

Case Report

Unilateral Sudden Deafness with Vertebrobasilar Arterial Hypoplasia: A Case Report

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Unilateral sudden deafness (SD) is often accompanied by vertigo and attributable to ipsilateral vestibular labyrinthitis or neuritis. Remission is often accompanied by episodic dizziness caused by residual problems with ipsilateral vestibular function. A 56-year-old woman visited our otorhinolaryngologic clinic complaining of episodic dizziness when she walked fast or when she stood up abruptly. She reported that she had been undergoing treatment for it from the time she experienced and was treated for SD six months earlier. Applied during purposely induced episodic dizziness, a caloric test found no abnormalities, though an optokinetic electronystagmogram and air conductive vibration-cervical vestibular evoked myogenic potential, indicating impairments of the bilateral upper and lower brainstem. Three-dimensional time-of-flight magnetic resonance angiography showed vertebrobasilar arterial hypoplasia. She received conservative medical treatment and was recommended a change in life-style. Over the following week, there were few recurrences of dizziness, and the following six months were uneventful. Although our patient's right SD could not be attributed to vertebrobasilar arterial hypoplasia, her subsequent episodic dizziness was caused by both SD-related mild residual right peripheral vestibular dysfunction and vertebrobasilar arterial hypoplasia-related bilateral central vestibular dysfunction. Therefore, episodic dizziness should not be routinely and completely attributed to peripheral vestibular dysfunction in a person who experiences SD, as it could also be caused by a central vascular anomaly related to central vestibular dysfunction.

Keywords: sudden deafness, episodic dizziness, vertebrobasilar arterial hypoplasia, hyperventilation, 3D TOF MRA

Introduction

Sudden deafness (SD) is defined as a hearing loss up to 30dB hearing level (dBHL) over 3 continuous audiogram frequencies occurring within 3 days. It is prevalent in adults between 40 and 60 years old, and it can possibly be caused by a central lesion, virus infection, vasculopathy, autoimmune disease or others^[1]. Patients who

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experience SD occasionally encounter vertigo, which is usually attributable to ipsilateral vestibular labyrinthitis or neuritis; remission is often accompanied by episodic dizziness (ED), which is attributable to residual problems with ipsilateral vestibular function, especially in patients with poor recovery. We encountered a patient with ED following unilateral SD. The ED was not completely occur as a result of unilateral peripheral vestibular dysfunction, but it more related to a vertebrobasilar arterial hypoplasia, demonstrated by three-dimensional time-of-flight magnetic resonance angiography (3D TOF MRA). Herein we report this rare case for its educational significance.

Case Report

A 56-year-old woman with a body mass index of 27.9 kg/m² had visited another hospital besides ours due to right tinnitus with vertigo, nausea and vomiting that had lasted three days, suggesting right SD. Right intratympanic injection of corticosteroids resolved the vertigo but not the tinnitus. Six months later, she came to our emergency department complaining that, since the time of the SD, she would experience one to two minutes of episodic dizziness whenever she walked fast or stood up abruptly. She said that it occurred up to two times a day. She did not complain of headaches, syncope, nausea, vomiting, photophobia, phonophobia, ataxia or other neurological symptoms. Over the following half year while being treated conservatively at the other hospital, the episodic dizziness recurred frequently, causing her to visit our clinic for a second opinion.

At our clinic, the patient was found to have a blood pressure of 110/55 mmHg and a heart rate of 82/min. Cardiac, otorhinolaryngological, and neurological physical examinations were normal. She had no gaze, positional or positioning nystagmus. Because her episodic dizziness was not resolved by Epley's maneuver or barbecue rotation, we decided to perform further tests, including Romberg, Mann, stepping, tandem gait, diadochokinesia, finger to nose tracing, head shaking and head thrust. No abnormalities were found. Hyperventilation with 20 deep respirations

and abruptly standing up from a squatting position induced the episodic dizziness, though an orthostatic hypotension test showed negative results. Electrocardiography and 24-hour Holter monitoring did not show any abnormalities, either. The data from all blood examinations, including some immunological studies, were within the normal range.

