CASE REPORT

A case of spontaneous intracranial hypotension: From Ménière-like syndrome to cerebral involvement

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KEYWORDS
Spontaneous intracranial hypotension; Cerebrospinal fluid; Ménière-like; Orthostatic headache; Hydrops ex vacuo

Introduction

Spontaneous intracranial hypotension (SIH) is a rare pathology, with annual incidence estimated at five per 100,000 of the population [1]. It results from CSF leakage, the cause of which is unknown but which may be associated with trauma in a context of fragile spinal meninges (meningeal diverticula, abnormal support tissue) [2]. The key symptom is orthostatic headache, but other less specific signs may be associated and may sometimes mislead diagnosis. Diagnosis is founded on a range of clinical and paraclinical data (Box 1) [3]. The present article reports a case of severe SIH, with initially misleading Ménière-like cochlear symptomatology.

Case report

A 47-year-old man consulted in ENT for tinnitus and hearing loss. History included mild spinal trauma sustained 2 months previously following a fall from a bicycle.

The patient had been suffering for 3 weeks from low-frequency tinnitus and a “blocked ear” sensation with “metallic noise”, especially in the left ear. He also,
Box 1: Diagnostic criteria for spontaneous intracranial hypotension [3].

Diagnostic criteria for spontaneous intracranial hypotension:

A) Evidence of spinal leakage of CSF (CSF in extrathecal space), or else;
B) Signs of intracranial hypotension on gadolinium-enhanced MRI (subdural effusion, diffuse meningeal contrast uptake, or signs of cerebral involvement);

And

At least one of the following:

1. CSF hypotension at lumbar puncture ($\leq 60$ mmHg);  
2. Meningeal diverticula;  
3. Symptom regression secondary to epidural blood-patch;

Or, failing criteria A and B

C) All of the following, or at least two in case of typical orthostatic headache:

1. CSF hypotension at lumbar puncture ($\leq 60$ mmHg);  
2. Meningeal diverticula;  
3. Symptom regression secondary to epidural blood-patch;

Exclusion criterion: iatrogenic or post-traumatic CSF leakage.

secondarily, complained of bifrontal retro-orbital morning headache.

Clinical ENT and vestibular examination was normal. Pure-tone audiometry found left perceptual hearing loss, predominating at low frequencies (Fig. 1).

Although no vertigo was reported, endolymphatic hydrops was suspected and exploratory treatment with acetazolamide (DIAMOX®) and betahistine (BETASERC®) was prescribed, and led to recovery of hearing but with persistent tinnitus.

Two weeks later, the patient was awoken by intense retro-orbital headache, exacerbated on orthostasis and accompanied by vomiting.

Emergency cerebromedullary MRI found bilateral subdural hematoma, with a chronic aspect, associated with diffuse meningeal uptake of contrast medium and incipient involvement of the cerebellar amygdalae. There were L3-L4 and S3-S4 meningeal cysts. Given such signs of intracranial hypotension, emergency epidural blood-patch was performed.

One hour later, there was sudden onset of somnolence with aphasia and stereotypic crumbling-type hand movements. A brain scan was immediately taken and found increased left subdural hematoma exerting a mass effect inducing deviation of medial structures. The patient was transferred the same day to neurosurgery for drainage of the hematoma under general anesthesia. Medical management to boost intracranial pressure comprised strict bed-rest in the Trendelenburg posture, adapted hydration, corticosteroid therapy and administration of caffeine [4].

Over the following 2 weeks, the right subdural hematoma increased, requiring iterative surgical drainage. At the 3rd intervention, an intrathecal lumbar catheter was fitted for continuous physiological saline perfusion to restore CSF pressure [5]. Postoperative myelo-CT found contrast medium leaking from the right D12 nerve root sheath toward the paravertebral muscles, confirming the diagnosis of dural tear (Figs. 2 and 3); the radiologists injected biologic glue [6] (TISSUCOL®) to seal off the leakage, leading to regression of all symptoms and radiologic abnormalities.

Discussion

Orthostatic headache is the key symptom of spontaneous intracranial hypotension. Other less specific symptoms, however, may predominate or be found in isolation, posing a real diagnostic challenge.

The auditory symptoms generally associated with SIH are a sensation of "blocked ear", auditory distortion, tinnitus and hearing loss, which are all highly suggestive when orthostatic [7]. Audiometry generally finds mild to moderate uni- or bilateral perceptual hearing loss, mainly affecting low frequencies; although it tends to be transitory, cases of deficit persisting after successful treatment of SIH have been reported.

The mechanism of these symptoms was first described by Carlberg et al. [8]. There is a balance of pressures between endo- and perilymphatic compartments, mediated by the CSF. Each compartment is in continuity with the CSF: the perilymph, via the cochlear aqueduct (if permeable) and the endolymph via the endolymphatic sac. The authors proved experimentally that intracranial pressure changes are transmitted to the inner ear by passive liquid transfer — rapid if the cochlear canal is permeable, and slow when it is non-functional. On this hydromechanical hypothesis, when CSF pressure falls perilymphatic pressure rapidly falls in parallel, inducing endolymphatic hydrops, clinically comparable to that found in Ménière’s disease, which the symptoms may mimic perfectly [9,10]. The physiopathology of this hydrops ex vacuo, however, remains entirely unknown.

In the present case, the diuretic treatment probably triggered the neurologic deterioration, which may have aggravated a latent intracranial hypotension. This highlights the importance, in case of a presentation of atypical hydrops, of complementary assessments, which were not performed in the present case: neurologic assessment, fundus examination, early MRI for differential diagnosis, and electrocochleography for positive diagnosis of hydrops. The reasons for the failure of the blood-patch are unclear: diffusion beyond the epidural space, or second dural break caused by the lumbar puncture?

Once the tear had been identified, myelo-CT-guided injection of biologic glue enabled complete and lasting resolution of symptoms.
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Figure 1  Tonal and vocal audiograms: left perceptual hearing loss. Above, pure-tone audiogram: RE = right ear; LE = left ear; blue, air conduction; red, bone conduction. Below, vocal audiogram: blue, left ear; red, right ear.

Figures 2 and 3  Myelo-CT: contrast medium leakage facing right D12 root sheath.
Conclusion

The present case is one of severe SIH. The failure of iterative neurosurgery to drain the subdural effusion raises doubts as to the role of this strategy in the management of severe SIH. It is not unusual for ENT specialists to be faced with this pathology, given the auditory symptoms that may perfectly mimic Ménière’s disease. Paraclinical examinations are indispensable before initiating diuretic treatment when the presentation is atypical of Ménière’s disease or incomplete. Headache, however mild, should in this context suggest a diagnosis of SIH, and cerebromedullary MRI should be performed in case of the slightest doubt.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References