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# Spontaneous Intracranial Hypotension Plus Cerebral Venous Thrombosis: A Case Report Study

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**ABSTRACT**

This is a case study of a 34-year-old woman who was admitted to hospital with a history of severe orthostatic headache. She was diagnosed as having spontaneous intracranial hypotension (SIH) by undetectable cerebrospinal fluid (CSF) pressure at lumbar puncture, and with evidence of diffuse dural enhancement of the brain detected by magnetic resonance imaging (MRI). However, the contrast-enhanced MRI of the spinal cord did not show a CSF leak site and she was treated conservatively. After a few days, the patient's recurrence of headache with continuous duration and progressive worsening led to further investigations by contrast-enhanced MRI, magnetic resonance venography (MRV) and computed tomography venography (CTV) that showed an extensive thrombosis in the superior sagittal sinus, left sigmoid sinus and both transverse sinuses. Then, the patient was treated successfully with heparin and oral anticoagulant. She had no neurological deficit after six months. SIH with concomitant intracranial cerebral venous thrombosis is a rare condition. We hypothesize that SIH may change cerebral blood-flow velocity and viscosity and can cause intracranial cerebral venous thrombosis.

**Keywords:** Intracranial Hypotension; Venous Thrombosis; Headache

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## Introduction

Spontaneous intracranial hypotension (SIH) is an uncommon but increasingly recognized syndrome which is produced by cerebrospinal fluid (CSF) leakage, usually from spinal dural sac tearing, resulting in CSF hypovolemia and hypotension (CSF pressure below 6 cm of $H_2O$). Orthostatic headache is usually resolved by recumbent position and it is the main symptom of SIH (1,2), but neck and interscapular pain, vestibulocochlear nerve-disease symptoms, and cranial nerve palsy

may also add to the clinical picture (3). In addition, SIH may occasionally be complicated by subdural effusion and also rarely by cerebral venous thrombosis (CVT) (1,4). One main mechanism which may be responsible for these uncommon complications is negative intracranial pressure caused by CSF hypotension, compensated by venous engorgement, subdural effusion, or even subdural hematoma (1,2,5-7). Another mechanism responsible for cerebral venous thrombosis is the loss of CSF absorption by cerebral venous sinuses, leading to increased blood viscosity in the venous system (5,8). On cranial MRI, SIH is characterized by diffuse pachymeningeal enhancement, engorgement of venous structure, sagging or downward displacement of the brain, and pituitary hyperemia (3,8-10).

Three common strategies for the treatment of SIH are hydration, bed rest, and epidural blood path (EBP) (3). When the disease is complicated by CVT, anticoagulant therapy is also recommended (2). In this case study, an uncommon combination of SIH and CVT with no other risk factors of thrombosis is being reported in a 34-year-old woman.

## Case Presentation

A 34-year-old woman without significant past medical history and without oral contraceptive consumption was admitted to the hospital with persistent pulsatile headache for seven days. She had bifrontal headache that was severe enough to awaken her and that suddenly became worse in the upright position and was relieved soon after lying down. Her physical examination detected no signs of photophobia, lacrimation, conjunctival injection, neck stiffness or neurological deficit. She had no history of previous dural puncture, surgical intervention, and trauma. Metabolic and biochemical blood tests, vasculitis and thrombophilia work-ups showed normal values during hospital course assessments.

Lumbar puncture was performed under sterile conditions that revealed an undetectable opening pressure of CSF, which had normal content except for its protein that was significantly high (325 mg/dl).

