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Case Report

Spontaneous intracranial hypotension complicated by cerebral venous thrombosis[☆]

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ABSTRACT

Spontaneous intracranial hypotension (SIH) is a well-known cause of orthostatic headache. Although subdural fluid collection is a usual complication of SIH, SIH as a risk factor for cerebral venous thrombosis (CVT) is not well-known. There are several mechanisms that could contribute to the development of CVT in SIH. Herein, we report a case of a 33-year-old woman with SIH complicated by CVT. She was treated with anticoagulation but did not receive a blood patch for the SIH, because there was resolution of orthostatic headache with bed rest and sufficient hydration. Follow-up magnetic resonance imaging showed resolution of the findings of SIH and CVT. Patients with SIH should be closely observed for any change in the headache pattern, which might suggest the development of CVT.

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Introduction

Spontaneous intracranial hypotension (SIH) is a well-known cause of orthostatic headache. It is a condition of negative intracranial pressure resulting from cerebrospinal fluid (CSF) leakage from the dural sac [1]. Although subdural fluid collection is a usual complication of SIH, little is known about SIH as a risk factor that can lead to cerebral venous thrombosis (CVT). Herein, we report the case of a 33-year-old woman with SIH complicated by CVT.

Case report

A 33-year-old woman was admitted at a local hospital because of nausea, vomiting, and headache. She had no pertinent past medical history or trauma, but she was taking oral contraceptives regularly. She initially underwent conservative observation because her symptoms improved. One month later, she experienced sudden onset of pain on the occipital and nape area, nausea, vomiting, and vertigo. Noncontrast head computed tomography showed a hyperdense area along the left transverse and superior sagittal sinuses (Fig. 1), which was

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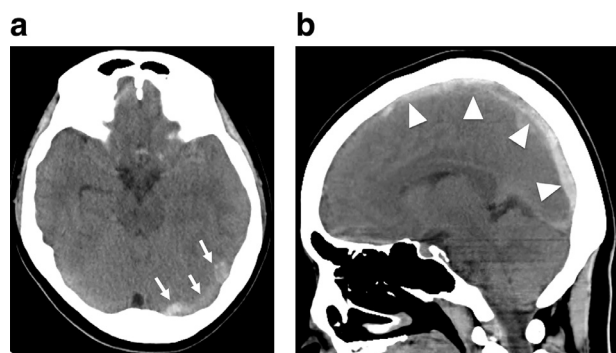


Fig. 1 – Noncontrast head computed tomography. (A) Axial and (B) sagittal views show a hyperdense area along the course of the left transverse sinus (arrows) and the superior sagittal sinus (arrowheads).

suspected to be CVT. She was transferred to our hospital for further examination.

At our hospital, general and neurologic examinations revealed orthostatic headache, dysarthria, hemiparesis, and hypoesthesia of the right upper extremity. Brain magnetic resonance imaging (MRI) showed fluid collections in the bilateral frontotemporal regions and swelling of the pituitary gland. Magnetic resonance venography (MRV) and diffusion-weighted imaging demonstrated thrombosis in the superior sagittal and left transverse sinuses with restricted diffusion (Fig. 2). In addition to CVT, SIH was suspected because of the presence of fluid collections in the bilateral frontotemporal regions and swelling of the pituitary gland.

To confirm the presence of CSF leakage, spinal MRI, and radionuclide cisternography were performed. Spinal MRI showed anterior epidural fluid collection at the level of C6–T3 spinal canal (Fig. 3). The radionuclide cisternography (not shown) revealed early accumulation of the tracer in the urinary bladder and delayed uptake in the cerebral convexity, which were suggestive of CSF circulation problems, but there was no epidural accumulation of the tracer. The CSF opening pressure before radionuclide cisternography was elevated at 20 cmH₂O. Tests for a hypercoagulable state using prothrombin time, activated partial thromboplastin time, and protein C were normal. Based on these, she was diagnosed with SIH complicated by CVT.

She was started on heparin, which was subsequently changed to oral anticoagulation, and was advised to discontinue the intake of oral contraceptives. She did not receive a blood patch for the SIH because there was resolution of the orthostatic headache with bed rest and sufficient hydration.

