

Pontine Ménière's syndrome – a mimicker of Ménière's disease?

Jiann-Jy Chen, MD, Dem-Lion Chen¹, Hsin-Feng Chang^{2,3}

ABSTRACT - Objectives: In one of 131 patients (48 men and 83 women) diagnosed with Ménière's disease at a neuro-otological clinic over a two-year period, the lesion was localized in the pons rather than the inner ear; herein, we report this rare curiosity. **Case description:** A 25-year-old man presented to our emergency department with vertigo and right tinnitus four times during a 5-year period. The clinical course fitted the diagnostic criteria of definite Ménière's disease and neuroimaging results showed no abnormalities. **Results:** A series of neurotologic studies confirmed right side retrocochlear neural hearing loss, marked depression of the optokinetic nystagmus-slow phase (pursuit) velocity in the leftward direction, and marked depression of the optokinetic nystagmus-fast phase (saccade) velocity in the rightward direction. The symptoms could be attributable to right-sided pontine impairment rather than right inner-ear endolymphatic hydrops. **Conclusion:** Currently, no known vertiginous disease could encode the relapsing symptoms of this patient. The pontine Ménière's syndrome might be a variation of pontine transient ischemia attack with caudal pontine tegmentum syndrome or a pathophysiology similar to that of vestibular migraine.

Key words: Ménière's disease; Ménière's syndrome; neural hearing loss; tinnitus; episodic vertigo

INTRODUCTION

Ménière's disease is diagnosed when there is histopathologic evidence of endolymphatic hydrops. Definite Ménière's disease is confirmed with the following 4 criteria: (a) at least two definitive spontaneous episodes of vertigo persisting for at least 20 minutes; (b) pure tone audiometry documented

hearing loss on at least one occasion; (c) tinnitus or aural fullness in the affected ear; and (d) exclusion

Department of Neurology, Neuro-Medical Scientific Center, Buddhist Tzu Chi General Hospital, Taichung Branch, Taichung, Taiwan

¹G-Home Clinic for Otorhinolaryngology and Neurology, Kaohsiung, Taiwan

²Department of Medical Education, China Medical University Hospital, Taichung, Taiwan

³Faculty of Chinese Medicine, Chinese Medical College, China Medical University, Taichung, Taiwan

Disclosure: This paper has not been presented at any conference or meeting. The authors declare no conflicts of interest.

of other causes (1). Using the criteria above, 131 persons (48 men and 83 women) were diagnosed with Ménière's disease at a neurotological clinic at a regional hospital in northern Taiwan over a two-year period (2008-2010). They all received the same diagnostic battery, including a history and physical examination, audiometric tests (pure tone audiometry, speech reception threshold, speech discrimination score [SDS] test, short increment sensitivity index [SiSi] test), tympanogram, electronystagmogram (saccade, smooth pursuit, optokinetic nystagmus [OKN] test, bithermal caloric test), extracranial and transcranial color-coded duplex sonography and brain magnetic resonance imaging/angiography. In one 25-year-old patient, the lesion was topographically localized in the pons rather than the inner ear, although the clinical course fitted the diagnostic criteria of definite Ménière's disease and neuroimaging results showed no abnormalities. Herein, we report this curious case.

CASE REPORT

A 25-year-old man presented to our emergency department with vertigo and right tinnitus four times over a 5-year period. The symptoms always remitted spontaneously in 3 or 4 hours, and then he felt dizzy with surrounding leftward tilting for several hours. He weighed 90 kg with a height of 175 cm and body mass index of 29.4 kg/m². He did not have hypertension, diabetes mellitus, heart disease, migraine or other systemic disease. He denied drinking alcohol and coffee, smoking, and areca chewing. He denied any familial hereditary disease, except for his mother who had vertigo of unknown etiology. His vital signs, physical and neurological examinations, and mental status were all normal, with the exception of spontaneous horizontal leftward beating nystagmus. His electrocardiogram, blood chemistry, lipid profile and thyroid function tests were all normal.

