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CASE REPORT

Spontaneous Intracranial Hypotension Syndrome with Bilateral Hearing Loss and Hyperacusia: A Case Report and Review of the Literature

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Objective: To present a case with bilateral hearing loss, hyperacusia, and dizziness as components of spontaneous intracranial hypotension mimicking labyrinthine hydrops.

Methods: Herein, in addition to a literature review, we present a case of spontaneous intracranial hypotension syndrome that was the cause of otologic symptoms, which was successfully treated with a high-volume epidural blood patch.

Results: A 40-year-old woman presented with orthostatic headache, emesis, dizziness, bilateral ear fullness, hyperacusia, and hearing loss. Audiography showed bilateral low-frequency hearing loss. Cranial magnetic resonance imaging (MRI) and MRI myelography showed increased pachymeningeal contrast enhancement, subdural collections, venous engorgement of the internal acoustic canals, and an extrathecal cerebrospinal (CSF) fluid leak. Lumbar puncture opening pressure in the lateral decubitus position was 50 mmH_2O. CSF protein, microscopic examination, and culture were normal. The patient was diagnosed with spontaneous intracranial hypotension syndrome. After conservative treatment failed a high-volume epidural blood patch was applied. The patient had no otologic complaints after this treatment and had normal audiogram results. Additionally, follow-up cranial MRI showed minimal pachymeningeal contrast enhancement and the absence of subdural collections.

Conclusion: Spontaneous intracranial hypotension syndrome, which is curable, should be considered in patients with orthostatic headache and symptoms mimicking labyrinthine hydrops.

Keywords: Spontaneous intracranial hypotension, orthostatic headache hearing loss, hyperacusia, labyrinthine hydrops-like symptoms, dizziness.

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Introduction

Spontaneous intracranial hypotension (SIH) is a syndrome characterized by orthostatic headache, low CSF pressure, and increased pachymeningeal contrast enhancement on cranial MRI. Although these are the primary features of the classic syndrome, some cases have non-orthostatic headache, normal cerebrospinal fluid (CSF) pressure, and no evidence of diffuse meningeal enhancement in MRI scans [1]. This syndrome was first described by Schaltenbrand in 1938 [2]; however, initial misdiagnosis is still common [3]. SIH is not a common disorder and occurs in 1 in 50,000 individuals, with a female-male ratio of approximately 2:1 [4]. Symptoms typically begin with a peak incidence at around 40 years of age, but children and the elderly may also be affected [5].

Various neurological scenarios can be observed and cranial nerve involvement has been reported. Nausea, vomiting, phonophobia, photophobia, diplopia, dysgeusia, neck pain and stiffness, tinnitus, hearing loss, and vertigo may be associated symptoms [5]. Herein we report a SIH case with hearing loss, hyperacusia, and dizziness, with pathognomonic MRI characteristics and orthostatic headache, which only responded to a high-volume epidural blood patch.

Case Report

A 40-year-old otherwise healthy woman experienced the sudden onset of severe headache 1 week before referring to our tertiary referral hospital. The patient complained of a frontal headache that was increasing in severity, and was more severe while in the upright

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position, but relieved by recumbency. The patient's medical history was non-contributory. She had bilateral aural fullness, tinnitus, hearing difficulties, hyperacusia, and mild dizziness. Photophobia, nausea, and vomiting were associated symptoms. She did not have blurred vision or diplopia. Physical and neurological examination results were within normal limits, except for mild nuchal stiffness. Vital signs were within normal limits. Laryngological examination revealed no pathology.

The patient had severe headache when she was in sitting position. Therefore, audiogram tests

were performed in semi-recumbent position.

Audiological evaluation showed a bilateral low frequency hearing loss. (Figure 1). Impedancemetric tests were normal. She did not have spontaneous, gaze-evoked, positional, or positioning nystagmus. The Romberg sign was negative. Past-pointing and Unterberger test results were unremarkable. CBC, sedimentation rate, CRP, and routine blood test results were within normal limits. Cranial computed tomography (CT) results were unremarkable.

