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Case Report

Bow Hunter's Syndrome Masquerading as Definite Ménière's Disease

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Abstract

Bow Hunter's syndrome (BHS) results from vertebral-basilar insufficiency secondary to mechanical stenosis or occlusion of the unilateral vertebral artery at the atlanto-axis during head rotation. It is sometimes difficult to detect BHS, which is often ignored at other processes. A 44-year-old woman had up to 13 episodes of acute vertigo with left tinnitus, which was related to head rotation. Pure tone audiometry documented left side hearing impairment, so definite Ménière's disease was impressed; however, electronystagmography, a caloric test, posturagraphy, a cervical vestibular evoked myogenic potential, color-coded duplex sonography and magnetic resonance imaging/angiography disproved this. Ménière's disease was impressed and was attributed to BHS, which was related to a vertebral-basilar artery anomaly. The patient was treated conservatively and changed her workstyle, and the symptoms did not recur over the following half year. Therefore, BHS can masquerade as Ménière's disease. We report this condition because of its rarity. [*Tzu Chi Med J* 2010;22(4):219–224]

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1. Introduction

Although certain cases of Ménière's disease (MD) are diagnosed when there is histopathologic evidence of endolymphatic hydrops, definite MD is confirmed when the following four clinical conditions occur: (1) at least two definitive spontaneous episodes of vertigo persisting for at least 20 minutes; (2) pure tone audiometry documents hearing loss on at least one occasion; (3) tinnitus or aural fullness in the affected ear; and (4) other causes are excluded [1]. Bow Hunter's syndrome (BHS), also called rotational vertebral artery syndrome, results from vertebral-basilar insufficiency (VBI) secondary to mechanical stenosis or occlusion of the unilateral vertebral artery (VA) at the atlanto-axis during head rotation [2,3]. It is sometimes difficult to detect BHS, which is always ignored at other processes. BHS was attributed to vertebral-basilar artery anomaly in one rare case [2]. It masquerades as definite MD quite rarely, so we report this case.

2. Case report

A 44-year-old woman was a factory worker and had been healthy until she was assigned to a new workstation at her factory in 2006, after which she visited our emergency department up to 13 times because

of acute vertigo with left tinnitus. The symptoms always occurred after she had rotated her head to the right side for a long period of time at work. She was not addicted to cigarettes, alcohol, or areca.

During each episode, changing position exacerbated the symptoms, and bed rest relieved them. She would stop work and visit our emergency room about 1 hour after the symptoms began. There was no headache, blurred vision, ataxia, phonophobia, photophobia or other neurologic signs. She was alert and oriented to time, place and person. Spontaneous clockwise rotational nystagmus was noted at each visit to the emergency room (Fig. 1A). Although the vertigo generally subsided in 3–4 hours, the dizziness and left tinnitus would gradually remit over the following week. Pure tone audiometry during the 1st, 8th, 9th and 13th attacks showed left side hearing impairment (Figs. 1B–E), but the short increment sensitivity index was bilaterally 0% over 1000 Hz, 2000 Hz and 4000 Hz, and speech discrimination scores bilaterally were 100%.

During remission after the 8th episode, she received a battery of studies. The neurological physical examination was normal. She did not have orthostatic hypotension. There was no gaze nystagmus, positional nystagmus, or positioning nystagmus. She refused a test of prolonged rightward rotation provocation of the head, because she feared a recurrence of her symptoms. Electronystagmography for pursuit, saccade, optokinetic nystagmus and optokinetic after nystagmus did not show any abnormalities, but a caloric test showed left vestibular paresis (Fig. 2A). Posturagraphy showed the center of gravity was posited rightward and backward (Fig. 2B). An air-conducted vibration cervical vestibular evoked myogenic potential (VEMP) test was normal (Fig. 2C). A hemodynamic decrease was detected in the basilar artery and left intracranial VA by transcranial color-coded duplex sonography when her head was rotated rightward and she incurred the same vertigo with clockwise rotational nystagmus (Table).

T1, T2 and fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging did not show any abnormality (Fig. 3A). Diffusion weighted imaging (DWI) and an apparent diffusion coefficient (ADC) map showed some remarkable findings in the left half of the midbrain and vermis (Figs. 3B and 3C). Time-of-flight magnetic resonance angiography (TOF MRA) showed that the bilateral proximal ends of the posterior cerebral artery were absent (Fig. 3D), and the basilar artery and right intracranial VA were both hypoplastic (Fig. 3E).

