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

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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, and 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%, which 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 pressure of 50 mmH2O (normal value 80-180 mmH2O), and magnetic resonance imaging cerebral venography suggested thrombosis of the supratentorial sinus, which was considered as cerebral venous sinus thrombosis due to Spontaneous intracranial hypotension syndrome after ruling out immunity, tumor, infection, abnormal coagulation mechanism, and hypercoagulable state, *etc*. She was given

rehydration and low-molecular heparin anticoagulation treatment for 15 days, and the magnetic resonance imaging cerebral venography suggested disappearance of thrombus and complete improvement of headache symptoms.

CONCLUSION

Spontaneous intracranial hypotension syndrome is one of the rare causes of cerebral venous sinus thrombosis, which is highly misdiagnosed or missed and deserves the attention of clinicians. 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 one of its rare causes, and lumbar puncture and cerebral venous sinus angiography are important diagnostic tools.

INTRODUCTION

Intracranial hypotension syndrome is a group of clinical syndromes characterized by postural headache and cerebrospinal fluid pressure < 60 mm H_2O. For the unexplained Intracranial hypotension syndrome called Spontaneous intracranial hypotension syndrome (SIH), the annual incidence is 4/100000[1]. Cerebral venous sinus thrombosis (CVST) refers to a special type of cerebrovascular disease caused by cerebral venous sinus or venous thrombosis caused by a variety of causes, which obstructs cerebral venous return and is accompanied by cerebrospinal fluid absorption disorders. It is rare in clinic and occurs frequently in young and middle-aged people, with an annual

incidence of 0.5/100000, accounting for 0%-3% of all strokes. Spontaneous intracranial hypotension syndrome is one of the rare causes of cerebral venous sinus thrombosis, with an incidence of only 1%-2%, which is easy to ignore, thus increasing the difficulty of 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 in order to provide reference for clinicians.

CASE PRESENTATION

Chief complaints

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

History of present illness

3 days ago, the patient had a headache after waking up in the morning, located in the temporal and occipital areas bilaterally, with persistent distending pain, which was severe and unbearable, accompanied by pulling pain in the back of the neck, and the pain appeared or worsened in sitting and standing positions, and could be significantly reduced in lying position, accompanied by nausea and vomiting several times, without loss of consciousness, fever, limb convulsions, incontinence, slurred speech, and weakness of the lateral limbs.

History of past illness

Nothing special.

Personal and family history

Nothing special.

Physical examination

Body temperature was 36.3℃, pulse rate was 64 beats/min, respiration was 19 breaths/min, and blood pressure was 137/79 mmhg. No significant abnormalities were seen in the heart, lungs and abdomen. He 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 four limbs were symmetrically present.

Laboratory examinations

Routine blood work, liver function, renal function, electrolytes, coagulogram, markers of myocardial injury, syphilis, HIV, glycosylated hemoglobin, thyroid function, lipids, rheumatoid factor, C-reactive protein, anti-O, erythrocyte sedimentation rate, immune panel, and tumor markers were not abnormal. The electrocardiogram suggested sinus bradycardia.

Imaging examinations

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

FURTHER DIAGNOSTIC WORK-UP

The pressure of 50 mmH_2O was measured by perfect lumbar puncture, the cerebrospinal fluid was colorless, and there were no significant biochemical and routine abnormalities. The cranial MRI enhancement scan suggested a localized filling defect in the right superior sagittal sinus strip with striped low signal in T1WI and T2WI, and the possibility of local thrombus was considered.

FINAL DIAGNOSIS

Combined with the clinical symptoms, medical history and ancillary examinations, the diagnosis was spontaneous intracranial hypotension syndrome with cerebral venous system thrombosis.

TREATMENT

The patient was admitted to the hospital because subarachnoid hemorrhage could not be excluded, and was given nimodipine to prevent vasospasm, rehydration and symptomatic analgesic treatment, and the patient's headache symptoms were reduced after 2 days of treatment. After the diagnosis of spontaneous intracranial hypotension syndrome with cerebral venous system thrombosis was clear, the patient was immediately adjusted to rehydration and low molecular weight heparin anticoagulation (5000iu q12h subcutaneous injection). After 13 days of treatment, the patient's headache symptoms completely disappeared, and he was discharged with oral warfarin anticoagulation and increased water intake (2000-3000 mL per day).

OUTCOME AND FOLLOW-UP

15 days after discharge, there was no postural headache, and the head MRI+MRV primary venous sinus thrombosis had disappeared. 3 mo later, the patient was followed up again, and he did not have any headache, so he asked him to stop taking anticoagulants.

