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Vestibular Neuritis and Labyrinthitis:

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SUMMARY

Vestibular Neuritis (or neuronitis) is a vestibular condition that is commonly caused by the inflammation of the vestibular branch of the 8th cranial nerve, which is responsible for carrying the signals of the balance organs of the inner ear (commonly known as vestibular labyrinth). Those signals allow the eyes and the head to move in a synchronous way, which is a major component of dynamic balance. The Bárány Society criteria from 2022 suggested the use of the term Acute Unilateral Vestibulopathy (AUVP) since causes other than inflammation can sometimes result in similar symptoms. AUVP and Vestibular Neuritis can be used interchangeably. It is one of the most common causes of vertigo due to an inner ear dysfunction and typically occurs between the ages of 30 and 60.[1,2]

The symptoms of neuritis can be severely debilitating and may be present in both an acute and chronic phase, leading to spinning or non-spinning vertigo, nystagmus, nausea, and vomiting without associated hearing loss.[3]

By definition, vertigo is an illusion of movement and, in cases of neuritis, it is typically a spinning movement, but non-spinning vertigo can also occur. Chronic symptoms typically continue if there is severe or irreversible damage to the affected nerve and mainly consist of imbalance, especially on irregular surfaces or in poorly lit environments. Patients also complain of dizziness related to head movement in some cases. This is due to the eyes not being able to follow head movement resulting from weakness of the vestibular nerve caused by the inflammation. Although there are no curative

treatments currently, supportive treatment should be initiated to target symptoms and hasten recovery.[4]

Labyrinthitis is caused by more extensive inflammation, typically involving both branches of the cochleovestibular nerve. Thus, patients may present with vertigo paired with hearing loss. Vestibular specialists agree that neuritis and labyrinthitis are not interchangeable terms. If no hearing loss is present, the condition should not be called labyrinthitis.

Vestibular neuritis is most likely related to a viral etiology, similar to Bell's palsy and some types of sudden sensorineural hearing loss. So far this has not been definitively proven but there are a lot of studies that provide evidence toward that common teaching.

Recent studies implicate Herpes Simplex Virus (HSV) to be the most common cause of vestibular neuritis.[5,6] A reactivation of the herpes zoster virus (shingles) is also a potential culprit. Although it usually comes with skin blisters either in the ear canal or the face, it can also present without any rash.[7]

Other viruses that may lead to vestibular neuritis include EBV, polio, mumps, measles, hepatitis, and influenza viruses. Patients are not contagious since the virus is not being shed, but rather the virus is reactivated deep in the body at the level of the balance nerve.

Bacterial infections and autoimmune conditions are infrequent causes of inflammation of the inner ear and should also be investigated. A disruption of blood flow through the anterior/superior vestibular artery (a tiny artery that nourishes the inner ear) can also lead to similar symptoms (see below for Differential Diagnosis).[2,8,9]

It is rare for a stroke to present with an isolated vertigo episode as the only symptom. However, a blood flow interruption to the vessel that nourishes the inner ear can cause the same symptoms as vestibular neuritis or labyrinthitis. This should be considered in individuals who are at high risk for stroke and an appropriate evaluation should be done (thorough clinical exam with or without imaging) before retaining the diagnosis of neuritis.

CAUSE
Infection or inflammation of the cochleovestibular nerve.

ANATOMY OF THE INNER EAR

The inner ear consists of the labyrinth that can be separated into the cochlea, and the vestibular labyrinth which encompasses three semicircular canals, the utricle and the saccule.

The cochlea is the major organ for transduction of sound from the inner ear to the brain. The utricle and saccule relay linear acceleration of the head through the movement of stones (otoliths). The utricle detects horizontal motion such as the movement of a car on a straight road, while the saccule detects vertical movement such as the movement in an elevator, including acceleration and deceleration during such movement.

In contrast, semicircular canals detect rotational acceleration of the head. These structures are crucial in maintaining orientation and position of the head. Nerve signals sent from these organs to

the brain bilaterally are used to interpret and maintain patient balance. They allow the eyes to follow head movements in a seamless way. This permits us to keep our eyes on target when we are moving, hence contributing to our dynamic balance.

Neuritis/Neuronitis occurs, specifically, when the vestibular portion of the labyrinth is inflamed. Inflammation leads to balance issues without involvement of the ability to hear.