All blood examinations were within the normal ranges, except for total cholesterol levels during the 8th and 11th episodes, which were 387 mg/dL and 413 mg/dL, respectively (normal range, 130–200 mg/dL). Therefore, symptom control was recommended in the event of recurrence. In addition, she was given rosuvastatin 10 mg/day, an HMG-CoA reductase inhibitor, and pentoxifylline 400 mg/day, a circulatory promoter. She was asked to avoid prolonged rightward head rotation at work. The patient's course was uneventful over the following half year. Pure tone audiometry showed hearing was normal bilaterally (Fig. 1F). Her total cholesterol was 293 mg/dL, and blood lipid control was still proceeding at her most recent follow-up.

3. Discussion

The patient's episodic vertigo with left tinnitus (Figs. 1B–E) could have been attributed to migrainous vertigo, MD or VBI. Migrainous vertigo was ruled out because she did not have migraines. The negative orthostatic hypotension test excluded dysautonomia. Since she feared a recurrence of symptoms, it was unreasonable to coerce her into a prolonged rightward head-rotation provocation test. The whole history matched the diagnosis of definite MD. However, symptomatic audiometry showed left side hearing impairment with a normal short increment sensitivity index and normal speech discrimination scores. This implies that the left cochlea and left cochlear nerve were healthy, so the hearing impairment should have resulted from impairment of the left auditory pathway, which is different from the simple inner ear injury of MD. Electrophysiological and imaging studies indicated another diagnosis.

During remission after the 13th episode, a cool-water caloric test in the left ear did not give rise to rightward caloric nystagmus, indicating that the left superior vestibular nerve and its corresponding vestibular system in the upper brainstem were impaired [4] (Fig. 2A). However, posturagraphy showed that the center of gravity was posited rightward and backward, indicating that left vestibular function predominated. Her cervical VEMP was normal, indicating that the inferior vestibular nerve and its corresponding sacculocollic reflex pathway in the lower brainstem were healthy [5]. DWI and an ADC map revealed possible ischemic cytotoxic edematous changes in the left-side vermis and left-side midbrain [6,7] (Figs. 3B and 3C), although the T1, T2 and FLAIR images did not show any abnormalities (Fig. 3A).

Application of digital subtraction angiography to a vertigo sufferer without any focal neurological signs is expensive and time-consuming. TOF MRA with color-coded duplex sonography is preferred because of its convenience, safety and accuracy. Defects of the posterior circle of Willis occur in 74.6% of adult Taiwanese, and 3.7% of them do not have the bilateral proximal ends of the posterior cerebral arteries [8]. This condition limits the circulatory communication of the anterior and posterior circulation, and so predisposes the patient to VBI (Fig. 3D). Although basilar artery hypoplasia is too rare to be well defined in the

