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# Apo- and Geotropic Horizontal Nystagmus Mimicking Benign Paroxysmal Positional Vertigo in Vestibular Migraine Patients: The Clinical Cases

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#### **ABSTRACT**

Symptoms of vestibular migraine (VM) are often indistinguishable from vestibular symptoms caused by other pathologies, such as benign paroxysmal positional vertigo (BPPV). Episodic positional vertigo is typical for both disorders. We observed two cases of apo- and geotropic positional nystagmus mimicking BPPV in patients previously diagnosed as having migraine (MIG). Positional nystagmus detected in our patients was not typical for horizontal canal BPPV (canalolithiasis/cupulolithiasis): it had low velocity, persisted as long as the provoking position was maintained, had no crescendo-decrescendo pattern and nearly symmetrical intensity on both sides. Multiple canalith repositioning maneuvers failed, although pseudo-BPPV incidences disappeared two weeks after starting intensive anti-MIG treatment. Previous history of migraine, accompanying migrainous features (One-sided headache, long duration, associated with nausea and/or vomiting, photophobia and phonophobia), nystagmus characteristics, ineffective response to positional maneuvers, contrary, favorable response to anti-MIG medications, make it much easier to differentiate these two disorders: VM and BPPV. Keywords: Apo- and geotropic positional nystagmus; Benign paroxysmal positional vertigo (BPPV); Dizziness; Vertigo; Vestibular migraine.

#### INTRODUCTION

estibular migraine (VM) is a disease established as a new medical entity at the beginning of the 21<sup>st</sup> century. However, the link between migraine (MIG) and vertigo was recognized by some neurologists in the 19th century. VM is the typical cause of spontaneous recurrent episodic vertigo, accounting for approximately 10% of patients with vertigo and dizziness.

The diagnosis of VM is often complicated because it does not have specific characteristic diagnostic signs. The diagnosis of VM mainly relies on clinical history and exclusion methods. The duration of attacks may vary from minutes to days. The VM clinic is diverse; patients report a broad spectrum of complaints during an attack. Subjective symptoms essential for diagnosing VM include vertigo and head motion-induced dizziness.<sup>1</sup> During VM attacks, patients frequently report subjective hearing loss in one or both ears, tinnitus, occipital pressure, neck stiffness, brain fog, feeling of tension, headmotion intolerance and/or cranial autonomic symptoms. Sometimes, the auditory complaints fluctuate, leading to the false diagnosis of Meniere's disease.

Different types of nystagmus should be observed in VM patients: position-dependent nystagmus and pathological spontaneous nystagmus. They can be detected during the ictal period and between the attacks of VM, although they appear less frequently in quiet periods.<sup>4,5</sup> Occasionally, recurrent episodes of positionally induced, short-lasting vertigo attacks imitate true BPPV. The diagnosis relies upon the distinctive

pattern of nystagmus observed during provocative maneuvers in the plane of the affected canal.

#### **CASES**

Case 1: Positional nystagmus in VM patient mimicking geotropic horizontal positioning nystagmus

A 35-year-old female was evaluated for positional vertigo, having a five-year history of MIG without aura associated with nausea, vomiting, light, and sound sensitivity. She complained about motion sickness since childhood and visual discomfort with looking at moving objects. During the attacks, she reported episodes of positional vertigo, provoked by turning her head side to side, that awakened her from sleep. She was feeling "drunk" all the time and was not able to drive or tolerate physical activity. She had the sensation of pressure in both ears without hearing loss.

The patient had been investigated during one of the ictal periods. No nystagmus was present with or without fixation, and no nystagmus occurred with hyperventilation or mastoid vibration. Gaze-holding, saccade accuracy, smooth pursuit, and optokinetic test results were also normal. The Video Head Impulse Test (vHIT) showed symmetrical canal gains without refixation saccades. Brain MRI did not reveal any abnormalities.

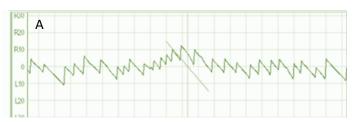
The Dix-Hallpike test on both sides did not provoke any nystagmus, and no nystagmus was observed in the supine position. Geotropic horizontal nystagmus was seen in the siderolling test. It was slightly more intensive on the right side than



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on the left. The nystagmus persisted as long as the position was maintained; it had no latency, fatigability, or associated vertigo sensation. Unlike lateral canalithiasis, the nystagmus did not have a crescendo-decrescendo slow phase velocity (SPV) profile (Fig.1).

