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Various symptoms and signs of vestibular paroxysmia in a tertiary neurotologic clinic: a retrospective comparative study

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Objectives: Vestibular paroxysmia (VP) is characterized by brief episodes of vertigo due to neurovascular cross-compression (NVCC) of the eighth cranial nerve. This study aimed to analyze the clinical features of VP patients in a tertiary neurotologic clinic using the 2008 and 2016 diagnostic criteria and to compare these features.

Methods: A retrospective review was conducted on patients diagnosed with definite or probable VP at the Asan Medical Center from May 2012 to May 2013. Patients underwent comprehensive evaluations including history taking, physical examination, audiometry, vestibular function tests, and magnetic resonance imaging (MRI). The 2008 and 2016 diagnostic criteria for VP were applied, and clinical characteristics were compared.

Results: Nineteen patients were included (14 females and five males; mean age, 57.9±14.5 years). According to the 2008 criteria, 17 patients were diagnosed with definite VP and two with probable VP; however, using the 2016 criteria, nine were definite and two were probable VP. NVCC was observed in 88.9% of patients under the 2016 criteria. MRI revealed NVCC predominantly involving the anterior inferior cerebellar artery. Patients with cerebellopontine angle (CPA) tumors presenting with VP-like symptoms responded to medication.

Conclusions: The 2016 diagnostic criteria for VP allow for a syndromic diagnosis based solely on clinical features. Neurovascular contact is commonly observed in VP patients, and CPA tumor can present VP-like symptoms and respond to carbamazepine. Although MRI is not included in the current criteria, it can be beneficial in diagnosis by identifying neurovascular contact and distinguishing CPA tumors.

Keywords: Vertigo; Vestibulocochlear nerve; Nerve compression syndrome; Carbamazepine; Prevalence; Magnetic resonance imaging

INTRODUCTION

Neurovascular cross-compression (NVCC) is when nearby blood vessels exert direct contact and compression on cranial nerves. Trigeminal neuralgia, hemifacial spasm, episodic vertigo, and glossopharyngeal neuralgia can result from neurovascular contact at the transition zones of the fifth, seventh, eighth, and ninth cranial nerves [1]. It is presumed that demyelination of axons and subsequent ephaptic transmission of action potentials are the causes of symptom onset [2]. Jannetta et al. [3] first described that NVCC affects the eighth cranial nerve, later termed "disabling positional vertigo". In 1986, Møller et al. [4] reported that 16 out of 21 patients who underwent microvascular decompression surgery showed improvement.

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In 1994, an attempt to establish diagnostic criteria for eighth nerve NVCC was made by Brandt and Dieterich [5], and it was termed vestibular paroxysmia (VP). The proposed diagnostic criteria were: (1) short attacks of spinning or non-spinning vertigo lasting from seconds to minutes; (2) attacks frequently dependent on a particular head position; (3) hyperacusis or tinnitus permanently or during the attack; and (4) auditory or vestibular deficits measurable by neurophysiological methods. For a diagnosis, three out of four criteria had to be fulfilled, and the patient had to respond to treatment with carbamazepine.

In 2008, Hüfner et al. [2] developed and subdivided criteria, including definite and probable VP (Fig. 1).

Recently, in 2016, the classification committee of the Bárány Society clarified the diagnostic criteria for VP after years of thorough discussion, presentation, and refinement [6]. These updates have made clinical diagnosis much easier and simpler (Fig. 2).

We aimed to analyze the various clinical features of VP patients experienced in a tertiary hospital using the 2008 criteria and the recent diagnostic criteria. Additionally, we report on the clinical features of patients with cerebellopontine angle (CPA) tumors who complained of symptoms similar to those of VP patients.

METHODS

Ethics Statement

The study protocol was approved by the Institutional Review Board (IRB) of Asan Medical Center (No. 2024-1082). Informed consent was waived by the IRB.

Definite vestibular paroxysmia
A. ≥10 attacks of spinning or non-spinning vertigo B. Duration <1 minute C. Stereotyped phenomenology[a] in a particular patient D. Response to a treatment with carbamazepine/oxcarbazepine E. Not better accounted for by another diagnosis
Probable vestibular paroxysmia
A. ≥5 attacks of spinning or non-spinning vertigo B. Duration <5 minutes C. Spontaneous occurrence or provoked by certain head-movements D. Stereotyped phenomenology[a] in a particular patient E. Not better accounted for by another diagnosis

Fig. 2. Diagnostic criteria for vestibular paroxysmia in 2016. [a]Stereotyped phenomenology: auditory symptoms (unilateral tinnitus or hyperacusis during the attacks), a horizontal+torsional nystagmus toward the affected ear.

