Evaluation of the clinical and imaging relationship of endolymphatic hydrops in Menière's disease

Original Article

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Abstract

Meniere's disease is characterized by episodes of audiovestibular symptoms. The presence of inner ear endolymphatic hydrops is a potential explanation for the clinical symptoms. This study aims to establish a relation between the imaging characteristics from the magnetic resonance with clinical data of patients with Meniere's disease. Among the study population, 84,2% presented with unilateral disease with average age at symptom onset of 50.6 years. Vestibular hydrops was identified in 100% of clinically affected ears and cochlear hydrops in 90.9% of the affected ears. The degree of hearing loss correlates with the degree of vestibular hydrops (p=0.022), but not with the degree of cochlear hydrops (p=0.524). No relationship was found between the time from symptom onset until the imaging study and the presence of hydrops. The presence of hydrops is not related to the results of vestibular tests, nor clinical findings.

Keywords: Hydrops; Menière

Introduction

Menière's disease is characterized by recurrent episodes of audiovestibular symptoms.

The diagnosis is clinical and based on the criteria established by the Barany Society in 2015, which include documented sensorineural hearing loss, episodes of vertigo lasting between 20 minutes and 12 hours and fluctuating aural symptoms such as aural fullness and/or tinnitus.[1]

Previous studies have reported prevalence rates of 34–190 cases per 100,000 population, primarily affecting individuals between the third and sixth decades of life.[2]

Although diagnosis is clinical, results of audiometric tests and magnetic resonance imaging (MRI) have been part of the diagnostic criteria since 2015. According to these criteria, pure-tone audiogram reveals sensorineural

hearing loss in the low-mid frequencies and can be used to identify the affected ear in Menière's disease.[1]

The presence of endolymphatic hydrops (EH) in the labyrinth is the potential cause of the symptoms in Menière's disease. EH, defined as dilation of the endolymphatic space due to endolymph accumulation, was first identified in histopathological studies of temporal bones.[3,4] Rupture of the Reissner's membrane or other parts of the membranous labyrinth due to excessive endolymphatic volume and/or pressure was initially proposed as the mechanism underlying the fluctuating nature of the symptoms.[5,6] Subsequently, endolymphatic movements have been suggested as the cause of these episodes. Excessively rapid drainage of the endolymph from the cochlear duct may lead to leakage in the endolymphatic duct and, consequently, the utricle, ultimately stretching the crista of the semicircular canals and triggering an episode.[7] However, EH has also been detected in up to 30% of asymptomatic individuals and is therefore not pathognomonic of the disease.[8]

Although vestibular tests are not specific for diagnosis, they are routinely used as part of the complementary evaluation of patients with Menière's disease. Caloric tests, which are used to assess low-frequency vestibulo-ocular reflex (VOR) function, are abnormal in approximately 84% of cases at some point during the disease course. In about 74% of patients, these tests can also demonstrate progressive hypofunction on the affected side.[9]

Vestibular evoked myogenic potentials (VEMPs) are used to assess otolithic function, including cervical VEMPs (cVEMPs) for the saccule and ocular VEMPs (oVEMPs) for the utricle. The interaural asymmetry ratio can also be used, as the response amplitudes tend to decrease and eventually disappear as the disease progresses.[10] Some authors have reported that the amplitude of cVEMP waves does not correlate with the audiometric threshold in the affected ears of patients with Menière's disease, as abnormal cVEMP thresholds have been observed in their healthy ears, thus compromising the validity of interaural amplitude comparison.[11]

A discrepancy between abnormal caloric test results and a normal video head impulse test (vHIT) has been reported as a marker of Menière's disease.[12]

Objective

This study aimed to correlate the MRI findings, which were obtained using an EH protocol, with clinical data and audiovestibular test results in patients diagnosed with Menière's disease.

Materials and methods

In this retrospective observational study, the inclusion criteria were a confirmed diagnosis of Menière's disease according to the Barany Society[1], prior MRI using an EH protocol, and at least one pure-tone audiogram. The only exclusion criterion was age under 18 years.

Data were collected through a review of individual medical records and included age at symptom onset; age at diagnosis; laterality, frequency, and characteristics of the vertigo episodes; initial symptoms; and presence of neurovegetative symptoms.

