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Case Report

Cerebral venous thrombosis following spontaneous intracranial hypotension diagnosed by craniotomy: A case report and diagnostic pitfalls

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ABSTRACT

Background: Cerebral vein and dural sinus thrombosis (CVT) is a rare but important complication of spontaneous intracranial hypotension (SIH). The diagnosis is difficult in cases lacking typical symptoms and typical imaging findings.

Case Description: A 29-year-old male patient with a seizure attack was admitted to our hospital. Based on the head imaging findings, we misdiagnosed the patient with primary cerebral parenchymal lesion and performed an open biopsy. However, during the procedure, the patient was diagnosed with low cerebrospinal fluid pressure and cerebral cortical vein thrombosis.

Conclusion: Thus, CVT due to SIH should be considered as a probable cause of secondary parenchymal lesions.

Keywords: Cerebral vein and dural sinus thrombosis, Isolated cortical venous thrombosis, Parenchymal lesion, Spontaneous intracranial hypotension

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is an unusual neurological situation caused by low cerebrospinal fluid (CSF) volume, and the symptoms are characterized by an orthostatic headache. The neurological imaging findings useful for its diagnosis include subdural hematoma or effusion, diffuse pachymeningeal enhancement, and engorgement of the venous structure.[1,4,13]

SIH is rarely known to cause cerebral vein and dural sinus thrombosis (CVT). The prevalence of CVT followed by SIH is approximately 2.1%,[13] and recalling the relationship between SIH and CVT is not easy when the disease is encountered. Furthermore, in clinical practice, some patients visit the hospital with atypical neurological manifestations and/or atypical imaging findings that commonly lead to a misdiagnosis of these conditions.

Herein, we present a case of brain parenchymal lesions due to CVT caused by SIH. We confirmed low CSF pressure and thrombosis of the cortical vein during the craniotomy procedure and made a final diagnosis.

CASE DESCRIPTION

A 29-year-old male with no relevant medical history was transported to our hospital twice for tonic–clonic seizures. Although he had no history of head/spinal trauma or surgery, he reported a 1-week history of generalized dull headache that was not orthostatic. On arrival at the hospital, his consciousness level was E4V5M6 on the Glasgow Coma Scale, with no abnormal neurological findings. Laboratory test results were negative for autoantibodies, coagulation disorders, infectious diseases, and tumor markers, and the possibility of a hypercoagulable state was excluded.

A head computed tomography scan performed after arrival at the hospital showed a low-density area in the left frontal lobe [Figure 1a]. Although no fresh infarction was detected [Figure 1b], subsequent head magnetic resonance imaging (MRI) revealed a left frontal lesion and a small subdural hematoma [Figure 1c], without diffuse dural thickening, cerebellar tonsillar herniation, or enlargement of the pituitary gland [Figure 1d-g]. Furthermore, gadolinium-enhanced MRI showed the contrast effect of the pia mater along the superior frontal sulcus [Figure 1d-g]. Magnetic resonance venography (MRV) showed that the main cerebral venous sinuses were patent [Figure 1h]. A lumbar puncture performed on the admission day could not yield an opening pressure because we experienced "dry tap," although we used X-ray fluoroscopic guidance.

The patient underwent open biopsy on the 5th day of admission because the imaging findings were suggestive of malignant brain tumors. After craniotomy, we had to change his head position from 15 to 20 cm above the heart to 0 cm and perform a tack-up suture in a hurry because we noticed that the dura mater separated away from the skull spontaneously and steadily because of low intracranial pressure [Figure 2a and b]. A dural incision led to the recognition of regional cortical vein thrombosis [Figure 2c and d]. No peculiar infection or malignant findings were noted in the pathological specimen. Based on these surgical findings, the patient was diagnosed with CVT following SIH.

Hydration and bed rest gradually improved his headache. A lumbar puncture performed 20 days after admission demonstrated an opening pressure of 170 mmH_2O, and he was discharged on the 33rd day of hospitalization with no neurological deficits.

