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Case Report

The Lyndsay-Hemenway Syndrome: Two Case Reports. Review and Comments

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Abstract

The syndrome of Lindsay and Hemenway i.e. vestibular neuritis with Benign Paroxysmal Positional Vertigo (BPPV) as a clinical sequel is believed to be not frequent. However the careful observation of the patient in the course of several months reveals a higher incidence in reality.

Aim of the study is the presentation of two case reports of patients with different clinical occurrences of the same syndrome.

In the first case the initial vestibular neuritis occurs a few months before the occurrence of BPPV and BPPV is resistant to treatment with repositioning maneuvers. In the second case BPPV occurs as an immediate sequel of vestibular neuritis and responds to treatment.

The syndrome of Lindsay and Hemenway is an interesting clinical entity the existence of which the clinician should be aware of, in order to be able to combine repositioning maneuvers for the treatment of BPPV with vestibular rehabilitation targeting the treatment of a defective central compensation due to unilateral labyrinthine paresis.

INTRODUCTION

The syndrome of Lindsay-Hemenway is vestibular neuritis having Benign Paroxysmal Positional Vertigo as a sequel. It is attributed to a thrombosis of the anterior vestibular artery irrigating the anterior and lateral semicircular canals and the utricle. Consequently ischemia in this territory can lead to classical symptoms of vestibular neuritis together with the detachment of otoconia from the gelatinous layer of the utricular macula. This latter event is the pathophysiological basis of Benign Paroxysmal Positional Vertigo. Given the fact that the function of the posterior semicircular canal is not abolished, because the canal ampulla is spared, a further translocation of otoconia into the canal lumen is the generative cause of clinical occurrence of Benign Paroxysmal Positional Vertigo.

Two case reports will be presented in this study. They concern two elderly males, but the clinical course of the syndrome is different in each one of them.

CASE REPORT 1

A 75 year old male patient with a strong sensation of imbalance was examined in a vertigo clinic seven months ago. He reported a progressive (during one month and a half before consultation) deterioration of his gait and sensation of dizziness at the movements of the head. He did not report any rotatory vertigo. His clinical examination is shown on (Table 1). He received cinnarizine oral solution (75 mg) once in the evening and followed a vestibular rehabilitation program with Cowthorne- Cooksey [1,2] exercises. A month later he reported a satisfactory improvement of his symptoms. Four months later he came back, complaining of a relapse of his unsteadiness with concomitant rotatory vertigo on lying and rolling over in bed. His symptoms were worse in the morning as he stood up from bed. The clinical findings are reported on table 1. The patient had a medical history of diabetes type I treated with insulin.

Lindsay-Hemenway (L-H) syndrome was suspected and the modified Epley maneuver (Canalith Repositioning Procedure, CRP) [3] was performed on the right side as a cure. He was reexamined five days later, to find a persisting positive Dix-Hallpike test [4] on the right. Epley maneuver was repeated, but it didn't succeed for a second time. Five days later, the Semont liberatory maneuver [5] was performed, three consecutive times at the same session. Ten days later, he was assessed with a persistent positive right Dix-Hallpike test, but Unterberger (stepping) test was improved (body did not turn at all) and he reported diminution of his symptoms. Due to persistence of positional vertigo he was investigated with CT scan of the brain, which was normal. He was given Brandt and Daroff [6] habituation exercises and was advised to walk up and down two or three steps of a staircase and to practice on tandem gait, in

Table 1: The clinical and laboratory investigations of patient 1. Empty cells signify no change comparing to previous.

