

## A thin line between Meniere's disease and spontaneous intracranial hypotension syndrome

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### ABSTRACT

**Aim** To point out the similarity of Meniere disease and spontaneous intracranial hypotension and difference of their treatment.

**Methods** A case of a 54-year-old male patient with previously diagnosed Meniere's disease and newly diagnosed spontaneous intracranial hypotension syndrome is presented. Additional neuro-radiological examination, Brain contrast-enhanced MRI and MR myelography were used for diagnosis.

**Results** Due to deterioration of vertigo, hearing loss and tinnitus in the right ear the patient was referred to the additional neuroradiological examination which confirmed the diagnosis of spontaneous intracranial hypotension syndrome. Brain contrast-enhanced MRI showed increased pachymeningeal contrast enhancement, and MR myelography identified the location of CSF leak. The patient was successfully treated conservatively.

**Conclusion** According to our knowledge this is the fifth case report of Meniere's disease and spontaneous intracranial hypotension coexistence. Both diseases have similar clinical presentation and initial treatment. We suggest procedures of additional examination when the treatment fails and initial diagnosis becomes questionable.

**Key words:** otogenic vertigo, intracranial hypotension, cerebrospinal fluid leakage

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## INTRODUCTION

Morbus Meniere is idiopathic inner ear disorder with characteristic clinical manifestation which includes relapse of vertigo, hearing loss, fullness in ear and tinnitus. Between attacks, hearing could normalize. Vertigo attacks usually last from 30 minutes up to several hours (1).

The cause of the symptoms is endolymphatic hydrops, an excess of fluid in the inner ear. Endolymph in membranous labyrinth bursts from its normal channels and goes to other areas (1). That causes swelling of the endolymphatic sac, which is responsible for the body's sense of balance (2).

The cause of the excess of fluid is still unknown. It can be related to scar tissue which causes obstruction in drainage (3) or to a virus infection, mainly herpes (4). The symptoms may be further exacerbated by excessive consumption of salt (3).

The diagnosis is based on patients' complaints, medical history, otorhinolaryngological (ENT) examination and audiometry. A dehydration test using glycerol is part of the differential diagnosis that distinguishes Meniere's from other inner ear disorders. If hearing improvement occurs (at least 10-15 dB in 3 frequencies) after applying glycerol, results can indicate hydrops of labyrinth (2).

In 1995 The American Academy of Otolaryngology-Head and Neck Surgery Committee on Hearing and Equilibrium (AAO HNS CHE) set criteria for degrees of Mb Ménière (5). In acute attack, therapy of Meniere disease include bed resting, antiemetics and antihistamin therapy (6).

It is believed that since high salt diet causes water retention, it can lead to an increase of fluid within the inner ear (7). Avoidance of alcohol, cigarette and caffeine is advisable. Patients who still suffer from frequent vertigo attacks after initial therapy should be treated with local application of dexamethason or gentamicin in the middle ear (8). Surgical procedures as vestibular neurectomy, endolymphatic sac surgery and labyrinthectomy are necessary in 15% of patients with Meniere's disease (9). Since there is no specific diagnostic test for Meniere's disease, diagnosis is made *per exclusionem*.

One of the diseases with similar symptoms is spontaneous intracranial hypotension (SIH), originally described by Schaltenbrand in 1938 (10). The SIH is a syndrome of low cerebrospinal fluid (CSF) pressure characterized by postural he-

adaches in patients without any history of dural puncture or penetrating trauma (11). First symptoms are usually neurological, such as nausea, double vision, tremor, but, rarely, there are also hearing problems such as tinnitus or deafness (12). The exact mechanism of SIH is unknown. Three main theories about SIH include: cerebrospinal fluid (CSF) leakage via small tears, reduced CSF production and CSF hyper absorption (13).

Main symptom of SIH is new onset headache with prompt relief when the patient lies down and prompt recurrence of pain while sitting or standing, very similar to headache after lumbar puncture (12). The reduction in CSF volume and pressure or altered distribution lead to increase of blood volume and finally, dilatation and engorgement of intracranial veins (14).