Definite vestibular paroxysmia
At least five attacks and the patient fulfills the following criteria A–E:
A. Vertigo spells lasting seconds to minutes. The individual attack subsides without specific therapeutic intervention
B. One or several of the following provoking factors of the attacks: 1. Rest 2. Certain head/body positions (not BPPV-specific positioning maneuvers) 3. Changes in head/body position (not BPPV-specific positioning maneuvers)
C. One or several of the following characteristics during the attacks: 1. No accompanying symptoms 2. Disturbance of stance 3. Disturbance of gait 4. Unilateral tinnitus 5. Unilateral pressure/numbness in or around the ear 6. Unilaterally reduced hearing
D. One or several of the following additional diagnostic criteria: 1. NVCC demonstrated on MRI (CISS sequence) 2. Hyperventilation-induced nystagmus as measured by ENG 3. Increase of vestibular deficit at follow-up investigations as measured by ENG 4. Treatment response to antiepileptics (not applicable at first consultation)
E. The symptoms cannot be explained by another disease
Probable vestibular paroxysmia
At least five attacks and the patient fulfills criterion A, and at least three of the criteria B–E

Fig. 1. Diagnostic criteria for vestibular paroxysmia in 2008.
BPPV, benign paroxysmal positional vertigo; MRI, magnetic resonance imaging; CISS, constructive interference in steady state; NVCC, neurovascular cross-compression; ENG, electronystagmography.

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Patients and Study Design

We retrospectively studied patients with definite or probable VP who were referred to the tertiary dizziness clinic at Asan Medical Center from May 2012 to May 2013, according to the 2008 criteria. The patients underwent history taking, physical examination, otoscopy, pure-tone audiometry (PTA), videonystagmography, caloric test, cervical vestibular evoked myogenic potential (cVEMP), and MRI.

Subsequently, we applied the latest VP diagnostic criteria of the Bárány Society to all the above VP patients based on previous diagnostic criteria and compared the characteristics.

Audiometry

Hearing thresholds were determined by PTA. The 1 kHz weighted pure-tone average was calculated by adding the threshold decibels at 500, 1,000 (double-weighted), and 2,000 Hz, then dividing by four.

Vestibular Function Test

For the subjective visual vertical (SVV) test, participants sat upright in a stabilized position with their heads supported by a neck rest. A computer monitor, positioned 100 cm away from the participants, displayed a faint white line against a dark background. The line, measuring 3 mm in width and 230 mm in length, was accompanied by a 3-mm diameter dot, centrally aligned with the line's axis of rotation. Participants were instructed to focus on the dot and adjust the visual rod to the perceived vertical alignment by using a handheld remote control. The rod initially appeared tilted at random angles, ranging from 10° to 40° clockwise or counterclockwise. Once participants identified the vertical position, they pressed a confirmation button, and the deviation from the true gravitational vertical was automatically recorded. The average of five measurements was calculated to determine the SVV tilt. Deviations greater than ±2.78° from the vertical axis were classified as abnormal [7].

The bithermal caloric test was used in our study patients and eye movements were recorded using a video-based system (ICS water caloric stimulator NCI-480; Otometrics). Constant flow of water at temperatures of 30 °C and 44 °C was irrigated into each ear for 30 seconds. In each irrigation of the ear and temperature, the maximum slow-phase eye velocities of the nystagmus were calculated. Canal paresis was defined as an asymmetry between the right-sided and left-sided responses of 20% or more, using the formula of Jongkees et al. [8].

cVEMPs were measured while participants remained seated, with their heads rotating away from the side receiving stimulation. Electrodes were positioned as follows: the active electrode was attached to the middle portion of the sternocleidomastoid muscle, the reference electrode was placed on the upper sternum, and the ground electrode was fixed on the forehead. VEMP responses were elicited using 500-Hz Blackman-modulated tone bursts, characterized by a 2-ms rise and fall time with a 1-ms plateau, delivered at a rate of nine stimuli per second via insert earphones. The stimulus intensity was set at 90-dB normal hearing level, and the resulting electromyographic signals were amplified and filtered using a bandpass range of 30–1,500 Hz with the GSI Audera system (Grason-Stadler). Peak-to-peak amplitudes of N1 and P1 waves were analyzed at maximum stimulation intensity. A VEMP result was classified as abnormal if the interaural amplitude difference ratio exceeded 40%, the interaural threshold difference was greater than 15 dB, or if no VEMP response was detected [9].