Simultaneously, all MRI scans, which were conducted using an EH protocol since 2020 were individually analyzed. This protocol involves volumetric acquisitions in T2 SPACE, T1 VIBE, and IR SPACE sequences performed before and after four hours of intravenous administration of gadolinium. The MRI scanner used had a field strength of 3 Tesla (Magnetom TIM TRIO, Siemens). Subsequently, the Baráth classification was used to assess the presence of vestibular and cochlear hydrops.[13] Complementary examinations, when available, were reviewed, including all pure-tone audiograms, with the degree of hearing loss classified according to the classification system proposed by the International Bureau for Audiophonology (BIAP),[14] along with caloric tests, vHIT, and VEMPs.

Audiological evaluation included assessment of pure-tone thresholds (GSI 16 audiometer)

for air conduction (0.125, 0.25, 0.5, 1, 2, 4, and 8 kHz) and bone conduction (0.25, 0.5, 1, 2, and 4 kHz) in a soundproof booth using an audiometer (Interacoustics A/S AT235 version 1.11®, Denmark).

Caloric tests were conducted using a video system for ocular response acquisition and analysis (Ulmer Videonystagmography v5, Synapsis®, France). Each external auditory canal was irrigated separately with 100 mL of water at 30°C and 44°C for 30 seconds, with a minimum interval of 5 minutes between the irrigations. A response asymmetry greater than 20% between the left and right ears was considered indicative of a pathology.

vHIT was conducted using a video-based system (vHIT GN, Otometrics®, Denmark) with the patient seated, and fixating on a target at a distance of 1.5 meters. Each plane was tested with a minimum of 20 impulses (angle of 15–20°, duration of 150–200 ms, peak velocity > 150°/s). Results were considered normal if the calculated VOR gain was greater than 0.7.

VEMPs were recorded using the Neuro-Audio system with software v2010 (Neurosoft®, Russia). Stimulation was delivered via air-conducted 500 Hz tone bursts at sound pressure level of 100–105 dB, 7 ms duration, five stimuli per second. The absence of a biphasic wave at 105 dB was interpreted as the absence of response. For cVEMPs, the muscular activity of the ipsilateral sternocleidomastoid muscle was recorded using surface electrodes. For oVEMPs, an electrode was placed in the infraorbital region to record the activity of the contralateral inferior oblique muscle corresponding to the stimulated ear.

Statistical analysis was performed using the SPSS software version 28. A p-value above 0.05 was considered statistically significant.

Results

The study population included 19 patients who were being followed up in the balance disorders clinic and diagnosed with Menière's disease according to the diagnostic criteria established by the Barany Society.[1]

All patients underwent a comprehensive neuro-otologic evaluation, and a detailed history was obtained. Additionally, each participants underwent MRI scans following an EH protocol. Table 1 shows the demographic characteristics of the study population.

The initial symptom was auditory (hearing loss, tinnitus, and/or aural fullness) in 36.5% of the cases; vestibular in 26.5%; and mixed in 36.8%. Regarding laterality, bilateral hearing loss was observed in four patients (21.1%). In 14 cases (73.7%), hearing loss involved both low frequencies (250 Hz–1 kHz) and high frequencies (> 2 kHz). Isolated involvement of low frequencies was identified in 15.8% cases, while exclusive high-frequency loss was noted in 5.3%. Most patients did not report headaches (n = 14, 73.7%). Headaches with migraine characteristics were reported by 21.1% of patients, while non-migraine headaches were reported by 5.3%. None of the patients were receiving any specific treatment for headache and/or migraine. No patients reported with daily or weekly episodes of rotary vertigo. Monthly episodes were reported by 26.3% of patients, while 73.7% experienced episodes at other intervals.