One month later, follow-up MRI showed resolution of the subdural fluid collection and pituitary gland swelling and MRV showed recanalization of the superior sagittal and left transverse sinuses (Fig. 4). Thereafter, oral anticoagulation was continued for 6 months. By far, after a follow-up period of 10 months, she has not experienced recurrence of the condition.

Table 1 – Diagnostic criteria for spontaneous spinal CSF leak and intracranial hypotension.

Criterion A, demonstration of a spinal CSF leak (ie, presence of extrathecal CSF), or, if criterion A not met:
Criterion B, cranial MR imaging changes of intracranial hypotension (ie, presence of subdural fluid collections, enhancement of the pachymeninges, or sagging of the brain), and the presence of at least 1 of the following:
1. low opening pressure (≤ 60 mmH ₂ O);
2. spinal meningeal diverticulum;
3. improvement of symptoms after epidural blood patching;
or, if criteria A and B not met:
Criterion C, the presence of all of the following or at least 2 of the following if typical orthostatic headaches are present:
1. low opening pressure (≤ 60 mmH ₂ O),
2. spinal meningeal diverticulum, and
3. improvement of symptoms after epidural blood patching.

Discussion

The incidence of SIH is estimated at 5 per 100,000 per year; women are affected more commonly than men, with a peak at around the age of 40 years [1]. The typical clinical manifestations of SIH include orthostatic headache, neck and interscapular pain, and cochleovestibular symptoms. In most cases, the CSF opening pressure is low (below 60 mmH₂O), but low CSF pressure is not always present in SIH. Typical MRI findings include subdural fluid collections, pachymeningeal enhancement, engorgement of venous structures, pituitary hyperemia, and sagging of the brain [1]. Schievink et al. proposed the diagnostic criteria for spontaneous spinal CSF leaks and intracranial hypotension in 2008 (Table 1) [2]. In the present case, based on criterion A, the patient was diagnosed with spontaneous spinal CSF leaks and SIH. In addition, MRV showed thrombosis of the superior sagittal and left transverse sinuses. Cerebral sinus thrombosis is known to lead to raised intracranial pressure [3], which had been thought to be the reason for an increased CSF opening pressure despite the presence of spinal CSF leaks.

Although CVT has been known to be associated with multiple factors, such as thrombophilia, inflammatory bowel disease, pregnancy, dehydration, infection, oral contraceptive use, substance abuse, and head trauma [4], little is known about the development of CVT in SIH. While not well-known, Schievink reported that the frequency of CVT among patients with SIH was about 2%, which was higher than the 0.0005% rate in the general population [5]. There were 3 reported mechanisms of SIH resulting in CVT. First, as dictated by the Monro-Kellie doctrine, the cumulative volume of intracranial blood, CSF, and brain remains constant. If the CSF volume decreases because of CSF leaks, volume compensation would be required; because the brain volume remains nearly constant, the volume of intracranial blood, especially the most expandable venous component, is the one that will be affected [6]. Therefore, SIH is associated with venous engorgement due to the loss of CSF. The dilatation of the intracranial veins and sinuses leads to slowing of venous blood flow velocity, which is one of the components of the Virchow's triad, and

