

## Spontaneous Intracranial Hypotension Due to CSF Leakage in a Healthy Adult Man: A Case Report

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**Received:** July 24, 2023; **Published:** January 11, 2024

### Abstract

Spontaneous intracranial hypotension (SIH) is a rare neurological disorder caused by cerebrospinal fluid (CSF) leakage, leading to reduced intracranial CSF pressure. This case report presents the clinical course and management of a healthy 44-year-old adult male who presented to the emergency department with a two-day history of fatigue and a severe headache. The primary symptom of SIH is an orthostatic headache that worsens upon standing and improves upon lying down. The patient underwent various imaging modalities, including brain and spinal computed tomography (CT) and magnetic resonance imaging (MRI), which confirmed the diagnosis of spontaneous intracranial hypotension due to CSF leakage. The treatment plan involved conservative measures, including bed rest, IV caffeine, and dexamethasone, followed by targeted epidural blood patching. Although the first epidural blood patch failed, a second attempt was successful in resolving the headache. The patient showed significant improvement in his symptoms and level of consciousness and was eventually discharged without any residual neurological deficits. This case highlights the importance of early diagnosis and appropriate management of SIH to prevent complications and improve patient outcomes. Clinicians should consider SIH in the differential diagnosis of postural headaches, especially in patients with a history of CSF leaks or connective tissue disorders. Early recognition and intervention are crucial to achieve favorable outcomes in patients with SIH.

**Keywords:** Headache; Spontaneous Intracranial Hypotension; CSF Leakage; Healthy Adult; Neurosurgery

### Introduction

Spontaneous intracranial hypotension (SIH) is an infrequent neurological illness [1] with an incidence rate that is half that of subarachnoid hemorrhage (SAH) [2]. Although its occurrence is not well proven, one paper made the assumption that it occurs in 1 in 50,000 people per year [3]. The majority of SIH instances are caused by a cerebrospinal fluid leak through a slit in the dura, which most usually occurs at the exit zones where the cervical spinal roots emerge into the subarachnoid space. SIH has been linked to connective tissue diseases including Marfan's syndrome [4]. Since an orthostatic headache is the primary symptom of this condition, it is often neglected in the diagnosis and frequently results in a misdiagnosis. The headache typically worsens within 15 minutes and gets worse during the day [5,6]. Fortunately, early detection is greatly aided by imaging modalities such as spinal and brain computed tomography (CT), magnetic resonance imaging (MRI), radionuclide cisternography, and myelography [7]. This condition has been treated with a variety of therapy modalities, from conservative management, such as bed rest, the use of analgesics, and increased fluid intake with caffeine, to

targeted therapy, including epidural blood patching, the injection of fibrin sealant at the site of the leak, and surgery [8]. We discuss a case of a generally healthy 44-year-old adult who presented to the emergency room with a history of 2 days of tiredness and a headache along with magnetic resonance imaging (MRI) findings that satisfy the diagnostic criteria for spontaneous intracranial hypotension.

### Case Presentation

The observation has taken place at Abdali Hospital, Jordan, on Tuesday 23 of November, 2021CE. At the emergency department (ED), physicians initially received a case, a 44-year-old Jordanian male who had been experiencing sleepiness and headache two days before the visit.

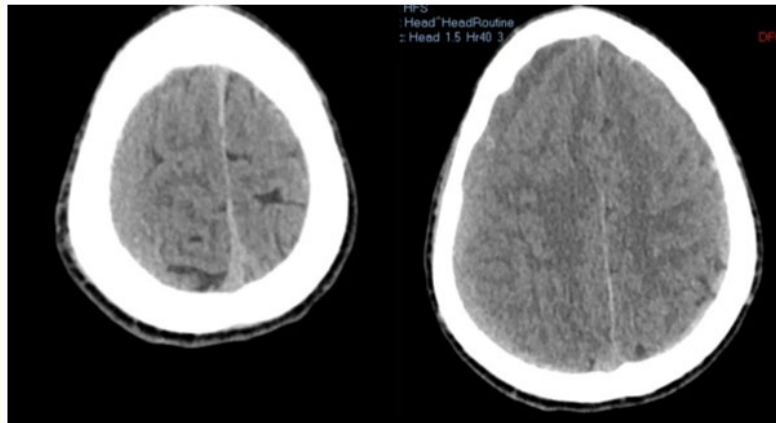
According to Hakeem, the hospital's software clinical reminders, the patient has been admitted to the neurology department to diagnose the reason/s behind the bad headache two months ago. As neurologists have prescribed Anesthesia, the patient has been able to cope with the pain for only 58 days. The second visit of the patient to the ER with the same symptom is the call for doctors to run more tests and figure out the cause.

