Clinical Reasoning: Labyrinthine hemorrhage

An unusual etiology for peripheral vertigo

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SECTION 1

A 65-year-old man presented with a 2-week history of worsening vertigo. The sensation of vertigo was constant and exacerbated by turning in bed, sitting, and standing. Exacerbations lasted 5–10 minutes and were intense enough to cause gait impairment. Two days prior to presentation, the vertigo had worsened in severity and was associated with one episode of nausea and vomiting. Review of systems was positive for a 3-month history of a whistling noise in the patient's left ear, a 2-week history of acute-on-chronic left-sided hearing impairment, and a recent upper respiratory infection.

The patient's medical history was notable for a left radical mastoidectomy performed 14 years prior to presentation for treatment of complications of chronic otitis media, which resulted in chronic left-sided hearing impairment. Medical history also included hepatitis C and remote alcohol and heroin abuse.

Questions for consideration:

1. What is the differential diagnosis for vertigo?
2. How do we clinically differentiate the peripheral causes of vertigo from central causes of vertigo?

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SECTION 2

The differential diagnoses of vertigo can encompass several etiologies (table e-1 at Neurology.org).¹⁻³ The priority in the neurologist's evaluation of vertigo is to distinguish peripheral from central causes. Peripheral vertigo indicates damage to the labyrinth, which consists of the cochlea and the vestibular system (semicircular canals, vestibule, vestibular nerve, and vestibular nuclei). Central vertigo indicates a lesion of the CNS, typically involving the cerebellum, brainstem, or their association pathways. A bedside examination, specifically evaluating nystagmus, ability to stand, and associated neurologic signs, can help distinguish peripheral causes of vertigo from central causes.⁴⁻⁷

A common cause of peripheral vertigo is benign paroxysmal positional vertigo (BPPV), which arises from free-floating otoconia that settle in a semicircular canal. Their shift during movement causes the cupula to create disconjugate stimuli that evoke vertigo. BPPV is characterized by paroxysmal nystagmus triggered by positional maneuvers. The Dix-Hallpike test is used to evaluate posterior canal BPPV. It is performed by rapidly moving the patient from an upright to a supine position with the head extended below the shoulders and turned 45° toward one side. The maneuver is then repeated with the head turned toward the opposite side. A positive result is a latent, brief (<30 seconds), combined torsional and vertical nystagmus when the head is turned to the affected side.⁴

A unilateral destructive labyrinthine lesion causes unopposed tonic vestibular input from the opposite side of the lesion, which affects the vestibulo-ocular reflex. This results in a combination of horizontal, torsional, and vertical nystagmus, with the horizontal component dominating as bilaterally intact vertical canals and otoliths balance and mitigate the vertical and torsional components.⁴ In contrast to BPPV, a destructive labyrinthine lesion leads to a constant, spontaneous, unidirectional nystagmus, with fast beats toward the same (unaffected) side regardless of gaze direction. A head impulse test can detect

labyrinthine dysfunction. It is performed by asking the patient to fixate on a reference point while the examiner rapidly moves the patient's head laterally to each side. Fixation during head rotation is impaired by the loss of vestibular input. Therefore, a positive head impulse test results in a corrective saccade to the reference point when the head is turned toward the affected ear.

Central vertigo can be characterized by purely torsional, pendular, or vertical nystagmus, and bidirectional gaze-evoked nystagmus.^{2,4,5} In addition, spontaneous nystagmus from central lesions is often persistent, with lack of suppression on fixation, and may continue for weeks to months.⁵ Alternate eye cover testing can uncover ocular vertical misalignment, or skew deviation, which has been linked to lateral medulla and pontine ischemic strokes.⁶ Impaired balance can be a feature of peripheral vertigo; however, the inability to stand or take a single step without fall is indicative of central vertigo.⁵ Associated symptoms can also help distinguish peripheral from central causes of vertigo.²⁻⁴ Diplopia, dysarthria, focal sensory, or strength deficits suggest a central etiology. Hearing loss, tinnitus, nausea, and vomiting suggest a peripheral etiology.

This patient's neurologic examination was notable for a subtle spontaneous rightward horizontal nystagmus with a torsional component on primary gaze. Fast beats were maintained toward the right side regardless of gaze direction. Hearing in the left ear was found to be significantly impaired. The patient was able to stand without impairment and demonstrated a steady gait. These features suggested a peripheral cause of vertigo. Vestibular neuritis was initially suspected given the patient's recent upper respiratory tract infection.

Questions for consideration:

1. Is there a role for neuroimaging in peripheral vertigo?
2. What other workup could be useful?

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SECTION 3

Neuroimaging, such as high-resolution brain MRI with special sequences evaluating the internal auditory canal, central brainstem nuclei, temporal bones, and auditory pathways, can be useful in supporting a suspected pathology of peripheral vertigo.^{7,8} For example, normal labyrinth contains perilymph and endolymph that is isointense with CSF on T1- and T2-weighted sequences.⁸ Hyperintensity in the labyrinth on unenhanced T1-weighted images can suggest labyrinthine hemorrhage.⁸ Meanwhile, loss of signal in unenhanced T2-weighted images could indicate filling with inflammatory cells, as in labyrinthitis, or buildup of intercellular methemoglobin and deoxyhemoglobin within erythrocytes, as in acute labyrinthine hemorrhage.