Labyrinthitis occurs when the entire labyrinth is inflamed or both branches of the cochleovestibular nerve is affected, and results in vertigo, nausea, vomiting and decreased hearing. However, pathophysiology is similar between these two conditions.

VIRAL INFECTION

Viral etiologies are considered the primary cause of vestibular neuritis. Given the dormancy of many of the HSV viruses and HZV viruses in neural ganglia, this group of viruses is considered the most common cause.[6] It is not uncommon for symptoms to occur soon after a systemic viral infection due to decreased immune function during this time. When a viral etiology is suspected, patients usually suffer from single-sided symptoms. Although latent viruses are considered the leading cause of vestibular neuritis, recurrent symptoms are extremely rare and warrant thorough workup for a possible central vestibular lesion.[3] In addition, recurrent symptoms suggestive of neuronitis should warrant suspicion of disorders such as vestibular migraine or Meniere's disease.

There is currently no clear-cut evidence that confirm that the Covid-19 virus or the Covid-19 vaccine cause neuronitis, labyrinthitis, or sudden hearing loss, although it has been previously reported.[10-12]

BACTERIAL INFECTION

We see bacterial infection more commonly with labyrinthitis.

There are two types of labyrinthitis: serous and suppurative. The former is more frequent and occurs in a setting of an otitis media (middle ear infection), whether acute or chronic. The inflammatory material can diffuse from the middle ear to the inner ear through the round window and oval window areas of the middle ear. The inflammation leads to disruption of the function of the cochlea and vestibular labyrinth but is subtle and milder than the other form. In suppurative labyrinthitis, the bacteria spreads into the fluid of the inner ear from a middle ear infection or from meningitis through cerebrospinal fluid. This typically occurs through natural openings of the inner ear such as the cochlear aqueduct or the internal auditory canal. Of note, a middle ear infection can undergo hematogenous spread (through blood) to inflame the meninges and cause meningitis. In suppurative labyrinthitis, the presentation is much more severe, and the risk of significant profound hearing loss is high.

SYMPTOMATIC PRESENTATION

About 25% of patients might experience prodromal symptoms of oscillating dizziness within two days prior to the onset of severe vertigo.[2,13,14]

This prodromal period can help guide clinicians towards vestibular neuritis over vertebrobasilar transient ischemic attack.[3] Upon visually fixating on a target, symptoms may decrease in severity. After onset of symptoms, patients experience acute prolonged vestibular symptoms with spinning or non spinning vertigo of moderate to severe intensity lasting at

least 24 hours. The vertigo is “continuous, persisting at rest and exacerbated by head or body movement.”[2]

On a physical exam, patients typically have a unidirectional horizontal-torsional nystagmus that typically beats away from the affected side (check this YouTube video for an example). Patients will also often complain of Oscillopsia (check this YouTube video for an example). Patients tend to fall towards the side of the lesion.[2]

CHRONIC PHASE

The acute phase symptoms typically resolve after a few days to weeks.[15] A minority of patients may also experience irreversible destruction of the cochleovestibular nerve. The chronic symptoms mostly consist of imbalance and short-lived episodes of disorientation with head movement related to the dysfunction of the vestibular ocular reflex. Patients with chronic symptoms experience significant decrease in their daily quality of life and often become frustrated regarding their symptoms. Given that onset of vertigo can happen at any time, patients become hesitant to perform daily activities (i.e., driving, being out in public, leaving their homes). AUVP/vestibular neuritis is the third most common trigger after BPPV and vestibular migraine of secondary functional dizziness, a disease now better known as Persistent Postural Perceptual Dizziness (PPPD).[2,16-19] Approximately 10-15% of people will develop BPPV following a vestibular neuronitis.[16,20]

DIAGNOSIS/TREATMENT

Vestibular neuritis can be broken down into three subtypes: superior, inferior, or total.[3] Superior neuritis is the most common, followed by total and inferior. The advent of vestibular testing has aided in more accurate diagnosis given that these

conditions are peripheral vestibulopathies. Vestibular Evoked Myogenic Potentials (cVEMP/oVEMP), Video Head Impulse Testing (vHIT), Ocular Tilt Reaction, Electronystagmography (ENG), and Videonystagmography (VNG) can all be utilized to pinpoint the lesion and the subtype.[4] An Audiogram may be required if patients experience hearing loss.