FIGURE 1. Geotropic horizontal nystagmus was recorded with videonystagmography in a vestibular migraine patient



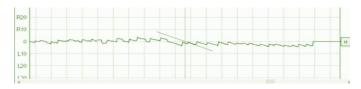


Explanations: A. The right-beating nystagmus registered during the roll test on the right side. SPV of nystagmus - 12°/s; B. The left-beating nystagmus registered during the roll test on the left side, SPV of nystagmus - 8°/s.

# Case 2: Positional nystagmus in VM patient mimicking apogeotropic horizontal positioning nystagmus

A 59-year-old male was evaluated for positional vertigo during the ictal period of MIG. Since his teenage years, he was diagnosed with MIG without aura, while since childhood, he suffered from motion sickness. In the past, he was successfully operated for an intestinal tumor and had no further complaints.

One day before the vestibular investigation, he began to feel acute vertigo, with nausea, vomiting, and neck pain. Spontaneous right-beating pure horizontal nystagmus was present without fixation; the direction and intensity of nystagmus were not changed during the horizontal headshaking test (Fig.2).



During the supine roll test, apogeotropic horizontal nystagmus was observed in the head-right and head-left positions; it was more intense in the head-left position (Fig. 3). There was no latency, fatigability, or associated vertigo, and

the nystagmus persisted as long as the position was maintained. An MRI was proposed and conducted the next day, and unremarkable results associated with vertigo were revealed.

FIGURE 3. Apogeotropic horizontal nystagmus elicited during right and left roll tests in vestibular migraine patient





Explanations: A. The left-beating nystagmus registered during the roll test on the right side, SPV of nystagmus -  $7^{\circ}$ /s; B. The right-beating nystagmus registered during the roll test on the left side, SPV of nystagmus -  $10^{\circ}$ /s.

In both patients, despite the different nystagmus characteristics, repositioning maneuvers were performed several times to no avail. The patients responded excellently to Flunarizine (5-10 mg/d) and Clonazepam (0.5-1 mg/d) therapy. In both patients, positional vertigo and nystagmus disappeared within two weeks after initiating the therapy.

#### **DISCUSSION**

BPPV, provoked by displaced otoconia from the otolith maculae, is the common cause of episodic, transient vertigo. As the head moves concerning gravity, the otoconia also moves, activating semicircular canal afferents and producing a false sense of head rotation and nystagmus. In the majority of cases, BPPV is idiopathic but may be provoked by head trauma or other ear pathologies.<sup>6</sup>

There are two main types of BPPV: (i) canalithiasis, when otoconia freely move within the semicircular canal-duct, and (ii) cupulolithiasis, when otoconia are attached to the cupula. The stimulus is transient in canalithiasis and sustained in cupulolithiasis.<sup>7,8</sup>

VM is considered the most common cause of episodic vertigo in adults and children. About 67% of the patients with VM reported spontaneous rotational vertigo, whereas 24% had positional one.<sup>9</sup> Recurrent episodic positional vertigo in VM patients may frequently be confused with BPPV.<sup>10</sup> Several previous authors reported that horizontal canal BPPV (HC-BPPV) had the highest prevalence in patients with acute VM.<sup>11</sup>

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In some cases, BPPV and VM presentations are similar, as both can be presented by similar positional nystagmus. 12

Studies have demonstrated a strong correlation between BPPV and migraines, with a threefold increased risk of developing BPPV in patients with migraine compared to nonmigraine individuals.<sup>13</sup>

The most frequently found objective vestibular signs in VM patients are geotropic and apogeotropic nystagmus, further compounding diagnostic confusion. Distinguishing HC-BPPV from VM is rather important since the treatment for either condition is very different.<sup>14</sup> The presence of MIG features such as headache, photophobia, phonophobia, and visual aura; moreover, well tolerated to anti/preventive MIG therapy supports the diagnosis of VM. However, it should be noted that there is also a high prevalence of MIG amongst those with benign positional vertigo, and the two conditions may cooccur.15

VM commonly affects females. The peak incidence of the attacks is about 40 years of age. Some patients are free from MIG attacks for years when VM first manifests itself. 16 Not infrequently, MIG headaches are replaced by vertigo attacks in women around menopause. As with migraine, the diagnosis of VM is based only on the disease history and the rule out of other medical disorders. The International Headache Society and the Bárány Society have recently collaborated to develop diagnostic criteria for VM (Tab.1).1,17,18