Audiometric evaluation is often useful in distinguishing between causes of peripheral vertigo and should be performed in cases of suspected hearing loss.⁹ Hearing loss is typically absent in vestibular neuritis, while it is present in conditions of labyrinthitis, Ménière disease, and internal auditory canal lesions. Specifically, patients with labyrinthine hemorrhage will demonstrate sensorineural hearing loss in addition to vestibular symptoms.⁸

High-resolution MRI with and without contrast, with dedicated images through the temporal bones, was performed on this patient to rule out other pathologies. Precontrast imaging demonstrated diminished CSF flow signal (figure 1) and an increased T1 signal

Figure 1 Diminished CSF flow signal in the vestibular system of the patient with labyrinthine hemorrhage

An axial multiplanar reconstruction noncontrast T2 DRIVE image through temporal bones shows diminished CSF flow signal in the left cochlea, vestibule, and semicircular canals.

in the left labyrinth (figure e-1). Audiometric evaluation showed profound mixed pure tone loss. These findings were suggestive of labyrinthine hemorrhage. Coagulopathy secondary to hepatic dysfunction was suspected to contribute to the etiology of labyrinthine hemorrhage in this patient.

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SECTION 4

Question for consideration:

1. What are the common therapeutic options for management of peripheral vertigo?

Management of peripheral vertigo can be symptomatic, specific, and rehabilitative.¹ Symptomatic relief can be accomplished by sedation with benzodiazepines, or by suppressing the amount of acetylcholine available for vestibular function with anticholinergics or antihistamines, such as meclizine, promethazine, or diphenhydramine.^{1,2}

Specific treatment, such as diuretics along with dietary salt restriction, is used to prevent the expansion of endolymphatic volume in Ménière disease.^{1,2} Antibiotics have a role in the treatment of bacterial labyrinthitis and otitis media-induced vertigo.² Glucocorticoid therapy has been employed in inflammation-induced vestibular dysfunction, vestibular neuritis, and Ménière disease.^{1,2} BPPV is frequently treated with canalith repositioning maneuvers.² Invasive procedures, such as chemical ablation with transtympanic gentamycin, have been used in Ménière disease, and surgical approaches have been used to treat severe perilymphatic fistulas and cholesteatomas, acoustic neuromas, and refractory Ménière disease.²

In cases of permanent vestibular injury, vestibular rehabilitation can provide compensatory skills. Rehabilitation can start after acute symptoms subside. Exercises necessarily evoke vertigo; therefore, vestibular suppressants should be avoided during rehabilitation.¹

Our patient was treated with lorazepam as needed, with symptomatic improvement of vertigo. He also completed a 7-day prednisone taper for management of sensorineural hearing loss. A 2-month audiometric re-evaluation showed improvement in both air and bone conduction responses, and the patient subjectively noted a return to baseline hearing.

DISCUSSION Labyrinthine hemorrhage is a rare cause of peripheral vertigo, with few cases described in the literature. Etiologies for spontaneous labyrinthine hemorrhages have included blood disorders, most commonly leukemia, or drug-induced coagulopathies.^{8,10} Further investigation of other etiologies may be affected by the limited ability for confirmatory pathologic studies.⁸ Given its rarity, there is no standard treatment for labyrinthine hemorrhage, although vertigo can be managed symptomatically, while the sensorineural hearing loss may be managed with corticosteroids. Prognosis varies, but permanent, severe impairment of both cochlear and vestibular function is possible. In described cases, vertigo, as well as sensorineural hearing loss, is a prominent characteristic.⁸

The evaluation of vertigo begins with a structured approach to distinguish peripheral from central causes of vertigo. Specific features of the medical history, including recent infections, prior surgeries, or trauma, should be obtained. Several algorithms have been developed to systematically assess physical features of vertigo. The bedside head impulse testing, nystagmus assessment, and test of skew deviation (HINTS) examination systematically assesses oculomotor findings that help distinguish peripheral and central causes of acute vestibular syndrome.⁶ A positive head impulse test, unilateral fast-phase nystagmus, and no skew deviation suggest a peripheral etiology. A prospective cross-sectional study by Katatah et al.⁶ showed that a benign HINTS examination within the first 24–48 hours of symptom onset is 96% specific in ruling out stroke. Meanwhile, the presence of any one of the following features is sensitive in identifying stroke as the cause of acute vestibular syndrome: a head impulse test without a corrective saccade, nystagmus that alternates direction in the fast phase, or positive skew deviation.⁶ A video demonstrating the HINTS examination can be found at content.lib.utah.edu/cdm/ref/collection/ehsl-dent/id/6.¹¹ Diagnostic tests, such as brain MRI with special sequences and audiometric evaluation, are also useful, especially in the setting of hearing loss. Labyrinthine hemorrhage is rare, but should be included in the differential diagnoses of peripheral vertigo, especially in the setting of dual sensorineural hearing loss and vestibular symptoms.

AUTHOR CONTRIBUTIONS

Simy K. Parikh: study concept and design, acquisition of data, analysis and interpretation, critical revision of the manuscript for important intellectual content. Samuel A. Frank: responsible for care, diagnosis, and treatment of the patient, critical revision of the manuscript for important intellectual content, study supervision.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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Neurology 2017;88:e14-e18

DOI 10.1212/WNL.0000000000003481

This information is current as of January 9, 2017

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