Computerized dynamic posturography (CDP) was utilized to evaluate postural stability, and sensory organization test (SOT) outcomes were analyzed using the Equitest System (NeuroCom International). The SOT consisted of six distinct conditions designed to assess the integration of visual, vestibular, and proprioceptive inputs for maintaining posture. These conditions were: (1) eyes open with stable surroundings and platform (SOT1); (2) eyes closed with a fixed platform (SOT2); (3) sway-referenced surroundings with a fixed platform (SOT3); (4) eyes open with fixed surroundings and a sway-referenced platform (SOT4); (5) eyes closed with a sway-referenced platform (SOT5); and (6) sway-referenced surroundings and platform (SOT6). Each condition was tested in three trials, resulting in a total of 18 trials.

Postural sway was quantified by measuring anterior-to-posterior center of gravity displacement, with equilibrium scores calculated as the percentage difference between observed sway and theoretical stability

limits (approximately 12.5°). Equilibrium scores ranged from 0% (indicating a fall) to 100% (indicating no sway). A composite score was derived by averaging equilibrium scores for SOT1 and SOT2 and combining these with scores from SOT3 through SOT6. Scores falling below the age-specific normative range provided by the CDP manufacturer were considered abnormal [10]. Additionally, an equilibrium score greater than or equal to the 95% confidence interval of the normative data was used to identify abnormalities in specific conditions.

Magnetic Resonance Imaging

MRI scans were acquired using standard clinical protocols on 3.0-tesla clinical scanners in 16 patients and 1.5-tesla in one patient. The patients have taken one of two types of MRI, which were the brain MRI and internal auditory canal (IAC) MRI. The brain MRI included T1- and proton-density/T2-weighted axial images with a slice thickness of 3–5 mm covering the whole brain. The IAC MRI included a constructive interference in steady state (CISS) sequence covering the brainstem and cerebellum, with a slice thickness of 0.6–1.2 mm. The CISS MRI is a heavily T2-weighted sequence and provides a strong contrast between the low-intensity nerves and the surrounding high-intensity cerebrospinal fluid (CSF). Neurovascular contact was defined and evaluated with contact being defined as a lack of CSF signal cleft between the nerve and implicated vessel [11]. Nerve angulation was defined as a change in direction or caliber of the Vestibulocochlear nerve at the point of neurovascular contact, as described by Sivarasan et al. [12].

RESULTS

Patient Population and Laboratory Examinations

Nineteen patients with VP were included: 14 females (73.7%) and five males (26.3%). Age ranged from 26 to 78 years, with a mean of 57.9±14.5 years. Duration of visits ranged from 0 to 120 months, with a mean of 40.8±39.1 months (Table 1).

Symptoms and Diagnosis of Vestibular Paroxysmia

All the patients experienced more than 10 episodes of attacks, with durations from a few seconds to minutes. Accompanying symptoms varied: auditory symptoms were observed in 17 patients (89.5%)—13 with tinnitus alone, three with ipsilateral hearing loss (2 of whom also had concurrent tinnitus), and one with hyperacusis accompanied by tinnitus. Vestibular symptoms were present in four patients (21.1%), and facial spasm was noted in five patients (26.3%). Thirteen patients (68.4%) experienced vertigo attacks provoked by certain head-movements. The clinically suspected side was determined based on unilateral symptoms or more prominent features on one side (Table 1).

According to the 2008 diagnostic criteria, 17 patients were classified as definite VP and two as probable VP. According to the 2016 criteria, nine patients were diagnosed with definite VP and two with probable VP. Notably, Patients 8 and 9, while initially classified as definite VP under both criteria based on their clinical presentations, were later reclassified as having CPA tumors following MRI findings (Table 1).

