

Vestibular Paroxysmia Associated with Paroxysmal Pulsatile Tinnitus: A Case Report and Review of the Literature

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Abstract-

Purpose: Vestibular paroxysmia is defined as paroxysmal, brief, and carbamazepine-responsive vertigo. Although neurovascular cross-compression (NVCC) of the vestibulocochlear nerve is believed to be the cause of vestibular paroxysmia, the mechanism remains controversial. Herein, we describe the case of a man with NVCC who presented with paroxysmal vertigo associated with paroxysmal pulsatile tinnitus.

Case Report: A 68-year-old man presented with paroxysmal vertigo for one month. Paroxysmal pulsatile tinnitus in the right ear occurred simultaneously with the vertigo. Magnetic resonance imaging demonstrated that the right anterior inferior cerebellar artery was compressing the right vestibulocochlear nerve. The vertigo and tinnitus completely disappeared within one week after treatment with carbamazepine.

Conclusion: The pulsatile nature of the patient's tinnitus implied that the auditory nerve was being compressed by a pulsating artery and was found to consolidate the causal relationship between NVCC and vestibular paroxysmia.

Key Words: vertigo, pulsatile tinnitus, vestibular paroxysmia, neurovascular compression

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INTRODUCTION

Vestibular paroxysmia is defined as paroxysmal and brief vertigo that is responsive to carbamazepine(1, 2). and although difficult to prove directly, it is thought to be caused by neurovascular cross-compression (NVCC) of the vestibulocochlear nerve and a corresponding offending vessel. However, this disorder is considered to be controversial by some researchers and clinicians.

Herein, we describe the case of a man with NVCC who presented with paroxysmal vertigo associated with paroxysmal pulsatile tinnitus. The pulsatile nature of the tinnitus supported the theory of neurovascular compression.

CASE REPORT

A 68-year-old man presented to our dizziness clinic

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with a 1-month history of paroxysmal vertigo. The vertigo occurred at a frequency of around 10 times per day, and each paroxysm lasted for a few seconds to 1 minute. Paroxysmal tinnitus in the right ear developed simultaneously with the vertigo. When the duration of these paroxysms increased, he noticed that the tinnitus was pulsatile and synchronized with his heartbeat. With an increase in the severity of vertigo, the pulsatile tinnitus became louder. These episodes were often induced with a change in position, but they also occurred at rest. Treatment with vestibular suppressants only partially improved the vertigo and tinnitus. In addition, the patient complained of mild unsteadiness during interictal periods.

Local otological findings were unremarkable. In neurological and neuro-otological examinations, only mild disequilibrium was noted in the tandem gait test, but he tested negative for the Romberg sign. There was no spontaneous or gaze-evoked nystagmus. The head impulse test yielded normal results, and the Dix-Hallpike test did not induce positional nystagmus. Other cranial nerve functions and coordination were normal.

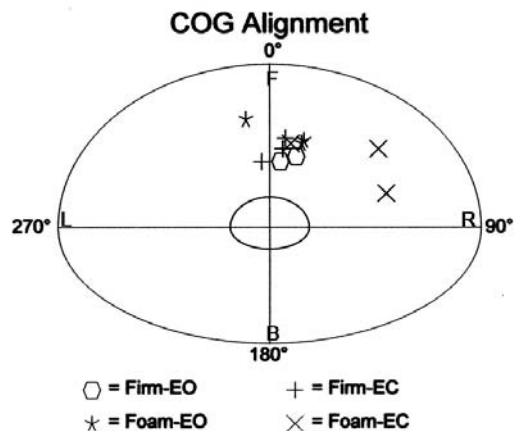


Figure 1. Computed posturography showed that the center of gravity of the patient had deviated anteriorly and rightward, especially in the foam-eye closed condition. (R: right side; L: left side; F: the front; B: the back; COG: center of gravity; Firm-EO: standing on a firm surface with eyes open; Firm-EC: standing on a firm surface with eyes closed; Foam-EO: standing on a foam surface with eyes open; Foam-EO: standing on a foam surface with eyes closed)

Pure-tone audiometry revealed bilateral symmetrical high-frequency sensorineural hearing loss. The caloric test recorded by video-nystagmography revealed 16% weakness in the right ear, but this was within the normal range as per our laboratory standards. Vestibular dysfunction was detected in the modified Clinical Test of Sensory Interaction on Balance (mCTSIB) by computed posturography. His center of gravity (COG) deviated anteriorly and rightward (Fig. 1).

