestibulopathy may overshadow those from central vestibular involvements. To define the characteristics of disorders involving both peripheral and central vestibular structures, we classified the combined vestibulopathies into four types according to their vestibular manifestations, and describe a typical case in each subtype. Infarction involving the territory of anterior inferior cerebellar artery is the most common cause of *acute unilateral* cases, whereas tumors involving the cerebellopontine angle should be of prime suspicion in patients with *chronic unilateral* ones. Wernicke encephalopathy was most common in patients with *acute bilateral* combined vestibulopathy while degenerative disorders should be considered in chronic bilateral ones. Since the head impulse test (HIT) is mostly positive in combined vestibulopathy, signs of central vestibular dysfunction other than negative HIT should be sought carefully even in patients with obvious clinical or laboratory features of peripheral vestibulopathy.

Keywords: Vertigo, vestibular disorders, infarction, tumor, Wernicke encephalopathy

1. Introduction

With recent progress in clinical neurotology, bedside diagnosis of peripheral and central vestibular disorders has been made easier with the head impulse test (HIT) [3,12,13] and HINTS (negative head impulse test, direction-changing nystagmus, and skew deviation) [16]. These clinical tools assist the clinician in the differentiation of central from peripheral vestibular disorders. HINTS aids the diagnosis of strokes. It has a sensitivity of 100% and specificity of 96% in patients with acute vestibular syndrome and at least one stroke risk factor [16]. However, there are several disorders that may involve both peripheral and central vestibular structures, and diagnosis of these combined central and peripheral vestibular disorders remains a challenge because the peripheral vestibular signs may overshadow the central ones [6,14]. The HIT is the most sensitive sign for diagnosis of a central vestibulopathy when it is negative [28], and the HINTS may not be enough to detect central lesions in combined peripheral and central vestibulopathy such as infarctions involving the territory of anterior inferior cerebellar artery (AICA) [14,24]. Furthermore, lesions restricted to the brainstem or cerebellum may show positive HIT or caloric paresis, which are considered typical findings of peripheral vestibular loss, making the distinc-