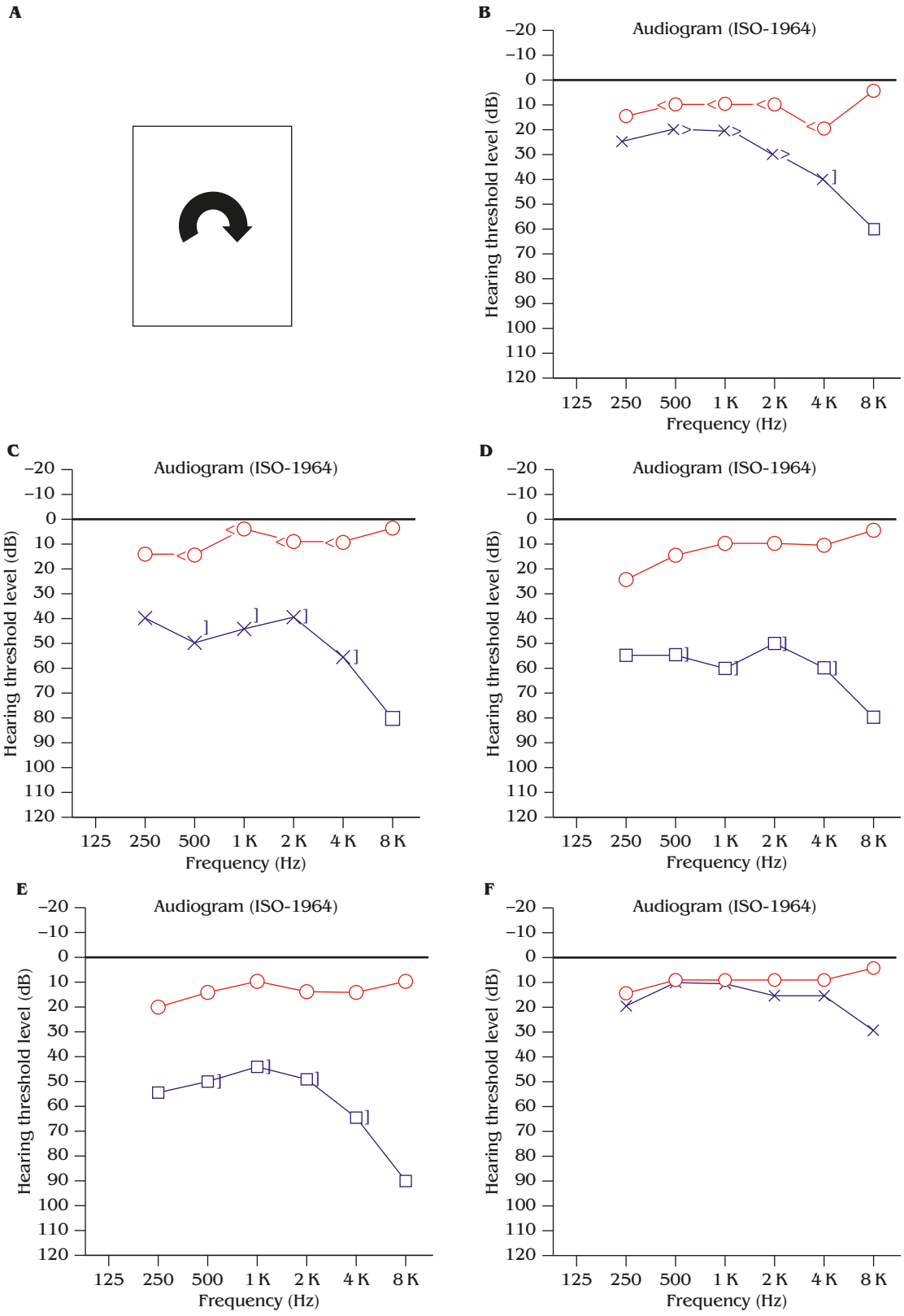

Fig. 1 — (A) Spontaneous clockwise rotational nystagmus is noted when the patient was symptomatic. (B) The average hearing threshold between 250 Hz and 8000 Hz was 12.5 dBHL (hearing level) in the right ear and 32.5 dBHL in the left ear during the 1st episode, (C) 10 dBHL and 51.7 dBHL during the 8th episode, (D) 12.5 dBHL and 60 dBHL during the 9th episode, and (E) 14.2 dBHL and 60.8 dBHL during the 13th episode. (F) The patient has normal hearing when asymptomatic. Keys for the audiogram: ○=right-ear air conduction; <=right-ear bone conduction; ×=left-ear air conduction; □=left-ear masking air conduction; >=left-ear bone conduction; }=left-ear masking bone conduction.

