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Cerebral Venous Thrombosis as a Complication of Spontaneous Intracranial Hypotension: A Case Report

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ABSTRACT

Spontaneous intracranial hypotension (SIH) is an uncommon condition caused by cerebrospinal fluid (CSF) leakage, which can lead to complications such as cerebral venous thrombosis (CVT), occurring in 1–2% of SIH patients. This case report describes the presentation, diagnosis, and management of SIH complicated by CVT. A 36-year-old woman with no comorbidities presented with a three-week history of severe, initially orthostatic headaches. Neurological examination revealed mild bilateral disc oedema, and imaging showed generalized brain oedema. MRI with MRV confirmed cerebral venous sinus thrombosis along with typical SIH features, including pachymeningeal enhancement and venous engorgement. She was treated with heparin infusion and isotonic saline, responding well without seizures or hemorrhagic complications.
This case highlights the risk of CVT in SIH and underscores the importance of early diagnosis through neuroimaging and lumbar puncture. The patient's favorable response to anticoagulation demonstrates effective management, though further studies are needed to establish definitive treatment guidelines for SIH-associated CVT.

Introduction

Spontaneous intracranial hypotension (SIH) is an uncommon condition marked by decreased cerebrospinal fluid (CSF) pressure, typically caused by CSF leakage. SIH is defined by a CSF pressure ≤6 cm H_2O and/or imaging evidence of CSF leak in patients without a history of lumbar puncture [1]. Headaches typically occur after moving from lying to standing, improving when recumbent [2]. Some patients may initially lack posture-related headaches or present with activity-induced or evening headaches [3, 4]. SIH most commonly affects young to middle-aged adults, with a higher prevalence in females, and is rarely reported during pregnancy [5].

According to the International Classification of Headache Disorders – 3rd edition (ICHD-3), SIH is identified mainly by orthostatic headache that improves with reclining and worsens with Valsalva maneuvers [6, 7]. Other common symptoms include

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tinnitus, auditory disturbances, and dizziness in 50% of patients, as well as nausea, vomiting, photophobia, and meningismus in 20% [8, 9]. In the absence of procedures or trauma, a diagnosis of headache due to spontaneous intracranial hypotension can be made when low CSF pressure (≤6 cm H_2O) and/or imaging signs of CSF leakage are present alongside orthostatic headaches [6]. However, many patients with intracranial "hypotension" show normal CSF pressure and neuroimaging [10].

Imaging findings commonly include subdural fluid collections resembling primary subdural hematomas, diffuse gadolinium-enhanced pachymeninges, venous congestion, increased pituitary blood flow, brain sagging, and occasionally enlarged vertebral venous plexuses with extradural fluid collections [11, 12]. CVT associated with SIH may lead to severe neurological complications, including subarachnoid hemorrhage and dural arteriovenous fistula. Brain downward shift due to low CSF pressure may rupture bridging veins, causing subdural hematomas [12, 13]. The relationship between SIH and CVT is not merely coincidental. Although partially clarified, the mechanistic link remains uncertain, complicating clinical evaluation and management [14]. CVT is a rare SIH complication, with a prevalence of 1–2% [15].

We present a case with no other known risk factors for thrombosis who exhibited clinical signs of SIH and subsequently developed CVT.

Case Presentation

A 36-year-old woman with no underlying medical conditions, no history of trauma, and no medication use presented to the hospital with a severe headache that had started three weeks prior to admission. Initially, the headache was mild and orthostatic, relieved by lying down and worsened by standing or sitting. However, over the following days, the intensity of the headache increased significantly, becoming persistent and unresponsive to outpatient treatments.

Upon presentation to the hospital, the patient underwent a thorough clinical examination. All neurological and general assessments were normal, except for bilateral mild disc oedema noted on fundoscopy. Given the nature of the headache, an initial brain CT scan was performed to assess for any acute intracranial pathology. The scan revealed generalized cerebral oedema, raising concern for a possible intracranial cause, and further imaging was recommended.

On the second day of hospitalization, the patient underwent MRI of the brain with MRV, including gadolinium contrast. The MRI revealed several significant findings: hyperintensity on T1-weighted images, abnormal signal voids on T2 sequences, and filling defects on post-contrast images, suggestive of venous thrombosis. Additionally, pachymeningeal enhancement was observed, indicating an inflammatory or pathological process involving the meninges (Figure 1). The MRV also demonstrated evidence of CVT, confirming the diagnosis of a venous thromboembolic event.

To rule out infectious or inflammatory causes such as tuberculosis and sarcoidosis, a lumbar puncture (LP) was performed to analyze the CSF. The opening pressure was recorded at 6 cm H_2O, which was low and consistent with SIH. CSF analysis was normal, with no signs of infection or inflammation. Given the absence of prior lumbar punctures, trauma, or surgery, and the findings of low CSF pressure, the diagnosis of SIH was established.