Table 1
Demographic characteristics of the study population

Characteristic	Value
Age at diagnosis in years (μ, SD)	54,2 (± 9,6)
Age at symptom onset (μ, SD)	50,5 (± 9,1)
Disease duration in years (μ, SD)	6,6 (± 5,1)
Unilateral MD (n, %)	16 (84,2%)
Female sex (n, %)	10 (52,5%)

μ - mean; SD - standard deviation; MD - Menière's disease

Approximately half of the patients (n = 10, 52.6%) reported neurovegetative symptoms during the episodes. In 84.2% of cases, there was no relevant family or medical history. A family history of hearing loss and/or vertigo was reported by 10.5% participants, while 5.3% (n = 1) had a family history of Menière's disease. Regarding comorbidities, 47.4% of patients had no associated conditions. Among those with comorbidities, dyslipidemia was present in 42.2%, hypertension in 30.6%, and diabetes mellitus in 10.5%. Table 2 shows the audiovestibular findings in the study population. Considering the retrospective design of the study, not all complementary diagnostic tests were available for analysis. Among the19 patients with Menière's disease, caloric test results were available for 15 cases, oVEMPs and vHIT for eight cases, and cVEMPs for nine cases. Regarding the MRI findings, vestibular EH was identified in 100% of the clinically affected ears, and cochlear hydrops in 90.9% ears. Unilateral vestibular hydrops was detected in 16 patients (84.2%): including one (5.3%) case classified as grade 0 in the Baráth scale, nine (47.4%) classified as grade 1, and nine (47.4%) classified as grade 2. Cochlear hydrops was classified as grade 0 in three cases (15.8%), grade 1 in seven (36.8%), and grade 2 in nine (47.4%) cases.

The Fisher's exact test revealed a statistically significant association between the laterality of hearing loss and laterality of cochlear and vestibular EH ($p < 0.001$). The degree of hearing loss was found to have a statistically significant association with the severity of vestibular hydrops ($p = 0.037$), but not with the degree of cochlear hydrops ($p = 0.250$).

The Kruskal–Wallis test revealed a significant difference between the time interval from symptom onset to MRI and degree of cochlear EH ($H [2, n = 19] = 7.618$, $p = 0.022$), but not for vestibular EH ($H [2, n=19] = 2.279$, $p = 0.320$). Post-hoc analysis using the Mann–Whitney U test indicated a significant difference in the median (Mdn) time from symptom onset to MRI between grade 1 (Mdn = 0.29 years) and grade 2 EH (Mdn = 5.00 years) ($U[N_1 = 7, N_2 =$

Table 2
Audiovestibular results of the study population. Only the affected ears were considered. In case of bilateral disease, both ears were considered.

Audiovestibular evaluation		N (%)
vHIT* (gain < 0.7)	LA SCC	0 (0%)
	RP SCC	1 (5,3%)
	RA SCC	1 (5,3%)
	LP SCC	1 (5,3%)
	RL SCC	0 (0%)
	LL SCC	0 (0%)
cVEMPs^	Interaural asymmetry (> 40%)	4 (44,4%)
	of which, absent in	2 (22,2%)
oVEMPs*	Interaural asymmetry (> 40%)	1 (5,3%)
Caloric tests'	RE deficit	4 (21,1%)
	LE deficit	4 (21,1%)
Audiogram	Mild hearing loss	2 (10,5%)
	Moderate hearing loss	10 (52,6%)
	Severe hearing loss	7 (36,8%)

RA – right anterior; LA – left anterior; SCC – semicircular canal; cVEMPs – cervical vestibular evoked myogenic potentials; RL – right lateral; LL – left lateral; RE – right ear; LE – left ear; oVEMPs – ocular vestibular evoked myogenic potentials; RP – right posterior; LP – left posterior; vHIT – video Head Impulse Test.
* Data from eight cases; ^ data from nine cases; ' data from 15 cases.

Table 3
Relationship between MRI findings, audiovestibular test results, and clinical data

		Degree of cochlear hydrops	Degree of vestibular hydrops
Audiovestibular evaluations			
Caloric asymmetry	RE	p=0,754*	p=1,000*
	LE	p=1,000*	p=1,000*
cVEMPs asymmetry		p=0,697*	p=1,000*
oVEMPs asymmetry		p=1,000*	p=0,250*
Degree of hearing loss		p=0,250*	**p=0,037***
vHIT	LA SCC	p=0,125*	p=1,000*
	RP SCC	p=0,125*	p=1,000*
	RA SCC	p=0,125*	p=1,000*
	LP SCC	p=0,500*	p=0,375*
	RL SCC	p=0,125*	p=1,000*
	LL SCC	p=0,125*	p=1,000*
Clinical data			
Initial symptoms		p=0,885*	p=0,458*
Frequency of vertigo episodes		p=0,393*	p=0,368*
Neurovegetative symptoms		p=1,000*	p=1,000*

RA – right anterior; LA – left anterior; SCC – semicircular canal; cVEMPs – cervical vestibular evoked myogenic potentials; RL – right lateral; LL – left lateral; RE – right ear; LE – left ear; oVEMPs – ocular vestibular evoked myogenic potentials; RP – right posterior; LP – left posterior; vHIT – video Head Impulse Test; MRI – magnetic resonance imaging.
* Fisher's exact test.

