



Rare cause of cerebral venous sinus thrombosis: Spontaneous intracranial hypotension syndrome: A case report

Pan Huang

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Pan Huang, Department of Neurology, People's Hospital of Deyang City, Deyang 618000, Sichuan Province, China

Corresponding author: Pan Huang, MD, Clinician, Department of Neurology, People's Hospital of Deyang City, No. 173 Section 1, North Taishan Road, Deyang 618000, Sichuan Province, China. 1032857970@qq.com

Abstract

BACKGROUND

Spontaneous intracranial hypotension syndrome is a relatively uncommon neurological disorder of unknown etiology with a good prognosis. Cerebral venous sinus thrombosis is a specific type of cerebrovascular disease caused by multiple etiologies of cerebral venous sinus or vein thrombosis that obstructs cerebral venous return and is associated with impaired cerebrospinal fluid absorption; this entity is rarely seen clinically. Spontaneous intracranial hypotension syndrome is one of the causes of cerebral venous sinus thrombosis, and the probability of their combined occurrence is only 1%-2%. As such, it is easily overlooked clinically, thus increasing the difficulty of diagnosis and treatment.

CASE SUMMARY

A 29-year-old young woman presented with postural headache. Lumbar puncture suggested a pressure of 50 mmH₂O (normal 80 mmH₂O-180 mmH₂O), and magnetic resonance imaging cerebral venography suggested thrombosis of the supratentorial sinus. These findings were considered indicative of cerebral venous sinus thrombosis due to spontaneous intracranial hypotension syndrome after ruling out immunological causes, tumor, infection, abnormal coagulation mechanism, and hypercoagulable state, *etc.* She was treated with rehydration and low-molecular heparin anticoagulation for 15 d, and follow-up magnetic resonance imaging cerebral venography suggested resolution of the thrombus. The patient had complete improvement of her headache symptoms.

CONCLUSION

Spontaneous intracranial hypotension syndrome is one of the rare causes of cerebral venous sinus thrombosis, which is frequently misdiagnosed or missed and deserves consideration by clinicians during differential diagnosis. Dehydration should be avoided in such patients, and early rehydration and anticoagulation therapy are effective treatment options.

Key Words: Spontaneous intracranial hypotension; Cerebral venous sinus; Thrombosis; Rehydration; Anticoagulation; Case report

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Core Tip: In patients with cerebral venous sinus thrombosis of unknown origin, spontaneous intracranial hypotension syndrome may be considered as a rare cause. Lumbar puncture and cerebral venous sinus angiography are important diagnostic tools.

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INTRODUCTION

Intracranial hypotension syndrome is a group of clinical syndromes characterized by postural headache and cerebrospinal fluid (CSF) pressure < 60 mmH₂O. Spontaneous intracranial hypotension syndrome (SIH) may be considered in cases of intracranial hypotension with unknown etiology. The annual incidence of SIH is estimated at 4/100000[1]. Cerebral venous sinus thrombosis (CVST) refers to a special type of cerebrovascular disease caused by cerebral venous sinus or venous thrombosis. CVST may arise due to a variety of causes, and obstructs cerebral venous return and is frequently accompanied by impaired CSF absorption. CVST is rare, and occurs frequently in young and middle-aged people with an annual incidence of 0.5/100000; it is also associated with 0%-3% of all strokes. Spontaneous intracranial hypotension syndrome is a rare cause of cerebral venous sinus thrombosis, with an incidence of only 1%-2%; clinically, it is easy to miss and presents challenges in diagnosis and treatment[2,3]. A patient with spontaneous intracranial hypotension syndrome with intracranial venous sinus thrombosis was recently admitted to our unit. Given that this case is rarely seen clinically and there are no clear treatment guidelines, the clinical presentation, imaging features, and diagnostic and treatment procedures are summarized here with the goal of providing a reference for clinicians.

CASE PRESENTATION

Chief complaints

The patient, a 29-year-old female, was admitted to the hospital with a 3-d postural headache.