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ISSN 0957-4271/14/$27.50

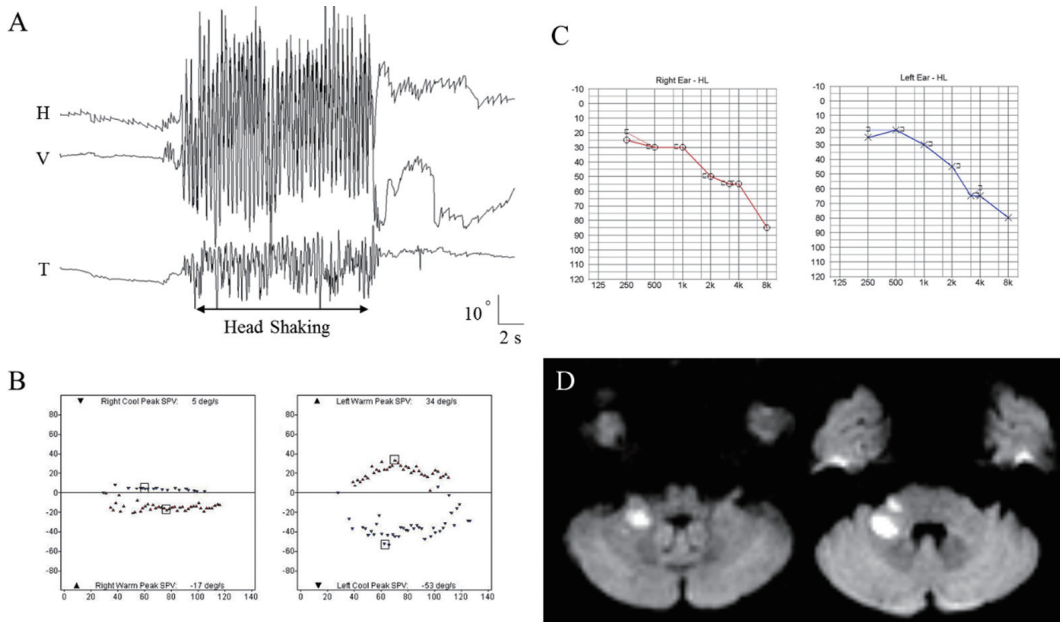

Fig. 1. Anterior inferior cerebellar artery infarction manifesting with acute unilateral combined vestibulopathy with hearing loss. (A) In the recording of eye movements using three dimensional video-oculography (SMI®, Teltow, Germany), the spontaneous horizontal-torsional nystagmus beating to the right changes into nystagmus beating to the left, counter-clockwise (upper pole of the eyes beating to the left ear), and downward after horizontal head shaking. Upward deflection indicates rightward, upward, and clockwise eye motion. (B) Bithermal caloric tests document right caloric paresis of 60%. (C) Pure tone audiometry shows right sensorineural hearing loss with pure tone average at 36.7 dB, compared with 31.7 dB in the left ear. (D) Diffusion-weighted MRI shows acute infarction involving right flocculus and middle cerebral peduncle supplied by the anterior inferior cerebellar artery. (Colours are visible in the online version of the article; http://dx.doi.org/10.3233/VES-140524)

tion between the central and peripheral vestibular lesions more difficult [10,17,29]. To define the characteristics of combined peripheral and central vestibular disorders, we classified the combined vestibulopathy into four subtypes according to their vestibular presentation, and describe a typical case in each subtype. Three of the patients (cases 1, 2, and 4) have been reported previously [7,14,17].

2. Case descriptions

2.1. *Case 1: Acute unilateral combined vestibulopathy with hearing loss: AICA infarction*

An 81-year-old woman with hypertension and diabetes developed acute spontaneous vertigo. She denied headache or hearing loss. On examination 7 days after symptom onset, she showed spontaneous horizontal-torsional nystagmus beating to the right with visual fixation. The nystagmus increased with prevention of visual fixation and in rightward gaze, but decreased on leftward gaze without changing directions. Skew deviation was not present, as determined with alternate cover test. Bedside HIT was positive when the head was turned to the right. After horizontal head shaking, the spontaneous nystagmus changed into left beating with downbeat and counterclockwise torsional (upper poles of the eye beating to the left ear) components (Fig. 1-A). She also had right caloric paresis (Fig. 1-B) and hearing loss (Fig. 1-C). Brain MRI showed acute right AICA infarction involving the right flocculus and middle cerebellar peduncle (Fig. 1-D).

2.2. *Case 2: Isolated vestibular nuclear infarction mimicking acute unilateral combined vestibulopathy without hearing loss*

A 34-year-old man with hypertension developed acute spontaneous vertigo with vomiting and imbalance. He had no other neurological or auditory symptoms. About 7 hours after symptom onset, examination showed spontaneous left beating horizontal-torsional nystagmus that increased during leftward gaze and with prevention of visual fixation, and changed into right beating during rightward gaze (Fig. 2-A). He had no head tilt or skew deviation. Bedside HIT was positive for right horizontal semicircular canal (HC). However, recording of HIT using a magnetic search coil

