# Spontaneous Intracranial Hypotension with Cerebrospinal Fluid Leakage at 2 Sites Treated by Epidural Blood Patch

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**Abstract-**

***Purpose:*** Spontaneous intracranial hypotension (SIH) is a relatively uncommon cause of headache, which usually presents as orthostatic cranial pain and is relieved by recumbency. The precise cause of spontaneous spinal cerebrospinal fluid leakage related SIH remains unknown.

***Case Report:*** We report the case of a 32-year-old man who presented with an orthostatic headache. Brain magnetic resonance imaging (MRI) revealed typical pachymeningeal enhancement. Radionuclide cisternography revealed leakages in the cervicothoracic and upper cervical areas. The patient was successfully treated by lumbar epidural blood patch (EBP).

***Conclusion:*** The diagnosis of SIH involves the assessment of the characteristic clinical presentations and non-invesive neuroimaging studies. The latest diagnostic criteria with more broadened spectrum due to variable manifestations are discussed. EBP is an effective treatment for SIH if conservative management fails.

**Key Words:** orthostatic headache, intracranial hypotension, magnetic resonance imaging, cisternography, blood patch

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## INTRODUCTION

Spontaneous intracranial hypotension is an uncommon cause of headache in adults, and the headache typically presents as an orthostatic headache that is relieved by recumbency. Persons with SIH may also experience symptoms such as dizziness, nuchal pain, nausea, vomiting, diplopia, blurred vision, or postural tremor[1-3]. Intracranial hypotension was first described in 1938 by George Schaltenbrand[4]. Thereafter, in the 1990s, MRI of the brain revealed a characteristic feature of intracranial hypotension-pachymeningeal enhancement[5]. Untreated SIH may be complicated with subdural hemorrhage or cerebral sinus thrombosis[6-8]. An epidural blood patch may be an effective treatment for SIH if conservative management fails[1,2,9-12]. We report the case of a man who presented with orthostatic headache; the findings of neuroimaging and radionuclide cisternogra-

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phy were characteristic of SIH, and the patient was successfully treated by a lumbar epidural blood patch.

## CASE REPORT

A 32-year-old man experienced occipital headache while performing work that involved lifting of heavy weight. The headache was throbbing and localized in the posterior region of the neck and the parietal areas of both sides. Subsequently, the headache intensified, and blurred vision, nausea, and vomiting occurred several days later. The patient stated that the headache aggravated when he assumed an upright posture (while sitting or standing) and alleviated when he lying down, and it was so severe that he was unable to work. When he was admitted to our department, he denied any history of a traumatic accident or lumbar puncture examination. Physical and neurological examinations were performed, and the results of both were normal. The results of brain computed tomography (CT) with and without enhancement were also normal. However, brain MRI, magnetic resonance (MR) angiography, and MR venography showed apparent intracranial pachymeningeal enhancement without any vascular abnormality (Fig. 1A). A cervical spine MRI revealed dural enhancement on the ventral side and collection of the retrospinal fluid at the upper cervical level (Fig. 1B). Moreover, a constrast-enhacement T1-weighted image revealed a paraspinal