Clinical and laboratory investigations of patient 1					
	Consultation 1: One month and a half after beginning of symptoms	Consultation 2: Four months after first consultation	Consultation 3: 5 days after 1st Epley treatment	Consultation 4: 5 days after 2nd Epley treatment	Consultation5 10 days after Semont treatment
Spontaneous nystagmus	Absent	Absent			
Vestibular Ocular Reflex (VOR) Sinusoidal testing	Reduced on the right	Normal			
VOR suppression	Normal	Normal			
Halmagyi Head Impulse Test	Positive on the right	Positive on the right			
Smooth pursuit	Normal	Normal			
Saccades	Normal	Normal			
Optokinetic nystagmus (OKN)	Normal	Normal			
Romberg test	Normal, but feels unsure	Normal			
Tandem Romberg	Unstable	Unstable			
Unterberger (Fukuda) Stepping test	Deviation of the body to the right	A small Deviation of the body to the right			Normal: No body deviation
Walking	Normal	Normal			
Tandem gait	Unstable	Unstable, turns toes out of the line to broaden the base of support			Unstable, turns toes out of the line to broaden the base of support
Dix-Hallpike test	Positive on the right	Positive on the right	Positive on the right	Positive on the right	Positive on the right
Supine Roll test	Negative	Negative			
Otoscopy	Normal	Normal			
Bithermal Caloric irrigation of the labyrinth	Important labyrinth paresis on the right	Important labyrinth paresis on the right			
Brain CT-scan					normal
Treatment	Cowthorne-Cooksey	Epley CRP	Epley CRP	Semont LP	Brandt-Daroff Step climbing Tandem gait

order to improve the feeling of moderate instability that he still reported. Two months later the patient reports a considerable improvement of both symptoms of instability and vertigo in bed.

CASE REPORT 2

An 80 year old patient was seen in a vertigo clinic, reporting a feeling of "dizziness inside his head" particularly when he moved his head or eyes. He also reported a slight feeling of rotatory vertigo on lying in bed. A month earlier he had experienced a strong vertiginous episode, for which he was seen in hospital casualties. Vertigo then was rotatory, strong and accompanied by serious nausea. He was prescribed dimenhydrinate capsules but he did not take more than two, due to stomach-ache. Rotatory vertigo stopped after three days, but he kept feeling dizzy. The patient's clinical examination findings are reported on (Table 2). Dix-Hallpike triggering position on the right revealed intense nystagmus with nausea. The patient was not able to undergo the complete maneuver of Epley, so he was prescribed dimenhydrinate capsules in order to reduce the symptoms and come back the next day to repeat the Dix-Hallpike test and continue with the therapeutic modified Epley maneuver. The therapeutic maneuver was performed the day after with success. On a following assessment ten days later, Dix-Hallpike was negative and the patient reported an important reduction of his dizziness. Clinical examination showed improvement of Unterberger test (he could make twenty steps, but still there was an obvious and repeatable turn of the body to the right side).

He was put on Cowthorne Cooksey exercises at home. Twenty days later, on re-examination he performed even better with a minor turn of the body at the Unterberger test.

DISCUSSION

The vascularization of the internal ear [7] [Figure 1], belongs to the vertebrobasilar system. The anterior inferior cebellar artery (AICA) derives from the basilar artery. The common labyrinthine artery (or internal auditory artery) derives from the AICA. The common labyrinthine artery divides into two rami: The common cochlear artery and the anterior vestibular artery. The anterior vestibular artery irrigates the anterior and lateral semicircular canal ampullae, the utricular macula and a part of the saccule. The common cochlear artery divides into the main

Table 2: The clinical and laboratory investigations of patient 2. Empty cells signify no change comparing to previous.

Clinical and laboratory investigations of patient 2				
	Consultation 1: One month after beginning of symptoms	Consultation 2 24hours later, with sedatives	Consultation 3: 10 days after first consultation (results after Epley)	Consultation 4 20 days later
Spontaneous nystagmus	Absent		Absent	
Vestibular Ocular Reflex (VOR) Sinusoidal testing	Reduced on the right		Normal	
VOR suppression	Normal		Normal	
Halmagyi Head Impulse Test	Positive on the right		Positive on the right	
Smooth pursuit	Normal		Normal	
Saccades	Normal		Normal	
Optokinetic nystagmus (OKN)	Normal		Normal	
Romberg test	Normal, but feels unsure		Normal	
Tandem Romberg	Unstable		Unstable	
Unterberger (Fukuda) Stepping test	Unstable after 10 steps		Deviation of the body to the right	Minor deviation to the right
Walking	Normal		Normal	
Tandem gait	Normal		Normal	
Dix-Hallpike test	Positive on the right, very nauseous, intense nystagmus	Positive on the right, no nausea	Negative	Negative
Supine Roll test	Negative		Negative	
Otoscopy	Normal		Normal	
Bithermal Caloric irrigation of the labyrinth	Important labyrinth paresis on the right		Important labyrinth paresis on the right	
Treatment	sedatives of labyrinth	Epley CRP	Cowthorne-Cooksey	