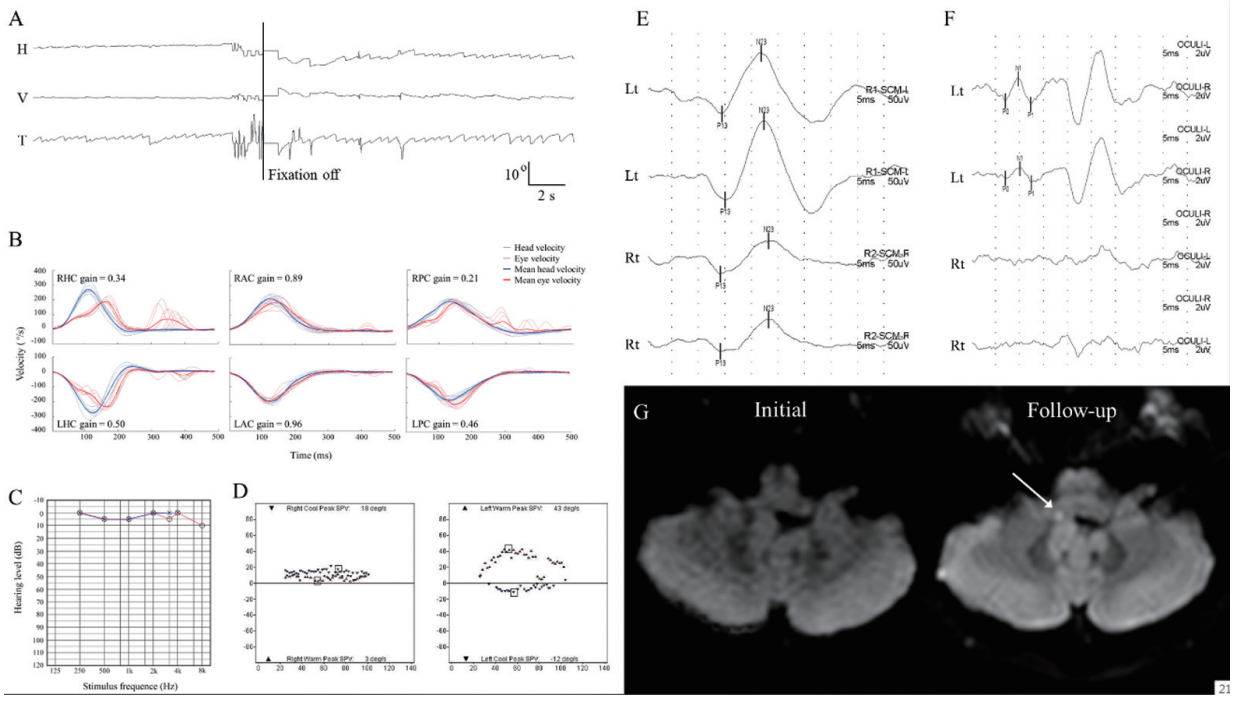

Fig. 2. Isolated vestibular nuclear infarction manifesting with acute unilateral combined vestibulopathy without hearing loss. (A) Three dimensional video-oculography (SMI®, Teltow, Germany) shows spontaneous nystagmus beating leftward, upward, and counter-clockwise, which increases with prevention of visual fixation. In each recording, the upward deflection indicates rightward, upward and clockwise torsional eye motion. (B) Recording of the head impulse test using a magnetic scleral coil technique documents reduced gains of the vestibulo-ocular reflex for the horizontal and posterior semicircular canals on both sides, but more for the right canals. (C) Pure tone audiometry is normal. (D) Bithermal caloric tests show right canal paresis of 57%. (E) Cervical vestibular-evoked myogenic potentials (VEMPs) in response to air-conducted tone burst sounds show decreased amplitudes during right ear stimulation with an interaural difference ratio of 39.7% (normal range $\leqslant$ 21.5%). (F) Ocular VEMPs elicited by vibratory stimuli shows no wave formation during right ear stimulation. (G) Diffusion-weighted MRI 4 hours after symptom onset is normal, but follow-up MRI 4 days later discloses acute infarction involving the right medial vestibular nucleus. AC: anterior canal, H: horizontal eye position, HC: horizontal canal, LHC: left horizontal canal, LAC: left anterior canal, LPC: left posterior canal, PC: posterior canal, RAC: right anterior canal, RHC: right horizontal canal, RPC: right posterior canal, T: torsional eye position, V: vertical eye position. (Colours are visible in the online version of the article; http://dx.doi.org/10.3233/VES-140524)

system documented decreased gains of the vestibulo-ocular reflex (VOR) for the HCs and posterior semicircular canals (PCs) on both sides, but more for the right HC and PC (Fig. 2-B). Pure tone and speech audiometry were normal (Fig. 2-C). Bithermal caloric irrigation showed right canal paresis (Fig. 2-D). Cervical and ocular vestibular evoked myogenic potentials were decreased or absent during stimulation of the right ear (Figs 2-E and F). Initial MRIs including diffusion-weighted images 4 hours after symptom onset were normal, but follow-up diffusion-weighted MRIs 4 days later disclosed a small acute infarction restricted to the right medial vestibular nucleus (Fig. 2-G).

2.3. Case 3: Chronic unilateral combined vestibulopathy with hearing loss: Intra-axial brainstem tumor

A 52-year-old man with progressive unsteadiness and rightward tilting sensation for 3 months developed right hearing loss 7 days before the presentation. He had undergone a craniotomy and tumor resection due to an anaplastic astrocytoma involving the left frontal cortex 3 years earlier. After the surgery, he had been healthy without any medical problems. Examination showed spontaneous horizontal-torsional nystagmus beating to the left. The nystagmus increased with prevention of visual fixation, during leftward gaze and after horizontal head-shaking, and changed into right beating during rightward gaze. Bedside HITs were positive for all three canals on the right side. He also showed rightward head tilt, ocular torsion (Fig. 3-A), and tilt of the subjective visual vertical, but no skew deviation. Bithermal caloric tests documented right canal paresis (Fig. 3-B) and audiometry showed a mild right hearing loss (Fig. 3-C). MRIs disclosed an ill-defined