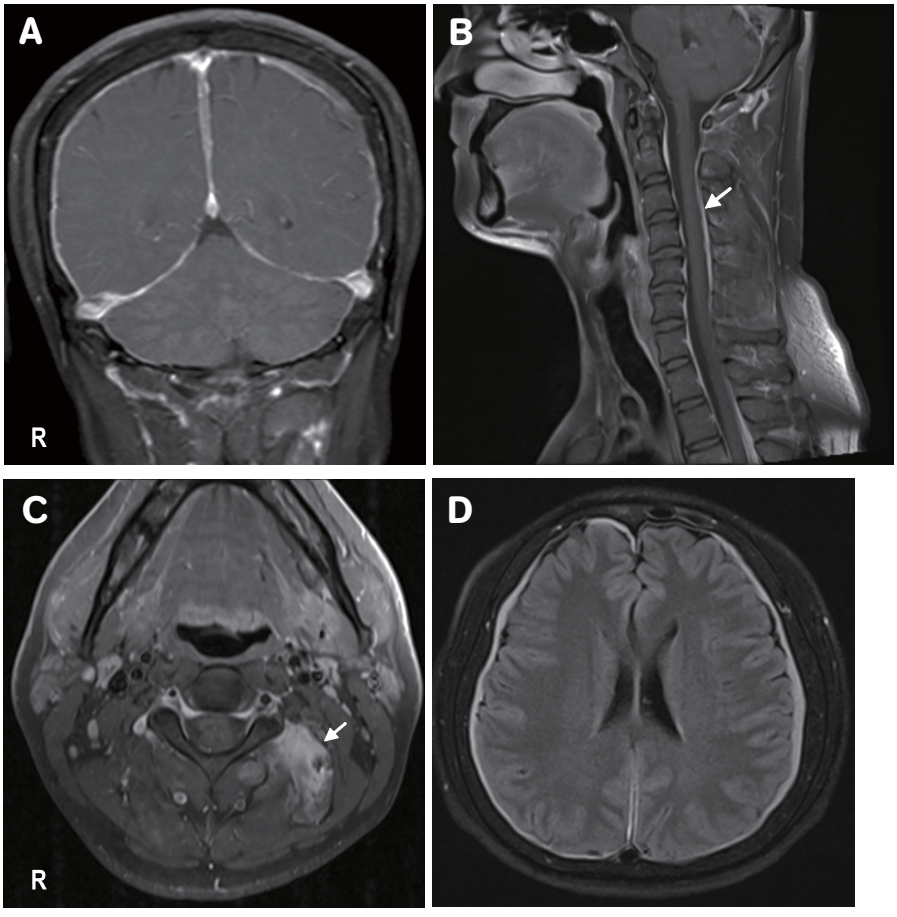


**Figure 1.** T1-weighted brain magnetic resonance image (MRI) in the coronal view with gadolinium enhancement shows diffuse pachymeningeal enhancement (A). T1-weighted spinal MRI in the sagittal view with enhancement shows ventral and dorsal dural enhancement (B, indicated by the white arrow). Spinal MRI in the axial view shows the paraspinal soft tissue enhanced lesion on the left side in the C1-C2 area (C, indicated by the white arrow). Brain MRI on fluid-attenuated inversion recovery (FLAIR) image in the axial view shows diffuse pachymeningeal hyperintensity (D, indicated by white arrows).

soft tissue lesion on the left side with high signal density at the C1-C2 level (Fig. 1C). A CT-guided biopsy was performed, and the results revealed tissue fragments and the absence of abscess or malignancy.

The patient's headache was relieved on the 3rd day of admission by conservative treatment, including bed rest, daily hydration with 2000 mL of normal saline, and administration of a nonsteroidal anti-inflammatory drug. However, the orthostatic headache recurred on day 8. Lumbar puncture for cerebrospinal fluid (CSF) studies and radionuclide cisternography were performed on day 11. The CSF studies revealed the following findings: clear CSF; leukocyte count, 10 /μL; total protein level, 93 mg/dL; and open pressure, 80 mmCSF. The radionuclide cisternography indicated CSF leakage in the upper cervical and cervicothoracic junction, and early visualization of radioactivity was noted in the kidney and urinary bladder (Fig. 2A, 2B). On day 13, EBP was performed because of the failure of conservative treatment. A total of 10 mL of the patient's own blood was slowly injected in the L2-L3 spinal epidural space. The headache improved immediately after EBP but recurred on the next day. Due to still orthostatic headache even after EBP , the patient ask transfer to medical center for further management. A follow-up spinal CT myelography in the medical center did not show CSF leaks or paraspinal lesions in the cervical area. The orthostatic headache finally resolved 7 days after the EBP.