At presentation in the emergency department with his 4th symptomatic attack, his judgment, orientation, memory, attention, and calculation were all intact. There was no headache, blurred vision, diplopia, dysphonia, aphasia, dysarthria, dysphagia, hemiparesis, hypesthesia/thermanesthesia, dysmetria, abasia or astasia. Extraocular movements and convergence were intact. On straight-ahead gaze, there was spontaneous horizontal leftward beating nystagmus, which was not suppressed by right-, up-, down- or leftward fixation (Fig. 1A). The clinical head-impulse test of the horizontal semicircu-

lar canals was normal. Romberg's test and Mann's test showed rightward tilting. Pure tone audiometry showed mild right high-tone hearing impairment (Fig. 1B). His SDS was 85% in the right ear and 100% in the left ear despite a 0% SiSi (1,000~4,000Hz) in both ears. Testing horizontal and vertical saccades and smooth pursuit eye movements revealed no further deficits. However, electronystagmogram showed marked depression of OKN-slow phase (pursuit) velocity in the leftward direction, and poor manifestation of the OKN-fast phase (saccade) velocity in the rightward direction (Fig. 1C). Bithermal caloric tests (warm [47 °C] and cool [27 °C] water, 20 seconds) demonstrated unilateral weakness of -37.5% (Fig. 1D) using Jongkee's formula, explaining right-sided weakness, and all visual suppressions were positive (Fig. 1D). Furthermore, emergency brain magnetic resonance imaging/angiography and extracranial and transcranial color-coded duplex sonography showed no abnormalities.

During the next three years, he experienced four additional episodes of the symptoms and received symptomatic therapy at our emergency department. During the inter-ictal uneventful period, physical examination, audiometric tests, tympanogram, electronystagmogram and bithermal caloric test results were normal.

DISCUSSION

Ménière's disease is frequently considered in the cases of relapsing unilateral tinnitus and vertigo. The symptoms are sometimes caused by an acoustic tumor or vestibular migraine; the former can be diagnosed by neuroimaging and the latter by a history of migraines (2,3). In addition, rotational vertebral artery syndrome (Bow-Hunter syndrome) should be considered if head rotation induces symptoms (4). According to the above criteria, only Ménière's disease might be considered in our patient because his clinical course fulfilled the diagnostic criteria of definite Ménière's disease, the neuroimaging results were normal, and he did not have migraines.

Speech discrimination loss is proportional to cochlear (sensory) hearing loss, whereas it is disproportionately greater in retrocochlear (neural) hearing loss. Use of an SDS test alone cannot discriminate between cochlear (sensory) and retrocochlear (neural) hearing loss. A recruitment test, such as an SiSi test or alternate binaural loudness balancing test, should also be performed (5). In our patient, mild right high-tone hearing impairment

