American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2020; 21: e925986
DOI: 10.12659/AJCR.925986

Received: 2020.05.14
Accepted: 2020.07.20
Available online: 2020.08.12
Published: 2020.09.15

Spontaneous Intracranial Hypotension and Its Management with a Cervical Epidural Blood Patch: A Case Report

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 25-year-old
Final Diagnosis: Spontaneous intracranial hypotension
Symptoms: Headache
Medication: —
Clinical Procedure: —
Specialty: Anesthesiology • Medicine, General and Internal • Neurology • Radiology

Objective: Rare disease

Background: Spontaneous intracranial hypotension (SIH) is a rare cause of postural headache. In most patients, the site of cerebrospinal fluid (CSF) leak is at the cervical or thoracic spinal level. The imaging modalities to establish the diagnosis of SIH include computed tomography (CT) and magnetic resonance imaging (MRI) of the brain, CT, and MRI myelography, and radionuclide cisternography. Treatment usually consists of conservative measures, but patients unresponsive to these treatments can be treated by epidural blood patch (EBP) administration at the site of CSF leak.

Case Report: A 25-year-old-man presented with headache aggravated upon sitting or standing and relieved by lying supine or consuming coffee. There was no history of recent trauma, lumbar puncture, or spinal anesthesia. His neurological examination was unremarkable. MRI of his head and entire spine showed features of intracranial hypotension with no obvious CSF leak. He was treated conservatively but his symptoms persisted. CT spinal myelography showed significant leakage of contrast medium at the retrospinal region between C1 and C2 spinous processes. The patient underwent cervical EBP administration under fluoroscopic guidance. His symptoms resolved completely and he remains asymptomatic more than 6 months later.

Conclusions: SIH is an important cause of postural headache. In patients with non-resolving symptoms, further investigations are warranted to identify potential CSF leak. Patients found to have a CSF leak at the level of the cervical spine can be safely and effectively treated by cervical EBP administration.

MeSH Keywords: Blood Patch, Epidural • Cerebrospinal Fluid • Headache • Intracranial Hypotension • Myelography

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/925986

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Background

Spontaneous intracranial hypotension (SIH) is a rare but well-recognized cause of headache. These headaches usually become worse on standing or sitting, but are relieved on lying down. Other symptoms can include nausea, neck stiffness, visual symptoms and cranial nerve palsies [1,2]. SIH has been reported to be associated with connective tissue diseases, including like Ehlers Danlos syndrome and Marfan syndrome [3]. Cerebrospinal fluid (CSF) leakage at the cervical and thoracic spine levels has been observed in most patients with SIH [2]. The imaging modalities helpful in establishing the diagnosis of SIH include computed tomography (CT) and magnetic resonance imaging (MRI) of the brain, CT and MRI myelography, and radionuclide cisternography [2,4,5]. Treatment options include conservative measures, such as bed rest, administration of analgesic agents, and increased fluid intake including caffeinated beverages. Patients unresponsive to conservative measures can be treated with epidural blood patch (EBP) administration at the site of CSF leakage. Cervical blood patch, if performed carefully under imaging guidance, is safe and extremely effective in treating SIH due to CSF leak at the cervical spine level [1,2,6]. To date, only a few case reports have described outcomes in patients administered EBP at the cervical level. The present study describes a 25-year-old-man with SIH due to CSF leak between C1 and C2 spinous processes. This patient was successfully managed with cervical EBP administration.

Case Report

A previously fit and healthy 25-year-old-man presented to the emergency department of Hamad General Hospital with a 4-week history of headaches, which started in the mornings soon after waking up and usually involved both frontal and occipital areas. These headaches, which were moderate to severe in intensity, were aggravated by sitting or standing and were relieved by lying supine or consuming coffee.

The patient did not report any infective symptoms, such as fever, chills, skin rash, photophobia, sore throat or cough. He had no history of migraine symptoms, such as blurred vision, flashes of light, nausea, or vomiting; and no symptoms suggesting focal neurological processes, such as vision loss, dizziness, loss of consciousness, seizures, motor symptoms or sensory symptoms. There was no history of recent trauma.

The patient was a non-smoker and he did not report any history of alcohol ingestion or illicit drug use. There was no history of recent lumbar puncture or spinal anesthesia. He was not using any prescribed medications. An MRI of his head at a private hospital in Doha, Qatar, revealed suspicion of intracranial hypotension. He was managed conservatively with bed rest and oral paracetamol for 2 weeks. The patient's symptoms improved, and the intensity of his headaches decreased. Two weeks later, he developed another episode of severe headache.