Pure tone audiometry (AC 40, Interacoustics, Denmark) found average hearing thresholds between 250 Hz and 4,000 Hz to be 14 dBHL in the left ear and 81 dBHL in the right ear (Fig. 1A). While purposely inducing episodic dizziness by hyperventilation, she was tested by electronystagmograms (NY-41, Rion, Japan) for pursuit, saccade and a caloric test (20°C water, 20 seconds). The results were normal. However, optokinetic electronystagmogram showed 50 seconds of rightward nystagmus, but no leftward nystagmus (Fig. 1B), and no subsequent bilateral optokinetic after-nystagmus. Air conductive vibration-cervical vestibular evoked myogenic

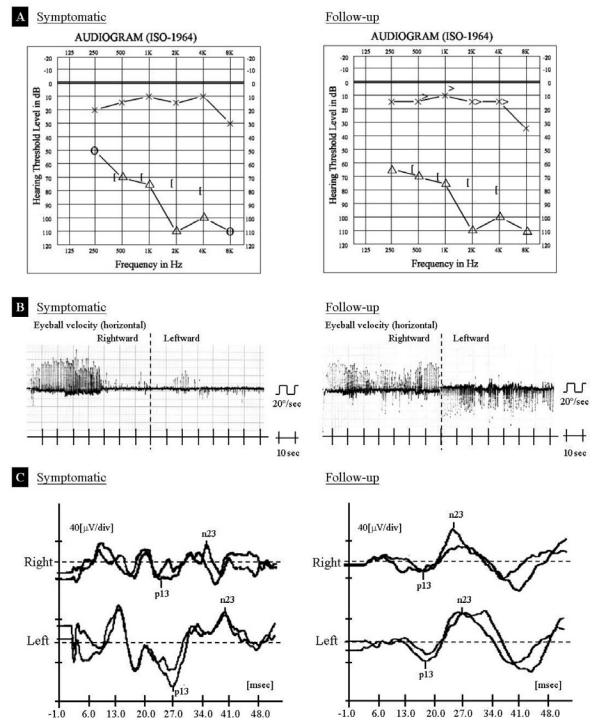


Fig. 1. A: pure tone audiometry. B: optokinetic electronystagmogram (Jung type). C: air conductive vibration-cervical vestibular evoked myogenic potential.

potential (ACV-cVEMP)(580-NAVPRO, Bio-Logic, USA), showing latencies (amplitude) of p13 and n23 for 23.10 msec (-14.79 μ V) and 35.00 msec (18.21 μ V) in the right ear, and for 27.10 msec (-42.19 μ V) and 40.0 msec (28.51 μ V) in the left ear (Fig. 1C). The interaural amplitude difference ratio was -0.36 (normal reference: -0.33~0.33).

Color-coded duplex ultrasonography (EnVisor, Philips, USA) showed bilateral common carotid arteries to be mildly atherosclerotic. Hyperventilation with 20 deep respirations per minute and lying supine at 45° reduced the blood flow in the bilateral vertebral arteries (Table 1). Magnetic resonance imaging (1.5 Tesla system, Picker Edge Eclipse, Picker 98 International, USA) was performed. 3D TOF MRA and true fast imaging in steady state precession (trueFISP) showed the inner and outer diameters of the upper 1.0 cm of the basilar artery to be 1.8 mm and 1.9 mm, respectively, the lower 0.5 cm of the basilar artery to be 2.4 mm and 2.9 mm, the right intracranial vertebral artery to be 1.5 mm and 1.7 mm, and the left vertebral artery to be 1.4 mm and 1.5 mm (Fig. 2A&B). The circle of Willis was complete and the bilateral posterior cerebral arteries were fetal-type (Fig. 2C).

We diagnosed her as having intracranial vertebrobasilar arterial hypoplasia, for which oral anti-platelet aspirin (Bokey 100 mg) was recommended. She was also advised to stand up slowly, to avoid standing abruptly after prolonged sitting, and to warm up before any vigorous exercise. Over the following week, her episodic dizziness seldom recurred. A skin rash was noted, so an oral circulatory promoter, piracetam (Eubrain 400mg), was prescribed as a replacement for oral aspirin. The following half year was uneventful

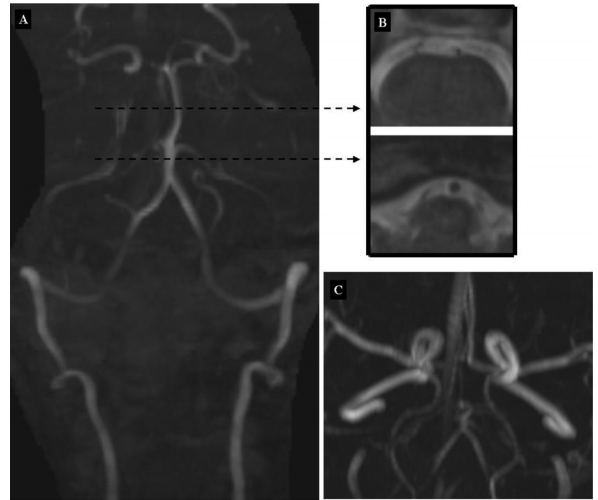


Fig. 2. A: three-dimensional time-of-flight magnetic resonance angiography (3D TOF MRA) of the vertebrobasilar artery (TR/TE/excitation: 29/6/1). B: axial T2 true fast imaging in steady state precession of the upper 1.0 cm and lower 0.5 cm of the basilar artery (TR/TE/excitation: 5,000/126/1). C: 3D TOF MRA of the circle of Willis.

