Journal of Neurology (2021) 268:2267–2272
https://doi.org/10.1007/s00415-021-10525-6

LETTER TO THE EDITORS

Vestibular paroxysmia associated with typewriter tinnitus: a case report and literature review

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Received: 9 February 2021 / Revised: 15 March 2021 / Accepted: 17 March 2021 / Published online: 21 March 2021

Dear Sirs,

Vestibular paroxysmia (VP) is characterized by brief and recurrent vertigo that respond well to carbamazepine or oxcarbazepine [1]. An assumed mechanism is a neurovascular cross-compression (NVCC) of the vestibular nerve offended by a vascular loop [2]. Typewriter tinnitus refers to unilateral staccato sounds, and has also been attributed to NVCC of the cochlear nerve [2, 3]. Association of these two conditions has rarely been recognized [4]. We herein describe a patient with typewriter tinnitus and VP due to a NVCC of the vestibulocochlear nerve with a literature review.

A 43-year-old man was referred for management of intermittent attacks of vertigo and staccato sounds in his right ear for 5 months. The attacks occurred 10–20 times a day, and each attack lasted less than 1 min. The tinnitus was always time-locked to the vertiginous spells with an abrupt onset and cease. The attacks were often triggered by bending his head and accompanied by oscillopsia and unsteady gait. He had no symptoms between the attacks, but showed spontaneous nystagmus beating leftward, upward, and counterclockwise (upper poles of the eyes beating to the left ear) without visual fixation (Fig. 1A). Horizontal head shaking increased the spontaneous left beating nystagmus, but hyperventilation, mastoid vibration or positional maneuvers did not alter the nystagmus. Video head impulse tests (HITs) showed a decreased gain of the vestibulo-ocular reflex (VOR) for right horizontal semicircular canal with overt saccades (Fig. 1b). The results of other audiovestibular function tests were all normal. Constructive interference in steady-state MRI documented a NVCC and mild angulation of the right vestibulocochlear nerve by a complex of the anterior and posterior inferior cerebellar arteries (Fig. 1c–e). Unfortunately, we did not have an opportunity to evaluate the patient during the attacks.

He started carbamazepine 200 mg daily with a minimal reduction of the tinnitus and subsequent discontinuation. Oxcarbazepine 300 mg twice a day was also ineffective. Re-initiation of carbamazepine at an escalated dose of 300 mg twice a day resulted in resolution of the symptoms, but the interictal spontaneous nystagmus persisted.

Our patient presented with paroxysmal attacks of vertigo and typewriter tinnitus that resolved with carbamazepine. The attacks were mostly spontaneous, but often triggered by head position changes, with an imaging evidence of NVCC. These findings are consistent with the diagnosis of VP according to the criteria proposed by the Barany Society [5]. Given the characteristics of typewriter tinnitus, the paroxysmal nature with an excellent response to carbamazepine, the tinnitus may be ascribed to NVCC affecting the cochlear nerve [2, 6]. Recent studies showed that the psychoacoustic characteristics and a good response to carbamazepine are more reliable for diagnosing NVCC of the cochlear nerve than the radiologic findings [4]. In our patients, the co-occurrence of typewriter tinnitus and VP and simultaneous resolution of these symptoms after carbamazepine trials indicates a common mechanism of NVCC in these paroxysmal disorders. The rare occurrence of typewriter tinnitus in comparison to VP, however, indicates a different susceptibility for ephaptic discharges between the vestibular and cochlear nerves. The larger size of cochlear nerve fibers than the vestibular ones may reduce the likelihood of ephaptic discharges in these nerve fibers [7, 8]. Otherwise, any difference in the reorganization thresholds, which is believed