On presentation to the emergency department at Hamad General Hospital, the patient's vital signs were normal, including a normal temperature, a pulse of 89 beats per min, a respiratory rate of 16 breaths per min, a blood pressure of 141/77 mmHg and oxygen saturation of 99% breathing ambient room air. His neurological examination revealed a Glasgow Coma Scale (GCS) score of 15/15, with normal cranial nerves and motor, sensory and cerebellar systems. There were no signs of meningeal irritation. Examinations of his cardiovascular, respiratory and abdominal organs were unremarkable.

The patient's laboratory investigations revealed that his complete blood count, urea concentration, creatinine concentration,

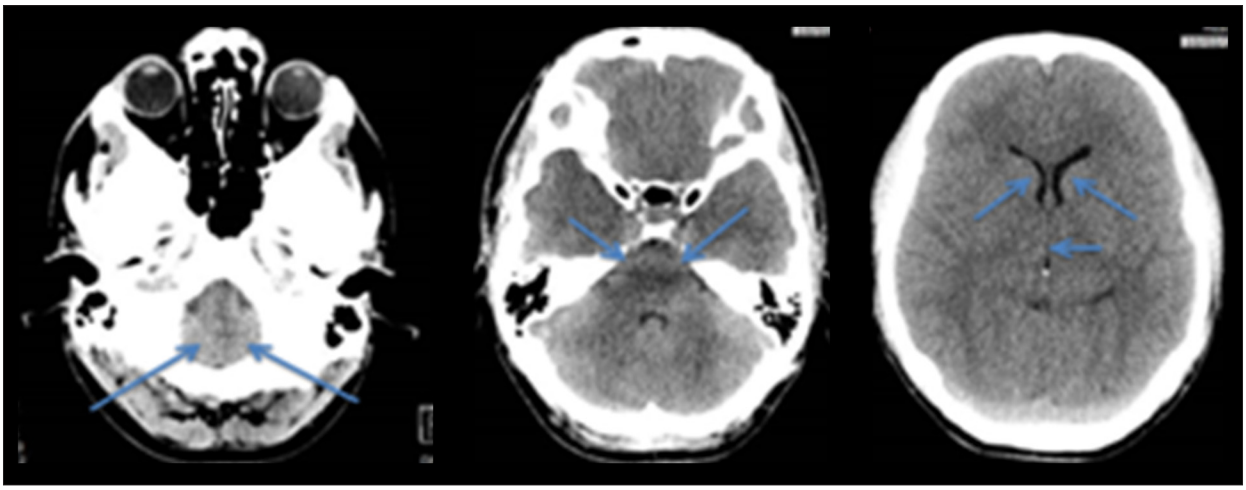

Figure 1. Axial non-contrast CT images showing sagging of the cerebellar tonsils into the foramen magnum, attenuation of the basal ventricles, and small lateral ventricles.

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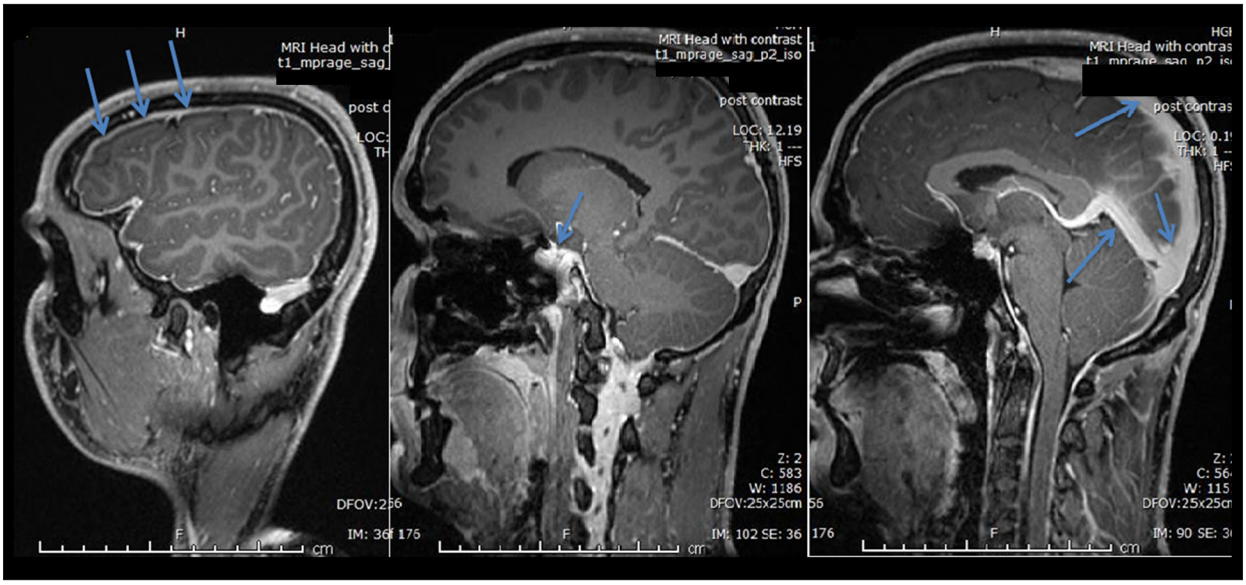

Figure 2. Sagittal MR images after intravenous injection of gadolinium, showing diffuse enhancement of the pachymeninges bilaterally, engorgement of the dural venous sinuses, hyperemia of the pituitary gland, and herniation of cerebellar tonsils.