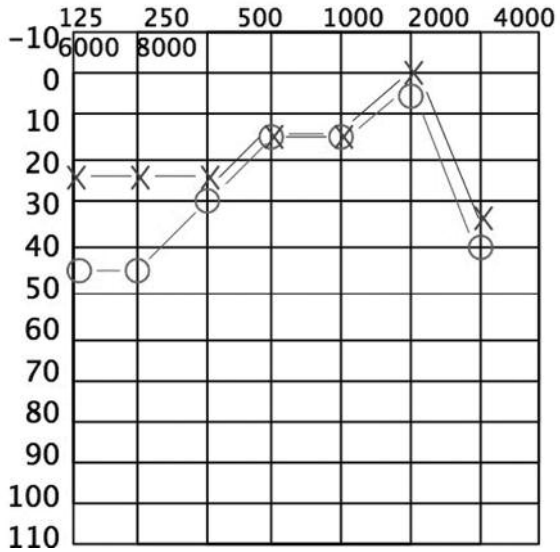

Figure 1. Audiogram shows bilateral threshold decrease in the low frequencies.

Magnetic resonance imaging (MRI) with gadolinium showed increased pachymeningeal contrast enhancement (Figures 2). The patient was treated conservatively with a 2-week period of strict bed rest and hydration; however, her headache did not respond well to this symptomatic therapy. With conservative treatment her hearing loss disappeared, but dizziness, hyperacusia, and tinnitus persisted. Follow-up pure tone audiogram results were within normal limits (Figure 3); however, repeat MRI showed bilateral frontal subdural effusion and increased contrast enhancement (Figure 4). Additionally, venous engorgement in the internal acoustic canals was observed (Figure 5). Lumbar puncture opening pressure in the lateral decubitus position was 50 mmH_2O (normal range: 60-120 mmH_2O). CSF cytology, protein, microscopic examination, and culture were normal. As such, we conducted MR myelography (spinal MRI following intrathecal injection of gadolinium), which showed extrathecal CSF (Figure 6). Then, a high-volume epidural blood patch was applied to 3 regions with 80 cc of autologous blood under fluoroscopic guidance. Following application of the epidural blood patches, most of the patient's complaints disappeared. Her headache and other otologic symptoms disappeared, but the patient reported minimal residual tinnitus. Two months post treatment there were no subdural collections and cranial MRI showed minimal pachymeningeal contrast enhancement.

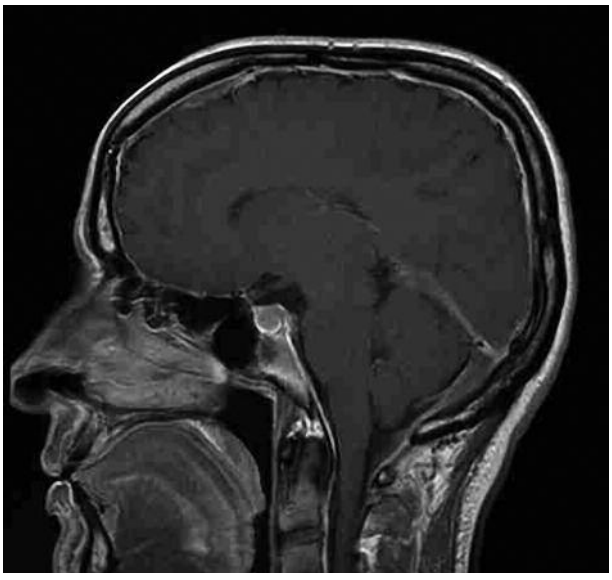

Figure 2. Post-gadolinium, sagittal T1-weighted MRI shows enhancement of meninges.

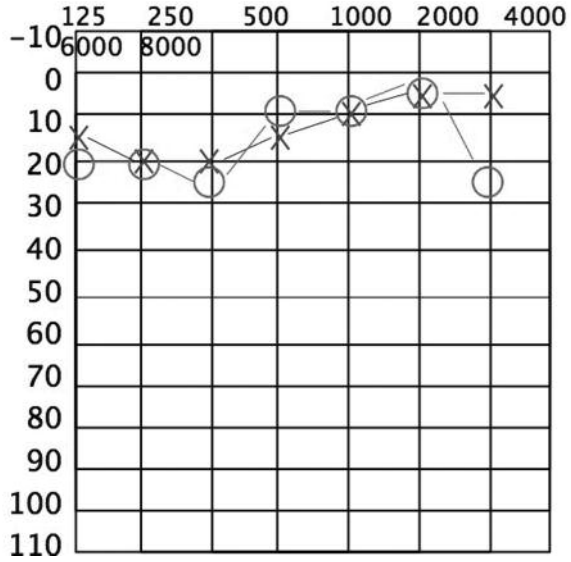

Figure 3. Follow-up audiogram shows bilateral normal hearing thresholds.

