Spontaneous intracranial hypotension combined with subarachnoid hemorrhage: report of 2 cases

Ya Cao
Chinese PLA General Hospital
Weinan Na
Chinese PLA General Hospital
Hui Su
Chinese PLA General Hospital
Xiaolin Wang
Chinese PLA General Hospital
Zhao Dong (dong_zhaozhao@126.com)
Chinese PLA General Hospital https://orcid.org/0000-0002-4581-2569
Shengyuan Yu
Chinese PLA General Hospital

Short report

Keywords: headache, low pressure syndrome, non-aneurysmal subarachnoid hemorrhage

Posted Date: April 2nd, 2020

DOI: https://doi.org/10.21203/rs.3.rs-19813/v1

Abstract

Background: Spontaneous intracranial hypotension (SIH) combined with subarachnoid hemorrhage (SAH) has rarely been reported. Herein, we report two patients with SIH who suffered from diffuse non-aneurysmal SAH and expanded the symptom spectrum of SIH.

Case report: A 55-year-old male was diagnosed with SIH based on orthostatic headache and diffuse pachymeningeal enhancement on brain MRI. One more month later, his headache was exacerbated, and brain CT showed diffuse SAH. Lumber puncture showed bloody CSF with a low CSF pressure of 20 mmH2O after a 30 ml intrathecal injection of saline. The patient was treated with a lumbar epidural blood patch and recovered. A 41-year-old male presented with orthostatic headache and nuchal pain. The brain CT scan confirmed the diagnosis of SAH. Brain MRI revealed diffuse dural thickening and bilateral frontoparietal subdural fluid collection. Lumber puncture showed bloody CSF with low CSF pressure. Then, an epidural blood patch was performed with satisfactory results.

Conclusion: Dilation and rupture of intracranial venous structures might play significant roles in SIH combined with SAH. We should be alert to SIH patients who develop a new persistent severe headache without relief after lying down or a suddenly changed state of consciousness.

Introduction

Spontaneous intracranial hypotension (SIH) is caused by spontaneous spinal cerebrospinal fluid (CSF) leaks, often at the spinal level and only rarely from the skull base [1]. SIH can lead to some complications, including subdural fluid collections, enhancement of the pachymeninges, engorgement of venous structures, pituitary hyperemia, and sagging of the brain [2]. However, complications, like subarachnoid hemorrhage (SAH), have rarely been reported. Herein, we report two cases in which SAH was caused by SIH and review the available literature that covers other reported cases, with the aim to expand the spectrum of SIH complications and discuss the underlying mechanism.

Case Report

Patient 1

A 55-year-old male suffered from orthostatic headache on the parietal and occipital areas on November 28th, 2019. Brain magnetic resonance imaging (MRI) with gadolinium showed diffuse pachymeningeal enhancement (Fig. 1a), and cervical MRI revealed a CSF leak on the atlantoaxial level (Fig. 1b) on December 12th. Then, the patient was diagnosed with SIH and treated with hyperhidratation and bed rest for 1 week in a local hospital, and the headache slightly improved. However, a new type of severe and persistent headache developed, accompanied by vomiting on January 9th, 2020, which led him to our hospital for further examinations. The first neurological evaluation revealed nuchal rigidity. Computed tomography (CT) angiography revealed no aneurysms or arteriovenous malformations, whereas brain CT

showed diffuse SAH (Fig. 1c). Lumbar puncture was performed on January 13th. First, we successfully performed a puncture at the L4-5 level and obtained bloody CSF, but the CSF pressure could not be measured; furthermore, the Queckenstedt-Stookey test was positive. For confirmation, we successfully reperformed the puncture at the L3-4 level, and the open pressure of the bloody CSF was 20 mmH_2O after a 30 ml intrathecal injection of saline with a positive Queckenstedt-Stookey test. Thus, the patient was diagnosed with SAH secondary to SIH and treated with nimodipine, an intravenous saline solution and an epidural blood patch (EBP) with 10 ml autologous blood injected at the C1–2 level on January 16th. On January 19th, another lumbar puncture was performed showing that the pressure of the CSF was 75 mmH_2O, and his brain CT was normal at discharge (Fig. 1d).

Patient 2

A previously healthy 41-year-old male with a 25-year history of smoking and drinking experienced a sudden onset of orthostatic cervical pain during sleep without headache, nausea or vomiting on May 16th, 2019. The patient was treated by a local hospital in a conservative manner with bed rest and painkillers, which provided limited benefits. On June 17th, the patient reported aggravation of the cervical pain and persistent bilateral headache in an upright position with mild nausea. Therefore, he was referred to our hospital for further evaluation. His neurological evaluation revealed nuchal rigidity and a positive meningeal irritation sign. The brain CT scan showed diffuse SAH (Fig. 2a). We performed a lumbar puncture on June 18, in which the open pressure was 60 mmH_2O, and the nearly uniform bloody CSF collected in three tubes was consistent with SAH. The total cell count was 25059×10^6/L (normal: 0–20×10^6/L). Brain and cervical MRI images showed dural thickening, bilateral frontoparietal subdural fluid collection (Fig. 2b) and CSF leakage at the C1-2 level (Fig. 2c). On June 24th, cerebral digital subtraction angiography (DSA) showed normal cerebral blood vessels without any aneurysmal malformation. The patient was treated with nimodipine, intravenous saline solution and EBP with 15 ml autologous blood injected at the C1–2 level on July 1st. After the procedure, his headache rapidly improved. He was asymptomatic at discharge.