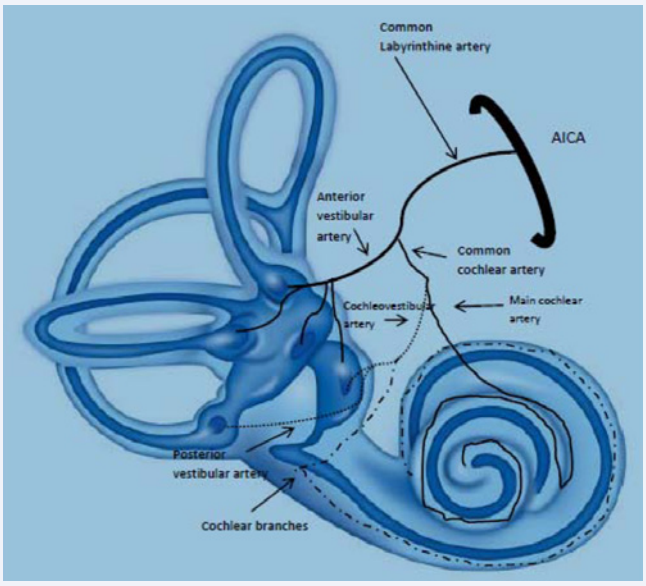

Figure 1 The arterial supply to the anterior and posterior labyrinth.

cochlear artery and the cochleovestibular artery, which gives off the posterior vestibular artery and cochlear branches.

The main cochlear artery irrigates the modiolus and three quarters of the cochlea. The basal quarter of the cochlea receives blood circulation from the cochlear branches of the cochleovestibular artery. The posterior vestibular artery irrigates the rest of the saccule and the posterior semicircular canal ampulla. All branches are terminal and a flow interruption leads to rapid functional loss. Lyndsay and Hemenway syndrome refers to interruption of the blood flow of the anterior vestibular artery.

The syndrome was described in 1956 by J.R. Lindsay and W.G. Hemenway [8], although H.F. Schuknecht discovered its vascular origin. It is a particular form of vestibular neuritis, beginning with major vertigo as described in the typical form, but which is followed by true BPPV of the posterior semicircular canal after an amount of time, ranging from a few days to several months. It is more frequent in patients over 60 years of age particularly if there are risk factors for arterial thrombosis. Hearing is spared, the labyrinth damage is major as measured in caloric irrigation, but in a few days to weeks, posterior canal BPPV is evident on the affected side. BPPV is a testimony of the utricular macular degeneration, which results in the dislocation of otoliths. At the same time however, BPPV marks the survival of posterior canal function. The explanation of the syndrome can be vascular thrombosis of the anterior vestibular artery, or a virus affecting the upper branch of the vestibular nerve, resulting in disruption of the homeostasis of the calcium carbonate crystals on the gelatinous macula of the utricle. Since the lower branch of the nerve is left intact, it can be excited by the dislodged otoconia, producing the symptoms of BPPV [Table 3].

A long term cohort study [9] found a 10% of patients with vestibular neuritis, presenting BPPV of the affected ear. It occurred predominantly to patients who had not fully recovered from the vestibular neuritis and was more difficult to treat, as it happened with the first patient of this study. Another study [10], seeking to detect L-H syndrome among patients consulting for BPPV, reported a prevalence of 16.3% of the syndrome among BPPV patients. Harada et al. (1993) [11], report that time interval between vestibular neuritis and BPPV onset ranged from 2 weeks to 20 years and assume that in case BPPV appears soon after vestibular neuritis, there is a possibility that the posterior canal has been spared from the beginning of the incident. In case it appears later, a possible initial lesion is assumed, followed by a progressive recovery of the posterior canal function later.

The present series of two patients is of particular value, because it is an example of the two possibilities of BPPV occurrence: either immediately after the symptoms of vestibular neuritis subside (one month time in the second case) or some time later (four months in the first case). Another particularity that has to be mentioned is the progressiveness of the initial symptoms of the first patient, who had diabetes i.e. an important risk of vascular thrombosis. Boffi [12] refers to the possibility of vestibular neuritis to occur without rotatory vertigo, but with a persistent feeling of unsteadiness. Pardal et al. [10], report the possibility of slowly progressive installation of the vestibular lesion preceding BPPV in Lindsay-Hemenway syndrome. One possible interpretation is a slow formation of a thrombus in the anterior vestibular artery, diminishing the patency of the vessel in a gradual fashion, leading to progressive loss of vestibular function.