Based on the patient's report the headache is as common, as a pressure that affects both frontal and occipital areas of the head. However, it is a state that continues the day course. Moreover, the position of the body has played a major role in the severity of the pain as it becomes intensive, sudden, and progressive upon standing and walking, but it remains partial or decreases upon laying down and analgesics. Therefore, a test to report the severity of headache has been done by using Visual Assessment Score (VAS); the severity rate was 6 -7 degrees.

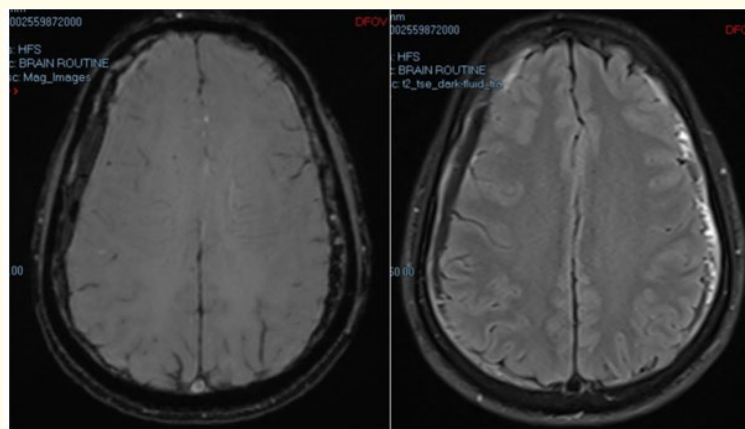
Then, an interview with the patient reveals that, first, the usual symptoms of systemic infection have not been experienced by the patient including ear discharge or pain, runny nose, sore throat, cough, diarrhea, pain during urination, and joint pain or swelling. Second, no signs of meningitis and sinusitis such as fever, chills, skin rash, photophobia, vomiting, nausea, neck stiffness, facial pain, teeth pain, or mucous in his pharynx in the morning have been noticed during that time. Third, neither the patient nor his family has a history of migraine or even reports seeing spots flashes, lines, numbness, paraesthesia, or unilateral weakness. Finally, there have not been any signs of focal neurological processes such as vision loss, diplopia, vertigo, dizziness, dysarthria, loss of consciousness, seizures, limb weakness or paralysis, ataxia or abnormal movement as well as a clean history of trauma, spinal procedures, or central nervous system tumors. Besides, the patient is a non-smoker, and he has not reported any history of alcohol ingestion or illicit drug use.

While the sleepiness, which refers to a low level of consciousness, has increased significantly two days prior to admission. In this way, more tests need to examine with the agreement of the patient. For instance, his vitals are measured with 105/70,51 and 97% for blood pressure, heart rate, and oxygen saturation respectively. Then a brain CT without contrast has been done. The finding of the test shows that there are distinctive and isodense subdural spaces that rise the possibility for bilateral convexity isodense subdural hematomas, mainly in the frontal regions and higher parietal regions bilaterally (Figure 1A). Also, the pathology has appeared larger on the right side with a thickness of about 9 mm, but only 5 mm on the left side. Also, MRI images for the brain and spinal cord have been done. Since MRI is vital in the diagnosis of spontaneous intracranial hypotension, Multiplanar T1W, T2W, FLAIR, SWI and DWI/ADC MRI map images throughout the brain and spinal cord have been requested and several imaging findings have been confirmed by the radiologists including (Figure 1B);

1. Sizable bilateral subdural fluid collection, measuring around 9 mm in maximum thickness on the right side and 5 mm on the left side and Pachymeningeal enhancement (Figure 1B).
2. Diffuse cerebral oedema with downward sagging of the brainstem and downward dropping of the posterior part of the splenium (Figure 1C).
3. Distended superior sagittal and transverse venous sinuses with no gross thrombosis.
4. Cut-off values for the mamillopontine distance with severe reduction in the interpedicular angle.