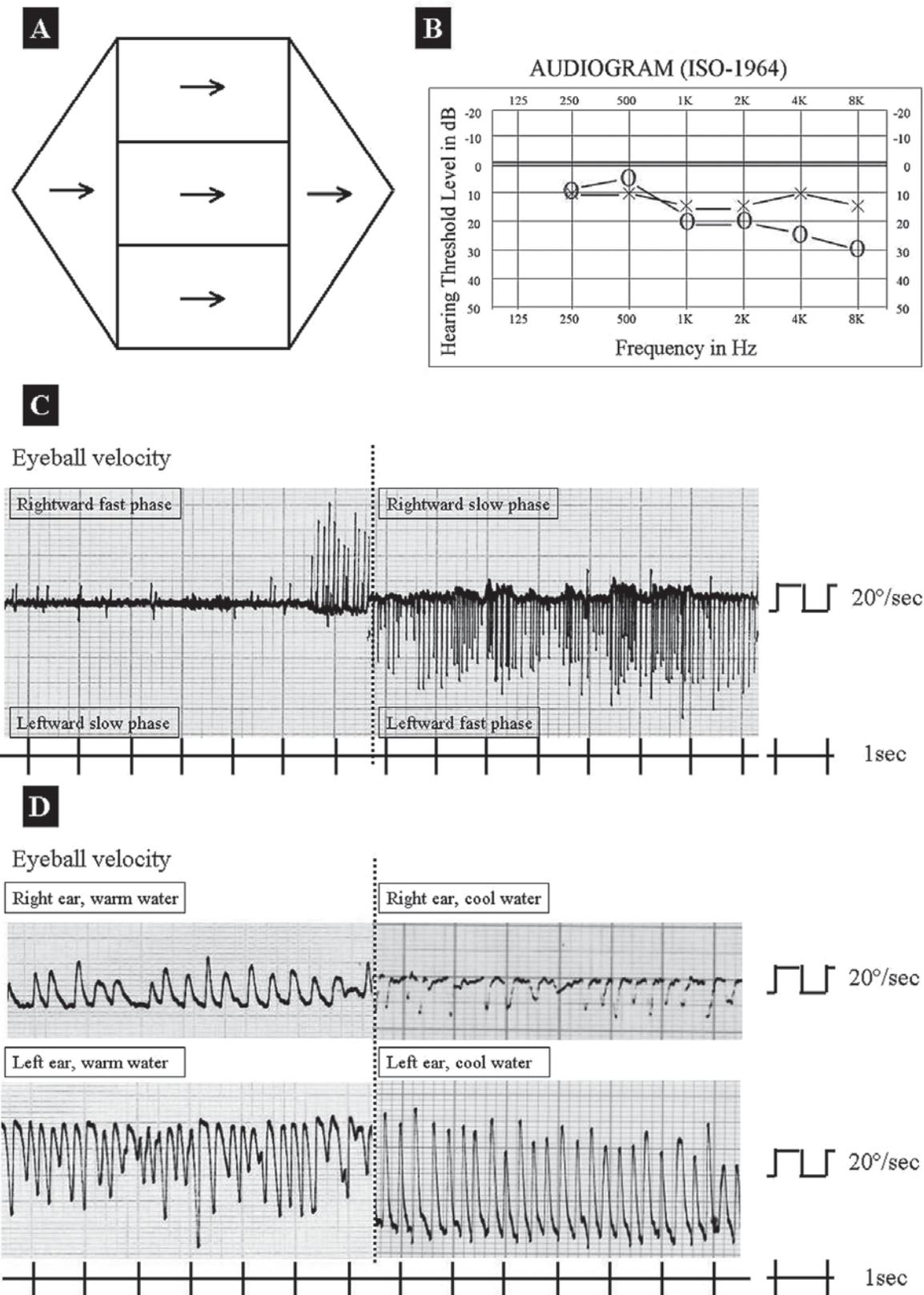


Fig. 1. (A) On straight-ahead gaze, a spontaneous horizontal leftward beating nystagmus is observed, which is not suppressed by right-, up-, down- or leftward fixation; (B) pure tone audiometry shows mild right high-tone hearing impairment; (C) an optokinetic electronystagmogram shows marked depression of the slow phase (pursuit) velocity in the leftward direction and poor manifestation of the fast phase (saccade) velocity in the rightward direction; (D) bithermal caloric tests shows that maximum slow-phase velocities of warm (47 °C) and cool (27 °C) caloric nystagmus in the right ear are both lesser than those in the left ear.