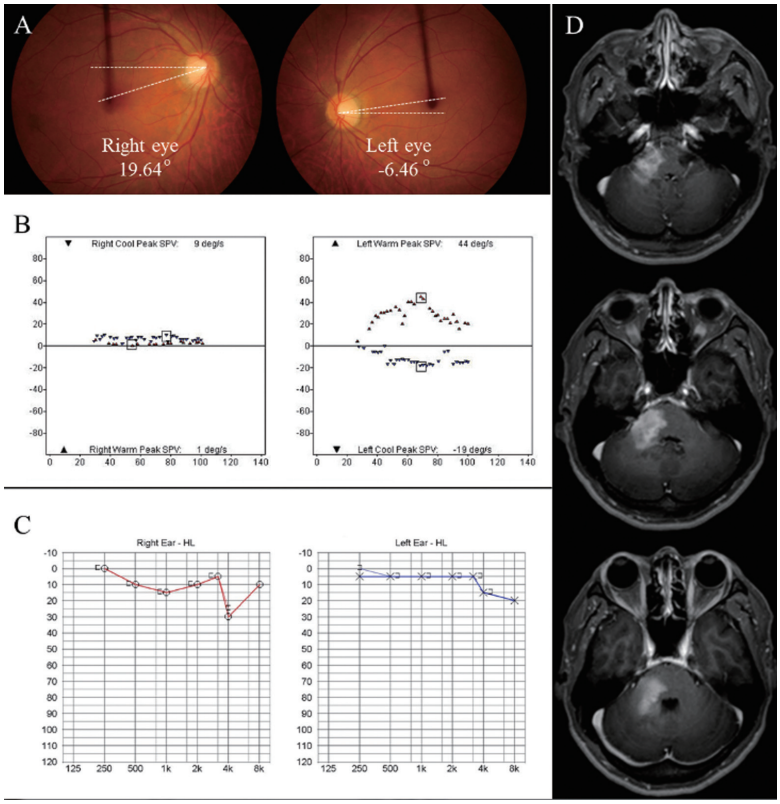

Fig. 3. Intra-axial brainstem tumor presenting with chronic unilateral combined vestibulopathy with hearing loss. (A) Fundus photographs showed rightward ocular torion (Right eye: 19.6° extorsion, Left eye: 6.5° intorsion). (B) Bithermal caloric tests showed 77% right caloricparesis. (C) Pure tone audiometry shows a mild sensorineural hearing loss in the right side with a pure tone average at 11.7 dB. (D) Gadolinum-enhanced T1-weighted MRIs show an ill-defined but well-enhanced tumor involving the right vestibular nucleus, middle cerebellar peduncle and vestibulocochlear nerve root-entry zone. (Colours are visible in the online version of the article; http://dx.doi.org/10.3233/VES-140524)

but well-enhanced intra-axial lesion in the area of right cerebellopontine angle (Fig. 3-D).

2.4. *Case 4: Acute bilateral combined vestibulopathy: Wernicke encephalopathy*

A 63-year-old woman developed dizziness and vomiting, and subsequently psychomotor slowing, apathy and ataxia two weeks after initiation of total parenteral nutrition due to Whipple's operation of the ampulla of Vater cancer. On examination, she was confused and unable to stand unaided due to severe truncal ataxia with limb dysmetria. Spontaneous nystagmus was absent in light and darkness in the straight (primary) ahead gaze, but gaze-evoked nystagmus (GEN) was evident in gaze to the left and right. Horizontal saccades were slow and delayed. Hearing was normal. She showed no responses to bithermal caloric irrigation of each ear (Fig. 4-A). Recording of the HITs using a magnetic search coil technique showed reduced gains of the VOR for both HCs, while the gains for all vertical semicircular canals were normal (Fig. 4-B). T2-weighted MRIs showed hyperintense lesions in the periaqueductal gray matter, medial thalami, and dorsal medulla, which were consistent with Wernicke encephalopathy (WE, Fig. 4-C). Three days after thiamine supplementation (100 mg intravenous daily), the abnormal neurological signs began to be resolved.

2.5. *Case 5: Chronic bilateral combined vestibulopathy without hearing loss: Cerebellar ataxia, neuropathy, and vestibular areflexia syndrome (CANVAS)*

An 81-year-old woman with hypertension presented with progressive imbalance for two years. She denied nausea, vomiting, tinnitus, hearing loss, dysphagia, or dysarthria. On examination, she showed spontaneous downbeat nystagmus and GEN (Fig. 5-A). Horizontal head shaking and straight head hanging in-