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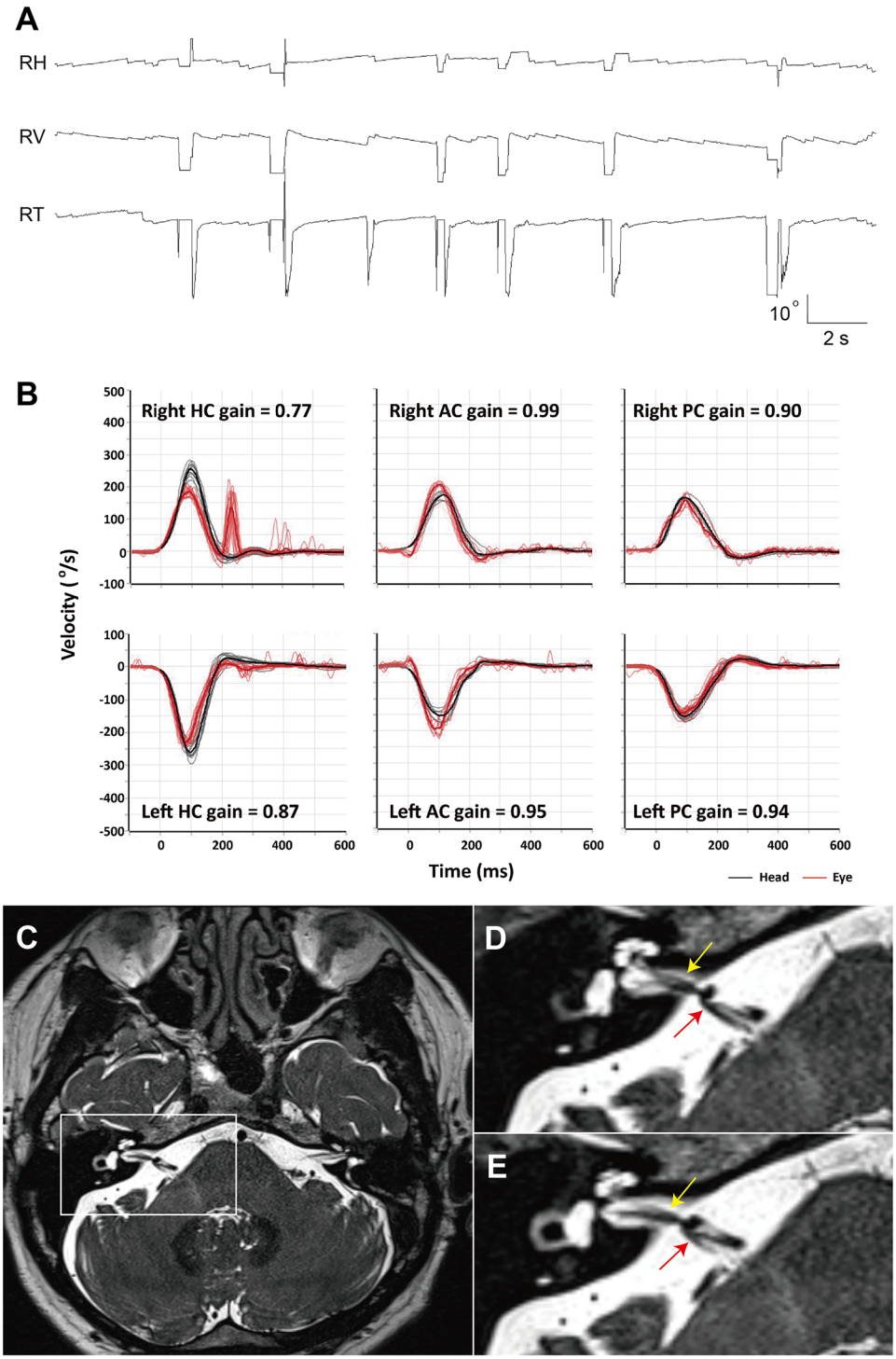

Fig. 1 **a** Video-oculography shows spontaneous nystagmus beating to the left (1.5°/s), upward (1.5°/s), and counterclockwise (1.5°/s) without visual fixation. RH = horizontal position of the right eye, RV = vertical position of the right eye, RT = torsional position of the right eye. Upward deflection in each trace indicates rightward, upward, and clockwise eye motion. **b** Video head impulse tests show overt saccades during stimulation of the right horizontal semicircular canal with a decreased gain at 0.77 (normal = 0.88–1.06). **c**–**e**. Constructive interference in steady-state images of magnetic resonance imaging of the brain and internal auditory canal (C) and magnified views (**d**, **e**, the white square in the Figure C) disclosed the AICA-PICA loop crossing the cisternal segment of the right vestibulocochlear nerve and causing a mild angulation of the nerve (red arrow: vascular complex of AICA-PICA, yellow arrow: the vestibulocochlear nerve)

to play a role in inducing symptoms in NVCC, may result in different occurrences of ephaptic discharges between the cochlear and vestibular nerves [9, 10].