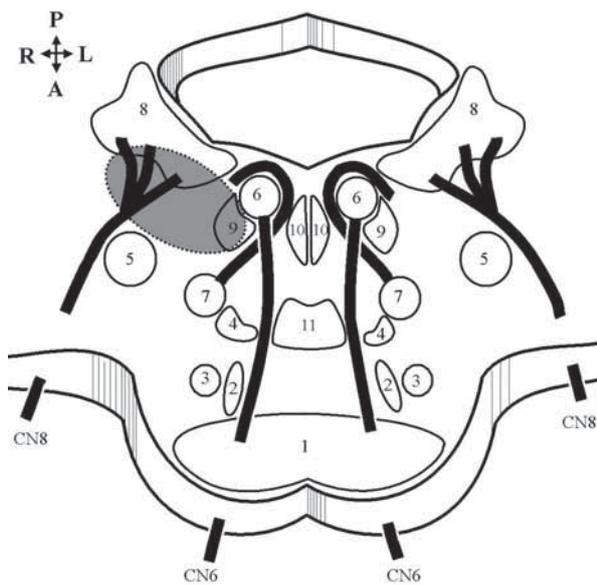


Fig. 2. The symptoms are attributable to a presumed lesion (gray circle) in the right pontomedullary junction, involving the right vestibular nucleus (No. 8), right paramedian pontine reticular formation (No. 9) and right auditory nerve (CN 8). No. 1: corticospinal tract; No. 2: lateral spinothalamic tract; No. 3: rubrospinal tract; No. 4: lateral lemniscus; No. 5: trigeminal spinal tract; No. 6: abducens nucleus; No. 7: facial nucleus; No. 10: medial longitudinal fasciculus; No. 11: trapezoid body; CN 6: abducens nerve and fasciculus.

with a 0% SiSi excluded hearing symptoms of cochlear origin, and the right-sided 85% SDS indicated a retrocochlear (neural) lesion of the right auditory nerve with ipsilateral neural hearing loss when he was symptomatic.

A directional preponderance of the OKN-slow phase (pursuit) velocity was found corresponding to spontaneous nystagmus. Unilateral peripheral vestibular hypofunction results in enhancement of the nystagmus OKN-slow phase (pursuit) velocity toward the side of the lesion and depression of the OKN-slow phase (pursuit) velocity toward the opposite horizontal direction (6). Theoretically, unilateral Ménière's disease with ipsilateral vestibular hyperfunction results in spontaneous nystagmus toward the lesion side, consisting of the fast phase toward the lesion side and slow phase toward the opposite side. Hence, patient with unilateral Ménière's disease should have depression of the OKN-slow phase (pursuit) velocity toward the lesion side and enhancement of the OKN-slow phase (pursuit) velocity toward the opposite horizontal direction. In our patient, right peripheral vestibular hypofunction on bithermal caloric tests and marked depression of the OKN-slow phase (pursuit) veloc-

ity in the leftward direction were explained by right peripheral vestibular hypofunction.

If extraocular movements are intact and there is no double vision, the horizontal pursuit and horizontal saccade tests focus on dysfunction of the ipsilateral paramedian pontine reticular formation (PPRF) (paraabducens nucleus or nucleus praepositus XII). The OKN test magnifies dysfunction of the ipsilateral PPRF if no parietal lobe lesion is found on neuroimaging (7). The bithermal caloric nystagmus test focuses on ipsilateral peripheral vestibular function and the visual suppression test focuses on the ipsilateral cerebellar flocculonodular lobe (8). In our patient, the poor manifestation of the OKN-fast phase (saccade) velocity could be explained by impairment of the right PPRF, representing ipsilateral central vestibulopathy. In addition, because of the leftward spontaneous nystagmus and right peripheral vestibular hypofunction, concomitant right peripheral vestibulopathy was considered. The bilateral positive visual suppression test showed that both cerebellar flocculonodular lobes were normal. Eventually, the episodic symptoms could be attributable to right-sided pontine impairment (Fig. 2) rather than ipsilateral inner-ear endolymphatic hydrops

CONCLUSION

The right retrocochlear (neural) hearing loss, impairments of OKN and ipsilateral peripheral vestibular hypofunction were not compatible with the pathophysiology of Ménière's disease. Besides, pure horizontal nystagmus is usually not found in a peripheral vestibular failure, and might indicate a central origin. Hence, our patient's symptoms were attributable to one-sided pontine impairment despite the normal neuroimaging results. Currently, no known vertiginous disease could encode our patient's relapsing symptoms. The pontine Ménière's syndrome might be a variation of pontine transient ischemia attack with caudal pontine tegmentum syndrome or a pathophysiology similar to that of vestibular migraine (9). The novel vertiginous disease herein might masquerade as Ménière's disease in most patients, with an incidence of 0.8% (1 of 131) among those fitting the diagnostic criteria of definite Ménière's disease.