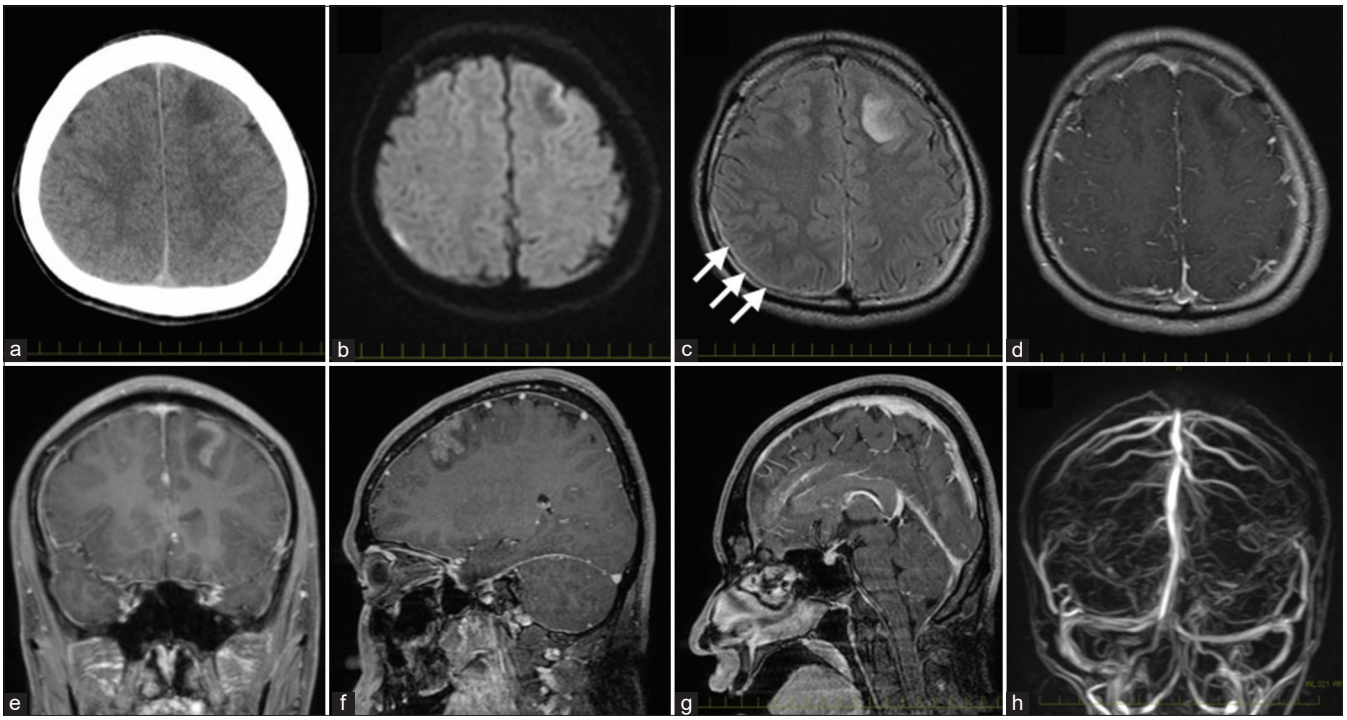

Figure 1: Neuroimaging findings of the patient. Head computed tomography performed on admission reveals a low-density area in the left frontal cortex (a). Axial image from diffusion-weighted magnetic resonance imaging (MRI) reveals no abnormalities that indicate acute cerebral ischemia (b). Fluid-attenuated inversion recovery (FLAIR) shows high-intensity lesions in the left frontal cortex and very thin subdural hematoma (arrows) (c). Gadolinium-enhanced MRI shows leptomeningeal enhancement along the superior frontal gyrus, which is involved in focal cortical hyperintensity in the left frontal lobe of FLAIR imaging (d-f). Note that dural thickening, pituitary swelling, and tonsil herniation are not observed (e and g). Magnetic resonance venography shows patency of the intracranial main venous system (h).

DISCUSSION

This case brings to light some important clinical issues. First, atypical clinical symptoms and imaging findings could make SIH diagnosis difficult, especially with brain parenchymal lesions. In the present case, it was difficult to suspect low CSF pressure during admission, because the patient presented atypical clinical (the onset was not an orthostatic headache but a seizure attack) and imaging findings (subdural hematoma or effusion was not clear, dura thickening was not obvious, pituitary swelling was not observed), and the parenchymal lesion was observed.