**Figure 1A:** A brain CT without contrast, shows distinctive and isodense subdural spaces that raise the possibility for bilateral convexity isodense subdural hematomas, mainly in the frontal regions and higher parietal regions bilaterally.



**Figure 1B:** SWI Mag-image on the left and T2W on the right showing a sizable bilateral subdural fluid collection, measuring around 9 mm in maximum thickness on the right side and 5 mm on the left side.

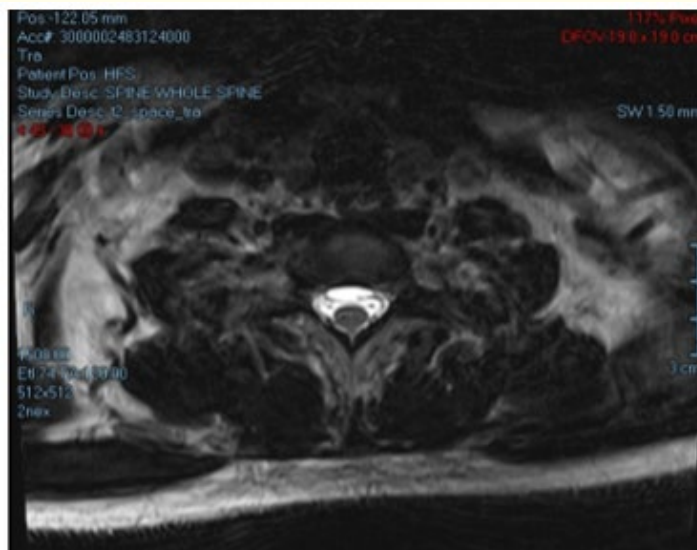


**Figure 1C:** Diffuse cerebral oedema with downward sagging of the brainstem and downward dropping of the posterior part of the splenium.

However, the spine image shows extradural cerebrospinal fluid (CSF) collection anterior to the cord extended from C7-T1 to T2, T3, indicating CSF leakage and small disc bulge in C1, C7 vertebrae (doesn't make sense in our case) (Figure 1D and 1E). obviously, these findings match severe spontaneous intracranial hypotension diagnostic criteria.



**Figure 1D:** Sagittal cut MRI shows extradural cerebrospinal fluid (CSF) collection anterior to the cord extended from C7-T1 àT2, T3, indicating CSF leakage.



**Figure 1E:** Axial cut MRI shows extradural cerebrospinal fluid (CSF) collection anterior to the cord extended from C7-T1 àT2, T3, indicating CSF leakage.

The new treatment has taken place in the intensive care unit. Several protocols including N/G tube insertion, dexamethasone 4 mg IV state 4 times, omeprazole 40 mg IV state, and acetazolamide 250 mg have been followed. Moreover, caffeine IV infusion protocol has been done by a specialized doctor as well a large blood patch has been obtained, due to the consultation with the anaesthesiologist.

After one day of the treatment, the patient’s level of consciousness has improved. However, the treatment has not been oriented yet as the patient is still sleepy. The following chart indicates the relevant vitals and examinations of the patient during the day.

