Spontaneous Intracranial Hypotension Syndrome: An Unknown and Benign Condition

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Abstract

Background: Spontaneous intracranial hypotension (SIH) is considered to be an uncommon disease caused by cerebrospinal fluid leakage. It is characterized by an orthostatic headache without history in the past of trauma or dural puncture. There is no consensus about level, volume and number of epidural blood patch (EBP) for SIH treatment in literature.

Methods: Our objective was to report a 38-year-old woman with SIH at two different levels (lumbar and cervical) as demonstrated by spine MRI and SPECT CT, treated by single lumbar low-volume EBP.

Results: The patient achieved good symptoms relief, with complete remission of headache, nausea and vomits. Besides, no complications occurred as consequence of the procedure and the discharged happened two days afterwards.

Conclusion: In spite of low volume used in the blood patch and of the existence of another CSF leak in a distant site, it is possible to obtain good results and at same time, to minimize complications due to higher blood volume patches application.

Keywords: Spontaneous Intracranial Hypotension; Epidural Blood Patch; Headache

Introduction

Spontaneous intracranial hypotension (SIH) is a rare well-recognized clinical syndrome that probably is undiagnosed cause of headache [1]. It is caused by a cerebrospinal fluid (CSF) leakage from spinal meningeal diverticula or dural rents along nerve sleeves [2]. Usually, SIH is manifested by a postural headache syndrome of acute or subacute onset, unrelated to dural puncture, surgery or trauma, although sometimes it is associated with underlying connective tissue disorders, manifests SIH. Meningeal diverticula and SIH have been described in association with Marfan’s syndrome, and autosomal dominant polycystic kidney disease, neurofibromatosis type 1 and Lehman syndrome [1,3] [3].

Clinically, SIH (first described by German neurologist Schaltenbrand, in 1938) mimics the presentation of the classic post-dural puncture headache syndrome, in which the mark is an orthostatic headache. The headache may be holocranial or localized to frontal or occipital regions. It probably is caused by CSF volume loss and subsequent traction on the pain-sensitive intracranial dura due to downward brain displacement [1]. Besides headache, other clinical features may include neck pain, nausea, vomiting, dizziness, diplopia, photo-
phobia, balance problems, vertigo and tinnitus, marked exacerbation by Valsalva manoeuvre, occur in patients with low CSF pressure (< 60mm H₂O) [3-5]. Among several treatment options such as bed rest, epidural blood patch (EBP) or surgical closure, EBP provides the most efficient treatment modality for SIH [6].

In 2013, the Headache Classification Committee of the International Headache Society proposed revised diagnostic criteria for SIH, as shown in Table 1 [7].

<table>
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<tr>
<th>Diagnostic criteria for spontaneous intracranial hypotension proposed by Headache Classification Committee of the International Headache Society 2013</th>
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<tr>
<td>A. Any headache fulfilling criterion C</td>
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<tr>
<td>B. Low CSF pressure (&lt; 60 mmHg) and/or evidence of CSF leakage on imaging</td>
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<tr>
<td>C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery</td>
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<td>D. Not better accounted for by another ICHD-3 diagnosis</td>
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*Table 1: The opening pressure depends on the patient’s position. For the healthy patient in the prone flat position, it is around 3 cm higher than in the lateral decubitus position.*


Reproduced from [7](7).

Although several articles have described EBP for treatment of SIH, the exact volume, level and number of EBP are still controversial. We report one patient with two identified sites for CSF leaks that received a single lumbar autologous EBP with a good outcome.

**Case Report**

**Presentation and examination**

A 38-years-old female reported a long history of occipital headache of gradual progression and chronic onset (since the childhood). Besides, nausea and vomiting was present and all those symptoms were worse in standing or sitting position and got better when she lying down. She had had previous history of three epidurals anesthesia, but no complications occurred during the procedures. All the symptoms got worse 4 years after the last delivery (5 years ago). No history of trauma was mentioned.

Physical examination revealed no abnormalities. No signs of CSF leak were noted. She had no neurological deficits and the neurological examination was normal, except for a slight horizontal nystagmus. She complained of headache 8/10 on the visual analog scale (VAS). Routine hematological and biochemical tests were normal. Low cerebrospinal fluid pressure (6 mm H₂O) was seen through the spinal tap.