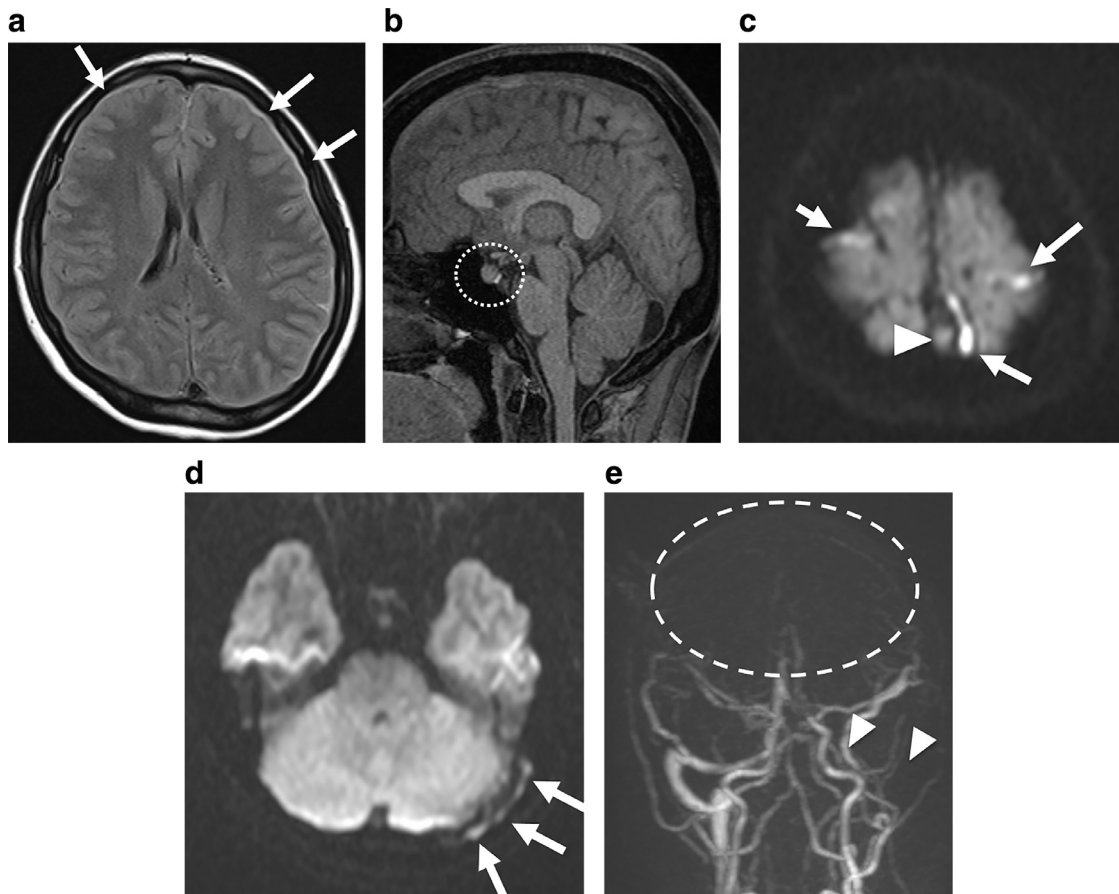


Fig. 2 – Brain magnetic resonance imaging and magnetic resonance venography. (A) Axial fluid-attenuated inversion recovery image shows thin subdural collections along the bifrontal convexities (arrows). (B) Sagittal fat-suppression T1-weighted image shows an edematous pituitary gland with a convex superior margin (dotted circle). (C) Axial DWI ($b = 1000$) shows hyperintense areas along the cortical veins (arrows) and superior sagittal sinus (arrowhead), which indicate venous thrombi. (D) Axial DWI ($b = 1000$) shows a hyperintense area along the left transverse sinus (arrows), which indicate a venous thrombus. (E) A maximum intensity projection image of magnetic resonance venography shows defects in the cortical vein, superior sagittal sinus (dotted circle), and the left transverse sinus (arrowheads).

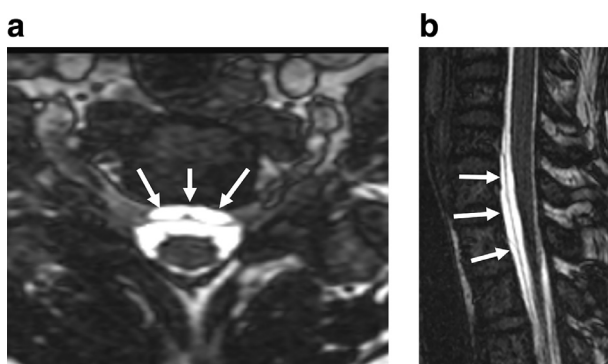


Fig. 3 – Spinal magnetic resonance imaging. Axial (A) and sagittal (B) reconstruction images of true fast imaging with steady precession imaging show anterior epidural fluid collection at the level of the C6-T3 spinal canal (arrows).

eventually results in a hypercoagulable state. Second, the loss of CSF buoyancy in SIH is associated with rostrocaudal sagging of the brain and traction on the cerebral veins and sinuses, which may lead to mechanical distortion of the vessel wall and increased propensity for thrombus formation [5,7–9]. Third, CSF leakage reduces absorption of CSF into the cerebral venous sinuses and consequently increases blood viscosity and hypercoagulability in the venous compartment [10].