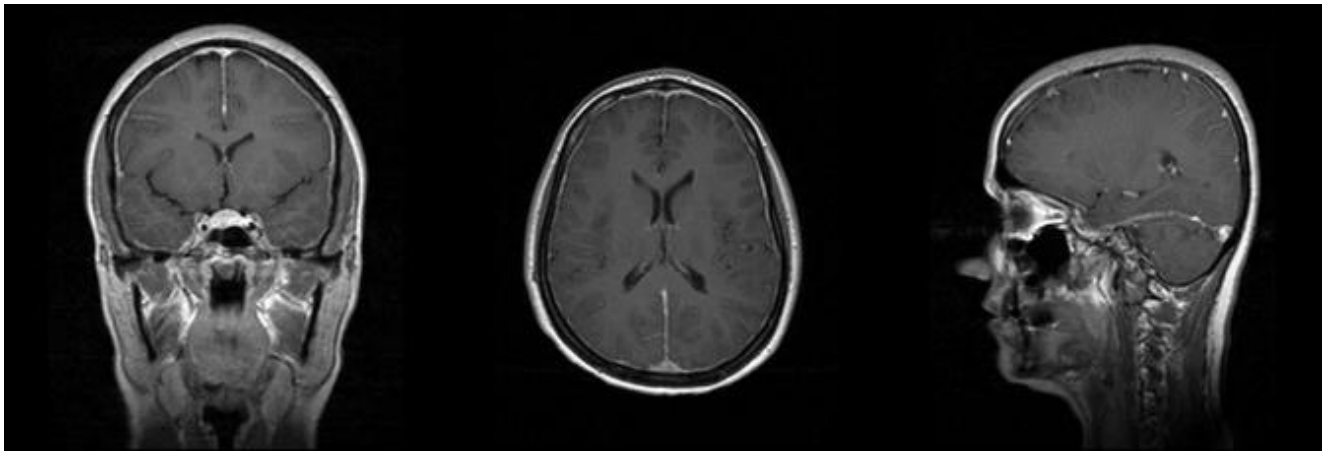

**Figure 1.** Diffuse pachymeningeal enhancement in the patients' brain MRI

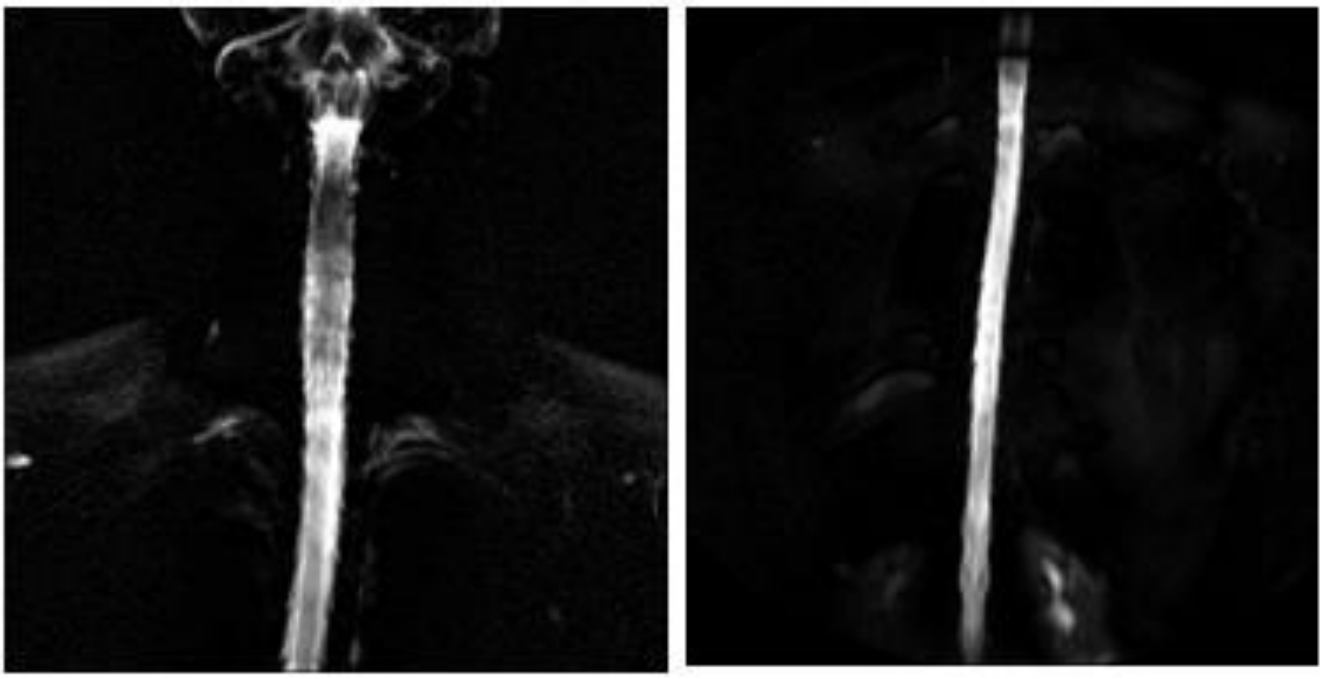

**Figure 2.** Contrast-enhanced spinal MRI did not show CSF leakage

Cerebral MRI was performed that demonstrated diffuse pachymeningeal enhancement (figure 1), but the spinal contrast-enhanced MRI did not show the leakage site (figure 2) so the patient with diagnosis of SIH was treated conservatively with complete bed rest, hydration and pain-relief drugs. After a few days, recurrence of the patient's headache with continuous duration and progressive worsening led to further investigations by post contrast-enhanced MRI, magnetic resonance venography (MRV), computed tomography venography (CTV) images, and gradient echo image (GRE) which revealed filling defect in the posterior half of the superior sagittal sinus, both transverse sinuses, left sigmoid sinus and posterior aspect of the rectus sinus, indicating acute sinus venous thrombosis (figure 3).

