Spontaneous Intracranial Hypotension with Magnetic Resonance Localisation of Spinal Cerebrospinal Fluid Leak

*Yahya H. Al-Brashdi,1 Sameer Raniga,2 Sharma R. Revati2

**Abstract:** To increase the awareness of spontaneous intracranial hypotension (SIH), we report in this paper a middle-aged woman who presented with an intractable headache that worsened in sitting and standing positions (a postural headache). Magnetic resonance imaging (MRI) of the spine demonstrated a cerebrospinal fluid (CSF) leak at the level of the cervical spine, in addition to typical features in a brain MRI, including symmetrical subdural collections, circumferential dural enhancement and features of midbrain sagging. The patient underwent a surgical repair of the cervical CSF leak which resulted in a dramatic symptomatic improvement that was confirmed by follow-up imaging.

**Keywords:** Hypotension, spontaneous intracranial; Cerebrospinal Fluid; Hematoma, subdural; Case Report; Oman.

**Case Report**

A middle-aged, 50-year-old woman, with no previous medical complaints, presented with a three-month history of headaches that were aggravated while sitting or standing but relieved by lying down. The headaches increased in severity one month prior to presentation, during which time it was associated with nausea and vomiting. She gave no history of other symptoms such as neck pain or
visual complications. A neurological examination was completely normal.

An MRI of the brain showed small bilateral cerebral subdural collections which were more conspicuous in FLAIR images [Figure 1A]. Post-contrast images showed bilateral circumferential pachymeningeal dural enhancement even involving the tentorium cerebelli [Figure 1B]. MRI features of brain sagging were also noted, including a closed pontomesencephalic angle, a reduced space between the dorsum sella and the mammillary body, and squaring and fattening of the midbrain. Reduction of the space between the sella turcica and the optic chiasm, abnormal positioning of the pituitary stalk (so that it was almost covering the sella) and buckling of the medulla were also seen [Figure 1C]. An MRI of the whole spine was also done, including a deep three-dimensional constructive interference in steady state (3D-CISS) sequence which showed a left-sided cervical CSF leak and accumulation at the level of the C2 vertebral body [Figure 1D]. No spinal subdural collection or other spinal CSF leaks were noticed. On the basis of these MR findings, SIH was diagnosed, and no additional imaging modalities were required to confirm the diagnosis.¹

Due to unavailable local expertise in performing an epidural blood patch, surgical exploration of the cervical spine at the C1, C2 and C3 levels was performed. It revealed a tiny area of CSF leak at the 3 o'clock position at the level of the C2 vertebral body. Tissue glue was applied with absorbable gelatin powder (GELFOAM®, Pfizer, Inc., New York, USA) placed over it. Dramatic improvement in the patient’s symptoms was noticed after surgery.

A follow-up MRI of the brain and spine was done after two months, which showed an interval size reduction of the subdural collections and dural enhancement, except for a small residual subdural collection in the right hemisphere [Figure 2A]. A 3D-CISS sequence of the cervical spine was repeated in the follow-up series showing the absence of a cervical CSF leak [Figure 2B].

Discussion

SIH is defined as a syndrome characterised by reduced CSF pressure that occurs in the absence of dural puncture, surgery, or trauma. It has been postulated that the most likely pathogenesis is an occult CSF leak leading to a decrease in CSF...
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The main presenting feature in those patients is headache, which is aggravated by sitting or standing, and relieved by lying down.\(^1\)\(^-\)\(^3\) There might be associated symptoms including nausea, vomiting or neck stiffness, with other features suggestive of cranial nerve neuropathies including vertigo and photophobia. Arterial cerebral infarcts are rare but potentially life-threatening complications of SIH.\(^4\) A lumbar puncture usually reveals a low opening pressure of less than 60 mm H\(_2\)O, or the measurement of pressure may not be possible.\(^1\)

The typical MRI findings of SIH are well-established in the literature, including symmetrical bilateral subdural collections/effusions which are isointense to CSF in standard MR sequences.\(^1\) These collections show high signal intensity in FLAIR images which can be attributed to dural thickening.\(^1\)\(^,\)\(^3\)\(^,\)\(^5\) The most important intracranial MRI finding is diffuse pachymeningeal enhancement in T1-weighted post-contrast images.\(^1\)\(^,\)\(^3\)\(^,\)\(^6\) Differential diagnoses include meningitis, meningeal metastasis, chronic subdural haematoma, post-surgical dural thickening and dural sinus thrombosis.\(^7\) The patient in this study showed bilateral subdural collections with evidence of a similar pattern of meningeal enhancement [Figures 1A and 1B]. The cause of this dural enhancement is believed to be related to vascular dilatation, mainly venous congestion, as has been described by Fishman and Dillon.\(^1\)\(^-\)\(^3\)

According to the Monro-Kellie rule, the CSF volume fluctuates with intracranial blood volume in an intact skull.\(^1\)\(^,\)\(^3\) Features of downward brain displacement can be seen upon MRI, including a closed pontomesencephalic angle, reduction of the space between the \textit{dorsum sella} and mammillary body, the \textit{sella} almost being covered by the pituitary stalk, and fattening of the midbrain with buckling of the \textit{medulla}. Other imaging findings are enlargement of the pituitary gland, prominence of the spinal epidural venous plexus, engorgement of the cerebral venous sinuses (venous distension sign, etc.), venous sinus thrombosis and isolated cortical vein thrombosis.\(^3\) Our patient demonstrated all the features of brain sagging, indicating the presence of intracranial hypotension.