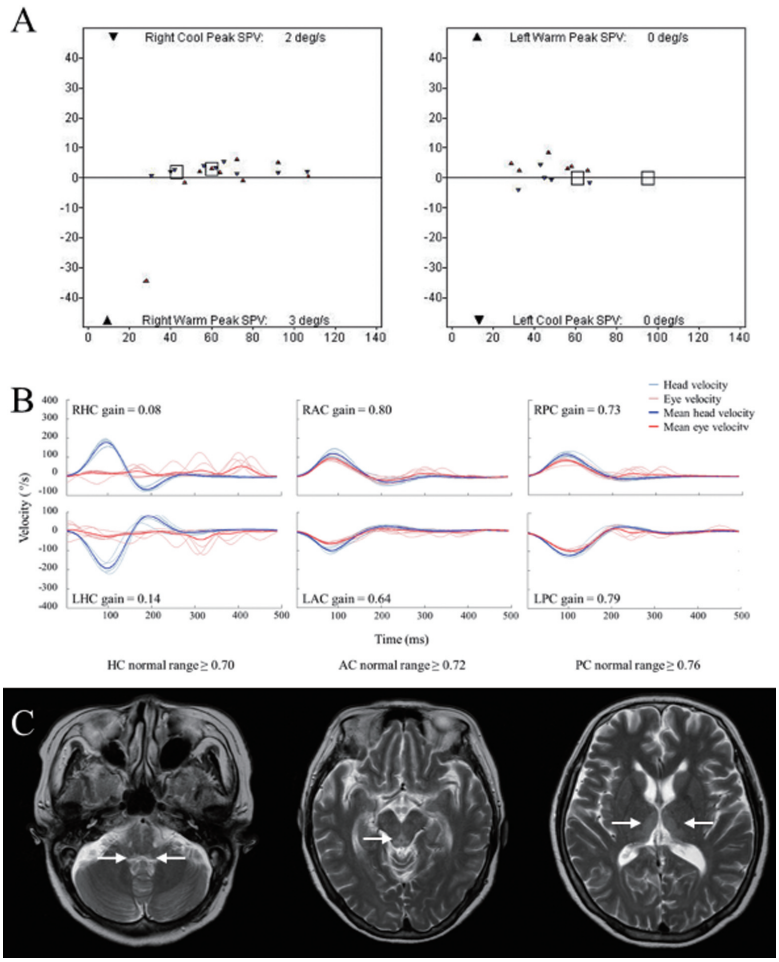

Fig. 4. Acute bilateral combined vestibulopathy: Wernicke encephalopathy. (A) Bithermal caloric tests showed almost no responses on each side. (B) Recording of head impulse tests shows reduced VOR gains for both horizontal canals, but normal gains for the vertical canals in each ear. (C) T2-weighted MRIs revealed high signal lesions in the bilateral periaqueductal gray matter, medial thalami, and dorsal medulla (arrows). AC: anterior canal, HC: horizontal canal, LHC: left horizontal canal, LAC: left anterior canal, LPC: left posterior canal, PC: posterior canal, RAC: right anterior canal, RHC: right horizontal canal, RPC: right posterior canal. (Colours are visible in the online version of the article; http://dx.doi.org/10.3233/VES-140524)

creased the downbeat nystagmus. Apogeotropic nystagmus was induced during head turning to either side while supine. Smooth pursuit and visual enhancement of the VOR were impaired in both horizontal and vertical directions. Bedside HITs were positive for both HCs. Recording of the HITs using a magnetic search coil technique documented reduced gains of the VOR for all semicircular canals (Fig. 5-B). Responses to bithermal caloric irrigation were mildly decreased on both sides, but were within normal range (sum = 43°/s). Rotatory chair test showed decreased gains and increased phase leads of the VOR during sinusoidal harmonic accelerations, and decreased time constants during step velocity rotation. Nerve conduction studies documented a mild sensorimotor polyneuropathy in all four extremities. Genetic analyses for spinocerebellar ataxia type 1, 2, 3, 6, and 7 were all negative. An MRI scan of her brain disclosed mild cerebellar atrophy (Fig. 5-C).

2.6. *Case 6: Chronic bilateral combined vestibulopathy with hearing loss: Hemosiderosis*

A 42-year-old woman had suffered from slowly progressive unsteadiness for seven years and hearing loss for four years. She had been diagnosed with cervical spinal chordoma 16 years earlier and had undergone several operations for tumor resection over the following three years. Since then, she had been left with mild