and she had no recurrence of episodic dizziness. Medication was halted and follow-up studies were performed. Pure tone audiometry revealed that the average of hearing thresholds to be 14 dBHL in the left ear and 85 dBHL in the right ear (Fig. 1A). Optokinetic electronystagmography showed good results (Fig. 1B). ACV-cVEMP showed the latencies (amplitude) of p13 and n23 to be 16.48 msec (-14.26 μ V) and 24.39 msec (45.70 μ V), respectively, in the right ear and 17.63 msec (-36.07 μ V) and 26.37 msec (54.58 μ V) in the left ear. The interaural amplitude difference ratio was -0.20 (Fig. 1C).

Table 1. Color-coded Doppler Ultrasonography

	Lying horizontal				Hyperventilation				Lying supine 45°			
	VA		ICA		VA		ICA		VA		ICA	
	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left
Inner diameter (mm)	2.39	3.00	4.86	4.97	2.32	2.81	4.81	4.85	2.38	2.95	4.85	4.95
Mean velocity (cm/sec)	15.9	19.1	22.7	31.7	16.1	20.1	23.1	32.2	14.8	18.5	22.6	28.5
Mean flow (mL/min)	42.8	81.0	252.7	369.0	40.8	74.8	251.9	365.8	39.5	75.9	250.5	329.1
Sum (mL/min)	123.8		621.7		115.6		617.7		115.4		579.6	
Resistance index	0.60	0.58	0.59	0.53	0.61	0.57	0.69	0.54	0.58	0.57	0.58	0.53

VA=vertebral artery. ICA=internal carotid artery.

Discussion

Episodic dizziness has been attributed to cervical abnormalities, peripheral vertigo, migrainous vertigo, vertebrobasilar insufficiency, dysautonomia causing orthostatic hypotension, intracranial hypotension, intracranial hypertension and other causes. The normal results for patient's blood examinations, including some immunological studies, ruled out various potential underlying causes such as systemic and autoimmune disease. Because there was no positioning nystagmus and a poor response to canalith repositioning procedures, benign paroxysmal positional vertigo was unlikely^[2,3]. No other focal neurological signs accompanied her episodic dizziness; thus, vertebrobasilar insufficiency (presence of at least two focal signs) was unlikely^[4], but transient ischemic attack was still suspected^[5].

Prolonged hyperventilation or standing up abruptly after being in a prolonged squatting position usually causes dizziness or other symptoms of discomfort in healthy persons. The results would be considered positive if her dizziness occurred along with these actions. In this patient, the symptom was related to decreased hemodynamics secondary to postural change and cerebral vasoconstriction as shown by positive results after both hyperventilation and standing abruptly. Autonomic dysfunction was unlikely as indicated by the negative orthostatic hypotension test [6]. Cardiovascular syncope was also unlikely because her heart auscultation and electrocardiogram were normal and she had no history of heart disease. After hyperventilation or assuming a 45° supine position, the inner diameter of the bilateral vertebral arteries decreased and resulted in reduced blood flow (Table 1). The ultrasonography results explained the clinical presentation induced by fast walking or abruptly standing up.

When her episodic dizziness was induced, the optokinetic electronystagmogram showed poor manifestations, with the leftward nystagmus being poorer than the rightward nystagmus. The pursuit, saccade, optokinetic-after nystagmus and caloric tests were all normal, ruling out impairment of

the vestibule, vestibular nerve, vestibular nucleus, cerebrum and cerebral cortex. ACV-cVEMP in both ears showed prolonged latencies of p13 and n23 with an abnormal interaural amplitude difference ratio, indicating bilateral lower-brainstem central vestibular dysfunction and mild right peripheral vestibular dysfunction^[7,8].

3D TOF MRA, which is safer than digital subtraction angiography, provides insight into the characteristics of blood flow and provides data making the measurement of the internal diameter of an examined vessel possible^[9]. T2 trueFISP depicts the external diameter of an examined vessel. Therefore, 3D TOF MRA with axial T2 trueFISP can discriminate congenital hypoplasia, in which the inner and outer diameters are both small, from acquired stenosis. In the latter, the vessel has a small inner diameter and a normal outer diameter^[10]. Except for the possibility of a possible central lesion, we do not recommend routine magnetic resonance imaging in SD. In our patient, the imaging demonstrated that the intracranial vertebrobasilar artery was hypoplastic but both extracranial vertebral arteries were not (Fig. 2A&B).