With the confirmed diagnosis of SIH and CVT, the patient was started on anticoagulation therapy to manage the CVT. Heparin infusion was initiated, and the patient was closely monitored for any signs of complications such as hemorrhage or seizures. In addition to anticoagulation, the patient received hydration with isotonic saline to support CSF volume and pressure, which is crucial in managing SIH.

By the fifth day, the patient showed significant improvement, with a reduction in headache severity. She reported a marked decrease in pain intensity, and her overall clinical condition improved. Given the positive response to anticoagulation and hydration, specific interventions for SIH, such as an epidural blood patch, were not required. The patient's neurological examination remained normal, and she did not experience any new symptoms.

The patient continued to improve with no signs of complications such as seizures, infarction, or hemorrhage. A follow-up CT scan was performed to ensure there were no new changes in the brain parenchyma or signs of worsening cerebral oedema. The scan confirmed stable findings. By the end of the first week, the patient was stable and asymptomatic. She was discharged from the hospital in good general condition, with no further neurological deficits.

She was prescribed warfarin for long-term anticoagulation therapy due to the provoked nature

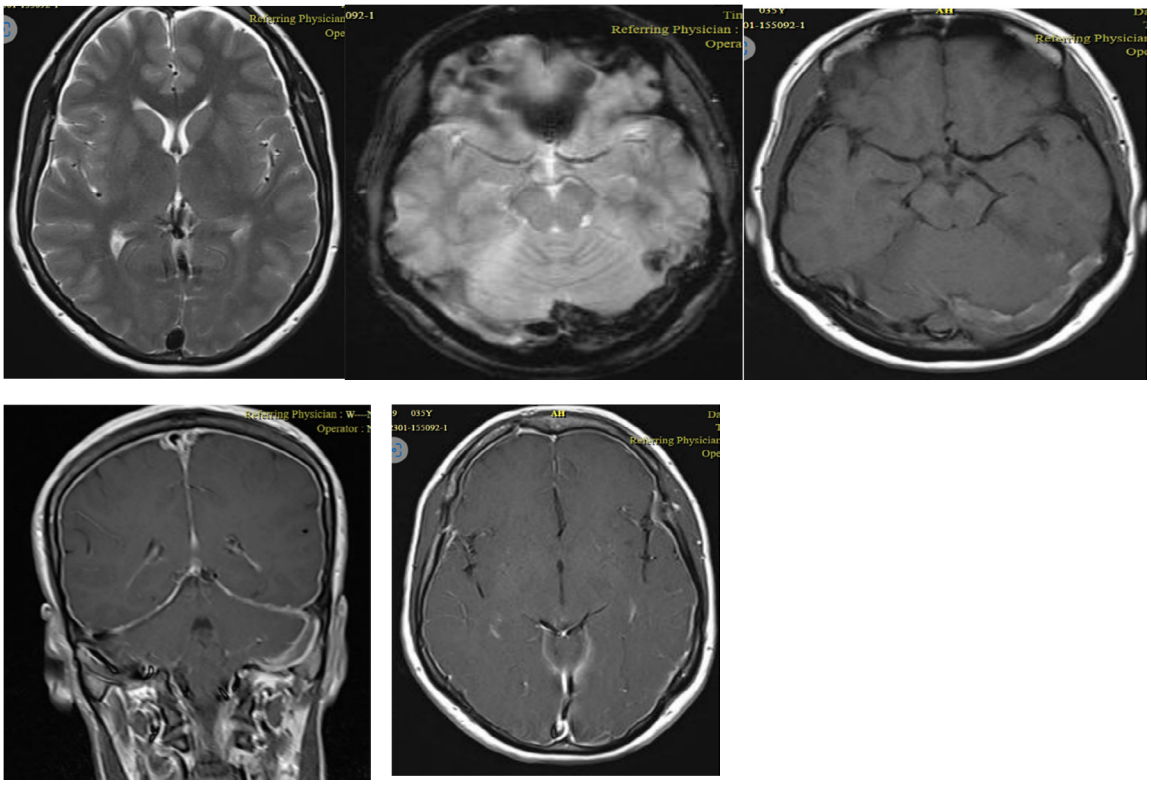

Fig. 1. MRI sequences show an abnormal signal void in the T2 sequence, a blooming artifact in the GRE sequence, hyperintensity in the T1 sequence, and pachymeningeal enhancement in the post-contrast sequences.

of the CVT and scheduled for regular follow-up appointments. The patient continued anticoagulation therapy for a total of six months. During this period, she remained asymptomatic and did not experience any complications. Follow-up imaging at the three-month mark showed no signs of CVT recurrence, and the patient remained stable.