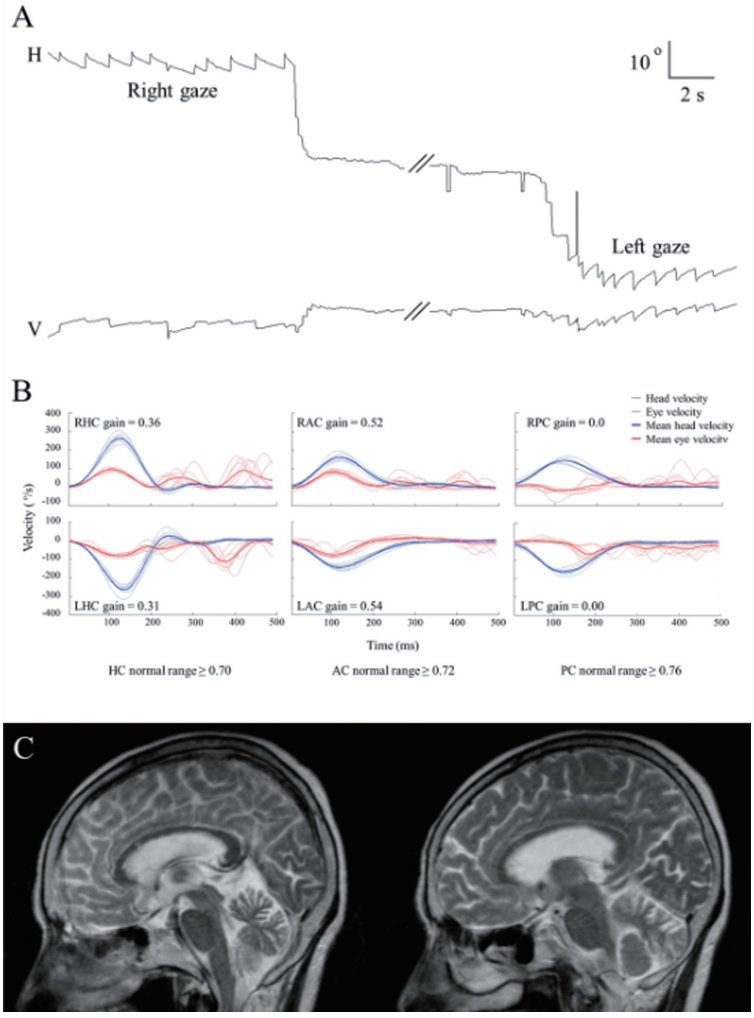

Fig. 5. Chronic bilateral combined vestibulopathy without hearing loss: Cerebellar ataxia, neuropathy, and vestibular areflexia syndrome (CANVAS). (A) Video-oculography (SMI®, Teltow, Germany) records horizontal gaze-evoked nystagmus with downbeat nystagmus, especially, during leftward gaze. (B) The VOR gains were reduced for all six semicircular canals during the recording of head impulse tests using a magnetic search coil technique. (C) T2-weighted sagittal MRI shows mild cerebellar atrophy. AC: anterior canal, H: horizontal eye position, HC: horizontal canal, LHC: left horizontal canal, LAC: left anterior canal, LPC: left posterior canal, PC: posterior canal, RAC: right anterior canal, RHC: right horizontal canal, RPC: right posterior canal, V: vertical eye position. (Colours are visible in the online version of the article; http://dx.doi.org/10.3233/VES-140524)

quadriparesis and hyperaesthesia. Seven years before the examination, however, she began to have truncal and limb ataxia that got worsened and finally made her wheelchair bound one year before. Neurological examination showed downbeat and GEN during lateral gazes, giving the appearance of side-pocket nystagmus (Fig. 6-A). Bedside HITs were positive for both HCs. She also showed bilateral caloric paresis (Fig. 6-B) and severe hearing loss (Fig. 6-C). An MRI head scan showed superficial hemosiderosis involving the whole cerebellum, brainstem, and fourth ventricle (Fig. 6-D and E).

3. Discussion

We described six cases of combined central and peripheral vestibulopathy according to their patterns of presentation.

3.1. Acute unilateral combined vestibulopathy

Since the AICA supplies both the peripheral labyrinth and central vestibular structures [1,2], the vestibular symptoms and signs in an AICA infarction may result from both peripheral and central vestibular le-