As standardized, conservative measures were used, she underwent to bed rest, intravenous fluid hydration analgesics (Tylenol®) and caffeine within 2 weeks, which had no benefits. As the conservative management has failed, then epidural blood patch was the next step.

**Radiologic aspects**

Magnetic resonance imaging (MRI) of neuro axis showed diffuse dural enhancement, mainly in the head, in T1-weighted image, as well as cerebellar tonsillar herniation. All the suprasellar and preptontine cisterns were effaced, since the structures were downward displaced. However, no subdural collections were seen.

Cervical and lumbar MRI showed gross appearance of CSF leak in approximately C3-C4 and L4-L5 levels, led us suspect the diagnosis of spontaneous intracranial hypotension, in spite of we could not localize any meningeal diverticulum.
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SPECT CT/Radionuclide cisternography revealed focal areas of increased activity on bilateral regions of the lumbar paraspinal region, meaning CSF accumulations and thus, suggesting CSF leaking in that point. Besides, visualization of the root sleeves, considered an abnormal finding, was possible. Therefore, the presence of early activity of the bladder was marked.

**Blood patch**

A 21-gauge Tuohy needle, using a midline approach with the patient in the lateral decubitus was chosen. Appropriately, anti-sepsis was done and fluoroscopic guidance was helpful in localize the interspace L4-L5. In the meantime, blood was took out from the antecubital vein, keeping strict anti-sepsis. The epidural space was identified using the loss of resistance technique and all the 15 mL of autologous blood was slowly injected, without any complain from the patient. No additional injection was performed even one had already known about the presence of cervical CSF leak. The patient received a dose of 2g intravenous cefazolin as standard antibiotic prophylaxis.

**Post-procedure course**

Postoperatively, she was kept in the supine position for two days. Her neurologic examination remained normal and she had a good recovery. In the same day after the procedure, relief from the headache occurred. Her symptoms ameliorated, and she was discharged two days afterwards, keeping the same conservative therapies and bed rest for 15 days more. Spine CT and MRI of the head (imaging finds have improved? Blood patch visualized in CT scan or MRI?)

The patient is still asymptomatic without any headache or symptoms, as previously mentioned, so far.

![Figure 1: T1 axial MRI demonstrating gadolinium-enhanced in pachymeninge (white arrow).](image1.png)

![Figure 2: T1 sagittal MRI demonstrates tonsillar herniation (white arrow) and decrease of mamillopontine distance, meaning sagging of the brain.](image2.png)

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Figure 3: T2 coronal cervical spine MRI shows CSF leak in the left C3-C4 interspace.

Figure 4: A 4 hours radionuclide cisternography with evidence of early visualization of bladder activity (black arrow) and CSF leakage at the lumbar level (arrowhead).

Figure 5: SPECT/CT localizes foci of increased uptake, suggesting cervical spine CSF leak.

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Discussion

Despite the advance in image methods over the last years, SIH is still misdiagnosed. Spontaneous CSF leak is by far, the most common cause of SIH and for this reason, many times a challenge for the diagnose [1].

The pathophysiology of spontaneous CSF leak is not completely understood, although the presence of meningeal diverticulum have been postulated and seems to be a prerequisite. That aspect might represent a weakness at the spinal meninges, creating a favorable condition to the CSF leaks and, as consequence the origin of intracranial hypotension [1].

The incidence of SIH is approximately 5 per 100,000 [4]. Symptom’s onset is usually in the fourth or fifth decade of life and occurs in females twice as often as males. The most common sites for CSF leaks are the lower cervical and upper thoracic spine [4].

As general stand of care, bed rest, IV hydration, analgesics and caffeine are the mainstay of the treatment. However, whether the conservative treatments fail, others methods should be try. With this regard, blood patch figures as effective treatment and must be the next option [9].

In 1992, Rando and Fishman reported two cases of SIH in which radionuclide cisternography demonstrated localized CSF leaks. They hypothesized the CSF leak was due to the spontaneous rupture of a spinal arachnoid (Tarlov) cyst [8]. Since then, the spontaneous spinal CSF leaks are thought to be the most common cause of SIH. Although the exact etiology of spontaneous spinal CSF leaks is still unknown, but they probably involve weakness of the meninges surrounding the spinal cord [1].

The incidence of SIH is approximately 2 - 5 per 100,000 [4,9,10] (9) (10). Symptom onset is usually in the fourth or fifth decade of life and occurs in females twice as often as males (2:1) [9,7]. The most common sites for CSF leaks are the lower cervical and upper thoracic spine [4].