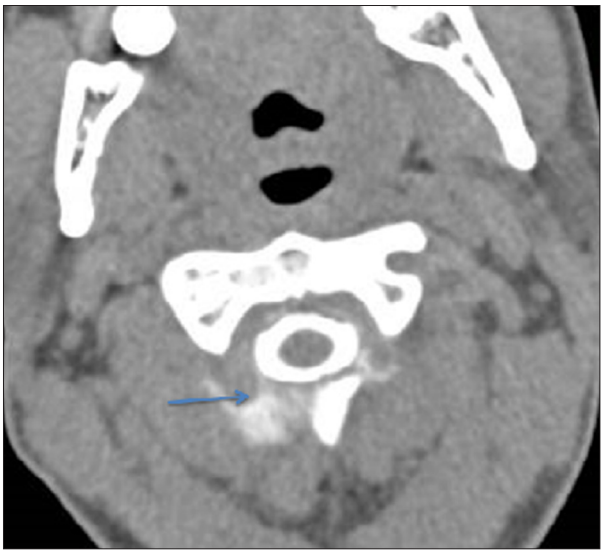

Figure 3. CT myelogram showing contrast fluid oozing posterolaterally on the right side, indicating the location of CSF leakage.

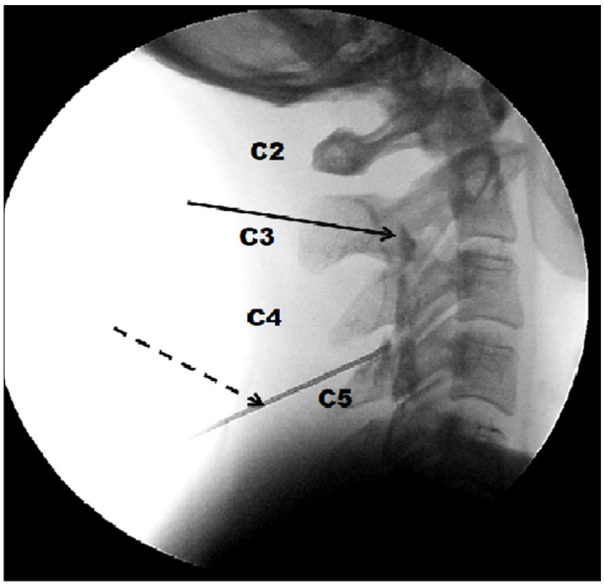

Figure 4. Contrast in epidural space (solid arrow) and the 18-gauge Tuohy needle (dotted arrow).

and electrolytes were normal. He was also normal on liver function tests, inflammatory markers, blood cultures, coagulation profile and thyroid function tests. Tests for autoimmune, rheumatological and connective tissue diseases were also normal.

A plain CT scan of the head showed sagging of the cerebellar tonsils into the foramen magnum (Figure 1). MRI of the head and spine with contrast showed downward herniation of the cerebellar "tonsils" crowding the structures in the foramen magnum and indenting the posterolateral aspect of the cervicomedullary junction associated with relatively small size cerebral ventricular system and tight extra-axial CSF spaces. Mild diffuse meningeal thickening and enhancement were observed, findings consistent with intracranial hypotension (Figure 2). However, no definite site of CSF leakage was observed along the skull base or spinal column. CT myelography of the spine showed significant leakage of contrast at the retrospinal region between the C1 and C2 spinous processes (Figure 3).

After obtaining written informed consent from this patient, he underwent cervical EBP administration under fluoroscopic guidance. The patient was placed on the operating room table

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in the prone position. Sterile conditions were maintained and his vital signs were monitored during the entire procedure. A radial arterial line was inserted to ensure easy access to autologous blood in the prone position. Because the C1–C2 intervertebral space is difficult for EBP to access, a lower intervertebral space, the C4–C5 space was localized under fluoroscopic guidance and blood was allowed to seep higher and seal the leak. Under local anesthesia, a 18 G Tuohy needle was advanced to the epidural space with loss of resistance to the fluid technique. A 1 ml aliquot of Omnipaque dye was injected into the epidural space to confirm the position of the needle, followed by injection of 10 ml of autologous blood into the C4-C5 epidural space under fluoroscopic guidance (Figure 4).

After cervical EBP administration, the patient's headaches resolved. He was discharged the following day with oral analgesic agents and was advised bed rest. More than 6 months after the procedure, he remains asymptomatic.