In 2008, Schievink \textit{et al.} described three criteria for diagnosing SIH.\(^8\) Criterion A is considered when spinal CSF leak is demonstrated in any imaging modality by the presence of extrathecal CSF. Our patient met this criterion according to her spinal MRI results. If no spinal CSF leak can be shown, criterion B can be considered if the patient has brain features of intracranial hypotension in addition to a low opening pressure (<60 mmHg), spinal meningeal \textit{diverticulum} or improvement of symptoms by epidural blood patch. If criteria A and B are not met, criterion C can be considered in the presence of all or at least two of the following: low opening pressure, spinal meningeal \textit{diverticulum} and symptomatic improvement following epidural blood patch.

An MRI of the spine can also be helpful, although it is not necessary in the diagnosis of SIH. It can...
demonstrate the presence of subdural collections with the ability to localise the site of the CSF leak in some cases. In this case report, a deep 3D-CISS sequence was very beneficial in pinpointing the level of the CSF leak at the C2 cervical spine, with it being more obvious in the left side. Intrathecal contrast-enhanced MR cisternography has increased both the sensitivity and specificity for detecting cranial and spinal CSF leaks with other clinical entities. Despite these clinical values, intrathecal injection of gadolinium has not yet been approved by the United States Food and Drug Administration, due to the risk of behavioural and neurological disturbances.9

A new developing MRI technique in evaluating a spinal CSF leak is the use of a spinal subtraction MRI, in which the T1-weighted scan is subtracted from the T2-weighted scan to reveal the extradural CSF collection. Bonetto et al. were able to show the extradural CSF collection in all of the SIH-included patients in their study in 2011.10

In cases where the MRI of the spine is negative in regard to the site of CSF leak, conventional and digital subtraction myelography, CT myelography or RC can be used. Digital subtraction myelography has been described as a valuable diagnostic tool to demonstrate rapid spinal CSF leak.11 Hoxworth et al. included 11 patients in their study in 2012; 6 were diagnosed with SIH and 5 with superficial siderosis. The site of the leak was demonstrated in 9 of the 11 patients (82%) using subtraction myelography. However, in addition to the radiation exposure, a few limitations were encountered with the digital subtraction myelography. These included the possibility of motion artifacts, limited assessment of CSF leaks at the level of the cervical and upper thoracic spine and reduced sensitivity in locating slow CSF leaks. In RC, a delay in radionuclide upward movement is usually seen, indicating low CSF pressure.1 However, low spatial resolution and the lack of cross-sectional images in RC make it less sensitive for spinal CSF leaks.9 In spite of their diagnostic values, these imaging modalities all use ionising radiation, except for the MRI. In addition, an MRI has better soft tissue differentiation with no radiation issues.

Most cases of SIH resolve spontaneously;7 however, in cases where treatment is required, management consists mainly of an epidural blood patch with other conservative measures.1–3 A CT-guided epidural blood patch is considered the most effective tool for treating patients with SIH after which dramatic improvement is usually observed.12,4 Conservative management includes bed rest, analgesics, use of sedatives and oral caffeine, and intravenous hydration.1–4 Mohammed et al. reported a young male diagnosed with SIH who became symptom-free after conservative treatment only. Surgical intervention is sometimes required when a blood patch fails or if subdural haematomas present with acute clinical deterioration.7 A patient is considered improved after the procedure when the patient’s symptoms disappear and by the resolution or near-resolution of subdural collections, or by pachymeningeal enhancement.6 It was noted that our patient improved significantly after surgery, when a follow-up MRI scan revealed the almost near-resolution of the previously noted imaging features.

**Conclusion**

Intracranial hypotension can be caused by previous lumbar punctures, trauma and previous spinal surgeries leading, in some cases, to life-threatening complications. However, SIH has no precipitating factors, with postural headache as the main presenting symptom. Although CT myelography or RC can be used in the diagnosis of SIH, MRI has the advantage of being a non-invasive modality with no ionising radiation involved. Despite the fact that a good number of MRI studies done for SIH turn out to be normal, our patient’s MRI clearly demonstrated the site of the spinal CSF leak in addition to the intracranial manifestations. Conservative measures and epidural blood patch should be the primary treatment strategies used in such patients prior to any surgical procedures.

**References**