The differential diagnosis of the syndrome comprises a wide variety of diseases [10]. Practically the clinician has to do differential diagnosis for two clinical entities. Multiple sclerosis and ischemic or tumoric lesions of the cerebellum have to be excluded with caution. Exclusion of central lesions will be based on the identification of benign (peripheral)characteristics of positional vertigo, the presence of a positive Halmagyi Head Impulse Test, the ability of the patient to stand with eyes closed (Romberg test) [Table3]. In general this is possible by the careful registration of medical history of the patient and clinical examination. Caloric irrigation should show lateral canal paresis (or important hypofunction), Playtone Audiometry should not be showing alterations of the hearing function of the patient. Air Conducted Cervical Vestibular evoked myogenic potentials (AC-VEMP) should be either present or absent in the initial incident, but should reappear in the case of BPPV. This is the case because air conducted VEMP measure the function of the saccule, which should be recovering function together with the posterior semicircular canal (or be spared from the beginning) as both structures are irrigated from the posterior vestibular

Table 3: The clinical and laboratory findings of Vestibular Neuritis in the very acute phase and Benign Paroxysmal Positional Vertigo (BPPV), presented as separate clinical entities. In patients with L-H syndrome, as BPPV occurs after some time, central compensation of the vestibular imbalance taking place in the mean time, can alter some of the clinical findings, i.e. disappearance of spontaneous nystagmus, VOR gain increase at the side of the lesion (Sinusoidal testing), Body deviation towards the healthy side at the Fukuda stepping test.

Clinical findings	Vestibular Neuritis	BPPV
Nystagmus Occurence	Non dependent on head positions (spontaneous)	Dependent on head position relative to gravity (positioning/positional)
Nystagmus Direction	Horizontal (with torsional component when all canal function is abolished unilaterally)	Torsional upbeating (PC-BPPV) or Horizontal(LC-BPPV)
	One directional	One directional (PC-BPPV) Direction changing on contralateral side(LC-BPPV)
Nystagmus Duration	Continuous (lasting from hours to days)	Paroxysmal (Beginning-crescendo-subsiding) lasting up to 40(PC-BPPV) or 60 seconds (LC-BPPV)
Nystagmus Intensity	Intensity affected by direction of gaze (Alexander's law)	Rotational or vertical component more obvious according to gaze direction(PC-BPPV)
Fixation	More pronounced without fixation	More pronounced without fixation
Vestibular Ocular Reflex (VOR) Sinusoidal testing	Reduced on side of lesion	Normal
Halmagyi Head Impulse Test	Positive on side of lesion	Normal
Romberg test	Body leaning to side of lesion with tendency to fall	Usually Normal
Unterberger (Fukuda) Stepping test	Body deviates to side of lesion	Usually normal
Dix-Hallpike test/Supine roll test	Negative (spontaneous nystagmus still present)	Positive on side of lesion (PC-BPPV) Positive bilaterally with direction changing horizontal nystagmus(LC-BPPV)
Laboratory findings		
Bithermal Caloric irrigation	Canal Paresis	Normal for PC-BPPV Occasionally hypofunction in case of LC-BPPV
AC-VEMP	Absent or present	Absent or present

PC-BPPV: Posterior Canal BPPV; LC-BPPV: Lateral Canal BPPV; AC-VEMP: Air Conducted Cervical Vestibular Evoked Myogenic Potentials.

artery. In cases of elderly people in whom degeneration of the saccular macula could have already taken place, AC- VEMP should be absent [13]. Imaging with CT scan or MRI is justified if treatment for BPPV is not successful and/or if clinical findings are suspicious for a central lesion. Due to the ischemic nature of underlying pathology, a full cardiovascular control is also mandatory in these patients.

CONCLUSION

Lyndsay-Hemenway syndrome is a clinical entity associating vestibular neuritis with BPPV. It manifests with a variety of symptoms, but in the majority of cases it is characterized by abrupt intense vertigo and nystagmus, followed by posterior canal BPPV after a variable amount of time. Clinicians dealing with vertigo should be aware of this clinical entity in order to diagnose it and reassure the patients accordingly and to help them with treatment combining vestibular rehabilitation and otoconia repositioning maneuvers.

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