Discussion

SIH is an infrequent cause of daily persistent orthostatic headache with an incidence of 5 per 100 000 persons per year [7]. The site of CSF leakage in these patients has been found to be predominantly in the cervical and cervico-thoracic regions. A study of sites of CSF leakage in 56 patients with SIH reported that 6, 16, 8, and 10 patients had leakage in the cervical, cervico-thoracic, mid-lower thoracic, and lumbar regions, respectively, whereas the site of leakage was unknown in 17 patients [8].

Intracranial hypotension is thought to be due to a decrease in CSF volume with a resultant decrease in CSF pressure. Dural weakness, as observed in patients with connective tissue diseases, can predispose patients to developing dural defects, leading to CSF leakage, low CSF volume, and postural headache [9]. Patients with connective tissue diseases are also prone to develop meningeal diverticula, which can rupture in response to minimal unnoticed trauma, leading to CSF leak [10]. SIH may also be caused by a CSF-venous fistula between the spinal subarachnoid space and a paraspinal vein, leading to persistent CSF loss [11].

MRI with gadolinium is regarded as the imaging modality of choice for the initial evaluation of patients with suspected SIH. This method was found to show diffuse non-nodular, pachymeningeal enhancement in up to 80% of patients. Other intracranial findings may include engorged dural venous sinuses and/or plexa, sagging of the brain, subdural hygromas/hematomas, enlargement of the pituitary gland, and ventricular collapse [7,12]. MRI of the spine may show distention of the spinal epidural veins, epidural fluid collection and fluid collection along the nerve root sleeves [13]. CSF leakage can be confirmed by indium-111 cisternography, with findings including parathecal activity, renal uptake in less than 4 hours, and delayed activity over the cerebral convexity [12]. CT/MR myelography can also show CSF leakage as split or parallel contrast flow [12].

Three criteria, involving clinical features, CSF pressure on lumbar puncture, brain/spine imaging and response to EBP procedures, are required for the diagnosis of SIH. Criterion A involves evidence of extra-thecal CSF leakage on spinal imaging. Criterion B, which is assessed if criterion A is not met, involves cranial MR imaging findings of SIH with 1) low opening pressure, 2) spinal meningeal diverticulum, or 3) improvement of symptoms after EBP. If criteria A and B are not met, SIH can be diagnosed by criterion C, which involves all 3 findings mentioned in criterion B or the presence of at least 2 of these accompanied by orthostatic headache [14].

Initially, SIH is treated by conservative methods, including bed rest, oral hydration, and caffeine intake, to improve clinical symptoms. If symptoms do not improve, EBP is recommended to alleviate the symptoms [15].

The exact mechanism by which EBP relieves postural headache in patients with intracranial hypotension is not fully understood. The "plug" hypothesis suggests that EBP results in the formation of a gelatinous plug at the site of dural leakage, preventing further loss of CSF fluid. The subsequent production of CSF restores CSF pressure, improving headache. The "pressure patch" hypothesis suggests that injected blood increases epidural pressure, which compresses the dura mater and elevates subarachnoid CSF pressure [16].

Few studies to date have addressed outcomes in patients undergoing cervical EBP. The success rates of thoraco-lumbar EBP are low in patients with SIH due to CSF leak at the cervical or cervico-thoracic junction. It is therefore important to apply EBP at the site of CSF leak even if the site of CSF leak is at a higher spinal level. Although a patient with SIH due to CSF leakage at the C7–T1 level failed to respond to EBP treatment at the T9–T10 level, a repeat EBP performed by targeting the site of leakage at the C7–T1 level resolved this patient's symptoms [17]. Cervical EBP at the C6-7 level successfully resolved symptoms in a patient with SIH [18]. Moreover, cervical EBP was successful in treating SIH due to a CSF leak at the C2 level after lumbar EBP failed twice [19].

Image guidance has been found to increase the effectiveness of cervical EBP. A clinical study compared outcomes in 31 patients who underwent targeted EBP under fluoroscopic guidance and 25 patients who underwent blind EBP at the lumbar level for management of CSF leakage [8]. Only 4 of the 31 patients in targeted EBP group showed rates of symptom resolution failure in these 2 groups were 19.0% (4/31) and 48%

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(12/25), respectively ($p<0.05$), indicating that targeted EBP at the site of CSF leakage under imaging guidance was significantly more effective, with less need for repeat procedures, as blind EBP at the lumbar level.

Conclusions

SIH, which may be of traumatic or idiopathic origin, is an important cause of postural headache. MRI of the brain and/or spine and CT and/or MRI myelography are the imaging modalities that help in establishing the diagnosis and identifying the site of CSF leakage. Cervical EBP administration is safe and effective for treatment of patients with CSF leakage at the level of the cervical spine.

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