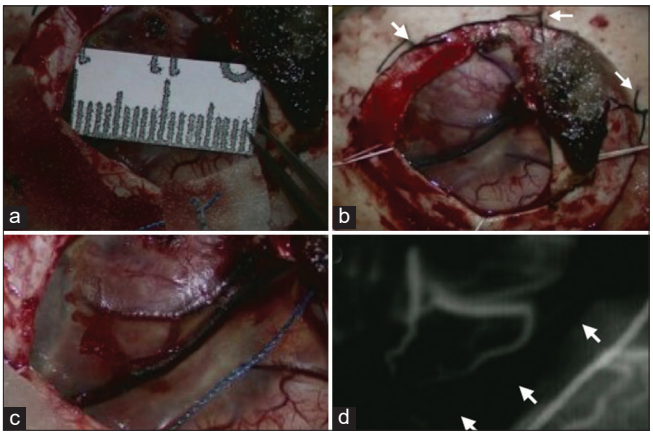

Figure 2: Intraoperative photographs of the patient. The fluid-attenuated inversion recovery high-intensity lesion is exposed through small craniotomy (a). Note that a tack-up suture is placed (arrows) since the dura mater separates away from the skull spontaneously (a and b), although the brain does not look sunken in the photographs because we lowered the patient's head position to increase the intracranial pressure. Magnified intraoperative photo shows that the cortical vein is discolored (black) (c), and indocyanine green video angiography shows the absence of flow that signifies thrombosis of the vein (d).

It should be noted that SIH should be considered even when orthostatic headache is absent or unknown. Moreover, seizures are quite common as the first symptom of CVT onset. According to past literature in which 42 case reports were reviewed,[14] 31 patients (73.8%) developed orthostatic headache, but 11 patients (26.2%) developed headache without orthostatic change. Furthermore, 12 patients (28.6%) had a seizure attack, as seen in our case. About one-third of CVT patients have reported experiencing seizures before they are diagnosed with CVT, though it was not limited to CVT due to SIH.[2,3]

Although it is known that SIH can lead to CVT, CVT is uncommon, and there are only few cases of concurrent cerebral parenchymal lesions. This is because anastomosis develops in the cerebral vein, and the anastomosis portion acts as collateral blood circulation when venous return is impaired. To the best of our knowledge, there have been only nine cases of secondary cerebral parenchymal lesions [Table 1].[4,5,7-13]

The second clinical issue to bear in mind is that "isolated" cortical venous thrombosis could be complicated by SIH. However, in the past reports, most CVT caused by SIH often has widespread thrombosis mainly in the superior sagittal sinus, transverse sinus, and sigmoid sinuses. In addition,

Table 1: Clinical features of 10 patients who developed parenchymal complications with cerebral venous thrombosis due to spontaneous intracranial hypotension.

Age, sex	Symptoms			Parenchymal	CVT	Author, year
	Seizure	OH	Other	Lesions	Location	
36 M	+	+	NOH, nausea,	ICH	CV	Lan *et al.*, 2007[8]
40 M	–	–	NOH, paresthesia	VI	SSS, StS, TS	Savoiardo *et al.*, 2006[12]
42 M	+	+	Dizziness, nausea, tinnitus, diplopia	ICH	SSS, TS	Haritanti *et al.*, 2009[5]
43 M	+	+	Diplopia	VI	SSS, TS, SS	Schievink and Maya, 2008[13]
43 M	+	–	NOH, dizziness, nausea, vomit	ICH	SSS, CV	Perry *et al.*, 2018[9]
45 F	+	+	Numbness	VI	CV	Lai *et al.*, 2007[7]
54 M	–	+	Dizziness, numbness, hemianopsia, hemiparesis, hemihypesthesia	ICH	CV	Garcia-Carreira *et al.*, 2014[4]
60 F	–	+	Hemiplegia	ICH	CV	Richard *et al.*, 2007[11]
75 M	+	–	Epilepticus state	ICH	SSS	Rice *et al.*, 2013[10]
29 M	+	–	NOH	VI	CV	Present Case

CV: Cortical vein, CVT: Cortical vein dural sinus thrombosis, ICH: Intracerebral hemorrhage, NOH: Nonorthostatic headache, OH: Orthostatic headache, SSS: Superior sagittal sinus, SS; Sigmoid sinus, StS: Straight sinus, TS: Transverse sinus, VI: Venous infarction

most cases of cortical venous thrombosis were reported to cooccur with superior sagittal sinus thrombosis.[15] Therefore, it is often possible to make a diagnosis using a venous MR angiography. However, isolated CVT may not be detected even by MRV, especially in the case of isolated small cortical venous thrombosis, as observed in our case. In the past, only four cases of cortical isolated CVT due to SIH have been reported [Table 1].[4,7,11,8]