Carotid duplex did not show atherosclerosis, and electroencephalography (EEG) did not show epileptiform discharges even during vertigo attacks. High-resolution computed tomography (HRCT) of the temporal bone yielded unremarkable results for pulsatile tinnitus. Brain magnetic resonance imaging (MRI) was performed for possible vertebrobasilar insufficiency, however, no structural lesions were found in the brainstem or cerebellum. Magnetic resonance angiography revealed no stenosis or apparent atherosclerosis in the vertebrobasilar arteries, however, steady-state MRI (fast imaging employing steady-state acquisition [FIESTA]) revealed that the right anterior inferior cerebellar artery (AICA) had apparently compressed and displaced the right vestibulocochlear nerve (Fig. 2).

On the basis of the imaging findings, the patient was diagnosed with vestibular paroxysmia, and carba-

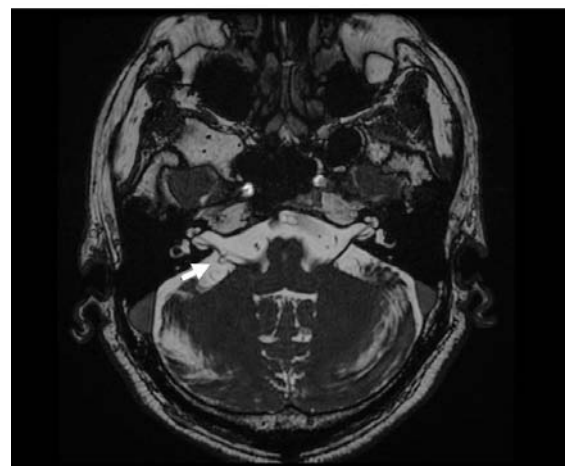


Figure 2. Steady-state (FIESTA) MRI revealed neurovascular cross-compression between the right AICA and right vestibulocochlear nerve (white arrow).

mazepine (200 mg daily) treatment was administered. His tandem gait had almost returned to normal the day after the treatment. A follow-up examination after 1 week revealed that he was almost completely free of the paroxysmal vertigo and pulsatile tinnitus and had only a mild floating sensation. We repeated posturography 2 weeks later and it showed normal results either in mCTISB or COG. However, he subsequently developed skin eruptions and we therefore replaced carbamazepine treatment with gabapentin (600 mg daily). The patient discontinued the treatment after he was free of vertigo for 2 months. However, vertigo reoccurred within 1 week of drug withdrawal, and gabapentin treatment was reinitiated which again relieved the vertigo for another one month.

DISCUSSION

The patient presented with brief and frequent paroxysms of vertigo and tinnitus, which were eliminated by carbamazepine. Before the diagnosis was established, many diseases were excluded. Benign positional paroxysmal vertigo and vertebrobasilar artery insufficiency, which are the most common causes of paroxysmal and brief vertigo, were excluded on the basis of the findings of the Dix-Hallpike test and magnetic resonance angiography. Vestibular migraine, Meniere's disease, and perilymphatic fistula were ruled out on the basis of the clinical history, audiometric findings, and his responsiveness to carbamazepine. Vestibular epilepsy, a rare central vestibular disorder which may be responsive to carbamazepine, was excluded because ictal EEG did not show epileptiform discharges. Etiologies of pulsatile tinnitus⁽³⁾ including dural arteriovenous fistulas, glomus tympanicum, carotid atherosclerosis, aberrant carotid artery, and high jugular bulb, were excluded on the basis of the local otological findings and results of carotid duplex, temporal bone HRCT, and brain MRI. In accordance with the pattern of the patient's vertigo, the associated tinnitus and the efficacy of carbamazepine, NVCC of the right vestibulocochlear nerve and AICA proven by MRI may have been the etiology in this patient.