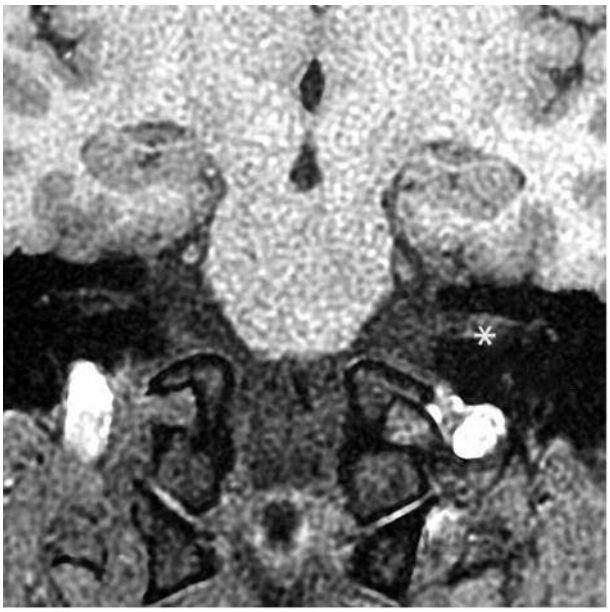

Figure 5. Coronal T1-weighted MRI after iv Omniscan injection shows venous engorgement of the internal acoustic canal.

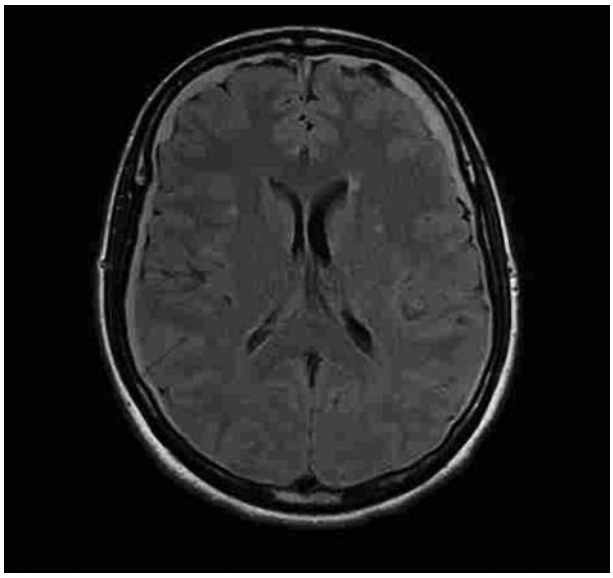

Figure 4. Axial FLAIR MRI sequence shows subdural effusion.

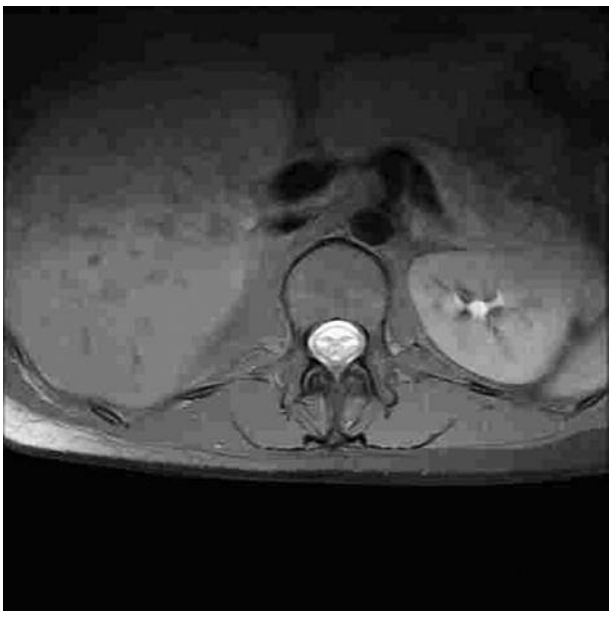

Figure 6. Axial T1-weighted MRI after intrathecal Magnevist injection shows epidural contrast leakage.

Discussion

SIH syndrome is caused by spontaneous CSF leakage (in the absence of a medical history suggesting a cause for CSF loss, such as trauma, lumbar puncture, etc). Although comprehensive population-based epidemiologic studies have not been conducted, the estimated incidence of SIH was reported as 5 per 100,000, with a female predominance [5].