The third important clinical issue suggested by our case is that the diagnosis of SIH and CVT can be made with craniotomy. To the best of our knowledge, the present case is the first in which low CSF pressure and CVT were confirmed by intraoperative findings, regardless of the imaging findings. In our case, cerebral parenchymal lesion with leptomeningeal enhancement raised suspicion of neoplastic diseases, and open biopsy was performed. Pathological diagnosis was negative for tumors, but intraoperative findings confirmed coexisting low CSF pressure and venous thrombosis. CVT that does not occur with SIH is reported to cause high intracranial pressure levels.[6] The intracranial pressure level is low in a patient with CVT associated with SIH. Clinicians need to be aware of this paradox for the correct diagnosis of SIH and CVT.

CONCLUSION

Initial misdiagnosis of CVT following SIH is quite common, especially when the patient presents atypical neurological manifestations and/or atypical image findings. It is important to note that SIH could cause CVT and secondary changes in the brain parenchyma.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ferrante E, Rubino F, Beretta F, Regna-Gladin C, Ferrante MM. Treatment and outcome of subdural hematoma in patients with spontaneous intracranial hypotension: A report of 35 cases. Acta Neurol Belg 2018;118:61-70.
2. Ferro JM, Canhão P, Bousser MG, Stam J, Barinagarrementeria F. Early seizures in cerebral vein and dural sinus thrombosis: Risk factors and role of antiepileptics. Stroke 2008;39:1152-8.
3. Ferro JM, Canhão P, Stam J, Bousser MG, Barinagarrementeria F. Prognosis of cerebral vein and dural sinus thrombosis: Results of the international study on cerebral vein and Dural sinus thrombosis (ISCVT). Stroke 2004;35:664-70.
4. Garcia-Carreira MC, Verge DC, Branera J, Zauner M, Herrero JE, Tio E, *et al*. Cerebral venous thrombosis in two patients with spontaneous intracranial hypotension. Case Rep Neurol Med 2014;2014:528268.
5. Haritanti A, Karacostas D, Drevelengas A, Kanellopoulos V, Paraskevopoulou E, Lefkopoulos A, *et al*. Spontaneous intracranial hypotension: Clinical and neuroimaging findings in six cases with literature review. Eur J Radiol 2009;69:253-9.
6. Kimber J. Cerebral venous sinus thrombosis. QJM 2002;95: 137-42.
7. Lai PH, Li JY, Lo YK, Wu MT, Liang HL, Chen CK. A case of spontaneous intracranial hypotension complicated by isolated cortical vein thrombosis and cerebral venous infarction. Cephalalgia 2007;27:87-90.
8. Lan MY, Chang YY, Liu JS. Delayed cerebral venous thrombosis in a patient with spontaneous intracranial hypotension. Cephalalgia 2007;27:1176-8.
9. Perry A, Graffeo CS, Brinjikji W, Copeland WR, Rabinstein AA, Link MJ. Spontaneous occult intracranial hypotension precipitating life-threatening cerebral venous thrombosis: Case report. J Neurosurg Spine 2018;28:669-78.
10. Rice CM, Renowden SA, Sandeman DR, Cottrell DA. Spontaneous intracranial hypotension and venous sinus thrombosis. Pract Neurol 2013;13:120-4.
11. Richard S, Kremer S, Lacour JC, Vespignani H, Boyer P, Ducrocq X. Cerebral venous thrombosis caused by spontaneous intracranial hypotension: Two cases. Eur J 2007;14:1296-8.
12. Savoiardo M, Armenise S, Spagnolo P, de Simone T, Mandelli ML, Marcone A, *et al*. Dural sinus thrombosis in spontaneous intracranial hypotension: Hypotheses on possible mechanisms. J Neurol 2006;253:1197-202.
13. Schievink WI, Maya MM. Cerebral venous thrombosis in spontaneous intracranial hypotension. Headache 2008;48:1511-9.
14. Yamamoto A, Hattammaru Y, Uezono S. Spontaneous intracranial hypotension associated with cerebral venous thrombosis detected by a sudden seizure: A case report. JA Clin Rep 2020;6:59.
15. Zhang D, Wang J, Zhang Q, He F, Hu X. Cerebral venous thrombosis in spontaneous intracranial hypotension: A report on 4 **case**s and a review of the literature. Headache 2018;58:1244-55.

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