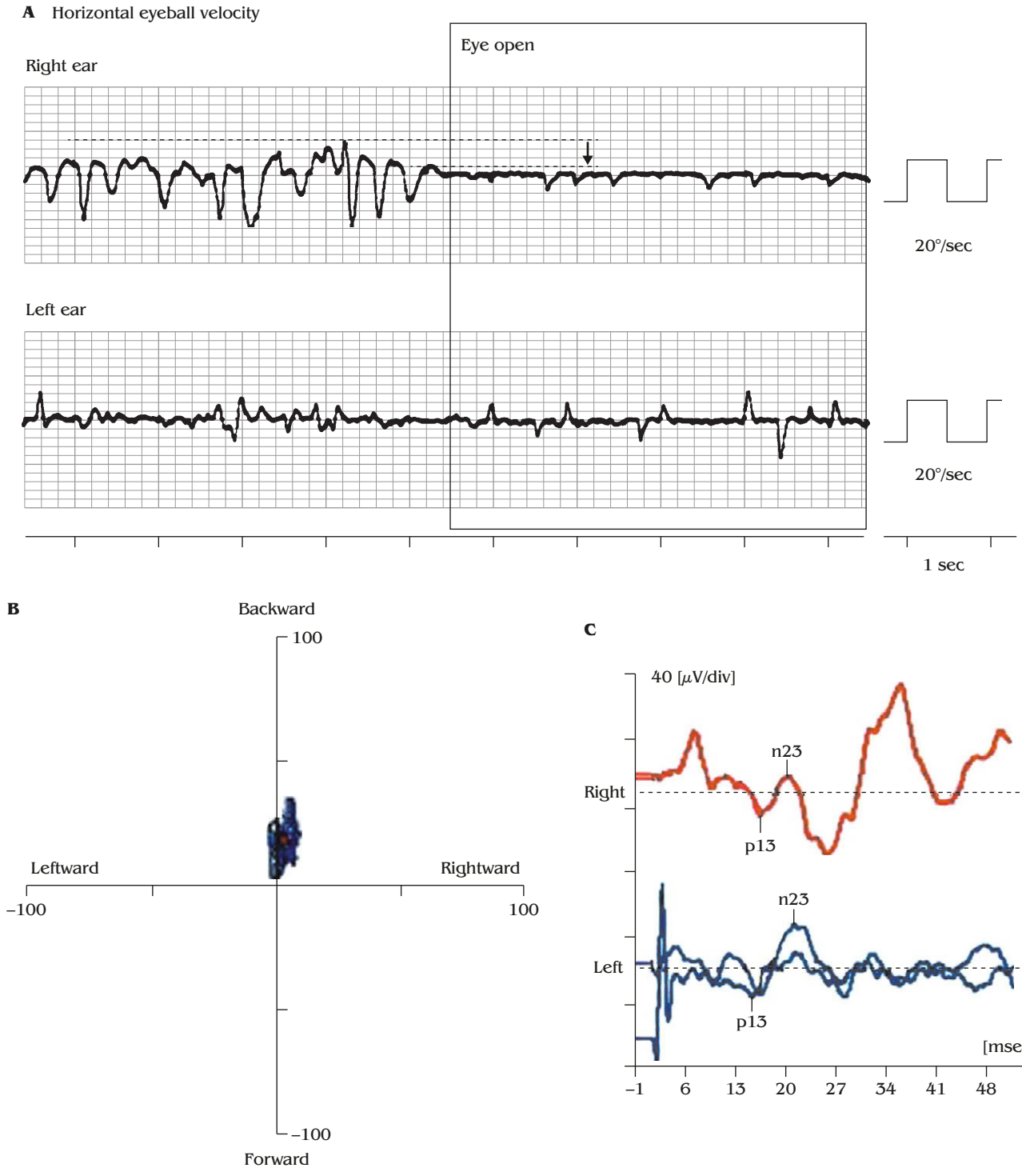

Fig. 2 — (A) Tap water at 20°C was applied in a caloric test. Leftward caloric nystagmus is induced by the right caloric test, with a positive visual suppression test (arrow). However, no significant rightward caloric nystagmus was induced by the left caloric test. (B) Posturagraphy shows that the center of gravity is posited right- and backward, rather than at the midpoint. (C) The latencies of the cervical VEMP p13 and n23 are within the normal ranges, respectively, at 17.11 msec and 21.06 msec in the right ear, and 17.11 msec and 21.89 msec in the left.

literature, it is considered if the artery diameter is much shorter than the common range (3.0–5.5 mm) [9,10]. This condition is often accompanied by VA hypoplasia (diameter <2.0 mm on TOF MRA) [11,12], and so predisposes the patient to VBI, ischemic stroke or even transient ischemic attacks. In addition, asymmetric blood flow in the VA might gradually cause tortuosity of the basilar artery (Fig. 3E) [13].

In our patient, color-coded duplex sonography showed lower blood flow in the basilar artery and left intracranial VA during rightward head rotation than leftward (Table). However, blood flow in the right intracranial VA decreased only slightly during leftward head rotation. We suggest that the rightward head rotation incurred insufficiency of the basilar artery, the left VA and their branches, indicative of BHS related to hypoperfusion of the posterior circulation. The right half of the brainstem and the right cerebellar hemisphere were still supplied by the right VA and its posterior inferior cerebellar artery branch, so they escaped hypoperfusion (Figs. 3B and 3C).