Results of Audio-Vestibular Tests

Patients 4, 6, 8, 9, and 19 had either unilateral hearing loss or a difference of more than 10 dB in PTA between ears. The caloric test was performed on 13 patients (mean, 21.9±27.4%) and showed horizontal canal dysfunction in three patients (23.1%), two of them had IAC mass. The cVEMP test was performed on 17 patients (mean, 22.8±16.7%) and four patients showed otolith dysfunction (23.5%); one had an IAC mass on the dysfunction side. Binocular SVV measurements were performed on 16 patients, with normal results in the majority (81.3%); slight deviations were observed in 18.8% (mean, 1.74±1.11°). Among three pa tients with abnormal SVV results, two with IAC masses had deviations matching the side of the mass. SOT results were obtained from 14 patients (mean composite score, 63.6%±16.7%), three patients had abnormal results. Patient 9 showed a particularly low score indicative of a vestibular dysfunction pattern (Table 1).

Magnetic Resonance Imaging Findings

Seventeen patients underwent MRI scans; five had brain MRI, and 12 took IAC MRI with CISS sequence (Fig. 2). NVCC was found in 15 patients, seven had NVCC on both sides, six had unilateral NVCC, and two had CPA tumors. Patient 8 had a left-sided 1.8-cm pe-

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Table 1. Clinical characteristics and results of audio-vestibular tests of patients, along with the application of both the 2008 and 2016 diagnostic criteria for vestibular paroxysmia

Patient No.	Sex	Age (yr)	Duration of visit (mo)	Diagnostic criteria		PTA		Dizziness		Medication		Lateralizing features	Clinically suspected side	SVV	Caloric test	cVEMP	SOT
				2008	2016	Right	Left	Attacks	Duration	Dose (mg)	Improvement						
1	F	53	36.5	Definite	Definite	13	15	2[a]	2	0x 400	Yes	Left tinnitus	Left	−2.38	5.7	26	-
2	F	74	120.5	Definite	Definite	20	18	2[a]	2	C 400	Yes	Left tinnitus, Right otolith dysfunction	Left	−1.43	5	−48	70
3	F	69	9.0	Definite	Definite	26	25	2[a]	2	0x 300	Yes	Both tinnitus	-[b]	−0.38	−15	22	76
4	F	78	10.3	Definite	Definite	56	30	2[a]	2	C 400	Yes	Right tinnitus, canal paresis, hearing loss, facial spasm	Right	2.24	−43	2	50
5	F	65	70.5	Definite	Definite	10	11	2[a]	2	C 600	Yes	Left otolith dysfunction, facial spasm	Left	2.58	-	57.2	-
6	F	61	16.9	Definite	Definite	6	23	2[a]	2	0x 600	Yes	Left hearing loss	Left	–	-	17	74
7	F	70	12.0	Definite	Definite	10	15	2	2	C 400	Yes	Right tinnitus, otolith dysfunction	Right	–	-	−55	-
8	M	71	32.8	Definite	Definite	10	23	2[a]	2	0x 900	Yes	Left tinnitus[c]	Left	−3.64	68	-	70
9	F	50	90.9	Definite	Definite	26	5	2	2	0x 900	Yes	Right tinnitus[c]	Right	3.80	−92	−53.4	42
10	F	56	2.1	Definite	Probable	33	41	2[a]	1	0x 900	No	Both tinnitus	-[b]	0.94	10.2	-	18
11	F	62	99.8	Definite		13	11	2[a]	0	C 600	Yes	Left tinnitus	Left	1.59	−18	−9	66
12	M	28	31.8	Definite		11	13	2	0	0x 300	Yes	Left tinnitus, facial spasm	Left	0.44	−3	25.5	-
13	F	51	83.4	Definite		10	8	2	0	C 600	Yes	Right tinnitus	Right	–	-	−12	88
14	M	69	1.8	Definite		15	20	2[a]	0	0x 900	Yes	Both tinnitus	-[b]	−0.68	−3.9	0	75
15	M	42	3.5	Definite		6	8	2[a]	0	C 600	Yes	Right tinnitus, Left hyperacusis	Right	1.29	−16.3	−1.2	68
16	F	26	23.5	Definite		1	5	2	0	0x 900	No	Both tinnitus	-[b]	0.73	−2.9	7.3	-
17	F	63	0	Definite[d]	Probable	20	18	2	2	0x 600	-	Left tinnitus, facial spasm	Left	3.24	−1.5	26	60
18	F	43	103.8	Probable		6	8	2[a]	0	0x 600	Yes	Right tinnitus	Right	0.44	-	−16	70
19	M	70	26.5	Probable		31	6	2[a]	0	0x 600	No	Right tinnitus, hearing loss, facial spasm	Right	−1.98	-	10.5	64

Attacks: 0, <5 attacks; 1, ≥5 attacks; 2, ≥10 attacks. Duration: 0, ≥5 minutes; 1, <5 minutes; 2, <1 minutes.