DISCUSSION

The clinical presentation of this patient was postural change-related headache, and Intracranial hypotension syndrome was considered after perfecting lumbar puncture and related examinations, while further etiological search revealed: (1) The patient had no history of infection or fever during the onset of the disease, and the cerebrospinal fluid pressure was low on lumbar puncture, but the cerebrospinal fluid biochemical routine was normal, so infectious meningitis could be excluded; (2) The patient did not show nodule-like enhancement on enhancement MRI, and there were no features of hypertrophic cranial pachymeningitis, and there were no abnormalities on immunological examination, which could exclude hypertrophic cranial pachymeningitis; (3) The patient had an acute course, no bloody cerebrospinal fluid, and no subarachnoid hyperdensity on computed tomography, no aneurysm on computed tomographic angiography of the head and neck, and no recent history of head trauma, so subarachnoid hemorrhage was not considered; (4) The patient denies the history of cranial trauma or surgery, history of lumbar puncture, myelography, subarachnoid block, history of poisoning and history of dehydration as possible causes of secondary hypocranial pressure; and (5) Haematological disorders are also a cause of venous sinus thrombosis, however, after screening for relevant haematological tests this cause was also ruled out[4]. Although it was not clear whether cerebrospinal fluid leakage existed in this patient, the cerebrospinal fluid pressure of lumbar puncture was less than 60 mmH_2O, which could still be considered as spontaneous intracranial hypotension syndrome. spontaneous intracranial hypotension syndrome is a rare clinical neurological disorder, often caused by cerebrospinal fluid leakage. The typical clinical manifestations are postural headache due to reduced intracranial pressure pulling on the meninges, and imaging often shows dilatation of the cerebral venous system, dural enhancement, subdural effusion, brain tissue prolapse, and myelography shows epidural cerebrospinal fluid accumulation[5-7]. In spontaneous intracranial hypotension syndrome, the core etiology is a decrease in cerebrospinal fluid volume rather than a decrease in cerebrospinal fluid pressure, so normal or elevated cerebrospinal fluid pressure measurements cannot be used as a basis for excluding

hypocranial pressure syndrome because cerebrospinal fluid pressure during lumbar puncture in the lateral position does not reflect intracranial pressure in the upright position, nor does it provide information on cerebrospinal fluid dynamics during postural changes[8].

In addition to spontaneous intracranial hypotension syndrome, further refinement of head and neck angiography in this patient suggested cerebral venous sinus thrombosis, 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 cerebral venous sinus thrombosis[3]. The pathophysiological mechanism mainly includes the following three hypotheses: (1) Monro-Kellie theory, due to the loss of cerebrospinal fluid, the compensatory blood volume in the venous cavity increases, causing the cerebral venous system to dilate, resulting in the slowing down of venous blood flow and stagnation, which promotes thrombosis[10]. Kranz *et al*[11] found that patients with low cranial pressure had dilated cerebral venous sinuses and that their cross-sectional area was 70% larger than the corresponding section in normal subjects, thus supporting the theory; (2) The theory of abnormal cerebrospinal fluid buoyancy, that is, the loss of cerebrospinal fluid buoyancy, resulting in intracranial tissue structure sagging, pulling the cerebral veins and venous sinuses to disrupt venous hemodynamics or even stagnation to promote thrombosis[12]; and (3) Cerebral venous blood component, cerebrospinal fluid loss can reduce the absorption of cerebrospinal fluid by the venous system, resulting in increased viscosity and hypercoagulation of blood in the cerebral venous sinus cavity, which increases the risk of thrombosis[13].

Treatment of spontaneous intracranial hypotension syndrome includes conservative treatment (bed rest, massive fluid replacement, oral caffeine, hormones), epidural blood patching, epidural saline injection, surgical treatment (cerebrospinal

fluid leak repair and other cause-specific procedures), and various treatments for complications[3]. One study showed a conservative treatment success rate of approximately 24.47%, where hormones may be a potential first-line treatment option. The use of hormones may significantly improve the clinical symptoms of spontaneous intracranial hypotension syndrome and may even reduce the probability of invasive epidural blood patching[14,15]. In addition, hormonal therapy may also be effective in improving postural headache in some patients who fail epidural blood patch treatment[16]. However, the duration, type, dose and route of administration of hormones are still inconclusive, and the majority of patients can have their symptoms relieved within a few days after administration[17]. Hormones may exert their pharmacological effects by: (1) Ameliorating brain edema and inhibiting inflammatory responses caused by brain tissue prolapse; (2) Inhibiting the inflammatory response of the meninges as well as the cerebrospinal fluid cells or proteins, thus reducing cerebrospinal fluid leakage; (3) Inhibit excessive absorption of cerebrospinal fluid; and (4) Causes reabsorption of cerebrospinal fluid from the epidural space and increases the volume of cerebrospinal fluid[18].

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

CONCLUSION

In conclusion, spontaneous intracranial hypotension syndrome can often lead to the rare complication of cerebral venous sinus thrombosis, which is highly misdiagnosed or missed in clinical practice. For patients with postural headache with focal neurological deficits or imaging findings of venous sinus thrombosis should be alerted to the possibility of co-existence of both diseases, and lumbar puncture and cerebral venous sinus angiography are important adjunctive examinations. Patients whose clinical symptoms and imaging improve after conservative medications such as rehydration and anticoagulation may not undergo epidural hemorrhage patching, but should be alert to the occurrence of intracranial hemorrhage.

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1	Zhong Luo, Jian Wang, Piao Cao, Lin‐Hai Zhang, Ren‐Fang Hao, Tao Liang. "Report of a family with Huntington's disease", Ibrain, 2022 Crossref	15 words — 1%

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