In our case, the initial MRI showed findings of both SIH and CVT. Schievink reported that of 20 cases of SIH complicated by CVT, 17 (85%) showed initial MRI findings of both SIH and CVT; 3 (15%) clearly demonstrated that SIH preceded the development of CVT; and none showed clinical or radiographic evidence of CVT preceding the development of SIH [5]. Although there is no reliable predictor of CVT, any new neurologic finding in the disease course of intracranial hypotension should raise the suspicion of CVT [9]. In the present case, the patient's symptoms suddenly worsened a few days before she was admitted to our hospital, which suggested that SIH was complicated by CVT. Moreover, she had a history of oral contraceptive intake, which was shown in a recent meta-analysis to be

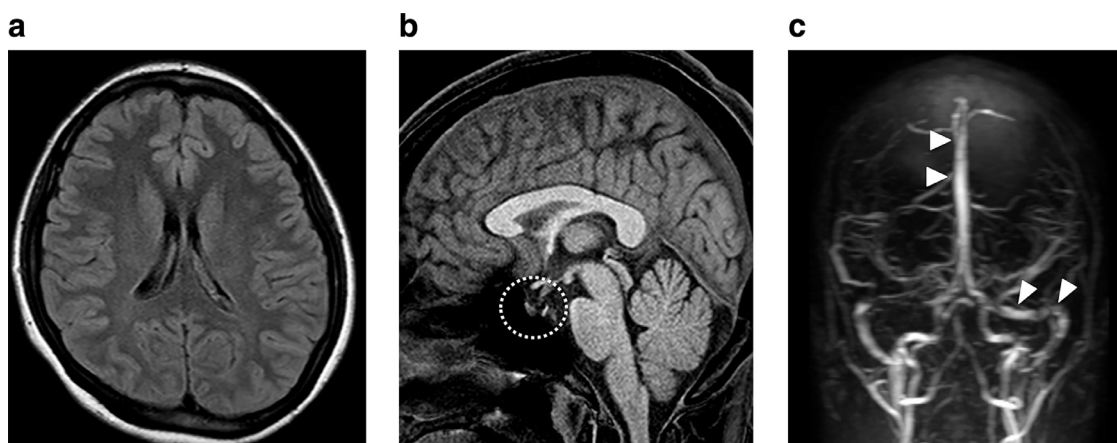


Fig. 4 – Brain magnetic resonance imaging and magnetic resonance venography. (A) Axial fluid-attenuated inversion recovery image shows resolution of the bilateral frontotemporal subdural hyperintensity. (B) Sagittal fat-suppression T1-weighted image shows a normal-sized pituitary gland (dotted circle). (C) A maximum intensity projection image of magnetic resonance venography shows recanalization of the superior sagittal and left transverse sinuses (arrowheads).

significantly associated with CVT [11]. Thrombosis is generally accepted as a multicausal disease. In a previous report, approximately half of the women had more than 1 thrombotic risk factor [12]. Therefore, in addition to SIH, the use of oral contraceptives likely contributed to the development of CVT in this case.

Many cases of SIH resolve spontaneously without any specific therapy [1]. When the symptoms do not improve with conservative treatment, several options are available. The mainstay of treatment is an epidural blood patch, which is effective in relieving symptoms in about one-third of patients and can be repeated. The other options include percutaneous placement of fibrin sealant and surgical repair. These treatments require identification of the exact site of the CSF leak. The consensus for noninvasive treatment of CVT is anticoagulation. Two randomized studies concluded that heparin treatment was safe and associated with relevant reduction of the risk of death and dependency, although not statistically significant [13,14]. In the present case, the patients had 2 coexisting conditions (ie, SIH and CVT). In 33 reported cases, 22 were treated conservatively for SIH and were given anticoagulation for CVT [8].

In conclusion, we encountered a rare and important case of SIH complicated by CVT. Patients with SIH should be closely observed for any change in the headache pattern, which might suggest the development of CVT.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.radcr.2018.05.014](https://doi.org/10.1016/j.radcr.2018.05.014).

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