Spontaneous intracranial hypotension (SIH) is a condition in which intracranial pressure (ICP) decreases owing to leakage of cerebrospinal fluid (CSF) in patients without any history of procedures or trauma. Patients typically complain of orthostatic headaches that worsen when standing but improve when lying down. Complications of decreased ICP in the nervous system include subdural hematoma, subdural effusion, cranial nerve palsy, and changes in consciousness. In the past decade, several cases of cerebral venous thrombosis (CVT) due to SIH have been reported, with an estimated occurrence rate of 1–2% among patients with decreased intracranial pressure. However, the causal relationship between SIH and CVT as a potential risk factor is not well understood. According to the Monro-Kellie principle, this is thought to be caused by compensatory expansion of the cerebral venous system, damage to the intravenous wall due to changes in cerebral buoyancy, or increased venous blood viscosity. Although anticoagulation therapy is typically the first choice of treatment for patients diagnosed with CVT, considering the potential risk of intracerebral hemorrhage in patients with CVT secondary to SIH is important. We report the case of a patient who developed CVT as a complication of SIH and discuss its mechanisms and treatment options. Early identification and appropriate treatment can lead to successful outcomes and the prevention of potential complications.

CASE

A 30-year-old man with no previous history of headaches visited the emergency department because of a headache that had occurred daily for the past two weeks. This was a positional headache—it occurred when sitting or standing but disappeared when lying down. There was no history of trauma or surgery before the onset of the headache. He complained of numbness and a decreased temperature sensation in the left side of the face and upper extremity six days earlier. No muscle weakness was observed. Neurological examinations did not reveal any specific findings. Based on these symptoms, SIH was suspected.
A cord sign in the right transverse sinus was observed on the computed tomography scan (Fig. 1A). The cord sign and hyperattenuation of the thrombus are considered imaging markers of CVT. Brain magnetic resonance imaging revealed right transverse and superior sagittal sinus (SSS) thrombosis (Fig. 1B). In addition to CVT, SIH was suspected because of diffuse pachymeningeal enhancement consistent with intracranial hypotension (Fig. 1C). MR venography (MRV) revealed a filling defect in the right transverse and SSS, confirming the diagnosis of CVT (Fig. 1D). To detect a hypercoagulable state, prothrombin time, activated partial thromboplastin time, and protein C and S activities were checked, and the results were normal. All tests related to the occurrence of CVT, including those for rheumatoid factor, lupus anticoagulant, anticardiolipin antibody, antineutrophil cytoplasmic antibody, and antiphospholipid antibody, were also normal.

Given the confirmed diagnosis of orthostatic headache caused by SIH, we did not perform a CSF study to measure ICP. The patient was treated with low-molecular-weight heparin and switched to oral anticoagulation therapy. The patient did not receive an epidural blood patch (EBP) for SIH because orthostatic headache improved with intravenous hydration and bed rest. Follow-up MRV after two weeks showed partial recanalization of the right transverse sinus and SSS (Fig. 1E). Two months after discharge, the patient did not experience any recurrence of the headaches or other neurological symptoms.

**DISCUSSION**

Although the mechanism of CVT caused by decreased ICP is unclear, the incidence of CVT in patients with SIH is approximately 2%, which is significantly higher than the rate of 0.0005% in the general population. Three possible mechanisms have been identified for the development of CVT due to SIH. The first mechanism is based on the Monro-Kellie principle, which states that the total volume of the intracranial blood, brain, and CSF is constant. If the volume of CSF decreases due to leaks, the brain compensates by increasing the volume of intracranial blood, particularly the venous component. This leads to venous engorgement, slows down the blood flow velocity, and increases the risk of thrombosis. The second is related to the loss of CSF buoyancy in SIH, which causes the brain to sag and pull on the cerebral veins and sinuses, leading to mechanical distortion of the vessel wall and increased

![Fig. 1. Brain computed tomography (CT), magnetic resonance imaging (MRI), and MR venography (MRV). (A) Initial noncontrast CT suggesting a cord sign with hyperdensity of the right transverse sinus (arrow). (B) Brain MR fluid-attenuated inversion recovery imaging showing a right transverse and superior sagittal sinus thrombosis (SSS) (arrow). (C) Brain MR T1-weighted post gadolinium imaging revealing diffuse pachymeningeal enhancement, consistent with intracranial hypotension. (D) Brain MR T1-weighted image (T1WI) and MRV demonstrating a filling defect in the right transverse and SSS, confirming the diagnosis of CVT (arrow). (E) Follow-up brain MR T1WI and MRV after two weeks showing partial recanalization of the right transverse sinus and SSS (arrow).](http://www.j-nn.org)
thrombus formation. The third is due to reduced absorption of CSF into the cerebral venous sinuses, resulting in increased BV and hypercoagulability in the venous compartment.

Regarding treatment, anticoagulation therapy is typically the first choice of treatment when a patient is diagnosed with CVT. However, in patients with CVT due to SIH, considering the potential risk of intracerebral hemorrhage (ICH) that may arise from SIH is important. Currently, no established treatment guidelines exist for CVT concomitant with SIH. A literature review of case reports found that approximately half of the patients received anticoagulation therapy alone, while others underwent EBP or a combination of both treatments. Though most patients in all three groups showed good progress, there was an increased risk of ICH in patients who received anticoagulation therapy alone. Therefore, EBP may be a reasonable treatment option for patients at a high risk of bleeding. This should be considered when deciding the treatment plan for CVT in combination with SIH.

In summary, we report a rare case of CVT secondary to SIH. These symptoms and imaging findings highlight the importance of considering both CVT and SIH in patients presenting with positional headaches. It is crucial to monitor patients with SIH for alterations in their headaches or neurological symptoms that could indicate the emergence of CVT. Early identification and appropriate treatment can lead to successful outcomes and the prevention of potential complications.

Ethics Statement
This study was approved by the Clinical Trial Review Committee of Sanggye Paik Hospital (Approval No. SGPAIK 2022-03-011-001).

Availability of Data and Material
All data related to this study are included in the main text.

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Conflicts of Interest
No potential conflicts of interest relevant to this article was reported.

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