Table — Transcranial color-coded duplex sonogram (EnVisor; Philips, Andover, MA, USA)

	Rightward head rotation			Leftward head rotation		
	Right VA	Left VA	BA	Right VA	Left VA	BA
Average velocity (cm/sec)	24.2	12.2	15.0	23.1	25.5	24.3
Resistance index	0.53	0.65	0.55	0.58	0.55	0.50

VA=vertebral artery; BA=basilar artery.

Positive findings on DWI/ADC raise fears of possible ischemic stroke. This patient should have VBI and ischemic infarction over the left half of the posterior circulation territory (Figs. 3B and 3C). We think that the left brainstem auditory pathway and the left vestibular system (central and peripheral) were also injured, but the left inferior vestibular nerve was spared; thus, reversible left hearing impairment occurred (Figs. 1B–E). Although the left side vestibular system was impaired, the left inferior vestibular nerve still worked, and its input could not be well inhibitory modulated by the left vestibular nucleus, just like the left posterior semicircular canal was continually excited. Therefore, spontaneous clockwise rotational nystagmus occurred at the onset of symptoms (Fig. 1A), and left vestibular function predominated in remission (Fig. 2B).

In conclusion, BHS is attributed to a vertebral-basilar artery anomaly, and occasionally masquerades

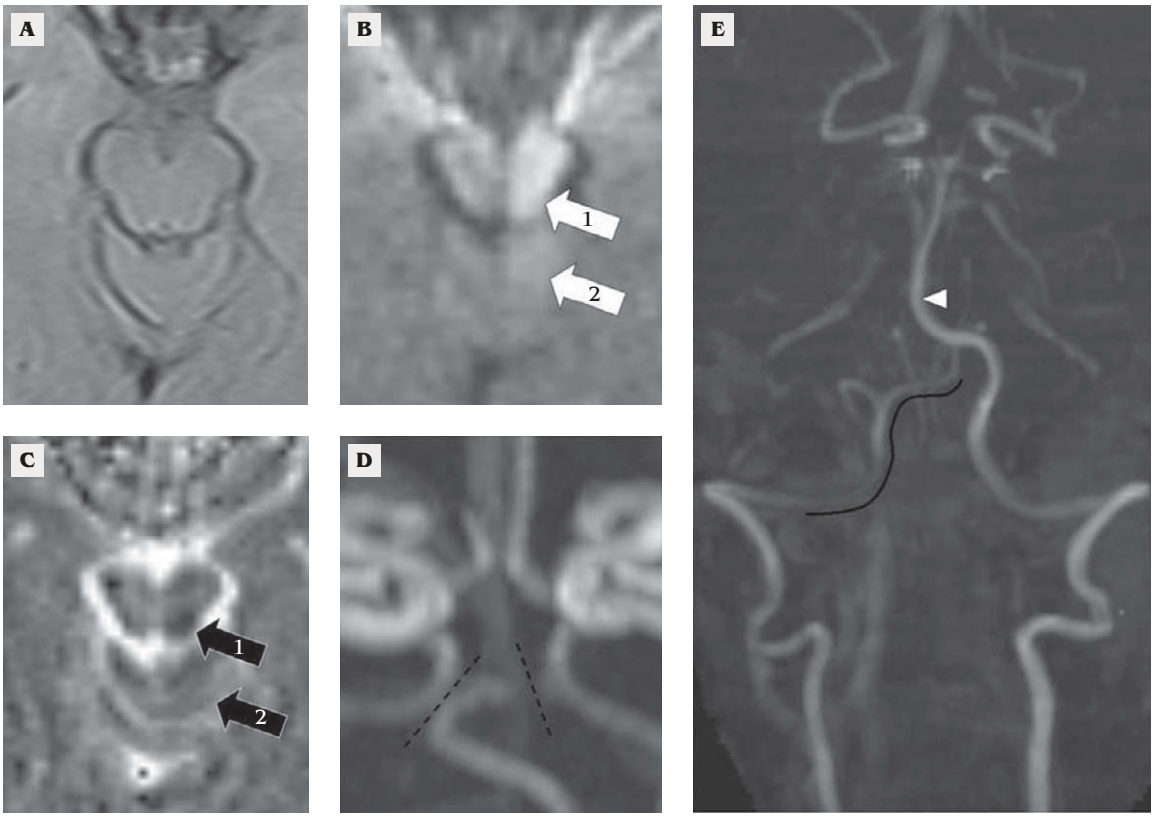

Fig. 3 — (A) FLAIR (TR/TE/excitation: 6000/84/1) shows no abnormalities in the midbrain and the vermis. (B) DWI (TR/TE/excitation: 8374/148/1) shows that the left half of the midbrain is hyperintense, especially over the vestibular nucleus (arrow 1). In addition, the left half of the vermis is mildly hyperintense (arrow 2). (C) An ADC map (TR/TE/excitation: 8374/148/1) shows that the left vestibular nucleus is hypointense (arrow 1), and the left vermis is also mildly hypointense (arrow 2). (D) TOF MRA (TR/TE/excitation: 29/6/1) shows that the bilateral proximal ends of the posterior cerebral arteries are absent. There is a defect over the posterior circle of Willis. (E) The basilar artery is hypoplastic (diameter, 1.7 mm) and tortuous (arrowhead). The intracranial segment (black curved line) of the right VA is hypoplastic (diameter, 1.7 mm), so the left VA mainly supplies the basilar artery (diameter, 2.3 mm).

as MD. Although our patient's symptoms could be controlled conservatively in the event of recurrence, we could do nothing for the vertebral-basilar artery anomaly; therefore, antiplatelet and blood lipid control was recommended. In addition, she was advised to change her work style, avoiding long-term head rightward rotation, in order to reduce transient BHS, and prevent Bow Hunter's stroke (14). Her course was uneventful over the following half year. However, she still has hyperlipidemia (total cholesterol, 293 mg/dL), so blood lipid control is continuing.