REFERENCES

1. American Academy of Otolaryngology, Committee on Hearing and Equilibrium. Guidelines for the diagnosis and evaluation of therapy in

- Ménière's disease. *Otolaryngol Head Neck Surg* 1995; 113: 181-5.
2. Lempert T, Olesen J, Furman J *et al.*. Vestibular migraine: diagnostic criteria. *J Vestib Res* 2012; 22: 167-72.
 3. Gorman EF, Bag AK, Palmer CA. Man with posterior fossa tumors 15 years apart. *Brain Pathol* 2012; 22: 117-20.
 4. Chen JJ, Chen DL. Bow Hunter's syndrome masquerading as definite Ménière's disease. *Tzu Chi Med J* 2010; 22: 219-24.
 5. Priede VM, Coles RR. Speech discrimination tests in investigation of sensorineural hearing loss. *J Laryngol Otol* 1976;90: 1081-92.
 6. Szirmai A, Keller B. Electronystagmographic analysis of optokinetic and smooth pursuit eye movement disorders in vestibular lesions. *Int Tinnitus J* 2011; 16: 174-9.
 7. Baehr M, Frotscher M. Cerebellum. In: Baehr M, Frotscher M. Duus' topical diagnosis in neurology, 5th edition. New York: Thieme, 2012, 158-68.
 8. Parker W. ENG analysis of 150 cases of posterior fossa disease. *Laryngoscope* 1977; 87: 575-87.
 9. Teggi R, Fabiano B, Recanati P, Limardo P, Bussi M. Case reports on two patients with episodic vertigo, fluctuating hearing loss and migraine responding to prophylactic drugs for migraine. Ménière's disease or migraine-associated vertigo? *Acta Otorhinolaryngol Ital* 2010; 30: 217.

Address for correspondence: Jiann-Jy Chen, MD. Department of Neurology, Neuro-Medical Scientific Center, Buddhist Tzu Chi General Hospital, Taichung Branch No. 88, Section 1, Fongsing Road, Tanzih District, Taichung City 42743, Taiwan; E-mail: jiannjy@yahoo.com.tw

Ménièreov sindrom na ponsu – oponašatelj Ménièreove bolesti?

SAŽETAK – U jednog od 131 bolesnika (48 muškaraca, 83 žene) kojima je u jednoj neurološkoj klinici u razdoblju od više od dvije godine postavljena dijagnoza Ménièreove bolesti lezija je bila lokalizirana u ponsu, a ne u unutarnjem uhu. Slučaj bolesnika opisujemo zbog rijetkosti. Dvadesetpetogodišnji muškarac došao je u naš hitni odjel 4 puta s vrtoglavicom i tinitusom desnog uha u razdoblju od preko pet godina. Klinički tijek je odgovarao dijagnostičkim kriterijima nedvojbene Ménièreove bolesti, a rezultati neuroslikovne pretrage nisu pokazivali nenormalnosti. Niz neurotoksikoloških studija potvrdio je neuralni gubitak sluha desno retrokohlearno, izraženu depresiju brzine sporofaznog optokinetičkog nistagmusa u smjeru lijevo i izraženu depresiju brzofaznog optokinetičkog nistagmusa (sakadni) u smjeru desno. Simptomi bi se mogli pripisati više desnostranom oštećenju ponsa nego endolimfatičkom hidropsu desnog unutarnjeg uha. Zaključuje se da se danas nijedna poznata vrtoglavica ne bi mogla pripisati ponavljajućim simptomima tog pacijenta. Ménièreov sindrom ponsa mogao bi biti varijacija prolazne ishemijske atake u ponsu s kaudalnim sindromom tegmentuma ponsa ili patofiziološka promjena slična vestibularnoj migreni.

Ključne riječi: Ménièreova bolest, Ménièreov sindrom, neurološki gubitak sluha, tinitus, epizodni vertigo