

Audiovestibular symptoms in spontaneous intracranial hypotension: Cues from two cases

Marianna Manuelli, Andrea Migliorelli, Marco Farneti¹, Laura Negossi, Chiara Bianchini, Stefano Pelucchi, Francesco Stomeo, Andrea Ciorba

Department of ENT and Audiology, University Hospital of Ferrara, Ferrara, Italy, ¹Department of Head and Neck, Neurosurgery Unit, University Hospital of Ferrara, Ferrara, Italy

Abstract

Spontaneous intracranial hypotension (SIH) is a rare condition that usually presents with posterior headache. A high percentage of patients with SIH experience audiovestibular symptoms, including vertigo, tinnitus, and hearing fluctuations. This pattern of symptoms is very similar to that observed in Ménière's disease; therefore, in selected cases, it is necessary to suspect SIH to formulate the correct diagnosis. Two patients with SIH and audiovestibular symptoms were studied at the Otolaryngology and Audiology Department of the University Hospital of Ferrara. Both the patients underwent audiometric testing, vestibular assessment, brain and spine magnetic resonance imaging (MRI) with contrast, neurosurgical evaluation, and treatment. A 75-year-old patient and a 53-year-old patient were referred to our center for vertigo exacerbated by postural changes, auditory fluctuations, and headache. In both the cases, contrast-enhanced brain MRI allowed to formulate the diagnosis of SIH, which was then confirmed by spinal MRI. Conservative treatment was attempted in both the patients, first, without therapeutic success; therefore, an epidural blood patch was performed with good control of symptoms. SIH is a rare condition that may present with symptoms similar to Ménière's disease; in selected cases, it is necessary to suspect an SIH, and a multidisciplinary approach, including a neuroradiological and a neurosurgical evaluation, is fundamental for the correct and timely diagnosis and intervention.

Keywords: Headache, hearing loss, spontaneous intracranial hypotension, vertigo

Address for correspondence: Prof. Andrea Ciorba, Department of Neuroscience and Rehabilitation, ENT and Audiology Unit, University Hospital of Ferrara, Via A. Moro 8 (Cona), Ferrara 44124, Italy.

E-mail: andrea.ciorba@unife.it

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is a rare condition characterized by orthostatic headache mainly located at the back of the neck, which affects approximately 2–5 people per 100,000, with peak incidence around the fourth decade.^[1-3] SIH is defined by a cerebrospinal fluid (CSF) pressure <60 mm H₂O in a patient with no

recent record of head trauma, neurosurgery, or lumbar puncture.^[4,5] The exact mechanism of SIH is unknown, but three different possible etiopathogenic mechanisms have been hypothesized, namely: loss of CSF through small tears, reduced CSF production, and CSF hyperabsorption.^[6-8] Headache, as a consequence of CSF hypotension, arises

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in orthostatism and worsens throughout the day while improving when supine.^[1,3]

About half of the patients also experience auditory symptoms such as tinnitus, fluctuating hearing loss, and fullness, which may initially lead to a suspected diagnosis of Ménière's disease.^[1]

The currently most widely accepted mechanism to explain hearing loss in SIH suggests that a drop in perilymphatic pressure may cause a compensatory increase in endolymphatic pressure, resulting in hearing loss similar to that reported in Ménière's disease.^[2,9]

Vestibular manifestations of SIH include vertigo and dizziness associated with headache that usually occur in the orthostatic position and resolve on assuming a supine position.^[1]

Contrast-enhanced brain magnetic resonance imaging (MRI) is the most reliable diagnostic method for diagnosing SIH, showing diffuse, symmetrical, nonnodular meningeal enhancement.^[3] For diagnostic purposes, it is also essential to perform a spine MRI scan to exclude CSF leakage, which can occur mainly in the thoracic region or at the cervicothoracic junction.^[10]

Treatment initially involves conservative measures such as bed rest, caffeine, and oral hydration. In cases not responsive to these measures, epidural blood patching or percutaneous placement of fibrin sealant may be performed.^[3] In case epidural blood patch proves ineffective, continuous saline infusion can be used to treat SIH.^[11]

Once remission of symptoms has been achieved, brain MRI shows no pathological alterations.^[12]

Audiological symptoms are present in a high percentage of cases, but the clinical presentation, which is extremely suggestive of Ménière's disease, can lead to delayed diagnosis or misdiagnosis.^[13] It is therefore essential to be familiar with this rare condition to identify the clinical picture and propose an effective and timely treatment.