Our patient showed the findings of right peripheral vestibular hypofunction (left beating spontaneous nystagmus and positive HITs for the right horizontal canal) between the attacks. These interictal findings indicate chronic vestibular hypofunction from NVCC, inducing demyelination and resultant conduction block in addition to ephaptic discharges [11]. Indeed, patients with VP have showed various patterns of audiovestibular abnormalities between the attacks [1, 12–15]. Preserved audiometric function in our patient indicates that cochlear damage is not mandatory for generation of tinnitus [6, 16].

Table 1 Summary of the findings in patients with combined vestibular paroxysmia and typewriter tinnitus

No	Age (yrs)	Sex	Side of NVCC	Affected vessel	Disease duration	Symptom duration	Attack frequency	Symptom provocation	Accompanying symptoms	Tinnitus	HL	SN	Triggered nystagmus	Vestibular dysfunction			Treatment (response)
														CP	**VEMP**	**BAEP**	
1 [2]	54	M	R	N/A	0.3 years	N/A	N/A	Noise	None	Ear-clicking	R	None	None	N/A	N/A	N/A	CBZ (PR)
2 [17]	54	M	L	N/A	1.5 years	10–15 s	Every minute	Right ear down	PI, oscillopsia, subjective visual horizontal tilt	Coin dropping into a tin can	WNL	None	Position(LB), HS(RB)	N/A	N/A	N/A	CBZ (PR)
3 [17]	82	M	L	N/A	10 years	10 s	N/A	Sneezing, loud sound, or turnig over in bed	PI, oscillopsia, subjective visual horizontal tilt	Staccato	B, HFHL	None	Position(RB), HS(RB), Vib(RB), HV(RB)	L	L	L	CBZ (CR)
4 [17]	81	F	R	N/A	19 years	15 s	N/A	Loud sound, movement	PI oscillopsia	Machine gun	B, HFHL	LB (ictal and interictal)	HV(LB)	N/A	N/A	B	CBZ (PR)
5 [17]	49	M	R	N/A	1 year	10–20 s	Every minute	None	PI, Pressure sense in ear (interictal)	Crackling	WNL	None	Vib(RB)	N/A	N/A	R	CBZ (CR)
6 [18]	37	F	B	Labyrin-thine artery	N/A	N/A	N/A	Morning time, burning sense when exposed to loud sound	Oscillopsia	Ringing and staccato	WNL	N/A	Gaze, position	R	WNL	WNL	N/A

Table 1 (continued)