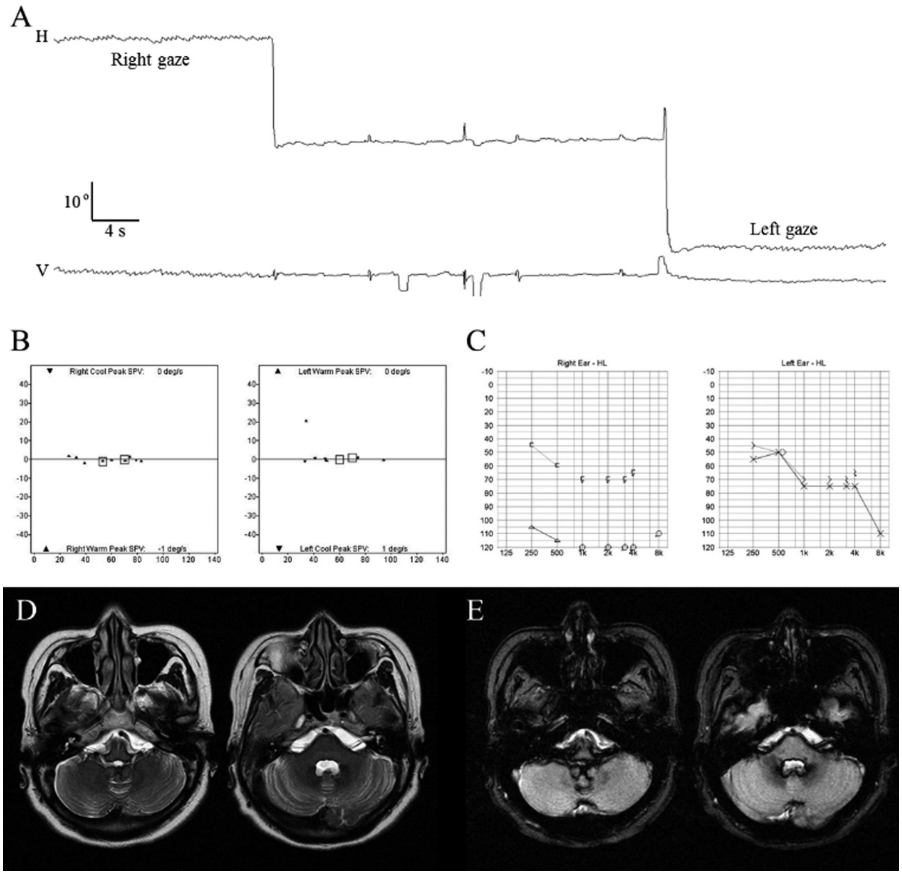

Fig. 6. Chronic bilateral combined vestibulopathy with hearing loss: Hemosiderosis. (A) Video-oculography (SMI®, Teltow, Germany) shows gaze-evoked and downbeat nystagmus on leftward and rightward gaze (B) Bithermal caloric tests show severe hypofunction on both sides. (C) Pure tone audiometry revealed severe bilateral sensorineural hearing loss, especially on the right side. (D) T2-weighted MRIs show superficial hemosiderosis in the lining of the fourth ventricle, brainstem and both vestibulocochlear nerves. (E) Gradient-echo MRIs also show dark signals lining the same structures. H: horizontal eye position, V: vertical eye position.

sions [24]. Diagnosis of an AICA infarction remains a challenge, especially when signs and symptoms other than those from an inner ear infarction are absent or inconspicuous [19,23]. Recognition of an AICA infarction as an occasional cause of an isolated audiovestibular loss is important because it requires evaluation of the cerebral vasculature and stroke risk factors [6,20]. Furthermore, patients with an AICA infarction should receive proper treatments including acute managements and lifetime prophylaxis for stroke [6]. In acute unilateral vestibulopathy, the HINTS is known to be useful in diagnosing a central vestibulopathy [28]. However, since the HIT is mostly positive in the combined vestibulopathy, the HINTS may not be sufficiently robust to detect central lesions in patients with combined central and peripheral vestibulopathy like our first patient. Our findings are supported by a previous study which reported HINTS failed to detect central lesions in 5 of 18 patients with AICA infarction [14]. Therefore, detection of central lesions may require additional tests such as horizontal head shaking nystagmus (HSN) that has detected a central lesion in three of five patients who had had an AICA infarct and a negative HINTS [14].

The bedside (positive HIT) and laboratory (canal paresis) findings of peripheral vestibulopathy in combined vestibular disorders should be differentiated from those observed in lesions involving of the flocculi and vestibular nuclei [17,29]. Since the medial region of the vestibular nucleus is the immediate recipient of peripheral vestibular projections, and is involved in the central processing of these vestibular signals; hence lesions involving the vestibular nuclei may cause features of both central and peripheral vestibular symptoms [17]. In clinical practice, the vestibular nuclei are most frequently involved in lateral medullary infarc-

tion (Wallenberg syndrome), and patients with Wallenberg syndrome may occasionally show features of peripheral vestibular dysfunction [8]. However, most patients with Wallenberg syndrome seldom poses any diagnostic difficulty due to associated neurological findings [18]. In contrast, patients with an isolated vestibular nuclear infarct may mimic vestibular neuritis, but they mostly have the findings of both peripheral (positive HIT or canal paresis) and central (GEN) vestibular dysfunction [17]. An isolated unilateral floccular infarct may also mimic a combined peripheral and central vestibulopathy by showing discrepancy in the abnormality of VOR gains between low and high frequency stimuli using HIT, caloric test, and rotatory chair test [29].