The upper 1.0 cm of the basilar artery averages 4.1 mm, although there is a range of 3.0 to 5.5 mm in autopsy^[11]. High-resolution computed tomography shows the average diameter of the basilar artery to be 3.17 ± 1.3 mm^[12]. Although basilar arterial hypoplasia is not defined in the literature, it can be diagnosed by 3D TOF MRA if the neuro-radiologist discovers that the outer and inner diameters to be below usual range^[13,14]. Basilar arterial hypoplasia with bilateral vertebral arterial hypoplasia (3D TOF MRA shows external and inner diameters < 2.0 mm^[15]) is the so-called vertebrobasilar arterial hypoplasia, a disorder which is seldom symptomatic, but occasionally leading to vertebrobasilar insufficiency secondary to hypoperfusion, thrombo-embolism, or atherosclerosis^[13]. Although rheological hemodynamic disturbance is implicated in the pathogenesis of SD^[16,17], vertebrobasilar insufficiency tends to be related to bilateral SD^[18,19]. Therefore, our patient's unilateral SD was unlikely related to the vertebrobasilar arterial hypoplasia.

We suggest a certain primitive trigeminal artery did not degenerate as scheduled when the bilateral longitudinal neural arteries were fusing medially during her fifth embryologic week. The function and development of the vertebrobasilar artery were thereby limited, and the intracranial vertebrobasilar arterial hypoplasia remained after the trigeminal artery degenerated^[13]. Most patients with intracranial vertebrobasilar arterial hypoplasia have no symptoms, but are predisposed to vertebrobasilar insufficiency or ischemic stroke^[14]. We do not think that age-related atherosclerosis altered the hemodynamics of the vertebrobasilar artery to become symptomatic until our patient experienced SD. During sudden exertion or when standing abruptly, it retarded the posterior circulation and induced episodic dizziness via brainstem impairment without the occurrence of syncope or ischemic stroke.

We could do nothing about the patient's vertebrobasilar arterial hypoplasia, so the patient was first prescribed oral antiplatelet aspirin and then oral circulatory promoter piracetam to reduce blood flow resistance. Life-style changes were also recommended so that she could avoid inducing her symptoms. Stenting or angioplasty would be a last resort if such measures failed^[20, 21]. The following six months was uneventful, so the medication was halted. Although her hearing did not improve (Fig. 1A), the results on the electronystagmogram for optokinetic nystagmus (Fig. 1B) and ACV-cVEMP (Fig. 1C) improved, indicating the brainstem had recovered.

Although our patient's unilateral SD was unlikely caused by vertebrobasilar arterial hypoplasia, her following episodic dizziness was caused by vertebrobasilar arterial hypoplasia related to bilateral central vestibular dysfunction, superimposed on the mildly residual right peripheral vestibular dysfunction. In conclusion, one cannot routinely and thoroughly attribute episodic dizziness subsequent to SD to peripheral vestibular dysfunction, and central vascular anomaly related to central vestibular dysfunction might also be considered.

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一病例報告單側突發性耳聾合併椎基底動脈發育不全

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突發性耳聾發作時常常會伴隨著眩暈，可歸咎於同側前庭迷路炎或前庭神經炎；突發性耳聾發作後常常會出現陣發性的頭暈，可歸咎於同側前庭功能尚未完全恢復。一56歲女性，自從罹患右側突發性耳聾後，屢屢因快速走路或突然站起就會陣發頭暈，病程已經6個月。當她有症狀時，兩耳溫差測試無異常發現，但視運動性眼振和頸部前庭誘發肌性電位檢查顯示有腦幹的障礙，三維飛行時間效應血管磁振造影顯示椎基底動脈發育不全。建議藥物保守治療和生活習慣調整。往後1週，頭暈就很少發作了。往後6個月，情況依然穩定。本個案之右側突發性耳聾很難歸咎於椎基底動脈發育不全，但是，爾後的陣發性頭暈不僅可歸咎於部分的右側週邊前庭功能未恢復，亦可歸咎於椎基底動脈發育不全所致之中樞性前庭功能失調。因此，當遇到突發性耳聾患者合併陣發性頭暈時，我們不能常規將之全然視為週邊性前庭功能失調，也要考慮是否有中樞神經血管變異所致之中樞性前庭功能失調。

關鍵詞：突發性耳聾、陣發性頭暈、椎基底動脈發育不全、換氣過度、三維飛行時間效應血管磁振造影

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