History of present illness

Three days prior to admission, the patient had a headache after waking up in the morning. The headache was located in the bilateral temporal and occipital areas, with persistent, severe, and unbearable distending pain. The headache was accompanied by a pulling pain in the back of the neck, which worsened with sitting or standing positions, and improved in a lying position. The patient also endorsed nausea and vomiting several times. She denied loss of consciousness, fever, limb convulsions, incontinence, slurred speech, and limb weakness.

History of past illness

The patient had no pertinent past illness history.

Personal and family history

The patient had no pertinent personal or family history.

Physical examination

At admission, the patient's vital signs were as follows: Temperature 36.3°C, pulse 64 beats/min, respiration rate 19 breaths/min, and blood pressure 137/79 mmHg. No significant abnormalities were seen in the heart, lungs, or abdomen. The patient was clearly conscious, fluent in speech, and had normal orientation. The pupils were round and equal in size (about 3 mm in diameter) with a sensitive light reflex and no nystagmus. There was no facial tongue palsy and the pharyngeal reflex was present. The muscle tone of the extremities was normal and the muscle strength was grade 5. Sensory

examination was normal. Bilateral pathological signs were negative, meningeal stimulation signs were negative, and tendon reflexes of all 4 limbs were present and symmetrical.

Laboratory examinations

The following blood tests were performed: Liver function, renal function, serum electrolytes, thyroid function, coagulopathy, markers of myocardial injury, syphilis, HIV, glycosylated hemoglobin, lipids, rheumatoid factor, C-reactive protein, antistreptolysin-O, erythrocyte sedimentation rate, immune panel, and tumor markers. All laboratory values were within reference ranges. An electrocardiogram demonstrated sinus bradycardia.

Imaging examinations

Computed tomography of the head suggested a nodular, linear high-density shadow in the right parietal cerebral cortex, superior parietal sagittal sinus with a differential of venous embolism or vascular malformation with thrombosis (Figure 1). No abnormality was seen in brain parenchyma, and magnetic resonance imaging (MRI) with magnetic resonance venography (MRV) was recommended (Figure 2). Computed tomographic venography of the head and neck suggested a hypodense filling defect in the cortical veins of the right parietal sagittal sinus, consistent with venous thrombosis (Figure 3).

FURTHER DIAGNOSTIC WORK-UP

An opening pressure of 50 mmH₂O was measured by uncomplicated lumbar puncture, and the CSF was colorless with no significant laboratory abnormalities. A cranial MRI enhancement scan demonstrated a localized filling defect in the right superior sagittal sinus strip with striped low signal in weighted sequences (T1WI and T2WI), suggesting a possible focal thrombus.

FINAL DIAGNOSIS

Taking into consideration the clinical symptoms, absence of medical history, the physical exam findings, and the laboratory and radiographic findings, a final diagnosis of spontaneous intracranial hypotension syndrome with cerebral venous system thrombosis was made.

TREATMENT

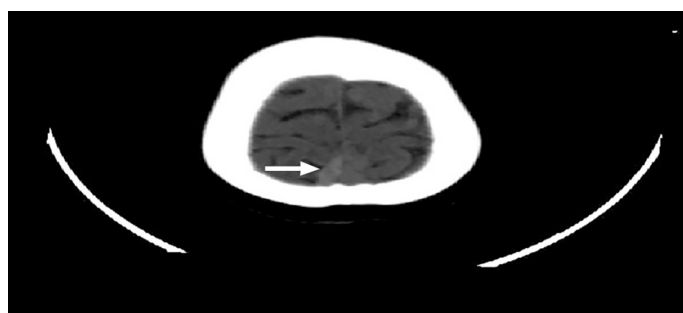
The patient was admitted to the hospital as subarachnoid hemorrhage could not be excluded. Initial treatment included nimodipine to prevent vasospasm, rehydration, and symptomatic analgesia. The patient's headache symptoms were partially resolved after 2 d of treatment. After the diagnosis of spontaneous intracranial hypotension syndrome with cerebral venous system thrombosis was clear, the treatment was adjusted to include rehydration and anticoagulation with low molecular weight heparin (5000 IU every 12 h *via* subcutaneous injection). After 13 d of treatment, the patient's headache symptoms completely resolved, and she was discharged on oral warfarin anticoagulation and recommended to increase her water intake to 2000 mL-3000 mL per d).