SVV, subjective visual vertical test; cVEMP, cervical vestibular evoked myogenic potentials; SOT, sensory organization test; M, male; F, female; C, carbamazepine; Ox, oxcarbazepine.

[a]Patients have dizziness attacks provoked by certain head movement. [b]The patients who didn't have a clinically suspected side either had tinnitus in both ears or couldn't determine which side the tinnitus was coming from. [c]Patients 8 and 9 had a tumor in the left and right cerebellopontine angle, respectively. [d]Patient 17 had neurovascular cross-compression on brain magnetic resonance imaging.

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trous meningioma, and patient nine had a right-sided 3-cm meningioma. Directional concordance of the NVCC with the clinically suspected side was found in 11 patients (78.6% among those with a clinically suspected side, 75.0% excluding patients with IAC mass). The anterior inferior cerebellar artery (AICA) was contacted with the vestibulocochlear nerve in 11 patients (13 sites), and the posterior inferior cerebellar artery in three patients; clinically suspected sides were all AICA. The mean distance from the brainstem to the point of contact in VP cases was 8.08±2.58 mm (12 patients, 16 sites). The mean distance with directional concordance to the clinically suspected side was 8.01±2.66 mm (nine patients). Nerve angulation was found in three patients (15.8%), with directional concordance in two patients (66.7%) (Table 2).

Comparative Analysis of Vestibular Paroxysmia according to the 2008 and 2016 Criteria

Compared to the 2008 criteria, the 2016 criteria for diagnosing VP became more specific and stringent to enhance reliability and repeatability. The most significant change is the introduction of a definite time standard. In our study, applying the 2016 criteria, eight patients were excluded because their episodes exceeded the required duration (>5 minutes). Additionally, two patients were reclassified as probable VP due to a lack of improvement after medication (Table 1).

The overall proportion of NVCC, excluding patients who did not undergo MRI, was 82.3% based on the 2008 criteria and 88.9% based on the 2016 criteria. The NVCC on the same side as the stereotyped phenomenon or hearing loss was observed in 78.6% of cases under the 2008 criteria and 87.5% under the 2016 criteria, excluding patients who did not have a clinically suspected side. The distance between the brainstem and the compressing vessels matching the clinically suspected side was 8.01±2.66 mm for the 2008 criteria and 7.54±2.62 mm for the 2016 criteria. Nerve angulation of the eighth cranial nerve was observed in three patients, but one

Table 2. MRI modalities used, the presence of neurovascular cross-compression or nerve angulation, and their directional concordance with clinical lateralizing features

Patient No.	Diagnostic criteria		Mode of MRI	Clinically suspected side	Neurovascular cross-compression	Directional concordance	Causing vessels & distance (mm)	Nerve angulation
	2008	2016						
1	Definite	Definite	IAC	Left	Both	○	Right PICA, 10.51; left AICA, 10.73	-
2	Definite	Definite	Brain	Left	-	×		-
3	Definite	Definite	-	-				
4	Definite	Definite	IAC	Right	Both	○	Right AICA, 6.87; left PICA, 10.05	Left
5	Definite	Definite	-	Left				
6	Definite	Definite	IAC	Left	Left	○	Left AICA, 3.0	-
7	Definite	Definite	IAC	Right	Right	○	Right AICA, 7.87	Right
8	Definite	Definite	IAC	Left	Left CPA mass	○		
9	Definite	Definite	IAC	Right	Right CPA mass	○		
10	Definite	Probable	IAC	-	Left		Left PICA, 4.37	-
11	Definite		Brain	Left	Right	×	Right AICA, 4.45	-
12	Definite		IAC	Left	Both	○	Right AICA, 8.99; left AICA, 10.85	-
13	Definite		IAC	Right	Both	○	Right AICA, 7.87; left AICA, 8.57	-
14	Definite		IAC	-	-			-
15	Definite		Brain	Right	Right	○	Right AICA, 11.02	-
16	Definite		IAC	-	Left		Left AICA, 10.21	-
17	Definite	Probable	Brain	Left	Left	○	Left AICA, 9.23	-
18	Probable		Brain	Right	-	×		-
19	Probable		IAC	Right	Right	○	Right AICA, 4.63	Right

MRI, magnetic resonance imaging; IAC, internal auditory canal; CPA, cerebellopontine angle; AICA, anterior inferior cerebellar artery; PICA, posterior inferior cerebellar artery.