No	Age (yrs)	Sex	Side of NVCC	Affected vessel	Disease duration	Symptom duration	Attack frequency	Symptom provocation	Accompanying symptoms	Tinnitus	HL	SN	Triggered nystagmus	Vestibular dysfunction			Treatment (response)
														CP	**VEMP**	**BAEP**	
7 [3]	54	M	L	AICA	N/A	10–15 s	Every minute	Position	PI, oscillopsia	Staccato and coin dropping into a tin can	WNL	N/A	N/A	WNL	WNL	WNL	CBZ (CR), MVD (CR)
8 [19]	54	F	R	N/A	N/A	5–30 s	Every 5–10 min	Position	Oscillopsia	Ear-clicking	B, HFHL	RB, torsional (ictal)	N/A	N/A	R	N/A	CBZ (CR)
9 [20]	70	F	L	VBD	10 years	N/A	N/A	N/A	HFS on the left side	Typewriter	L, LFHL	N/A	N/A	N/A	N/A	N/A	MVD (PR)
10 [20]	71	F	R	VBD	20 days	less than 1 min	10–20 times a day	N/A	None	Typewriter	N/A	None	WNL	WNL	N/A	N/A	OXC (CR)
11 [4]	52	F	L	AICA	0.5 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	WNL	Abnormal cVEMP	N/A	CBZ (PR)
12 [4]	47	M	L	AICA	0.75 months	N/A	N/A	N/A	Facial spasm	Typewriter	WNL	N/A	N/A	N/A	N/A	N/A	CBZ (PR)
13 [4]	43	F	R	AICA	7 years	N/A	N/A	N/A	N/A	Typewriter	R	N/A	N/A	WNL	WNL	N/A	CBZ (PR)
14 [4]	58	F	R	AICA	3 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	WNL	WNL	N/A	CBZ (PR)
15 [4]	72	F	L	AICA	5 months	N/A	N/A	N/A	N/A	Typewriter	L	N/A	N/A	WNL	N/A	N/A	CBZ (PR)
16 [4]	51	F	R	AICA	2 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	WNL	N/A	N/A	CBZ (PR)
17 [4]	56	F	L	AICA	4 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	WNL	WNL	N/A	CBZ (PR)
18 [4]	23	F	R	AICA	1 month	N/A	N/A	N/A	Facial spasm	Typewriter	WNL	N/A	N/A	WNL	WNL	N/A	CBZ (PR)
19 [4]	66	F	R	AICA	2 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	N/A	N/A	N/A	CBZ (PR)
20 [4]	45	M	R	AICA	6 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	N/A	WNL	N/A	CBZ (PR)

Table 1 (continued)

No	Age (yrs)	Sex	Side of NVCC	Affected vessel	Disease duration	Symptom duration	Attack frequency	Symptom provocation	Accompanying symptoms	Tinnitus	HL	SN	Triggered nystagmus	Vestibular dysfunction			Treatment (response)
														CP	VEMP	BAEP	
21 [4]	54	F	L	AICA	1.5 months	N/A	N/A	N/A	N/A	Typewriter	WNL	N/A	N/A	N/A	N/A	N/A	CBZ (CR)

AICA anterior-inferior cerebellar artery; *B* bilaterally involved; *BAEP* brainstem auditory evoked potentials; *CBZ* carbamazepine; *CP* caloric paresis; *CR* complete remission; *F* female; *HFHL* high-frequency hearing loss; *HL* hearing loss; *HS* head-shaking; *L* left; *LB* left-beating nystagmus; *LD* lying-down position; *LFHL* low-frequency hearing loss; *M* Male; *MVD* microvascular decompression; *N/A* not available data; *NVCC* neurovascular cross-compression; *OXC* oxcarbazepine; *PI* postural imbalance; *PR* partial remission; *R* right; *RB* right-beating nystagmus; *s* seconds; *SN* spontaneous nystagmus; *VBD* vertebrobasilar dolichoectasia; *VEMP* vestibular myogenic evoked potentials; *Vib* skull-vibration; *WNL* within normal limit

We were able to find 21 patients with combined VP and typewriter tinnitus in the literature (**Table 1**) [2, 3, 4, 17–20]. Except one with bilateral NVCC on MRIs, all patients showed an imaging evidence of NVCC on the side of tinnitus. Of the 15 patients who had audio-vestibular function tests, 6 showed abnormal results on the symptomatic side. Most patients (19/21, 90%) showed an excellent response to carbamazepine or oxcarbazepine. This report highlights the etiological association of VP and typewriter tinnitus, and proper titration of carbamazepine or oxcarbazepine in managing the patients with these disorders.

Author contributions YJK analyzed and interpreted the data, and wrote the manuscript. J-YC and HJK analyzed and interpreted the data, and revised the manuscript. J-SK designed and conceptualized the study, interpreted the data, and revised the manuscript.

Funding This study was supported by Basic Science Research Program through the National Research Foundation of Korea (NRF) funded by the Ministry of Education, Science and Technology (no. NRF-2016R1D1A1B04935568).

Declarations

Conflicts of interest Drs. Koo, HJ Kim and Choi report no disclosure.

Ethical approval This study followed the tenets of the Declaration of Helsinki and was performed according to the guidelines of Institutional Review Board of Seoul National University Bundang Hospital (B-2102/666–101).