The etiology of SIH remains unknown. Most CSF leaks occur at the thoracic and cervicothoracic junction via arachnoid membrane rupture [6]. Additionally, an underlying connective tissue disorder or dural weakness has been suspected to be the cause [7].

The exact mechanism of SIH also remains unknown. Initial proposed theories for SIH included prolonged CSF leakage via small tears, reduced CSF production, and hyperabsorption of CSF [8]. Mokri proposed the term CSF hypovolemia should be used for this syndrome instead of intracranial hypotension and low CSF pressure [1]. On the other hand, Schievenk suggested the term spontaneous spinal CSF leak was more appropriate than CSF hypovolemia [5]. Due to the reduction in CSF volume and pressure, or altered distribution, blood volume increases, leading to dilatation and engorgement of meningeal veins [9]. Diffuse engorgement of intracranial venous structures can be seen in patients with SIH [10]. In addition, venous engorgement in the internal acoustic canal might contribute to the clinical picture, as reported in a recently published case [11].

Moreover, minor head trauma, a fall, an inciting event, or sports activities that result in dural tears of the vulnerable dural nerve sheath could be contributing factors in SIH. There is a history of minor trauma in 33% of SIH patients [12]. The International Classification of Headache Disorders (ICHD-2) revised 2004 diagnostic criteria for SIH are orthostatic headache and pachymeningeal enhancement on cranial MRI, and the recommended treatment is an epidural blood patch. [13]. Due to the broad spectrum of clinical and radiographic manifestations of this syndrome, Schievink et al. published new diagnostic criteria for spontaneous CSF leakage and intracranial hypotension in 2008 (Table) (3).

Table 3. The distribution of SNHLs and CHLs in RAG and CG

Criterion A	Demonstration of a spinal CSF leak (i.e., presence of extrathecal CSF). When criterion A is not met use criterion B.
Criterion B	Cranial MRI changes in intracranial hypotension (i.e. presence of subdural collections, enhancement of the pachymeninges, or sagging of the brain) and the presence of at least 1 of the following: low opening pressure (<60 mmH_2O) spinal meningeal diverticulum improvement of symptoms after epidural blood patching. When criteria A and B are not met use criterion C.
Criterion C	The presence of all of the following or at least 2 of the following if typical orthostatic headache is present: 1) low opening pressure (<60 mmH_2O) 2) spinal meningeal diverticulum 3) improvement of symptoms after epidural blood patching.

The cardinal symptom of SIH is orthostatic headache of sudden or gradual onset. This intense, throbbing, or dull pain is due to displacement of pain-sensitive structures [14]. Headache pain is relieved by recumbency within minutes. Upright posture, head movement, coughing, sneezing, or high altitudes may exacerbate this symptom [15]. Associated clinical symptoms include neck pain and stiffness, tinnitus, nausea, photophobia, audio-vestibular complaints, blurred vision, diplopia, and dysgeusia [5].

Audio-vestibular symptoms consist of hearing loss, hyperacusia, tinnitus, dizziness, and vertigo [11,15-18]. CSF, perilymph, and endolymph pressure are normally equal [19]. The loss of CSF decreases CSF pressure. This decrease is thought to be due to transmission of abnormal CSF pressure to the perilymph via a patent cochlear aqueduct, producing perilymphatic hypotonia, which results in relative endolymphatic hypertension and compensatory expansion of the endolymphatic compartment, mimicking endolymphatic hydrops [11,17,20]. It has been suggested that neurosurgery, lumbar punctures, spinal anesthesia, myelography, and traumatic events may cause CSF leakage, thus decreasing CSF and perilymphatic fluid pressure, which translates into transient endolymphatic hydrops [20-23].

Portier et al. classified hydrops into 2 groups: 1. Syndromes of endolymphatic origin (Meniere's disease, autoimmune processes) and 2. Syndromes of perilymphatic origin, in which loss of perilymph induces compensatory expansion of the endolymphatic space (spontaneous or traumatic perilymphatic fistula and intracranial hypotension with a patent cochlear aqueduct) [20]. The incidence of auditory symptoms in a series of 30 patients with SIH was as follows: tinnitus: 20%; plugged ear: 20%; hearing loss: 3% [16]. The low incidence of documented audio-vestibular symptoms in SIH patients might be attributed to the lack of a patent cochlear aqueduct [17,24].