Examination	Rate/Result
Blood pressure	126/76 millimeters of mercury (mmHg)
Pulse	60 beats each minute (bpm)
Respiratory	18 breaths per minute
Oxygen saturation	95 percent (%)
HEENT examination	1. No pallor or cyanosis 2. Ear, nose, throat, neck/lymph nodes were normal 3. Cranial nerves function were normal
Chest examination	Good air entry bilaterally
Abdominal examination	Distended, soft lax abdomen/no tenderness
CVS examination	S1 normal/S2 normal
limbs examination	No lower limb edema
Neurology examination	The patient was alert and oriented to person, place, and time with normal speech. Memory was normal and thought process was intact good muscle tone. Strength was 5/5 bilaterally at the deltoid, biceps, triceps, quadriceps, and hamstrings. Biceps, brachioradialis, triceps, patellar, and Achilles reflexes were 2/4 bilaterally. No clonus. Plantar reflex was downward bilaterally the sensation was intact bilaterally to pain and light touch. Two-point discrimination was intact. Finger-to-nose and heel-to-shin test were normal bilaterally. Balances with eyes closed (Romberg). Rapid alternating movements were normal. Gait was steady with a normal base. Coordination was intact as measured by heel walk and toe walk.

**Table**

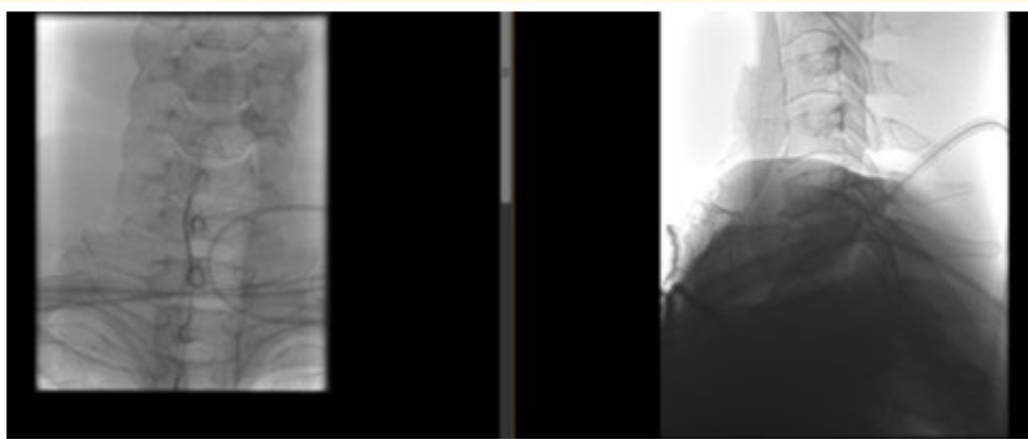
As the patient is still inserted with the N/G tube as well as laying in Trendelenburg position, several doses have been given including dexamethasone 4 mg, IV state 4 times, IVF NS 0.9% 100 cc/hr., diamox 2 doses, GI and DVT prophylaxis (Omeprazole 40 mg IV state, rised 40 mg 1\*1, and low molecular weight heparin), and pneumatic compression. Also, acetazolamide 250 mg PO and caffeine IV infusion protocol (350 mg then 300 mg) have been given to prepare for a targeted blood patch at the site of leak instead of neurosurgical intervention with glue.

The following day, the patient is still clinically the same, conscious, and alert, and his vitals are stable. The NG tube is still in place and patient was kept NPO and placed on reverse Trendelenburg position; to avoid the risk of aspiration. As the patient complaints of hiccoughs, IVF D /0.9% saline rate 70 ml/hour and dexamethasone 4 mg Iv Q 6 hours also Gabatres 300 mg PO Q 24 hours help him to recover.

While the specialists keep checking the patient, they have every reason to believe the treatment is going to work. The follow-up includes taking a cervical spine Ct, thoracic spine Ct, and heavy weighted MRI t2 of cervical and thoracic spine. Also, feeding at supine

to 30 degrees elevation with aspiration precaution has been included after removing the NG tube. However, the patient is still in reverse Trendelenburg position as the team is monitoring the level of consciousness Q 4 hours by Interventional Radiologist consultation.

