VESTIBULAR MIGRAINE

Robert W. Baloh, MD
University of California at Los Angeles

Case Illustration. A 37 y/o woman reported recurrent episodes of vertigo dating back to her early 20's (2 to 5 per year). The attacks typically last 3 to 6 hours but it can take several days before she returns to normal. During the attack her ears feel full, the room spins, her balance is off and she usually vomits. Her husband noticed that "her eyes jump back and forth". After one severe bout she developed brief episodes of positional vertigo (seconds) when turning over in bed and extending her head back to look up that recurred over 4 to 5 months. She also describes life-long sensitivity to motion both self and surround motion.

Background. Vestibular migraine (VM), also known as migrainous vertigo or migraine-associated vertigo, is characterized by recurrent vestibular attacks often accompanied by migraine headaches. It is a prevalent presenting complaint to physicians in primary care, otolaryngology, and neurology. Epidemiologic data suggest that VM may affect 1% of the general population and 10% of patients seeking treatment for recurrent attacks of vertigo. Yet, the clinical spectrum of VM and its underlying pathophysiological mechanisms remain unclear, with much debate about the causal relationship of vestibular symptoms and headache, no evidence-based guidelines for clinical management, limited characterization of its disease burden, and no information about its negative impact on health-related quality of life. Currently there is no proven therapy for VM. Vestibular suppressants such as meclizine and dimenhydrinate are often used for symptomatic relief, but they do not prevent attacks and may have sedating side effects. Uncontrolled case reports suggest that migraine abortive and prophylactic medications may be effective for treating VM.

What is vestibular migraine (VM)? The observation that migraine and dizziness often occur together dates back to the nineteenth century. However, only in recent years have vestibular symptoms been recognized as part of the migraine syndrome. Vestibular migraine (VM), also known as migrainous vertigo and migraine-associated vertigo, affects about 1% of the general population and 10% of patients seen in dizziness and headache clinics. Episodic vestibular symptoms occurs in about 25% of unselected migraine patients. Vestibular symptoms can occur during headaches, but often occur during headache-free intervals. Only about one quarter of patients reliably experience headaches during vestibular attacks. Vestibular attacks in patients with VM typically last from minutes to days. If examined during an attack, patients with VM may show spontaneous or positional nystagmus that can have central or peripheral features, raising concerns about other neurotologic illnesses. These factors have made it difficult to characterize the relationship between migraine and vestibular symptoms. As a result, a causal relationship has not been proven. This problem has been used as an argument against the concept of VM, but it is no different than the situation with other transient symptoms that accompany migraine. For example, scintillating scotomata are universally accepted as migrainous phenomena, yet their pathophysiologic link with headache is still unknown after hundreds of years of clinical observation. Conventional migraine and VM share numerous features. Both have a marked female preponderance (2-3:1). Both can be precipitated by alcohol, lack of sleep, fasting, certain foods, and emotional stress. Patients with VM have past or present histories of migraine and often have family histories of both migraine and vestibular symptoms. Finally, case reports and case series suggest that migraine abortive and prophylactic agents, including triptans, may be effective for treating VM.

Neurotologic conditions such as Meniere's disease and benign paroxysmal positional vertigo (BPPV) share some symptoms with VM. Recurrent vertigo without ear symptoms can be the first presentation of Meniere's disease; however, with Meniere's disease, tinnitus and hearing loss invariably occur within a year of onset. Vertigo attacks in BPPV are positional and last no more than 1-2 minutes. Vertigo attacks in VM may have a positional element, but symptoms persist even when patients are still and typically last longer than 2 minutes. VM may coexist with Meniere's disease, BPPV, chronic subjective dizziness, anxiety, and depression. Coexisting conditions may alter its clinical presentation and morbidity. Recent advances in our understanding of the genetics and pathophysiology of migraine provide new hope for elucidating the link between periodic headaches and vestibular symptoms.

The (3rd) edition of International Classification of Headache Disorders (ICHD-III) includes the diagnosis of VM based on consensus criteria developed by a joint committee of the International Headache Society and the Barany Society (Table 1). These criteria define the types of vestibular symptoms; minimum number, severity, and duration of attacks; association of vestibular and migrainous symptoms; requirement for a migraine diagnosis; and absence of other causes of symptoms.