References

1. Hüfner K, Barresi D, Glaser M, Linn J, Adrion C, Mansmann U, Brandt T, Strupp M (2008) Vestibular paroxysmia: diagnostic features and medical treatment. Neurology 71(13):1006–1014
2. Levine RA (2006) Typewriter tinnitus: a carbamazepine-responsive syndrome related to auditory nerve vascular compression. Orl 68(1):43–47
3. Mathiesen T, Brantberg K (2015) Microvascular decompression for typewriter tinnitus-case report. Acta Neurochir 157(2):333–336
4. Sunwoo W, Jeon YJ, Bae YJ, Jang JH, Koo J-W, Song J-J (2017) Typewriter tinnitus revisited: The typical symptoms and the initial response to carbamazepine are the most reliable diagnostic clues. Sci Rep 7(1):1–8
5. Strupp M, Lopez-Escamez JA, Kim J-S, Straumann D, Jen JC, Carey J, Bisdorff A, Brandt T (2016) Vestibular paroxysmia: diagnostic criteria. J Vestib Res 26(5–6):409–415
6. Ryu H, Yamamoto S (2001) Neurovascular decompression of the eighth cranial nerve for intractable vertigo and tinnitus. Oper Tech Neurosurg 4(3):142–152
7. Gardner WJ (1966) Cross talk—the paradoxical transmission of a nerve impulse. Arch Neurol 14(2):149–156
8. Kim H-S, Kim D-I, Chung I-H, Lee W-S, Kim K-Y (1998) Topographical relationship of the facial and vestibulocochlear nerves in the subarachnoid space and internal auditory canal. Am J Neuroradiol 19(6):1155–1161

9. Schwaber MK, Whetsell WO (1992) Cochleovestibular nerve compression syndrome. II. Vestibular nerve histopathology and theory of pathophysiology. The Laryngoscope 102(9):1030–1036
10. Møller AR (1999) Vascular compression of cranial nerves: II: pathophysiology. Neurol Res 21(5):439–443
11. Brandt T, Strupp M, Dieterich M (2016) Vestibular paroxysmia: a treatable neurovascular cross-compression syndrome. J Neurol 263(1):90–96
12. Schwaber MK, Hall JW (1992) Cochleovestibular nerve compression syndrome. I. Clinical features and audiovestibular findings. The Laryngoscope 102(9):1020–1029
13. Markowski J, Gierek T, Kluczewska E, Witkowska M (2011) Assessment of vestibulocochlear organ function in patients meeting radiologic criteria of vascular compression syndrome of vestibulocochlear nerve–diagnosis of disabling positional vertigo. Med Sci Monit 17(3):CR169
14. Best C, Gawehn J, Krämer HH, Thömke F, Ibis T, Müller-Forell W, Dieterich M (2013) MRI and neurophysiology in vestibular paroxysmia: contradiction and correlation. J Neurol Neurosurg Psychiatry 84(12):1349–1356
15. Ihtijarevic B, Van Ombergen A, Celis L, Maes LK, Wuyts FL, Van de Heyning PH, Van Rompaey V (2019) Symptoms and signs in 22 patients with vestibular paroxysmia. Clin Otolaryngol 44(4):682
16. De Ridder D, Heijneman K, Haarman B, van der Loo E (2007) Tinnitus in vascular conflict of the eighth cranial nerve: a surgical pathophysiological approach to ABR changes. Prog Brain Res 166:401–411
17. Brantberg K (2010) Paroxysmal staccato tinnitus: a carbamazepine responsive hyperactivity dysfunction symptom of the eighth cranial nerve. J Neurol Neurosurg Psychiatry 81(4):451–455
18. Singh NK, Singh P, Usha M, Akshay M (2013) Audio-vestibular findings in Vestibular Paroxysmia. Indian J Otol 19(2):82
19. Young AS, Jonker B, Welgampola MS (2019) Vestibular paroxysmia presenting with irritative nystagmus. Neurology 92(15):723–724
20. Huh G, Bae YJ, Woo HJ, Park JH, Koo J-W, Song J-J (2020) Vestibulocochlear symptoms caused by vertebrobasilar dolichoectasia. Clin Exp Otorhinolaryngol 13(2):123