During the third day, the patient has not experienced any new complaints. Similarly, the vitals are 92/60 for blood pressure, 60 for heart rate, and 36.7 for temperature. The physical examination includes that the patient is conscious, alert, oriented, and kept in Reverse Trendelenburg position. The result of the chest examination shows good air entry, and no added sounds are heard. The CVS examination shows normal indicators in S1 and S2. In abdominal examination, there is a soft lax abdomen with no tenderness or organomegaly; LL examination indicates no edema. However, it is worth mentioning the failure of blood patch implantation which has been done by the anesthesiologist previously. Therefore, a targeted blood patch by an interventional radiologist has been planned to be done on Friday 26 of November (Figure 1F).



**Figure 1F:** Shows an inserted targeted blood patch at C6/7 level.

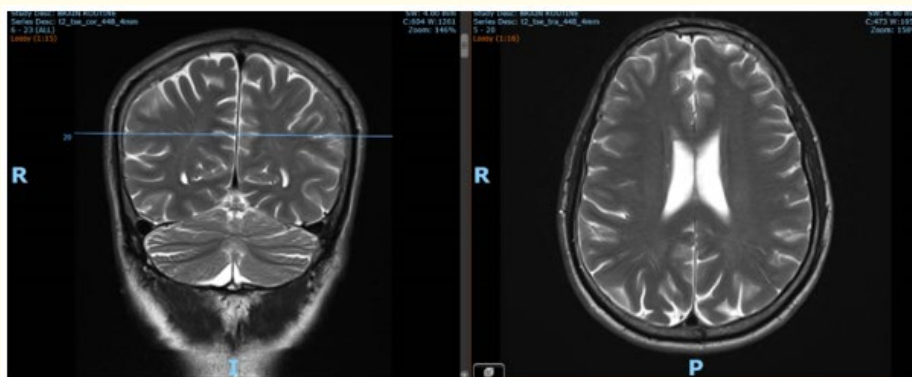
Gradually, the following medications have been given to the patient including dexamethasone 4 mg IV status 4 times, Gabators 300 mg PO Q 24 hours, IVF D/0.9 saline rate 100 ml/hour, and GI and DVT prophylaxis. In the 4<sup>th</sup> day after admission 27/11/2021, there has been no new complaint, and the examination has been unremarkable like the day before. The careful health care results in allowing the patient to sit in an upright position and ambulated with assistance multiple times and has tolerated his oral intake. The plan of treatment includes observation and follow-up the patient, control the pain as needed, medications as prescribed, encouraged oral intake, ambulation and deep breathing, and care of dressing.

A setback has taken place the following day when the patient complains of dizziness. However, general examination shows the patient is conscious, alert, and oriented 3 times more than the previous days and is not in pain or distress. Besides, vital signs are stable within normal limits. Therefore, the plan of treatment has continued as the previous day with more monitoring to observe the patient's regular vital signs, medications as well as dressing.

Finally, on Tuesday 29/11/2021 the headache completely resolved and the patient level of consciousness returned to its normal with GCS of 15. More than 1 year after observation, the patient is completely asymptomatic. Also, a brain and spine MRI images after three months have been done for follow-up which have showed, completely resolved brain lesion and a relatively resolved spine lesion (Figure 1G and 1H).



**Figure 1G:** Axial and sagittal spine MRI images which show well improved spinal lesion.



**Figure 1H:** An axial and coronal brain MRI images which show completely normal brain with resolved brain lesions.

## Discussion

With an annual frequency of 5 per 100 000 people, spontaneous intracranial hypotension (SIH) is a clinical disease that is frequently misdiagnosed. It mainly results from cerebrospinal fluid (CSF) leakage from the spinal cord levels, resulting in reduced CSF volume and accordingly, reduced intracranial CSF pressure. The most common site of CSF leakage has been proven to be in the cervical and cervicothoracic regions [9].

Numerous explanations for fluid leakage have been put forward, including dural weakness, which is more common in people with connective tissue illnesses and increases their risk of CSF leakage. These people are also susceptible to developing dural diverticula, which can rupture at any time as a result of minor trauma [10,11]. Furthermore, SIH may result from a CSF-venous fistula between a paraspinal vein and the spinal subarachnoid space, which causes ongoing CSF loss [12].

Patients with spontaneous intracranial hypotension typically present with orthostatic headache, nausea, and neck pain/stiffness [13]. Other unusual symptoms include decreased level of consciousness, dizziness, tinnitus, sleepiness, hearing disturbances, back pain, and vertigo.