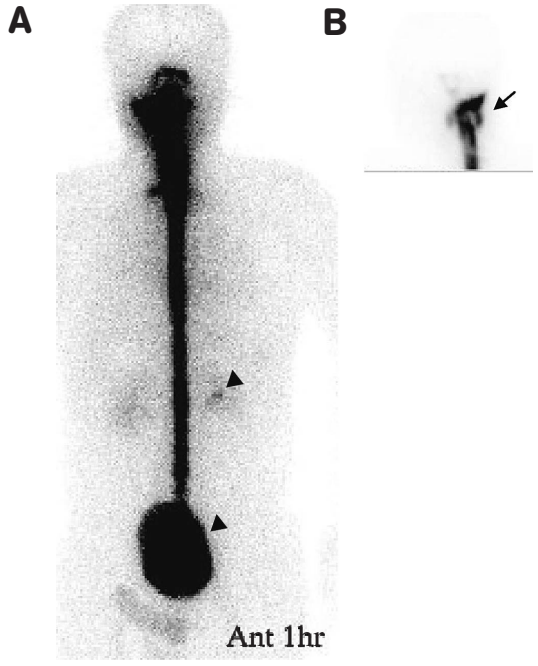


**Figure 2.** Radioisotope cisternography shows early visualization of radioactivity in the kidney and urinary bladder (A, indicated by the black arrow head). The absence of activity is noted over the cerebral convexities, and the leakage area is located in the upper cervical and the cervicothoracic level; B, indicated by the black arrow).

## DISCUSSION

Intracranial hypotension is a relatively uncommon but important cause of headache. Improvements in neuroimaging techniques have led to an increase in the identification rate of the cases of SIH with headache. However, some patients may be misdiagnosed with migraine, tension headache, viral meningitis, or malingering[11]. The diagnosis criteria for headache due to SIH, which indicate that the cardinal feature of SIH-related headache is a diffuse or dull headache that worsens within 15 min after sitting or standing, were established by the International Headache Society in 2004[12] (Table 1); the associated symptoms include neck stiffness, tinnitus, photophobia, nausea, and hyperacusia. However, the diagnostic criteria have been revised because of the broad spectrum of clinical presentations in patients with SIH (Table 2)[13]. A trivial trauma may precede the onset of symptoms[14]. The typical MRI findings in these patients are diffuse pachymeningeal gadolinium enhancement, subdural collection of fluid, and evidence of descent of the brain[1]. The pachymeningeal enhancement may be caused by dural vasodilation and concentration of gadolinium in the microvasculature and interstitial fluid of the dura mater[15]. Due to the more various clinical manifestations of the SIH, the recently developed diagnostic criteria included four diagnostic components: A, orthostatic headache; B, the presence of at least one of the following: low opening pressure, improvement of symptoms after EBP, demonstration of an active CSF leak, cranial MRI changes of intracranial hypotension; C, no recent history of dural puncture; and D, not

**Table 1.** International Headache Society's diagnostic criteria for spontaneous intracranial hypotension

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| A. Diffuse and/or dull headache that worsens within 15 minutes of sitting or standing, with at least one of the following and fulfilling criteria D |
| 1. Neck stiffness |
| 2. Tinnitus |
| 3. Hypacusia |
| 4. Photophobia |
| 5. Nausea |
| B. At least one of the following |
| 1. Evidence of low CSF pressure on MRI |
| 2. Evidence of CSF leakage on conventional myelography, CT myelography or cisternography |
| 3. CSF opening pressure < 60 mmH2O in sitting position |
| C. No history of dural puncture or other cause of CSF fistula |
| D. Headache resolves within 72 hours after epidural blood patching |