PATIENTS AND METHODS

Two patients with SIH and audiovestibular symptoms have been evaluated at the Otolaryngology and Audiology Department of the University Hospital of Ferrara between 2019 and 2025. Medical history and audiological symptom assessment, audiometric testing, vestibular assessment, brain and spine MRI with contrast, neurosurgical evaluation, and blood patching were performed in both the cases.

The present study was conducted in accordance with the Declaration of Helsinki (2008). It was conducted retrospectively through a systematic review of hospital case histories and therefore did not affect patient care in any way.

RESULTS

Case 1

The first case is a 75-year-old male patient with Parkinson's disease and type 2 diabetes mellitus. For 2 months, the patient had been experiencing vertigo and dizziness triggered by postural changes, particularly when moving from a cline to the orthostatic position. He also complained of headaches that preceded or were combined with vertiginous symptoms.

The patient underwent a bedside examination, which proved negative, showing no spontaneous nystagmus, a negative head-shaking test, and the absence of positional nystagmus during the McClure–Pagnini and Dix–Hallpike maneuvers.

Tonal audiometry showed moderate–severe bilateral asymmetrical sensorineural hearing loss, with a bilateral down-sloping curve, worse on the right side [Figure 1].

Caloric tests showed right vestibular hypofunction, while the auditory brainstem response showed a bilateral absence of wave I and bilateral increased values of III and V waves.

MRI scan of the brain and petrous bones with contrast enhancement was therefore performed, which showed meningeal enhancement after gadolinium administration in the absence of recent trauma, CSF fistulas, or lumbar punctures. The MRI images displayed in Figure 2 led to a suspected SIH diagnosis, which was then confirmed by spinal MRI.



Figure 1: Audiometry of case 1

The neurosurgical evaluation initially prescribed conservative treatment with oral steroids, bed rest, and adequate hydration, but this approach did not control the symptoms properly. A blood patch was therefore performed successfully in February 2019. Subsequent MRI scans proved negative, and the patient observed a regression of vestibular symptoms and stability of hearing, with excellent gain from hearing aids.

Case 2

A 53-year-old male patient with no significant medical history was investigated at another hospital for 8 years for suspected Ménière’s disease. The patient reported progressive bilateral hearing loss, worse on the left side, bilateral tinnitus, and sporadic episodes of vertigo associated with headache and triggered when moving from a cline to an orthostatic position. The bedside examination showed no abnormalities with the absence of spontaneous nystagmus, negative head-shaking test, and absence of positional nystagmus during the McClure–Pagnini and Dix–Hallpike maneuvers. Video head impulse test (vHIT) was negative with normal vestibulo-ocular reflex (VOR) gain for all semicircular canals, and caloric tests showed bilateral normal function.

Tonal audiometry revealed moderate–severe sensorineural hearing loss, worse on the left side [Figure 3].

The patient then underwent a brain MRI, which raised the diagnostic suspicion. Spinal MRI excluded the presence of fistulas. After neurosurgical evaluation, the patient was first treated conservatively, but the treatment was ineffective. Hence, he also underwent blood patching in December 2024, achieving good control of symptoms. The patient currently has no vertigo symptoms, tinnitus has improved, and hearing is stable.

DISCUSSION

SIH is a rare, not widely known, and challenging to diagnose disease, which implies a decreased CSF pressure, resulting in orthostatic headache.