TABLE 1. Diagnostic criteria for vestibular migraine proposed by the International Headache Society Classification Committee and the Committee for Classification of Vestibular Disorders of the Barany Society<sup>1</sup>

- At least 5 episodes with vestibular symptoms of moderate or severe A. intensity, lasting 5 min to 72 hours;
- Current or previous history of migraine with or without aura according to the International Classification of Headache Disorders (ICHD-3);
- C. One or more migraine features with at least 50% of the vestibular episodes:
  - headache with at least two of the following characteristics: one sided location, pulsating quality, moderate or severe pain intensity, aggravation by routine physical activity;
  - photophobia and phonophobia;
  - visual aura.
- Not better accounted for by another vestibular or ICHD diagnosis

The diagnosis of vestibular migraine (VM) relies on the history of migraine, recurrent vestibular symptoms, the temporal association between vestibular and motor neuron disease (MIG) symptoms, and the rule out of other causes of vestibular symptoms. A diagnosis of VM can be made based on moderate to severe vertigo and head motion-induced dizziness with nausea. Acute episodes can last from five minutes to seventy-two hours.1

The clinical presentation of VM is quite different. Various types of vertigo may occur during one attack of the VM. The accompanying nystagmus can be vertical, horizontal, or torsional. It tends to be without latency, low velocity  $(2-7^{\circ}/s)$ , persistent and sometimes appears in a non-expected plane. 11

Symptoms of VM are often indistinguishable from BPPV. Horizontal positional nystagmus may indicate posterior fossa lesions without other neurological signs. 19 Horizontal geotropic positional nystagmus has been described in acute VM and central cerebellar peduncle and lateral medulla lesions. This nystagmus is persistent rather than paroxysmal, making it easy to separate from canalithiasis. Horizontal apogeotropic nystagmus has been reported in central lesions, typically of the cerebellar vermis and modulus, VM, and more rarely in Ménière's disease and vestibular schwannomas. 20,21 The central type apogeotropic nystagmus can closely resemble cupulolithiasis. Differential diagnosis may rely on the presence of other neurological or aural signs and the response to repositioning maneuvers. Comparing the intensity of sitting and supine nystagmus helps differentiate the peripheral vestibular pathologies from central ones. More intense nystagmus in a supine position compared to sitting is typical for HC-BPPV cases, while a symmetrical response between sitting and supine is more characteristic of central lesions.<sup>20</sup>

We observed two cases with geotropic and apogeotropic horizontal nystagmus during VM attacks. Positional nystagmus patients was not typical of HC-BPPV (canalolithiasis/cupulolithiasis). With the supine roll test, the patient had low velocity, slightly asymmetric geotropic NY in one case, and apogeotropic NY in another, which persisted as long as the provoking position was maintained.

The typical nystagmus of HC-BPPV is characterized by a crescendo-decrescendo pattern, duration less than 60 seconds (longer in cupulolithiasis), fatigability, and an asymmetric intensity, which helps to identify the affected ear. Horizontal geotropic nystagmus of VM can be separated from lateral canalithiasis by its low velocity, persistent, and symmetrical nature, while horizontal apogeotropic nystagmus can closely resemble that of lateral cupulolithiasis. Differentiating between VM and BPPV was made more accessible by atypical positional nystagmus, which included migrainous symptoms during episodes and positional vertigo that was well tolerated by anti-MIG medication.

### **CONCLUSIONS**

We presented two cases of geotropic and apogeotropic horizontal nystagmus mimicking HC-BPPV during VM attacks. Nystagmus characteristics found in performing supine roll tests: low velocity, persisted as long as the provoking position was maintained, no crescendo-decrescendo pattern, and nearly symmetrical intensity on both sides, differed from HC-BPPV. Multiple canalith repositioning maneuvers failed, although, 2 weeks from starting intensive anti-MIG treatment, pseudo-BPPV features disappeared.

Sustained nystagmus with positional testing in young to middle-aged patients with accompanying or previous history of migrainous symptoms (One-sided headache, long duration, associated with nausea and/or vomiting, sensitivity to light and

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sound or to touch) Ineffectiveness of repositioning maneuvers, and contrary, effective response to anti-MIG medications, help to distinguish BPPV from VM.

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