In a recently published report Isildak et al. described venous engorgement in the internal acoustic canal veins, and speculated that audio-vestibular symptoms may be related to this condition [11]. In the presented case we also observed bilateral venous engorgement in the internal acoustic canal veins on MRI scans and our patient also had bilateral low tone hearing loss, tinnitus, hyperacusia, and dizziness. These 2 findings might be related to each other, and an irritation-like effect mimicking labyrinthine hydrops could be considered in patients with SIH, yet the exact mechanism is currently unknown.

The gold standard for diagnosis of SIH is gadolinium-enhanced cranial MRI and measurement of lumbar CSF opening pressure [12]. The characteristic abnormal features of SIH on cranial MRI are diffuse meningeal enhancement, subdural hematomas or hygromas, engorgement of venous structures, pituitary enlargement, sagging of the brain, and a decrease in the size of cisterns and ventricles [5]. Despite the fact that MRI results are abnormal in most patients with SIH, cranial MRI results are normal in up to 20% of cases [5].

Cranial CT results in patients with SIH are usually normal; however, subdural fluid collections, slit-shaped ventricles, and obliteration of subarachnoid cisterns have been reported [25]. Myelography with iodinated contrast, followed by thin-cut CT scanning or MRI are necessary to identify the location and extent of CSF leakage [7]. Single or multiple meningeal diverticula can be detected with myelography. Most CSF leaks occur at the cervicothoracic junction or along the thoracic spine nerve root [5]. Multiple simultaneous CSF leaks can also be observed. .

Radioisotope cisternography is a technique that detects the accumulation of a tracer outside the normal confines of the subarachnoidal spaces [26]. The most common finding is limited radioisotope activity over the cerebral convexities [27]. If myelography is non-diagnostic, then this technique is particularly useful for identifying the location of CSF leaks. Nevertheless, the exact site of CSF leakage remains undetected in 33% of patients [7]. Simultaneously, cotton pledgets may be placed in the nose for the detection of radioactivity, and also aids in the detection and localization of a possible rhinorrhea.

Patients with SIH typically have a CSF opening pressure <60 mmH_2O (normal range: 65-195 mmH_2O) [14]. The cell count is usually mildly elevated, but there may be lymphocytic pleocytosis of up to 200 cells mm–3.

Persistent headache unrelated to SIH, subarachnoid hemorrhage, arterial dissection, cerebral venous thrombosis, meningitis, pseudotumor cerebri, and post-trauma headache should be taken into account in the differential diagnosis [26]. Conservative therapy, epidural blood patch, and surgery are options to stop CSF leakage and restore CSF volume. First-line treatment, which is conservative therapy, simply includes hydration and bed rest in a horizontal position [26]. Caffeine intake, theophylline, and steroids were reported as treatment choices with limited effectiveness [5].

When conservative treatment methods fail, an epidural blood patch (EBP) is a widely used treatment option. This technique consists of infusion of 10-30 mL of autologous blood into the epidural space. Epidural blood patching procedures in the thoracic and cervical region necessitate radiographic guidance. Such treatment usually provides rapid relief, presumably via forming a dural tamponade and replacement of lost CSF, and is also diagnostic. Patients should be placed in the Trendelenburg position for 30-60 minutes. If a patient still complains of headache, then a high-volume (up to 100 mL) epidural blood patch is the choice for infusion [26]. This high-volume technique (80-mL infusion) was successfully used in the presented case. The epidural blood patch technique can be repeated after a minimum of 5 days if the headache persists.

Other therapeutic techniques include intrathecal saline infusion and placement of fibrin sealant. If all of the above conservative methods fail to provide relief, then

the next step is surgical treatment. Structural abnormality, meningeal diverticula, and dural defects may require surgical repair of the CSF leak. Fibrin sealant, suture ligations, and clipping constitute surgical options [26]. Schievink reported that 10% of patients had CSF leakage recurrence; however, there has been no long-term follow-up studies [5,26]. In addition, it has been suggested that patients with a focal spinal CSF leak and abnormal brain MRI results have an excellent prognosis, while those with normal initial MRI findings and a diffuse multilevel spinal CSF leak have a poor prognosis [5].

In conclusion, we reported a patient with SIH. SIH can be associated with hearing loss, hyperacusia, and dizziness, as in the presented case. A review of literature and the presented case highlight the fact that otological presentation of SIH may mimic labyrinthine hydrops. This rare and treatable clinical entity is under-recognized by clinicians and must be included in the differential diagnosis of audio-vestibular disorders.

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