Discussion

The common abnormalities of intracranial hypotension or CSF volume depletion, such as subdural fluid collections, meningeal enhancement, engorgement of venous structures, pituitary hyperemia and sagging of the brain [2], are based on the Monro–Kellie hypothesis, which indicates that the sum of the volumes of the brain, CSF, and intracranial blood is constant [3]. However, the clinical features of SIH are more variable, and the relationship between SAH and SIH is increasingly being investigated.

SIH may mimic SAH in clinical symptoms, radiological features and CSF findings. Approximately 14% of SIH patients suffered from thunderclap headache [4], CSF xanthochromia could also be observed in SIH [5, 6], and increased attenuation on CT was interpreted as a pseudo-SAH [7]. These abovementioned manifestations are easy to misdiagnose as SAH.

We presented two cases of clearly diagnosed non-aneurysmal SAH with clinical features of nuchal rigidity and meningeal irritation signs, the CT scans showed diffuse SAH, and lumber puncture revealed bloody CSF with low intracranial pressure, which is an uncommon complication secondary to SIH. In our cases, the duration between SAH and SIH was short, and bloody CSF was obtained immediately after SAH, implicating a closer relationship between SAH and SIH.

In the literature, there are 5 other cases in four studies of SAH secondary to SIH (Tab. 1) [8-11]. Tomokiyo et al reported the first case of SAH associated with SIH in Japan [8]. The patient complained of an orthostatic headache in January 2001 and presented with a sudden onset of severe nuchal pain 4 months later (on May 19th). On May 28th, a lumber puncture was performed, showing xanthochromic CSF with hypotension. Although the SAH appeared in the setting of SIH, the slightly prolonged duration and xanthochromic CSF are not sufficient evidence. Two other patients were accompanied by cortical venous thrombosis (CVT) [9, 10]. Schievink et al reported two cases of unresponsive patients with a long history of CSF leak diagnosed with SAH [11].

We speculate that the main reason for SAH secondary to SIH stems from an intravenous source for the following reasons: the patients' clinical performance did not have serious consequences, the CSF pressure was still below the normal range for SAH, the CT scans demonstrated a relatively low bleeding volume and no aneurysms were observed in the cerebral vascular angiographies, which is not very similar to the signs of artery-related SAH. Some articles speculated that as the CSF volume decreases, the intracranial venous structures dilate, and the venous blood flow velocity slows down, resulting in venous thrombosis being limited to a single cortical vein. Thus, SAH was induced by damage to the vessel wall due to blood and pressure accumulating backwards in the subarachnoid segment of the cortical vein [9, 10]. Even though venous thrombosis was not found in our two cases, we speculate that SIH might mechanically lead to congestive intracranial veins, which in turn rupture the cortical veins, basilar plexus or bridging veins, finally resulting in SAH.

In conclusion, special attention should be paid to orthostatic headache in patients with SAH-like CT scans. Moreover, we should be more vigilant about noting SIH patients' clinical features, such as a new persistent severe headache or a suddenly changed state of consciousness during a therapeutic procedure.

Abbreviations

CSF: cerebrospinal fluid; CT: computed tomography; CVT: cortical venous thrombosis; DSA: digital subtraction angiography; EBP: epidural blood patch; MRI: magnetic resonance imaging; SIH: spontaneous intracranial hypotension; SAH: subarachnoid hemorrhage

Declarations

Ethics approval and consent to participate

The ethics committee of the Chinese PLA General Hospital approved this study.

Consent for publication

Consent for publication was obtained from patients.

Availability of data and materials

The datasets used during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

Funding

This work was supported by the National Natural Science Foundation of China (grants 81671077, 81600952, 81771180, 81771200, 81901134 and 81901145), Beijing Natural Science Foundation Essential Research Project Z170002.

Authors contributions

Dr. YC was responsible for reviewing the literatures and writing the manuscript. Dr. WN was responsible for searching the literatures and collecting the clinical materials. Dr. HS and Dr. XW was responsible for giving instructions in treatment procedures. As co-corresponding author, Dr. ZD and SY had final responsibility for the decision to submit for publication. All authors read and approved the final manuscript.

Acknowledgements

The authors are most grateful for the extensive effort and contribution by doctors and nurses who participant in the therapeutic procedure.

Author information

[1] Department of Neurology, The First Medical Centre, Chinese PLA General Hospital, No.28 Fuxing Road, Haidian District, Beijing, China.

[2] School of Medicine, Nankai University, No.94 Weijin Road, Nankai District, Tianjin, China.