Accurate diagnosis of AUVP/Vestibular Neuritis is of the utmost importance. The first step is to differentiate it from an acute central vestibular syndrome which can often present with similar symptoms in patients with significant cardiovascular risk factors. The physical exam is critical in determining which patients require further workup to rule out a stroke.[21]

Vestibular migraine can be difficult to differentiate from a vestibular neuritis episode, but the repetitive, stereotypical nature of the episodes with or without migraine headaches often leads to the diagnosis.[22]

Other peripheral disorders to differentiate from AUVP/vestibular neuritis include:

- Cogan syndrome, which is an autoimmune disorder presenting with bilateral peripheral vestibular deficit, bilateral sensorineural hearing loss, and interstitial keratitis (painful red eye).[23]
- Cupulolithiasis of the horizontal

semicircular canal, which is a rare type of BPPV. It can be diagnosed on physical exam during positional maneuvers.[24,25]

- Herpes zoster oticus/Ramsay Hunt syndrome resulting in shingles infection in the territory of the 8th and facial nerve. This typically presents with blisters and a facial paralysis similar to Bell's palsy in Ramsay Hunt. Sometimes it can present exactly like vestibular neuritis without the additional symptoms.[26]
- Isolated acute unilateral utricular or saccular vestibulopathy, which presents with sudden onset of unsteadiness and is diagnosed with vestibular evoked myogenic potentials (VEMP) testing.
- Labyrinthitis resulting in ear pain, hearing loss, and possibly tinnitus.
- Meniere's disease, which is a stereotypical triad of symptoms. Recurrent episodes of vertigo with aural fullness, tinnitus, and low frequency hearing loss lead to the diagnosis.[27]
- Recurrent vestibulopathy or Vestibular Migraine resulting from recurrent episodes of AUVP. The etiology is unclear, and duration of symptoms is usually shorter than AUVP.[28]
- Stroke due to occlusion of the anterior inferior cerebellar artery rarely presents with isolated vertigo symptoms. Usually there are more symptoms and typical findings on the clinical exam.[21,29]
- Susac syndrome, which is a vasculitis that affects the inner ear and the eye leading to various ophthalmologic signs, but also hearing loss and sometimes a picture similar to an AUVP.[30]

TREATMENT

In the acute phase, patients experience the symptoms continuously, even at rest, with worsening of their symptoms on movement. Patients may also be inclined to lay with their healthy ear facing downwards, although there is no evidence to indicate this maneuver improves symptom severity.

Supportive treatment is the mainstay of the acute phase. Medications tailored to decrease nausea, vomiting, and vertigo should be utilized. However, caution must be taken to not prolong treatment with these medications as they can inhibit compensatory mechanisms employed by the brain. Common medications prescribed as supportive treatment include Meclizine, Promethazine Hydrochloride, Antivirals, Antibiotics, Diphenhydramine, Ondansetron, and Benzodiazepines. Unless contraindicated, we typically also prescribe high-dose steroids.

Evidence has shown that steroid use does improve canal paresis in the acute phase.31,32 However, no significant improvement has been seen in dizziness, chronic canal paresis, or symptomatic recovery.[2,31-33]

Some patients do not compensate completely and continue to suffer from chronic imbalance following an episode of vestibular neuronitis. Paraclinical investigations can help confirm the site of lesion, and timely referral to vestibular rehabilitation should be made to help the patient work on compensatory strategies. This can help the central nervous system compensate for peripheral vestibular insults.[34,35] Vestibular rehabilitation can only be effective if patients learn to compensate for their symptoms.

Exercises utilized in vestibular rehabilitation evoke moderate dizziness/unsteadiness, so patients should practice exercises for habituation, gaze stabilization, balance, and gait.[36]

Computerized Dynamic Posturography (CDP), a complementary test to VNG, may be used to establish a baseline for patients by testing for three sensory inputs: vestibular, somatosensory, and vision.[37] Exercises taught to combat chronic symptoms are encouraged by providers to be done frequently at home, even after completing physical therapy.

Vestibular rehabilitation is crucial in ensuring improvement of dynamic and chronic symptoms.[38] Central compensation may take months to years for patients to achieve. However, this is crucial to establish a better quality of life for patients experiencing debilitating symptoms. Patients should seek follow-up with their otolaryngologist or neurologist to ensure continuous improvement in symptoms.

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