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did not meet the 2016 diagnostic guidelines. The percentage of CPA tumors was 10.5% and 18.2% under the 2008 and 2016 criteria, respectively; all patients with CPA tumors showed improvement with oxcarbazepine treatment (Table 2).

In addition, there were four more patients who experienced facial twitching alone or facial twitching accompanied by tinnitus, without dizziness. One of them had a CPA tumor. Three patients showed directional concordance between the side of NVCC and the clinically suspected side. One patient exhibited facial nerve angulation, and all four patients had a response to drug therapy (Supplementary Table 1).

DISCUSSION

Diagnosis and Symptoms of Vestibular Paroxysmia

Transitioning from the 2008 to the 2016 diagnostic criteria for VP increased the specificity and stringency significantly. This shift resulted in seven patients no longer meeting the 2016 criteria due to prolonged episodes of dizziness (>5 minutes), highlighting the stricter temporal standards. Furthermore, about half of these patients exhibited very specific symptoms for VP, such as vertigo accompanied by tinnitus and facial spasms. Patients 8 and 9, while initially meeting all clinical diagnostic criteria for definite VP, were reclassified as CPA tumors after MRI findings that revealed CPA tumors presenting with VP-like symptoms. Additionally, two patients previously diagnosed with definite VP were reclassified as having probable VP under the 2016 criteria, despite confirmed NVCC on MRI scans.

Magnetic Resonance Imaging Findings of Vestibular Paroxysmia

To enhance practicality and applicability worldwide, MRI demonstration of NVCC was excluded from diagnostic criteria. T2-weighted CISS and fast imaging employing steady-state acquisition (FIESTA) MRI sequences are quite useful in identifying neurovascular relationships within the IAC and CPA. Sivarasan et al. [12] revealed that the predictive value of MRI (including CISS sequences) in identifying NVCC in VP patients compared with controls was significant (p=0.0049). In our study, most patients showed NVCC (82.3% and 88.9% based on 2008 and 2016 criteria, respectively). However, obtaining such MRI sequences is not feasible everywhere, and while sensitivity was reported as high (100%), specificity was shown to be relatively low in recent studies (65% and 55%, respectively) [11,12].

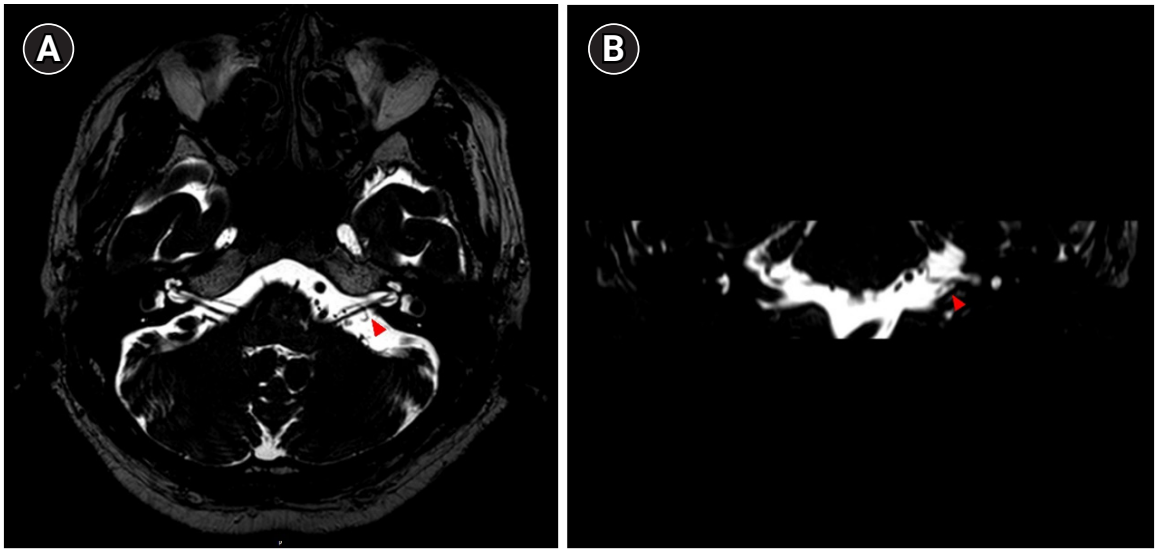

Fig. 3. (A) Axial constructive interference in steady state (CISS) sequence magnetic resonance imaging (MRI) of the internal auditory canal showing left-sided neurovascular cross-compression (arrowhead) of the vestibulocochlear nerve by the S-shaped posterior inferior cerebellar artery. (B) Coronal CISS sequence MRI of the internal auditory canal highlighting the left-sided neurovascular cross-compression (arrowhead) by the posterior inferior cerebellar artery.