History of lumbar puncture, clinical signs, and responses to epidural blood patching are used as the basis for the diagnostic criteria. The identification of extra-theal CSF on spinal imaging is criterion A of the diagnostic criteria. If criterion A is not satisfied, then criterion B-which is the presence of spontaneous intracranial hypotension as shown on cranial MR imaging-is required, with at least one of the following: Low opening pressure, a spinal meningeal diverticulum, or symptom relief following an epidural blood patch. Criterion C, the presence of all of the following or at least two; low opening pressure, spinal meningeal diverticulum, and improvement of symptoms after epidural blood patch if typical orthostatic headaches are present, must be met if criteria A and B are not satisfied [14].

Patients are often diagnosed primarily using criteria A, although a minority of patients are diagnosed according to criteria B and C. The diagnosis was made for our patient using criteria B. Indeed, this case was challenging to clinically diagnose because the patient had unusual complaints for longer than usual (2 - 3 weeks before presentation). Furthermore, our patient is a healthy adult who has no substantial medical history that could support the mechanism behind the CSF leak.

Brain and spinal magnetic resonance (MR) imaging with gadolinium was proved to be the most effective imaging modality for the diagnosis of SIH. Spinal MRI could demonstrate epidural vein enlargement, epidural fluid accumulation, and aberrant nerve root sleeve visibility. On the other hand, brain MR imaging shows brain stem sagging, enlargement of the pituitary gland, cerebellar tonsil descent, generalized pachymeningeal enhancement, and subdural fluid accumulation [15].

Diagnosing SIH requires LP and CSF analysis where we can find low CSF opening pressure, and it often shows mild elevation of total protein and increased cell count [16]. CSF leakage could be confirmed by brain imaging having CT-Myelography, the most accurate study demonstrating the exact site of spinal CSF leakage. It can also show the meningeal diverticula, extra- Dural, or arachnoidal fluid collection. While on MRI, there would be a diffuse pachymeningeal enhancement- the most common and most reliable head MRI abnormality in spontaneous CSF leaks-sagging of the brain, subdural fluid collection, enlargement of the pituitary, engorged cerebral veins, and decrease in the size of the ventricles. While if we do a spinal MRI, there will be an extra arachnoid fluid collection and extravasation, meningeal diverticula -which could be the actual site of the leak-, dural spinal enhancement, and engorgement of spinal epidural venous plexus [17]. In our patient, on whom we performed a CT scan, isodense subdural hematomas were noted in both cranial convexities, mainly in both frontal and high parietal regions. The subdural hematoma appeared more significant on the right side, measuring about 0.8 cm in thickness, with no considerable midline shift noted.

The patient had a GCS of 13 and normal vital signs. After confirming the diagnosis, the treatment is started traditionally conservatively with bed rest, oral hydration, and caffeine, besides analgesics such as NSAID and steroids. If conservative management and pharmacotherapy fail, we might use Epidural Blood Patch EBP (homologous blood) targeted or at a distant lumbar level (blindly); other epidural injections include fibrin or fibrin and blood. If there is no response to EBP, we go to surgery to repair the leak [18]. In our case, our patient who stayed for seven days in the hospital was treated the same as protocol with bed rest, IV caffeine, IV normal saline 0.9%, and dexamethasone; we also had the patient in reverse Trendelenburg position, NPO with NG tube, and acetazolamide was also given on the day of admission. EPB was performed but failed, so we performed a targeted EBP 2 days later. The SIH can be complicated by subdural hematomas -as seen in our case-brainstem compression, focal cranial nerve palsies, or cerebellar tonsillar herniation [19].

### Conclusion

SIH is an important differential for postural headache. This disorder can be diagnosed clinically, in addition to imaging modalities for the brain and spine including CT scan and MRI to identify the level of CSF leak. The majority of cases are managed conservatively. However, EBP can be employed in refractory patients and is thought to be equally safe.



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**Volume 16 Issue 1 January 2024**

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