OUTCOME AND FOLLOW-UP

Fifteen d after discharge, the patient had no further postural headache symptoms, and a follow-up head MRI with MRV suggested no residual venous sinus thrombosis (Figure 4). Three mo later, the patient was seen again in follow-up, and she again endorsed no further headache symptoms, and anticoagulant therapy was discontinued.

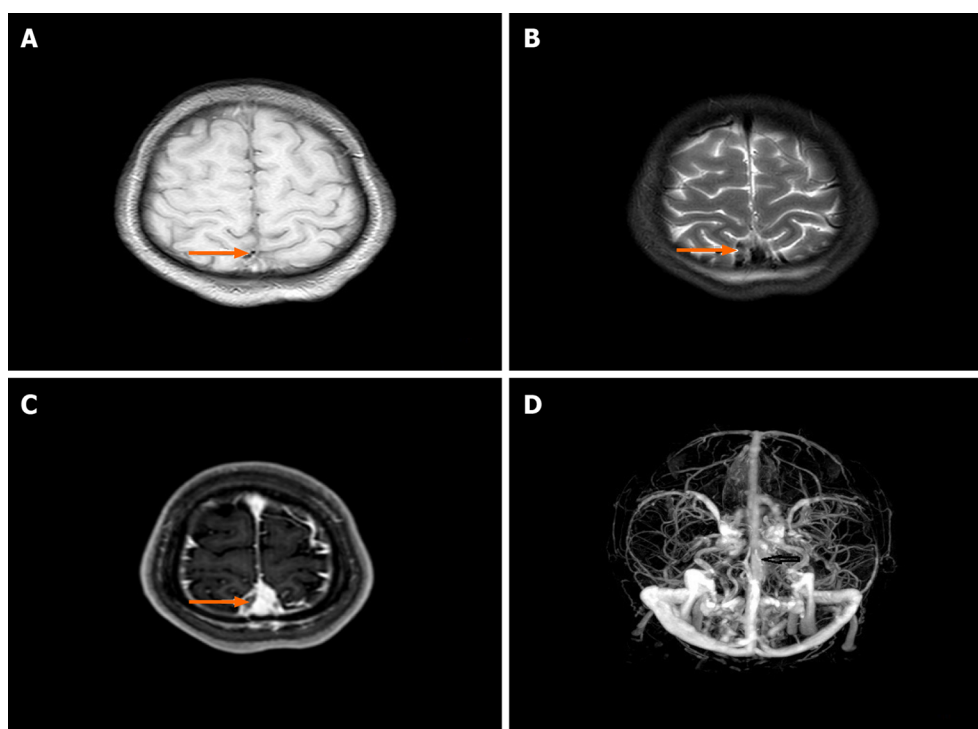
DISCUSSION

The presenting symptom of this patient was postural change-related headache. Intracranial hypotension syndrome was considered after uncomplicated lumbar puncture and related examinations were performed. The patient had no history of infection or fever during or near the onset of symptoms, the CSF opening pressure was low, and the CSF was normal on laboratory evaluation, so infectious meningitis could be excluded. There was no nodule-like enhancement on MRI, there were no features of hypertrophic cranial pachymeningitis, and there were no abnormalities on immunological examination,



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Figure 1 Head computed tomography with nodular, linear hyperdense shadow in the right parietal cerebral cortex and superior sagittal sinus (white arrow).