Additionally, Sivarasan et al. [12] suggested that nerve angulation caused by neurovascular contact can be a highly specific imaging feature for diagnosing VP (sensitivity, 44% and specificity 100%, $p = 0.0053$). This concept was first applied in trigeminal neuralgia. Antonini et al. [13] noted that identifying NVCC on MRI for the diagnosis of trigeminal neuralgia has high sensitivity but low specificity. They suggested finding neurovascular contact with nerve displacement and atrophy at the point of contact represents a highly specific feature (100% specificity). Therefore, considering that the vestibulocochlear nerve is particularly susceptible to mechanical irritation, MRI has a more significant role in diagnosing VP. In our study, the sensitivity was 28.6% and the specificity was 85.7% based on the 2016 criteria (excluding patients with CPA tumors and those who did not undergo MRI).

Episodic vertigo accompanied by auditory symptoms is characteristic of VP but also indicative of pathological pressure on the vestibulocochlear nerve [2,5,14]. The transition zone, an anatomic area between central and peripheral myelin portions, was originally thought to be the most vulnerable portion to mechanical irritation in symptomatic NVCC of cranial nerves [15]. Furthermore, a vestibulocochlear nerve has a long and distally located transition zone compared to other cranial nerves, meaning it has a longer central myelin portion, the longest among cranial nerves [16-18]. The most remote part of the transition zone from the brainstem measured at 9.28 to 13.84 mm [19]. For this reason, it was natural to consider the vestibulocochlear nerve particularly vulnerable to mechanical stimulation. However, recent studies demonstrated that neurovascular contact occurs in the central myelin portion, not in the transition zone [11,12,17]. Also in this study, the mean distance from the brainstem to the neurovascular contact point was 8.01±2.66 mm, indicating that the contact occurred in the central myelin portion [14].

Vestibular Paroxysmia-Like Symptoms in Patients with Cerebellopontine Angle Tumor

Conversely, tumor-induced symptoms share a similar pathophysiology with NVCC due to nerve irritation, so the improvement of symptoms with carbamazepine or oxcarbazepine was been observed in patients with CPA tumors [20]. Additionally, the 2016 diagnostic criteria for VP mention the need to consider cases involving CPA tumors [21]. In our study, there were three patients with CPA tumors (one in Supplementary Table 1) who exhibited characteristics of VP or facial twitching, and all of them responded to the medication.

Limitations of This Study

There are some limitations in this study. First, it was quite difficult to standardize the MRI protocol and follow-up durations for all patients due to the retrospective nature of the study. Additionally, some patients had relatively short follow-up periods, affecting the diagnostic process.

Conclusions

The recent diagnostic criteria for VP no longer require objective test results; thus, the diagnosis of VP is now purely syndromic. Given the pathophysiology of NVCC, various specific clinical symptoms—frequent attacks of short-duration vertigo, typewriter tinnitus, and facial twitch—are indicative of VP. CPA tumors can present VP-like symptoms and can respond to carbamazepine. While not included in the formal diagnostic criteria, appropriate imaging studies such as MRI using CISS or FIESTA sequences are beneficial in detecting CPA tumors, ruling out other conditions, and assessing NVCC and nerve to aid in accurate diagnosis.

ARTICLE INFORMATION

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Conflicts of Interest

Hong Ju Park is an Associate Editor of *Research in Vestibular Science* and was not involved in the review process of this article. All authors have no other conflicts of interest to declare.

Availability of Data and Materials

All data generated or analyzed during this study are included in this published article. For other data, these may be requested through the corresponding author.

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Authors' Contributions

Conceptualization: TC, HJP; Data curation, Visualization: TC, EHC; Formal analysis: TC; Methodology: TC, YK; Project administration: TC, HJP; Writing–original draft: TC; Writing–review & editing: TC, HJP.
All authors read and approved the final manuscript.

Supplementary Materials

Supplementary materials can be found via https://doi.org/10.21790/rvs.2024.021.

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