Neuroradiologic evaluation is crucial in the diagnosis and management of SIH. Recently, advances in diagnostic tools such as MRI, CT myelography and radionuclide cisternography dramatically facilitated the diagnosis of intracranial hypotension [1,2,5,6,11] (2) (1) (6) (11). The main imaging findings of SIH in CT and MRI scans reflect CSF volume loss and compensatory increased venous blood volume [1].

At present, radionuclide cisternography is considered the gold standard for the detection of CSF leakage when SIH is suspected [5]. It has demonstrated that direct findings of CSF leakage on radionuclide cisternography are defined as focal areas of increased activity on unilateral or bilateral regions of paraspinal area [5].

Importantly, the presence of until 4 indirect findings on radionuclide cisternography may help the diagnosis. Those are early visualization of bladder activity (1 - 3 hours after intrathecal injection of In-111 diethyl-enetriamine penta-acetic acid, 111In-DTPA), and no visualization of activity over the brain convexities (no remarkable accumulation around the brain convexities 24 hours after injection).

Additionally, rapid disappearance of spinal activity (1 - 5 hours after injection), and abnormal visualization of the root sleeves (asymmetric activity outlining the spinal nerve roots, any time after injection) might occur [5]. In a retrospective review, early visualization of bladder activity was seen in all 27 patients [5].

Treatment of SIH aims to maintain CSF volume [2]. Although autologous EBP is widely accepted as a major treatment modality for SIH and for post-dural puncture headaches, the mechanism responsible for its effect has not been determined. It may involve a temporary increase in epidural pressure, the sealing of a tear by coagulating injected blood, or the initiation of inflammatory reactions that promote dural tear healing [6]. Other speculative physiopathological hypothesis proposes the participation of the spinal venous drainage system as cause of epidural hypotension [12].

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Besides, the maintenance of low epidural pressure, by the inferior cava vein outflow to the heart, could be created by the presence of large 1-way valve collector veins in constitutionally predisposed individual. Then, the goal of blood patch procedure could be not to seal CSF leaks, but instead to help in reversing the CSF-blood gradient within the epidural space along the entire cord.

Autologous EBP is highly effective in the management of SIH and, in general, pain relief is obtained immediately [2]. Success rates for the first EBP for SIH range from 52% to 87%, although targeted EBP by spinal imaging appears to be more effective than blind EBP [6]. Generally, EBP can safely be performed in spite of complications, including backache and back stiffness have been documented [2].

Direct approaches have risks such as iatrogenic epidural hematoma, compression of the spinal cord and nerve roots, chemical meningitis, intrathecal blood injection, arachnoiditis, and may present difficulties in narrow cervical or thoracic epidural spaces [2,6] (6). Furthermore, only small volume of blood can be injected at the cervical or thoracic level as compared with the lumbar level, due to the risk of neurological complications [6].

Thus, blind epidural EBP through lumbar space represent a relatively wide and safe area [6]. Furthermore, blindly patched blood through lumbar route may spread to the cervico-thoracic level in the Trendelenburg position, and could restrict CSF leakage, balancing the CSF pressure [13]. In terms of efficacy, blind EBP for SIH might reach 90%, even when placed at a location far from the CSF leak’s site or when the CSF leakage site was not determined [6].

Concerning the volume and number of EBP, there is still not consensus. The data about the volume of autologous blood patch used is conflicting, ranging from 9 mL [6] to 70 mL [14] per procedure. Larger volumes of up to 100 mL could be injected for the second treatment if the first one is unsuccessful [10]. An alternative to high blood volume is the injection of blood with fibrin glue at the level of suspected leak(s) [7]. Still, some patients require to perform similar technique twice, even three times, depending on whether or not the symptoms returned [3,4] (3).

Conclusion

Spontaneous intracranial hypotension is a treatable cause of headache and more severe neurological symptoms such as encephalopathy [10]. No consensus has been reached regarding the optimal method between targeted and blind EBP [6]. In spite of some controversies about the appropriate blood volume to be injected, our case has shown good results using a single small one. Therefore, even when multiple CSF leak is present, is possible to achieve complete resolution of them performing one single application and at a distant site. Randomized prospective trials are still necessary to further evaluate the efficacy of different treatment modalities of SIH.

Conflicts of Interest and Source of Funding

The authors have no funding or conflicts of interest to disclose.

Bibliography