References

1. Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. *Headache*. 2013;53(7):1034–1053.

2. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. *JAMA*. 2006; 295(19): 2286–2296.
3. Mokri B. The Monro-Kellie hypothesis: applications in CSF volume depletion. *Neurology.* 2001;56(12):1746–1748.
4. Schievink WI, Wijdicks EF, Meyer FB, Sonntag VK. Spontaneous intracranial hypotension mimicking aneurysmal subarachnoid hemorrhage. *Neurosurgery*. 2001;48(3):513–517.
5. Schievink WI. Misdiagnosis of spontaneous intracranial hypotension. *Arch Neurol*. 2003;60(12):1713–1718.
6. Miyazawa K, Shiga Y, Hasegawa T, et al. CSF hypovolemia vs intracranial hypotension in "spontaneous intracranial hypotension syndrome". *Neurology*. 2003;60(6):941–947.
7. Ferrante E, Regna-Gladin C, Arpino I, et al. Pseudo-subarachnoid hemorrhage: a potential imaging pitfall associated with spontaneous intracranial hypotension. *Clin Neurol Neurosurg*. 2013;115(11):2324–2328.
8. Tomokiyo M, Nishihara K, Nakazawa K. A case of subarachnoid hemorrhage associated with spontaneous intracranial hypotension. *No Shinkei Geka*. 2006; 30 (8), 847-51.
9. Wang YF, Fuh JL, Lirng JF, Chang FC, Wang SJ. Spontaneous intracranial hypotension with isolated cortical vein thrombosis and subarachnoid haemorrhage. *Cephalalgia*. 2007; 27(12): 1413–1417.
10. Ferrante E, Citterio A, Valvassori L, Arpino I, Tiraboschi P. A case of convexity subarachnoid haemorrhage from intracranial hypotension treated with epidural blood patch. *Neurol Sci*. 2012; 33(3): 715–716.
11. Schievink WI, Maya MM. Diffuse non-aneurysmal SAH in spontaneous intracranial hypotension: Sequela of ventral CSF leak?. *Cephalalgia*. 2016;36(6):589–592.

Table

Due to technical limitations, Table 1 is only available for download from the Supplementary Files section.

Figures

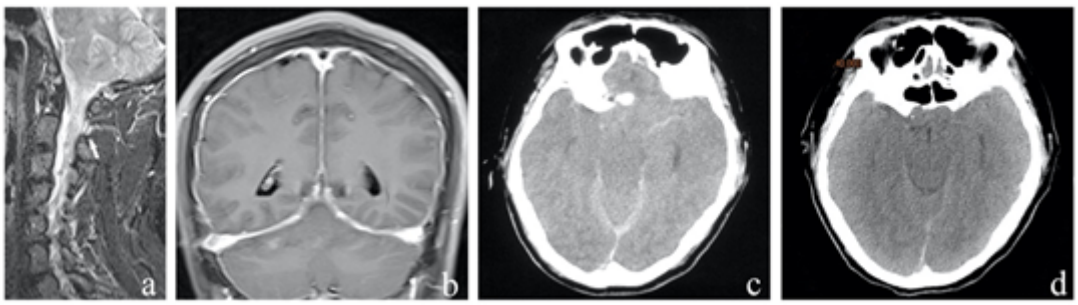

Figure 1

Sagittal T2-weighted MRI showing a CSF leakage on the C1-2 level ((a), arrow), MRI image showing diffuse pachymeningal enhancement (b); CT scan showing diffuse SAH (c) and normal image at discharge (d)

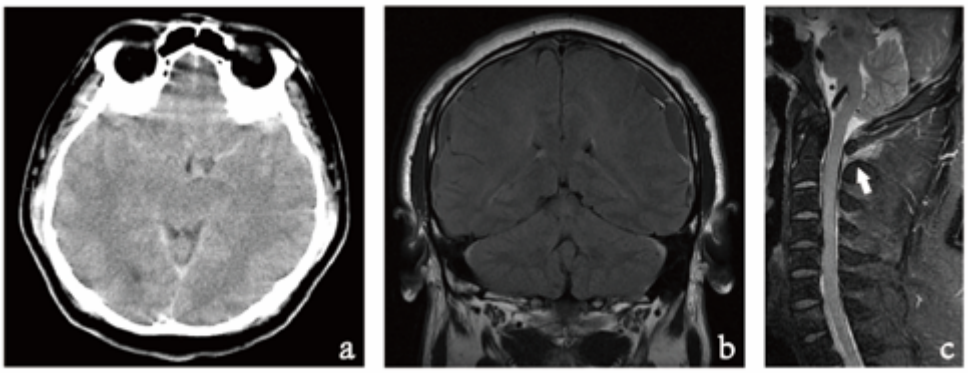

Figure 4

CT scan showing diffuse SAH (a); MRI image showing diffuse dura thickening and bilateral frontoparietal subdural fluid collection (b); Sagittal T2-weighted MRI showing a CSF leakage on the C1-2 level ((c), arrow).

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- 5Table.docx
- 5Table.docx
- 5Table.docx