Audio-vestibular symptoms are hearing loss, hyperacusis, tinnitus, dizziness and vertigo (15), all can be the result of venous engorgement in the internal acoustic canal (16). Another most common cause of these symptoms can be due to the loss of CSF, which decreases CSF pressure. If cochlear aqueduct is functional, lower CSF pressure will decrease pressure to perilymph, which leads to endolymphatic hypertension and expansion of endolymphatic compartment. This condition can mimic endolymphatic hydrops (17).

The most common site of the CSF leak in patients with SIH is the thoracic region (18). Several imaging techniques may be used to identify the exact area of leakage. Because the majority of CSF leaks produce only a small volume of fluid and are only intermittently active, radionuclide cisternography can be very useful in detecting and localizing them (19). A radioactive tracer, Indium-111, is introduced into the subarachnoid space via lumbar puncture, and its course is followed for 24-48 hours. Findings that are consistent with SIH include absence of tracer in the cerebral convexities and early presence of tracer in the kidneys and bladder (18). Rarely, a dural tear at the skull base may cause SIH; this should be suspected in individuals with symptoms of intracranial hypotension and CSF rhinorrhea (17). CT myelography and/or gadolinium-enhanced MRI scan may be of benefit if the site of CSF leakage is not identified by cisternography (18).

Initial treatment of SIH is similar to that of a post-lumbar puncture headache, combination of strict

bed rest and ample hydration. If the patient does not improve over the course of several days, then an epidural blood patch (EBP) on identified site of CSF leakage is the treatment of choice (20). For patients who remain symptomatic after EBP, surgical repair of the CSF leak should be considered. The aim of this study is to investigate similarity of clinical presentation of Meniere disease and spontaneous intracranial hypotension, where symptoms overlap and at what point should we introduce new examination to exclude differential diagnosis.

### PATIENTS AND METHODS

A 54 – year old male patient was admitted to our hospital for diagnostic work-up and treatment of Meniere's disease. Eighteen years before he had suffered from tinnitus in the right ear, vertigo and hearing loss in both ears with predominance of low frequency impairment in the right ear. After glycerol test and fluctuation of hearing by 20 dB, morbus Meniere was diagnosed. Radiographs of mastoid were also within normal limits. Other radiological work-up was not performed. He was treated conservatively with betahistinum, cinnemazine and diuretics.

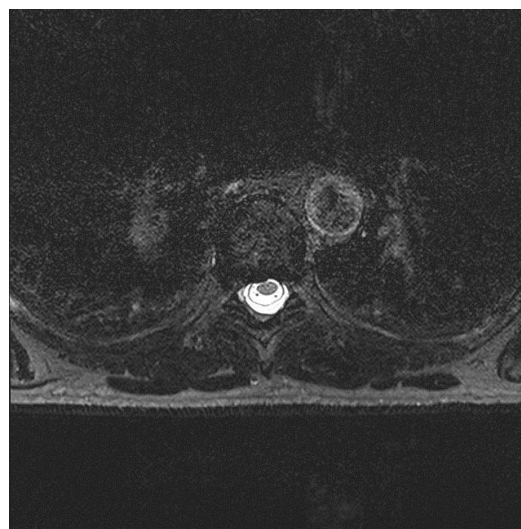
During the next few years, the patient suffered from headache, mostly in occipital and frontal region, combined with vertigo, nausea and vomiting. Intensity of these symptoms gradually increased. The patient started salt free diet and vertigo lessened. Other symptoms decreased when the patient lay down. When severe headache occurred, the patient noticed that it decreased if he performed "handstand" manoeuvre, but he stopped doing it when diplopia occurred.

In 2008 the patient reported severe hearing loss and lack of understanding speech. Audiogram revealed sensorineural hearing loss in the right ear at 60 – 70 dB, similar as in 1996, and aggravation of hearing loss in the left ear, between 40–85 dB. Speech perception was damaged, ability to understand speech in the left ear was 72% and in the right ear it was 36%. Behind-the-ear hearing aid was prescribed. Due to unsatisfying hearing condition, the patient changed the hearing aid. Speech perception test showed 25 % improvement with the new hearing aid.

In 2011 the patient started complaining of diplopia. At first it was double edged picture but soon

it turned to persistent everyday double picture displaced horizontally. Headache was still present. Ophthalmic examination showed vertical gaze nystagmus. Cranial computed tomography results were unremarkable.