9] = 9.00, z = -2.42, p = 0.015). These findings suggest that there is a shorter interval between symptom onset and MRI is cases with a lower degree of cochlear EH.

The Fisher's exact test did not show significant associations between cochlear or vestibular hydrops and the results of complementary vestibular tests, frequency of vertigo episodes, presence of vegetative symptoms, or type of initial symptoms (p > 0.050).

Discussion

Predominance in women, age, and disease course

As observed in our study population, previous studies have reported a slight predominance of Menière's disease in women,[2,15], although some authors have also described an equal distribution between the sexes.[16] The average age at onset ranges between the third and seventh decades of life.[17] The disease most commonly presents unilaterally; however, bilateral involvement may develop over time.[18]

Relationship between EH and auditory thresholds

Numerous studies have indicated that EH progression follows a non-random pattern. Hydropic changes progressively affect the cochlea, saccule, utricle, and finally, the semicircular canals.[19] Therefore, audiovestibular abnormalities are typically detected by an audiogram, followed by cVEMPs, oVEMPs, caloric tests, and vHIT.[19] Our results revealed a correlation between the severity of vestibular EH and worse thresholds in pure-tone audiometry, which aligns with the findings of previous studies.[20, 21]

Nonetheless, some individuals with normal audiograms may present with EH, while

those with severe hearing loss may not show any degree of EH. This discrepancy can be attributed to several factors, including the fluctuating nature of hearing loss and possibility of coexisting etiologies.[20]

Relationship between EH and vestibular testing

Saccular and cochlear dysfunction may occur concomitantly. Gürkov reported a negative correlation between saccular function assessed by cVEMP and the degree of vestibular EH.[21] In our sample, we observed a clear asymmetry in cVEMP results in approximately 40% of cases; however, this finding did not reach statistical significance. As described in the literature, abnormal caloric testing results do not correlate with the degree of EH.[10]

vHIT results, as expected, were largely normal or showed minimal alterations.[12] Future studies with larger sample sizes and a comprehensive battery of vestibular tests may enable more robust conclusions, particularly regarding cVEMP findings and saccular function in Menière's disease. Furthermore, establishing a standardized evaluation protocol with defined timelines for each test could make data collection more consistent.

Relationship between EH and disease duration

Some studies have indicated that the degree of cochlear and vestibular EH is proportional to the duration of the disease.[20, 21] In our sample, a higher degree of cochlear EH was observed in patients with a longer time interval between symptom onset and MRI. However, this pattern was not observed for vestibular hydrops, possibly due to the small sample size. An outpatient EH MRI protocol should be used for etiological investigation and disease characterization. At our institution, the current EH MRI protocol was implemented in 2020. Since then, some patients who had been under long-term follow-up for Menière's disease only underwent an MRI years after symptom onset, whereas others with more recent symptoms were scanned earlier in the disease course. Nevertheless, our data support the progressive nature of the disease, with the hydrops severity ranging from moderate to vestibular involvement over time.

Other results

Thus far, no study has demonstrated a relationship between MRI findings and presenting symptoms, presence of neurovegetative symptoms, or frequency of vertiginous episodes in patients with Menière's disease. Furthermore, most authors agree that EH should be considered as an adjunctive diagnostic finding rather than a direct cause of symptoms in Menière's disease.[22, 23]

Conclusion

Our study demonstrates that MRI conducted using an EH protocol may serve as a valuable complementary diagnostic tool in suspected cases of Menière's disease. Rather than being used solely to rule out differential diagnoses, this imaging modality can aid in the early identification of the affected ear, which is crucial in potentially bilateral cases, as in our sample vestibular hydrops was detected in 100% of the affected ears.

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols used at their working center regarding patient data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and the 2013 Helsinki Declaration of The World Medical Association.

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Availability of scientific data

There are no datasets available, or publicity related to this work.

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