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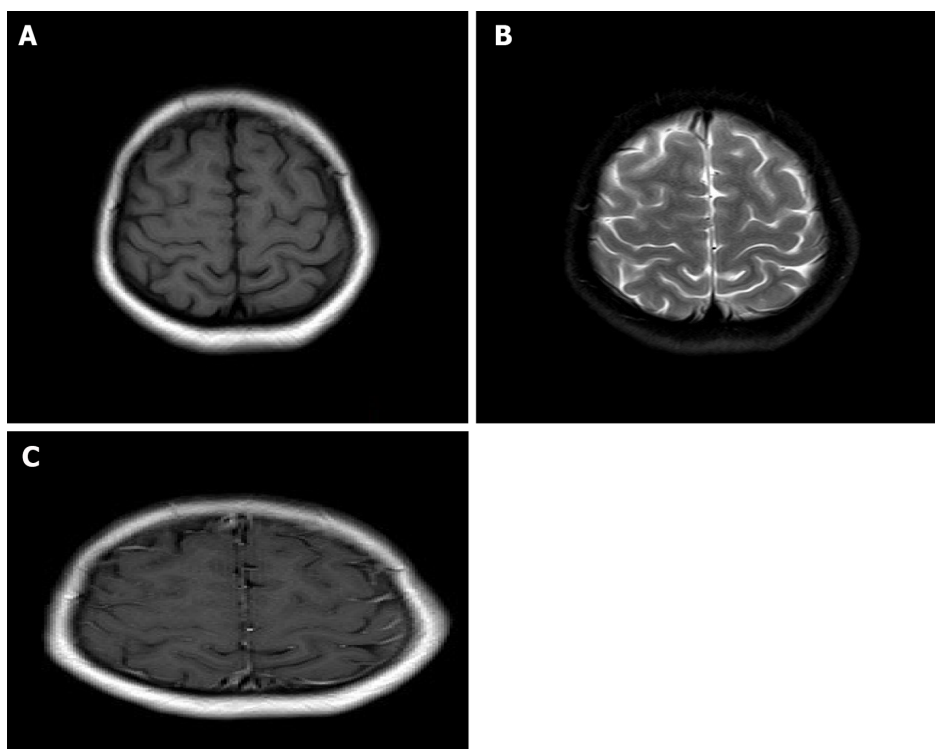
Figure 2 Head magnetic resonance imaging with magnetic resonance venography. A: A right superior sagittal sinus strip of low signal was seen on T1-weighted imaging; B: The right superior sagittal sinus strip of low signal was seen on T2-weighted imaging; C: T1-enhanced scan shows a low signal in the right superior sagittal sinus; D: The lesion is not clearly displayed on contrast-enhanced magnetic resonance venography.

so hypertrophic cranial pachymeningitis was excluded. The patient's symptoms followed an acute course, there was no blood in the CSF, no subarachnoid hyperdensity was seen on computed tomography, no aneurysm was seen on computed tomographic angiography of the head and neck, and there was no recent history of head trauma, so subarachnoid hemorrhage was not considered. The patient denied prior head trauma, cranial surgery, lumbar puncture, myelography, subarachnoid block, history of poisoning, and history of dehydration; therefore, secondary hypocranial pressure due to these etiologies was excluded. Hematological disorders are also a cause of venous sinus thrombosis; however, after a hematological workup, this cause was also ruled out[4]. Although it was not clear whether CSF leak existed in this patient, the opening pressure on lumbar puncture was less than 60 mmH₂O, which could still be considered indicative of SIH. SIH is a rare clinical neurological disorder frequently associated with CSF leakage. The typical clinical manifestation of SIH is postural headache due to reduced intracranial pressure which causes pulling on the meninges; imaging often shows dilatation of the cerebral venous system, dural enhancement, subdural effusion, and brain tissue prolapse, while myelography shows epidural CSF accumulation[5-7]. In SIH, the major etiology is a decrease in CSF volume rather than a decrease in pressure, so normal or elevated CSF pressure measurements cannot be used as a basis for excluding hypocranial pressure syndrome because CSF pressure measured during lumbar puncture in the lateral position does not reflect intracranial pressure in the upright position, nor



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Figure 3 Head and neck computed tomographic venography. A hypointense filling defect was seen in the parasagittal sinus cortical vein of the right parietal lobe, and venous thrombosis was considered (white arrow).