**Table 2.** Revised diagnostic criteria for headache due to spontaneous intracranial hypotension

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| A) Demonstration of a spinal CSF leak (i.e. presence of extrathecal CSF), or, if criterion A not met |
| B) Cranial MRI changes of intracranial hypotension (i.e. presence of subdural fluid collections, enhancement of the pachymeninges, or sagging of the brain) |
| And |
| The presence of at least one of the following: |
| 1. low opening pressure (≦60 $mmH_2O$) |
| 2. spinal meningeal diverticulum |
| 3. improvement of symptoms after epidural blood patching |
| or, if criteria A and B not met |
| C) The presence of all of the following or at least two of the following if typical orthostatic headaches are present |
| 1. low opening pressure (≦ 60 $mmH_2O$) |
| 2. spinal meningeal diverticulum |
| 3. improvement of symptoms after epidural blood patching |
| Patients with onset of symptoms following dural puncture or other penetrating spinal trauma are excluded. |
| CSF, cerebrospinal fluid; MRI, magnetic resonance imaging. |

attributable to another disorder[16]. Mainstay management of the headache includes conservative treatment by hydration and EBP. The treatment using EBP is not always effective within 72 h, and complete resolution of the symptoms may occur after second or more EBPs are performed[13].

During daily work, this patient had orthostatic headache along with neck pain, dizziness, and nausea, and the headache could be relieved by recumbency. The brain MRI showed apparent intracranial pachymeningeal enhancement. A FLAIR image revealed diffuse pachymeningeal hyperintensity (Fig. 1D). Both the diffuse pachymeningeal hyperintensity and diffuse pachymeningeal gadolinium enhancement noted by FLAIR MRI and the T1-weighted brain MRI, respectively, are indicative of SIH[17]. The region of CSF leakage may be indicated by extradural fluid collection on the ventral side, dural enhancement, and retrospinal CSF collection that are noted by the cervical spine MRI[18]. It has been reported that CSF leak in the C1-C2 region could be a false localizing sign in SIH[19]. However, the infiltrative lesion of the paraspinal soft tissue on the left side found in this patient could have occurred because of a leakage in the upper cervical area due to the pathological examination of the paraspinal lesion revealed only tissue fragments. Moreover, a spinal CT myelography performed after treatment by EBP revealed that the lesion had disappeared[20]. Although the outcome of SIH is mostly good, response to EBP may be inconsistent, this is why additional survey such as biopsy and CT myelography were performed in this patient. The recent noninvasive heavily T2-weighted magnetic resonance myelography was developed for localizing CSF leaks for patient with SIH, and the technique may arrange prior to CT myelopathy for clinically suspect SIH patients[21]. Radionuclide cisternography confirmed the CSF leakages of the patient in the upper cervical and cervicothoracic areas. Radionuclide cisternography is useful for better diagnosis while planning the management of SIH, and it may reveal the direct signs of SIH in up to 80% of the patients[22].

EBP was performed for treating this patient because hydration alone failed to relieve his headache. Although EBP remains the treatment of choice for SIH, even with a blind epidural blood patch[2,10], the first EBP performed

treats only about 57% of the cases; hence, a second or third EBP may be considered if the symptoms do not subside[9,10]. An EBP should be close to the region of the CSF leak to be effective[1] as called target EBP; however, a remote EBP distal to the CSF leak may also cause remission of the headache[1,9,10]. Su et al. reported that an SIH patient was treated by EBP on the lumbar area (L2); thereafter, a spinal MRI performed 1 h later revealed that most of the injected blood had spread from the lumbar area in the direction of the cranium[2]. Although EBP is a safe procedure, contraindications, including systemic sepsis, coagulopathy, and local infection at the puncture site, should be considered[23]. Radiculopathy or intracranial hypertension may occur after the EBP, but this occurrence is unusual[24].

SIH is an uncommon but not rare disorder in clinical practice; neurologists as well as general physicians should be aware of this syndrome. The diagnosis of SIH involves the assessment of the characteristic clinical presentations and neuroimaging studies. Untreated SIH may delay the course of resolution, or it may even be complicated with subdural hemorrhage. EBP is an effective treatment for SIH if conservative management fails.

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