Table 1. Diagnostic criteria for vestibular migraine (ICHD-III)

A	At least 5 episodes of vestibular symptoms[a] of moderate or severe intensity,[b] lasting 5 minutes to 72 hours.
B	Current or previous history of migraine with or without aura according to International Classification of Headache Disorders
C	One or more migraine features with at least 50% of the vestibular episodes.[c]
D	Not better accounted for by another vestibular or ICHD diagnosis.

a. *Vestibular symptoms qualifying for a diagnosis of vestibular migraine include: 1) spontaneous vertigo – false sensation of self motion or that the visual surround is spinning or flowing; 2) positional vertigo, occurring after a change in head position; 3) visually-induced vertigo, triggered by a complex or large moving visual stimulus; 4) head-motion induced vertigo, occurring during head motion; 5) head motion-induced dizziness with nausea, characterized by a sensation of disturbed spatial orientation.*

b. *Moderate, interferes with but does not prohibit daily activities; severe, prevents daily activities.*

c. *Associate migraine features: 1) headache with at least two of the following: unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity; 2) photophobia and phonophobia; 3) visual aura*

What is the public health burden of VM? Neuhauser and colleagues found a 1% prevalence of VM in the general population of Germany and concluded that VM is underdiagnosed, causing considerable personal and health-care burden. At a rate of 10%, VM is the third most common diagnosis in specialty neurotology centers. In our clinical practices, patients with VM have been symptomatic for an average of 4.5 years before referral and have seen 4-5 previous physicians for their symptoms. Most have missed significant work, school, or home obligations.

How is VM treated? Currently there is no proven therapy for VM. Vestibular suppressants such as meclizine and dimenhydrinate are frequently used for symptomatic relief of vestibular symptoms, but these medications do not prevent vestibular attacks and often have sedating side effects. Uncontrolled case reports suggest that commonly used migraine abortive and prophylactic drugs may be effective for treating vestibular attacks in patients with VM, but high placebo effects are well documented in controlled treatment trials for migraine headaches.

Future Directions. Now that diagnostic criteria for VM have been established (Table1) it is critical that these criteria be evaluated in controlled studies. Likely modification will be required as experience is developed with the criteria. For example, can patients have VM if they do not have migraine headaches? In the study described above the majority of patients with benign recurrent vertigo had a personal history of migraine. However, even those who did not have such a history occasionally experienced migraine features during some or all of their vertigo attacks. Moreover, they had a similar age of onset and duration of vertigo attacks as those with migraine and they usually had a family history of migraine. This raises the question of whether there is a fundamental difference between patients with recurrent vertigo with or without a history of migraine. Because there was a significantly large group of patients who experienced both vertigo spells with migraine features and vertigo spells without, we suspect that there may not be a fundamental difference between these two types of patients.

We know relatively little about the pathophysiology of vertigo and other vestibular symptoms with migraine. As noted above the serotonin system has been implicated in the production of both headaches and vertigo. Vasospasm could result in ischemia of the inner ear and brainstem and cerebellar pathways. Numerous studies over the years have documented a major genetic component in the pathogenesis of migraine. Recent identification of the genes for a rare subtype of migraine, hemiplegic migraine, may provide the first true molecular insight into the pathophysiology of migraine. Within families with these relatively rare mutations some members

have just headaches or vertigo suggesting that these more common migraine features have a similar pathophysiologic mechanism. Mutations in the calcium channel gene, CACNA1A, cause hemiplegic migraine and familial episodic ataxia, a disorder with episodic vertigo with features similar to VM.

Clearly, there is a need for controlled studies of potential therapies for VM. We are currently conducting a multicenter clinical trial of rizatriptan for aborting episodes of VM. Abortive and prophylactic therapies are needed for vestibular migraine (VM), but existing knowledge favors an abortive trial right now. The target of abortive therapy (discrete attacks) is clearer; available data implicate migrainous processes in discrete symptoms; case reports suggest that triptans may be effective; and comorbid conditions or co-administered medications are less confounding. In contrast, knowledge gaps make design of a prophylactic trial rather murky.

REFERENCES

Baloh RW, Kerber KA. Clinical Neurophysiology of the Vestibular System, 4th edition. New York, Oxford University Press, 2012.

Cha YH, Lee H, Santell LS, Baloh RW. Association of benign recurrent vertigo and migraine in 208 patients. Cephalalgia 2009;29:550-555.

Goadsby PJ, Lipton RB, Ferrari MD. Migraine--current understanding and treatment. N Engl J Med 2002;346:257-270.

Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalgia 2013;33(9):629-808.

Kayan A, Hood JD. Neuro-otological manifestations of migraine. Brain 1984;107 (Pt 4):1123-1142.

Neuhauser H, Leopold M, von Brevern M, Arnold G, Lempert T. The interrelations of migraine, vertigo, and migrainous vertigo. Neurology 2001;56:436-441.

Oh AK, Lee H, Jen JC, Corona S, Jacobson KM, Baloh RW. Familial benign recurrent vertigo. Am J Med Genet 2001;100:287-291.

von Brevern M, Zeise D, Neuhauser H, Clarke AH, Lempert T. Acute migrainous vertigo: clinical and oculographic findings. Brain 2005;128:365-374.