Throughout the treatment course, the patient's headache resolved, and she did not develop any further neurological symptoms, including seizures, infarction, or hemorrhage. She continued routine monitoring of her anticoagulant therapy and was advised to follow up regularly for ongoing assessment.

Discussion

CVT is a rare cerebrovascular disorder, affecting about 5 per million (0.0005%) in the general population. While its incidence is significantly higher in patients with SIH, it still remains relatively uncommon [16]. The mechanisms linking SIH to CVT are not fully understood; however, several distinct pathophysiological processes have been proposed. First, according to the Monro-Kellie doctrine [17], SIH causes compensatory venous congestion due to CSF loss, increasing thrombosis risk in the dural sinuses. The first reported case of CVT secondary to SIH, associated with severe venous engorgement and stasis, was described by Sopelana D et al. in 2004 [18]. Second, brain sagging in SIH may stretch venous endothelium and distort vessel walls, promoting a prothrombotic state [19]. Third, CSF volume depletion may reduce CSF absorption into cerebral venous sinuses, increasing blood viscosity—a recognized CVT risk factor. Overall, both morphological and dynamic alterations in the brain and CSF appear to elevate CVT risk in SIH patients [20].

Our patient presented with classic SIH features, including orthostatic headache and bilateral disc edema, confirmed by MRI and MRV. Imaging showed pachymeningeal enhancement and cerebral venous sinus thrombosis. We hypothesize that CVT development resulted from venous engorgement and increased blood viscosity due to CSF depletion, with altered hemodynamics and venous stasis contributing to thrombosis. No definitive guidelines exist for SIH-related CVT management, though anticoagulation—typically heparin followed by oral therapy—is commonly used [21]. Gaetano Risi et al. reported that CVT resolution was often incomplete

with anticoagulation or urokinase alone. Complete resolution was usually achieved through combined anticoagulation and epidural blood patch (EBP) therapy [22]. When EBP fails, dynamic fluoroscopy- or CT-based myelography may help identify and repair CSF leaks. Mechanical thrombectomy may be considered in severe sinus thrombosis, especially if SIH is complicated by subdural hematoma, which increases anticoagulation risk [23, 24].

Our patient received anticoagulation without complications. Although anticoagulation is standard for CVT in SIH, safety must be carefully monitored, as brain descent and subdural fluid accumulation may tear bridging veins, causing subdural hematoma [25]. In most of the previously documented cases, SIH was managed conservatively without the use of epidural blood patches (EBP), whereas CVT was treated with anticoagulant therapy [14, 26–32]. Addressing the underlying spinal CSF leak is likely the most effective approach to counteract the pathophysiological mechanisms contributing to the development of CVT. Despite these risks, our patient tolerated therapy well, without hemorrhage or hematoma. CVT is potentially fatal, with a case-fatality rate of up to 5% [33]. Successful treatment of the spinal CSF leak is likely the most effective approach to address the underlying pathophysiological mechanisms contributing to the development of CVT.

Conclusion

In conclusion, SIH is a recognized risk factor for CVT, though it occurs in only 1–2% of SIH patients. This case highlights the rare but clinically significant association between SIH and CVT, emphasizing the diagnostic and management challenges. While changes in headache patterns may suggest SIH, they are not reliable predictors of CVT. Clear treatment guidelines for CVT in the context of SIH are lacking, making management difficult. Addressing the underlying spinal CSF leak remains the most effective strategy to counteract the pathophysiological mechanisms leading to CVT. In our patient, anticoagulation was safely administered without complications, indicating it can be an effective treatment for SIH-related CVT when no contraindications exist. Further research is needed to clarify the pathophysiological link and establish standardized treatment protocols.

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Ethical Considerations

Ethics Approval and Consent to Participate

Ethics approval for this case report was not required according to the institution's policies on case reports. Written informed consent was obtained from the patient for participation, including publication of clinical data and images. The patient was informed about the purpose of the report, the information to be published, and potential risks and benefits. Participation was voluntary and could be withdrawn at any time without affecting medical care.

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Conflict of Interests

The authors have no conflict of interest to declare.

Author Contributions

Mohammad Ali Nahayati and Maryam Payere were the primary physicians responsible for patient care, follow-up, and treatment. Amir Khanmirzaei contributed to the study's conception and design, conducted data collection, prepared case reports, performed the literature review, and drafted the manuscript. Bahar Karimi assisted in manuscript drafting and critically reviewed it for intellectual content. Mohammad Ali Nahayati supported manuscript preparation, reviewed imaging and laboratory results, and study supervision. All authors reviewed and approved the final manuscript and take responsibility for all aspects of the work.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Data Availability

The data supporting the findings of this study are available from the corresponding author upon request. Data are not publicly available due to privacy considerations.

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