NVCC of the vestibulocochlear nerve was first

reported in 1975 by Jannetta, who termed this condition as disabling positional vertigo^(4,5). Surgical decompression is considered to be an effective treatment. However, the presentation of vertigo has not been described specifically, and the clinical diagnosis of this condition is mainly established by excluding other well-known vestibulopathies such as benign positional paroxysmal vertigo, Meniere's disease, and vestibular neuritis.

Brandt et al. described a vertigo syndrome which they termed vestibular paroxysmia in 1994^(1,2). The diagnostic criteria proposed for vestibular paroxysmia are as follows: (1) short attacks of rotational to-and-fro vertigo lasting seconds to minutes; (2) attacks frequently dependent on particular head positions; (3) hypoacusis or tinnitus permanently or during the attack; (4) measurable auditory or vestibular deficits by neurophysiological methods; and (5) efficacy of carbamazepine. The clinical manifestations including the onset, duration, and frequency of this condition are similar to those of trigeminal neuralgia and hemifacial spasm. A high incidence of NVCC has been reported in MRI studies, and the effectiveness of carbamazepine also implies the presence of NVCC.

In a study of 32 patients with vestibular paroxysmia in 2008⁽⁶⁾, 69% of the patients were found to have rotatory vertigo and 25% stated that they had a to-and-fro sensation. In addition, 28% of the patients suffered from vertigo even at rest, whereas 22% experienced vertigo while performing certain actions. The most common accompanying symptoms were unsteady gait (75%), nausea/vomiting (41%), and tinnitus (28%). Twenty-three of 24 patients were found to have NVCC on MRI. After carbamazepine or oxcarbazepine treatment, the frequency and intensity both reduced significantly in most patients with vestibular paroxysmia. The authors of the above-mentioned study further stated that vestibular paroxysmia is not uncommon and occurred in 4% of all patients who presented with dizziness to their dizziness unit. On the basis of this, we believe that vestibular paroxysmia is probably misdiagnosed by most physicians in dizziness clinics.

In a study of 100 MRI scans from patients with no vertigo⁽⁷⁾, contact between the vessel and nerve was

observed in 12.5% of the MRI scans. When both the nerve and vessel were seen concurrently in a single MRI slice, the contact rate was found to be 50%. Therefore, a large number of NVCCs that are detected on MRI may be asymptomatic. NVCC detected on the MRI scans of patients with vestibular paroxysmia is possibly a coincidental finding, and further studies are required to establish the pathophysiology of NVCC in patients with paroxysmal vertigo.

Paroxysmal tinnitus has also been linked to vascular compression of the auditory nerve⁽⁸⁻¹⁰⁾. This type of tinnitus is often staccato and intermittent and is described as “ear-clicking tinnitus” or “typewriter tinnitus”, and is partly associated with hemifacial spasms. Ryu et al. provided convincing evidence of a relationship between tinnitus and NVCC⁽¹¹⁾. In their study, during decompression surgery for hemifacial spasms, 10 patients having tinnitus ipsilateral to the hemifacial spasm underwent neurovascular decompression of the eighth nerve, and tinnitus was relieved after the operation in 8 of the 10 patients. In addition, another study showed that some types of tinnitus that are cured by neurovascular decompression are low-pitched and pulsatile⁽¹²⁾. Carbamazepine can also suppress most types of tinnitus in vestibular paroxysmia.

Our patient presented with paroxysmal vertigo that was associated with paroxysmal pulsatile tinnitus. The paroxysmal characteristic was compatible with the features of nerve compression, and the pulsatile tinnitus further indicated that the auditory nerve was compressed by a pulsating artery. The unique clinical features in our patient may support the hypothesis that vestibular paroxysmia is related to NVCC and that their association is not merely a coincidence.

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