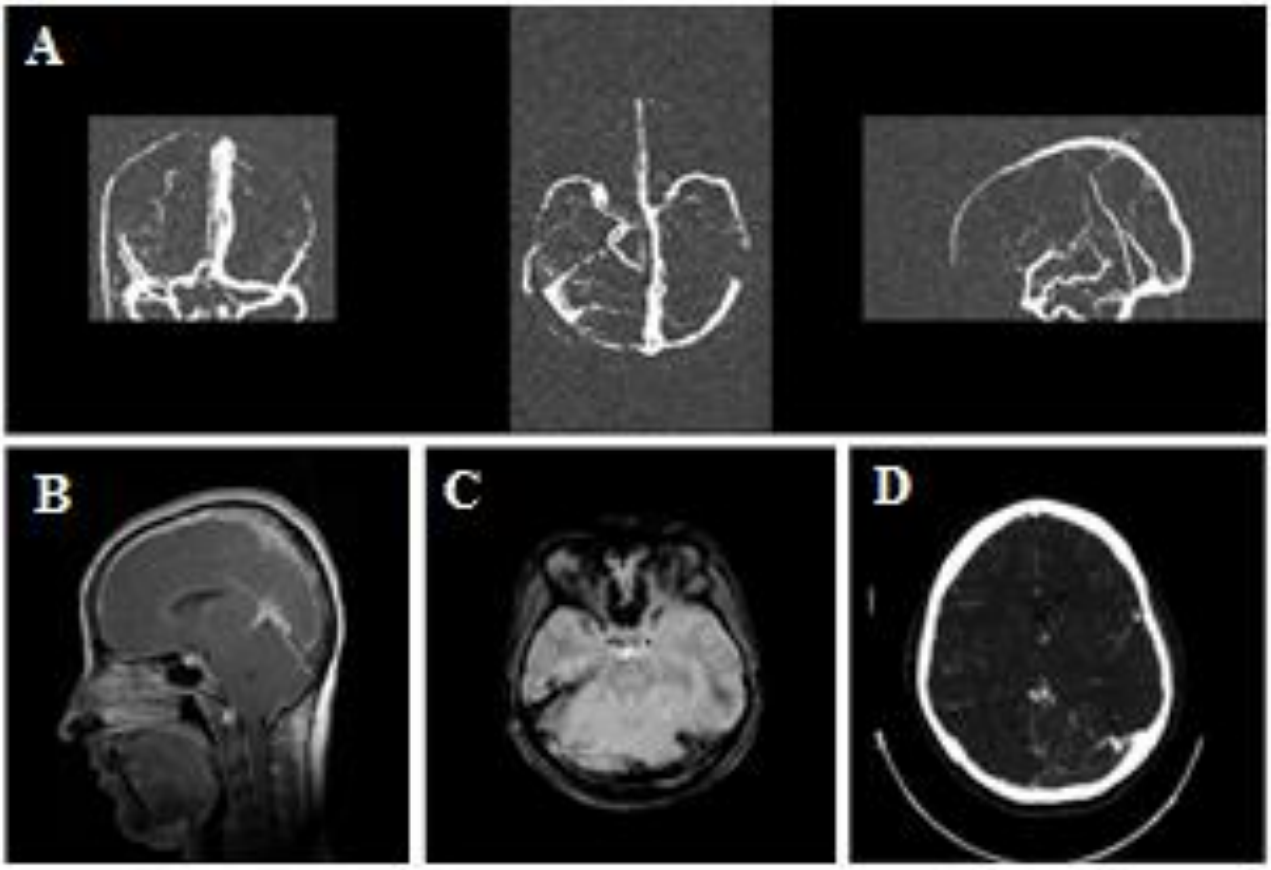


**Figure 3.** (A) Brain MRV, (B) contrast enhanced MRI, (C) Gradient Echo Image, and (D) CT venography revealed acute venous thrombosis

The patient was treated with intravenous heparin followed by oral anticoagulants, and the treatment improved her headache during the hospitalization period. The patient had no neurological deficit when she was discharged from the hospital and after six months when she visited the neurology clinic.

## Discussion

We reported a young adult woman with SIH complicated with CVT. In a systematic review conducted in 2008, Schievink *et al.* reported 141 patients with SIH and calculated a rate of CVT about 2% in these patients (11). Orthostatic headache, diffuse pachymeningeal enhancement in the brain MRI and a low opening CSF pressure (below 60 mm of $H_2O$), are known as diagnostic triads of SIH (12), which were found in the case of the present study. Similar to our case, an increase in CSF protein concentration with values up to 100 mg/dL is not an uncommon finding in SIH and there is a report of CSF protein concentration as high as 1,000 mg/dL by Mokri *et al.* in 2013 (3). However, our case had no other clinical manifestations of SIH such as back pain, dizziness, tinnitus, and cranial nerve palsy (3). In addition, this case had no other radiological hallmarks of CSF

hypotension such as downward displacement of the brain and brainstem and cerebellar tonsils, hypophysial enlargement, and subdural effusion or hematoma (13). Radioisotope cisternography, spinal MRI, and CT myelography are commonly used to explore the CSF leakage site in SIH. Paucity of radioisotope activity after 24 h over cerebral convexity and the presence of parathecal activity and/or meningeal diverticula in radioisotope cisternography are indirect and direct evidence of CSF extradural extravasation, respectively (3). Presence of a single or multiple meningeal diverticula and spinal dural enhancement in the MRI and CT myelogram are other radiologic clues of CSF leakage sites (14). However, spinal MRI and MR myelogram showed that there were no meningeal diverticula or parathecal CSF extravasation in our case (figure 2). Radioisotope cisternography was not performed in this patient. Several studies have shown that SIH is a risk factor for developing CVT (3,15,16). Increased blood viscosity secondary to loss of CSF absorption into cerebral venous sinuses, traction of the cerebral veins and sinuses due to sagging of the brain, and slowing of the venous blood velocity induced by cerebral veins engorgement, are three proposed mechanisms that can contribute to the development of CVT in the course of SIH (2). A change in the headache pattern from orthostatic to continuous (17), and the appearance of other signs and symptoms of CVT such as papilledema, seizure, and cortical infarction or hemorrhage are some clinical guides to diagnosis of this rare complication of SIH. Recurrence of headache with continuous and worsening quality led to further investigations and diagnosis of CVT in our patient. EBP is the treatment of choice in patients with no response to other managements (3). In the present study, traditional treatment of CVT by heparin followed with oral anticoagulants along with bed rest, hydration and analgesic medication resulted in complete resolution of all signs and symptoms with no need for EBP or other invasive procedures.

## Conclusion

SIH is a relatively uncommon medical problem and CVT is one of the rare and potentially harmful complications of this disorder which requires appropriate management.

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## Conflict of Interest

The authors have no conflict of interest.

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