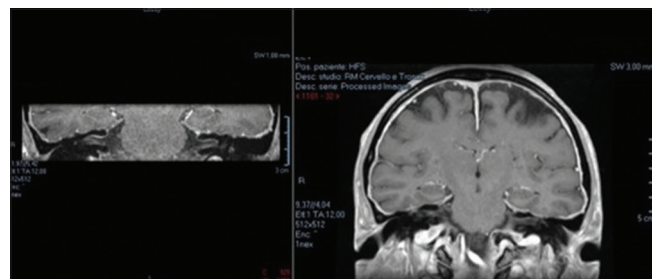


Figure 2: Magnetic resonance imaging of case 1 showing meningeal contrast enhancement

Other symptoms may also be observed, such as interscapular pain, diplopia, visual field alterations, cognitive and personality alterations, instability, tremor, and cervical radicular symptoms.^[14]

Audiovestibular symptoms such as fluctuating hearing loss, vertigo, and tinnitus affect more than half of the patients in association with headache. Since headache is a nonspecific symptom shared by many diseases, it is essential to recognize the features of this condition to successfully identify SIH cases.^[15]

In the first treated case, data concerning orthostatic headache were decisive and, together with the instrumental findings, allowed to include SIH in the differential diagnosis, even though the main suspicions at first were Meniere’s disease or a retrocochlear pathology. When collecting patients medical history, it is crucial to evaluate carefully the associated symptoms, their timing and onset, as in the presented case.

In the second case, the SIH diagnosis was challenging, as for 8 years, the patient received another diagnosis, and headaches were reported as sporadically. The experience of the previous case helped to raise the diagnostic suspicion and indicate the need for an MRI scan, including the spine. Eventually, also vestibular migraine could be cited among the differential diagnoses; however, the type of hearing loss, with a progressive feature in SIH, and the headache features in particular, have been reported to be useful to reach the appropriate diagnosis.^[13]

Both the cases analyzed attempted a conservative therapeutic approach with bed rest, oral steroids, and abundant hydration, but these measures did not allow symptom control. Therefore, according to the neurosurgical

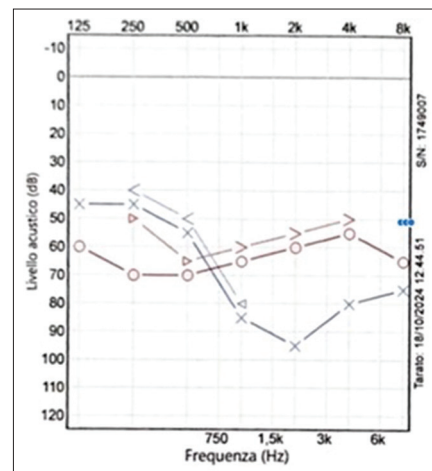


Figure 3: Audiometry of case 2

evaluation, both the patients were treated by epidural blood patching, achieving good control of symptoms, including audiovestibular symptoms.

Since SIH often presents with audiovestibular symptoms attributable to Ménière's disease, it is essential to conduct a thorough medical history, particularly regarding headache and its presentation, as the onset of symptoms in an upright position is highly suggestive; indeed, in the presented cases, particularly at the very beginning of the clinical signs presentation, the symptoms were very similar to those of Meniere's disease.^[16] In our opinion, a multidisciplinary approach including a neuroradiological and a neurosurgical evaluation is fundamental for the correct and timely diagnosis and intervention.^[7,15]

The main limitations of this study relate to the low number of cases identified. It will be therefore necessary to expand the study by collecting more cases on a multicenter basis and with an exhaustive and standardized diagnostic pathway to obtain as much information as possible on the disease and its pathophysiology.

CONCLUSIONS

SIH is a rare condition that can present with audiovestibular symptoms mimicking Meniere's disease. Anamnestic features can guide the diagnosis, which can be very complicated if not carefully considered.

Imaging assessment is essential and decisive in distinguishing SIH from other diseases, and its management must necessarily be multidisciplinary. Further studies with a larger number of cases will be necessary to investigate the disease also to better evaluate its pathophysiology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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