References

1. American Academy of Otolaryngology, Committee on Hearing and Equilibrium. Guidelines for the diagnosis and evaluation of therapy in Menière's disease. *Otolaryngol Head Neck Surg* 1995;113:181–5.
2. Marti S, Hegemann S, von Büdingen HC, Baumgartner RW, Straumann D. Rotational vertebral artery syndrome. 3D kinematics of nystagmus suggest bilateral labyrinthine dysfunction. *J Neurol* 2008;255:663–7.
3. Horowitz M, Jovin T, Balzar J, Welch W, Kassam A. Bow Hunter's syndrome in the setting of contralateral vertebral artery stenosis: evaluation and treatment options. *Spine* 2002;27:E495–8.
4. Su MC, Young YH. Neurotological aging change of the cerebellum. *J Taiwan Otolaryngol Head Neck Surg* 1990;30: 305–9.
5. Yang TL, Wu CH, Young YH. Normal value of vestibular evoked myogenic potential. *J Taiwan Otolaryngol Head Neck Surg* 2001;36:160–4.
6. Oppenheim C, Stanescu R, Dormont D, et al. False-negative diffusion-weighted MR findings in acute ischemic stroke. *AJNR Am J Neuroradiol* 2000;21:1434–40.
7. González RG, Schaefer PW, Buonanno FS, et al. Diffusion-weighted MR imaging: diagnostic accuracy in patients imaged within 6 hours of stroke symptom onset. *Radiology* 1999;210:155–62.
8. Chen HW, Yen PS, Lee CC, et al. Magnetic resonance angiographic evaluation of circle of Willis in general population: a morphologic study in 507 cases. *Chin J Radiol* 2004;29:223–9.
9. Seemant C, Timonthy L, Chen W. Ischemia in the territory of a hypoplastic vertebrobasilar system. *Neurology* 1999; 52:980–3.
10. Turgut N, Pekindil G, Utku U, Celik Y, Siengün S. Isolated hypoplasia of distal basilar artery: clinical and imaging findings. *Yeni Symposium* 2004;42:121–5.
11. Chuang YM, Huang YC, Hu HH, Yang CY. Toward a further elucidation: role of vertebral artery hypoplasia in acute ischemic stroke. *Eur Neurol* 2006;55:193–7.
12. Chuang YM, Hwang YC, Lin CP, Liu CY. Toward a further elucidation: role of vertebral artery hypoplasia in migraine with aura. *Eur Neurol* 2008;59:148–51.
13. Hong JM, Chung CS, Bang OY, Joo IS, Huh K. Vertebral artery dominance contributes to basilar artery curvature and peri-vertebrobasilar junctional infarcts. *J Neurol Neurosurg Psychiatry* 2009;80:1087–92.
14. Sorensen BF. Bow Hunter's stroke. *Neurosurgery* 1978; 2:259–61.