3.2. Chronic unilateral combined vestibulopathy

CPA tumors should be the prime suspicion in patients with chronic unilateral combined vestibulopathy. Tumors involving the CPA may be classified by their origin into intra- and extra-axial ones [4,5]. Although extra-axial vestibular schwannoma represents the majority of CPA tumors, other intra-axial tumors involving the lateral pons in the area of the vestibular root entry zone and vestibular nuclei, as in our patient, may cause combined vestibulopathy [10,15]. In a previous study, vestibular manifestation of CPA tumors included unilateral caloric paresis (87%), contralesional spontaneous nystagmus (27%), and gaze-evoked (12%) and Bruns nystagmus (14%) [15]. Bruns nystagmus is a bilateral nystagmus seen most often in CPA tumors and around 11% of vestibular schwanomma cases and mainly in those with larger tumors [26]. Bruns nystagmus is caused by gaze paretic nystagmus when looking towards the side of the lesion and vestibular nystagmus when looking away from the side of the lesion [25].

3.3. Acute bilateral combined vestibulopathy

In our experience, WE was most common in patients with acute bilateral combined vestibulopathy. WE is characterized by a triad of confusion, ataxia, and ophthalmoplegia due to deficiency of thiamine [30]. In WE, the vestibular nucleus and nucleus prepositus hypoglossi are frequently involved, and neurotological findings include GEN, central positional nystagmus, ophthalmoplegia, and caloric paresis [7,11,21]. A previous study documented bilateral caloric paresis and decreased gain of the horizontal VOR during HIT in patients with WE [7]. Unilateral infarction of the vestibular nucleus or flocculus may also mimic an acute bilateral combined vestibulopathy by producing a positive HIT in both horizontal canals [17,29]. In contrast, in isolated infarction in the vestibular nucleus or flocculus, caloric responses may be normal or decreased on one side only [17,29].

3.4. Chronic bilateral combined vestibulopathy

When hearing impairment was associated, superficial hemosiderosis was the most common disorder in chronic bilateral combined vestibulopathy, whereas CANVAS was most common when hearing was preserved. Superficial hemosiderosis is characterized by slowly progressive neurological deficits due to diffuse deposition of hemosiderin into the subpial space lining the cerebral cortices and ventricles, brainstem, cerebellar convolutions and cranial nerve roots due to chronic or repeated subarachnoid bleeding [32]. The vestibulocochelar nerve is the most vulnerable of all the cranial nerves because of its longer glial segment for hemosiderin deposition [22]. T2-weighted or gradient echo MRIs may show typical superficial siderosis in the brain or spinal cord, as in our patient. The cerebellum, vestibulocochlear nerve, and the vestibular nuclei may be affected by neural degeneration, giving rise to chronic progressive bilateral combined vestibulopathy.

Bilateral vestibulopathy may be associated with various causes of progressive cerebellar ataxia including autoimmune, nutritional (vitamin B 12, folate, etc.), familial (spinocerebellar ataxia, episodic ataxia type 2, etc.), or idiopathic origin [9]. When it is idiopathic, cerebellar ataxia with bilateral vestibulopathy (CABV) [27], or CANVAS, when peripheral neuropathy is associated, is termed [31]. A large number of patients with idiopathic bilateral vestibulopathy have associated cerebellar dysfunction and peripheral polyneuropathy [33].

4. Conclusion

AICA infarction is the most common cause of acute unilateral combined vestibulopathy while CPA tumors should be a prime suspicion in chronic lesion. In acute bilateral cases, WE should be considered. Hemosiderosis and CANVAS are most common in chronic bilateral combined vestibulopathy. Since the diagnosis of central vestibulopathy is crucial for treatments and prognosis of combined peripheral and central vestibular disorders, central signs should be sought through a

comprehensive bedside neurotological evaluation, including those patients with obvious clinical or laboratory features of peripheral vestibulopathy. The HINTS may not be enough to detect central lesions in patients with combined vestibulopathy. Despite the fact, vestibular nucleus or floccular lesions may mimic combined vestibulopathy, dissociation of the VOR abnormalities according to the stimulation frequency is the differentiating factor.

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