Magnetic resonance imaging of brain (MRI) showed increased pachymeningeal contrast enhancement. Nuclear magnetic resonance (NMR) myelography detected collected liquid between Th 6-9, which was 7 cm long. In front of 7<sup>th</sup> and 8<sup>th</sup> thoracic vertebra, a small "pocket" of liquor (4.3x7.1x4 mm) was noticed (Figure 1).



**Figure 1.** Nuclear magnetic resonance myelography of thoracic spine of the patient showing small pocket of liquor in front of the 7<sup>th</sup> and 8<sup>th</sup> thoracic vertebra (University Hospital Clinic Dubrava, Zagreb, Croatia, 2011)

Neurosurgeon suggested radiological check-up in 3 months. The CT scan myelography showed the same area filled with contrast fluid as the previous time, but the "pocket" which was filled with liquor had disappeared. With this finding, it was concluded that spontaneous closing of the gap had occurred and neurosurgeon suggested not to perform a surgery.

### DISCUSSION

According to our knowledge this is the fifth described case of Meniere's disease and spontaneous intracranial hypotension coexistence (21-24). Meniere's disease is diagnosed when a combination of typical audiovestibular symptoms occurs (5). Due to an unknown cause, unknown pathogenesis, large number of symptom combinations, in 1995 the AAO HNS CHE set criteria

for degrees of Mb Ménière (5): certain - definite disease with histopathological confirmation, definite - requires two or more definitive episodes of vertigo with hearing loss plus tinnitus and/or aural fullness, probable - only one definitive episode of vertigo and the other symptoms and signs, possible - definitive vertigo with no associated hearing loss.

There is no certain diagnostic test that can determine Meniere's disease and exclude other similar diseases (2).

Neurological symptoms are usually more severe than the audio vestibular so it is more likely that a patient will first undergo thorough neurological examination than ENT diagnostic workup (18). During neurological examination, CT scans will be performed in the beginning of the work up and therefore neurological disease that mimic Meniere's disease, such as vestibular schwannoma and superior canal dehiscence will be detected (3).

Initial treatments of Meniere's disease and spontaneous intracranial hypotension are similar - bed rest, antiemetics and antihistamin therapy. In most patients, symptoms disappear after the initial treatment (6). But, if that conservative treatment fails, the next step will involve a more radical approach. Radical treatment for Meniere's disease includes local application of dexamethason or

gentamicin in the middle ear (8). If that fails, the next step in treatment is a surgical procedure - vestibular neurectomy, enolymphatic sac surgery and labyrinthectomy (9). Destructive surgeries are irreversible and involve removing entire functionality of most, if not all, of the affected ear.

If initial treatment of SIH fails, surgical procedure is recommended - epidural blood patch (EBP) on an identified site of CSF leakage (13).

According to the results described in this paper we suggest that extended neuroradiological examination of brain is recommended in patients with Meniere's disease to exclude underlying spontaneous intracranial hypotension syndrome. A multidisciplinary approach is necessary in the treatment of such patients.

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#### TRANSPARENCY DECLARATION

Competing interests: None to declare

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## Tanka linija između Menierove bolesti i spontane intrakranijalne hipotenzije

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### SAŽETAK

**Cilj** Istaknuti sličnosti kliničkog tijeka i dijagnostike, te različitosti liječenja Menierove bolesti i spontane intrakranijalne hipotenzije.

**Metode** Prikazan je slučaj pacijenta s dijagnosticiranom Menierovom bolesti kod kojeg je nakon dodatne neurološke i radiološke obrade postavljena i dijagnoza spontane intrakranijalne hipotenzije.

**Rezultati** Magnetskom rezonancom mozga s kontrastom prikazano je pojačano nakupljanje kontrasta u području dure i arahnoidalne ovojnice. Mijelografijom je prikazano istjecanje likvora. Bolesnik je uspješno izliječen konzervativnom metodom.

**Zaključak** Pregledom literature pronađena su četiri do sada opisana slučaja prisutnosti Menierove bolesti i spontane intrakranijalne hipotenzije kod istog bolesnika. Obje bolesti imaju slične simptome i slično početno liječenje. Predloženi su postupci dodatne obrade u slučaju neuspješnog liječenja i upitne inicijalne dijagnoze.

**Ključneriječi:** otološki vertigo, intrakranijaska hipotenzija, otjecanje cerebrospinalne tekućine