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Figure 4 Review of head magnetic resonance imaging. A: No abnormal signal shadow was seen in the right sagittal sinus on T1-weighted imaging; B: No abnormal signal shadow was seen in the right sagittal sinus on T2-weighted imaging; C: No abnormal signal shadow was seen in the right sagittal sinus on T1-enhanced scan.

does it provide information on CSF dynamics during postural changes[8].

In addition to SIH, further refinement of head and neck angiography in this patient suggested CVST, which is a rare and potentially fatal cerebrovascular disease with an annual incidence of 0.5/100000, accounting for 0% to 3% of all ischemic strokes[9]. The order of occurrence of spontaneous intracranial hypotension syndrome and cerebral venous thrombosis has been uncertain, however, with the advancement of science and technology, it has been confirmed that spontaneous intracranial hypotension syndrome is one of the risk factors for CVST[3]. The pathophysiological mechanism is not definitively known, but several hypotheses have been proposed. First, the Monro-Kellie theory states that due to the loss of CSF, the compensatory blood volume in the venous cavity increases, causing the cerebral venous system to dilate resulting in the slowing and stagnation of venous blood flow promoting thrombosis[10]. In support of this theory, Kranz *et al*[11] found that patients with low intracranial pressure had dilated cerebral venous sinuses and that their cross-sectional area was 70% larger than the corresponding section in normal subjects. Another possibility is outlined by the theory of abnormal CSF buoyancy, which suggests that the loss of CSF buoyancy results in intracranial tissue structure sagging, pulling the cerebral veins and venous sinuses leading to disruption of venous

hemodynamics or even stagnation of venous blood flow[12]. A final possible mechanism to consider is that if a CSF loss occurs, this can reduce the absorption of CSF by the venous system, resulting in increased viscosity and hypercoagulability of blood in the cerebral venous sinus cavity, thus increasing the risk of thrombosis[13].

Treatment of SIH includes conservative approaches (*e.g.*, bed rest, massive fluid replacement, oral caffeine, hormone therapy), epidural blood patching, epidural saline injection, surgical treatments (*e.g.*, CSF leak repair or other cause-specific procedures), and various treatments for complications[3]. One study showed the success rate of conservative treatment to be approximately 24.47%, and suggested that hormone therapy may be a potential first-line treatment option. The use of hormones was proposed to significantly improve the clinical symptoms of SIH and reduce the probability of need for invasive epidural blood patching[14,15]. In addition, hormone therapy may also be effective in improving postural headache in some patients who fail epidural blood patch treatment[16]. However, the ideal duration, type, dose, and route of administration of hormones are still inconclusive; however, the majority of patients could have symptom relief within days of administration[17]. Hormones may exert their pharmacological effects by: (1) Ameliorating brain edema and inhibiting inflammation caused by brain herniation; (2) Inhibiting meningeal inflammation and that of CSF cells or proteins, thus reducing CSF leakage; (3) Inhibiting excessive absorption of CSF; and (4) Promoting reabsorption of CSF from the epidural space and increasing the overall CSF volume[18].

In cases where SIH symptoms persist or are associated with CVST, epidural hemopexy is recommended abroad to close the CSF leak before treating CVST[19,20]. Treatment of CVST includes anticoagulation and endovascular intervention, with anticoagulation being the first-line treatment option. Anticoagulation is an important treatment modality in patients with SIH with CVST. Endovascular therapy is only indicated in patients with CVST with severe thrombotic load, psychiatric abnormalities during the course of the disease, or worsening symptoms with systemic anticoagulants.

CONCLUSION

In conclusion, SIH can often lead to the rare complication of CVST, which is frequently misdiagnosed or missed in clinical practice. For patients with postural headache with focal neurological deficits or imaging findings of venous sinus thrombosis, clinicians should consider the possibility of coexistence of both diseases, and lumbar puncture and cerebral venous sinus angiography should be performed. Patients whose clinical symptoms and imaging improve after conservative management with rehydration and anticoagulation likely do not need epidural hemorrhage patching, but should be monitored for intracranial hemorrhage.

FOOTNOTES

Author contributions: Huang P performed all the work for this paper.

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Country/Territory of origin: China

ORCID number: Pan Huang 0000-0002-5927-0369.

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