Clinical Reasoning: A benign but potentially life-threatening headache

Dan Zhang, MD
Jin Wang, MD
Feifang He, PhD
Shuxu Yang, MD
Xiangyang Gong, MD,
PhD

Correspondence to
Dr. Wang:
wangjinjoy@hotmail.com

SECTION 1

A previously healthy 51-year-old man presented with a 1-week history of severe daily bilateral temporal headaches, associated with neck pain and nausea, with no fever or thunderclap-like onset. His symptoms were orthostatic, occurring within several minutes of assuming the upright position, and alleviated within 5 minutes after lying down. He had no history of lumbar puncture or head or spinal trauma. Cranial CT and CT angiography in a local hospital had normal results. Bed rest and hydration for 4 days failed

to relieve his symptoms. One day prior to admission at our hospital, he began to experience headaches even when he was lying flat. Upon admission, his neurologic examination was unremarkable except for neck stiffness.

Questions for consideration:

1. What is your differential diagnosis for his presentation?
2. What further workup would be helpful for this patient?

[GO TO SECTION 2](#)

From the Departments of Neurology (D.Z., J.W.), Anesthesiology (F.H.), Neurosurgery (S.Y.), and Radiology (X.G.), Sir Run Run Shaw Hospital, College of Medicine, Zhejiang University, China.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

SECTION 2

The hallmark of this patient's symptoms is orthostatic headache. Differential diagnoses include intracranial hypotension, postural orthostatic tachycardia syndrome (POTS), and sinusitis. Spontaneous intracranial hypotension (SIH) is caused by a spinal CSF leak without a secondary reason, such as trauma or dural puncture. Typically, orthostatic headache occurs because of decreased buoyancy of the CSF. Associated symptoms include neck pain, change in hearing, nausea, vomiting, tinnitus, diplopia, and even coma. POTS is a disorder in which the autonomic nervous system fails to compensate for upright body posture. Patients with POTS may experience excessive tachycardia, cerebral hypoperfusion, and neurologic dysfunction, including headache upon upright posture. Headache caused by sinusitis may also be positional, as sinus congestion will impede mucus drainage. Typically, however, the pain is localized to the involved sinus and may worsen when the affected person bends over or lies down. Vestibular

pathology like benign paroxysmal positional vertigo (BPPV) would also have body position–related symptoms like dizziness and spinning sensation. Some patients with BPPV may also have headache, associated with vertigo. Other possible causes of positional headache with syncope include mobile intraventricular tumor or intraventricular parasitic infection (Brun syndrome).

Cranial MRI scan with gadolinium demonstrated pachymeningeal enhancement, bilateral subdural hematomas (SDHs), and brain sagging (figure, A–C). Sequential heavily T2-weighted magnetic resonance myelography showed CSF leakage in the lower cervical region (figure, D). Based on these symptoms and neuroimages, the diagnosis of SIH was confirmed. A targeted epidural blood patch (EBP) was performed. Only 6 mL of autologous blood was injected, due to the patient's experience of chest discomfort during the injection.

The patient's symptoms partially improved after the EBP. The Visual Analog Scale for pain score decreased from 4–5 to 1–2. Seven days after the EBP, his headache

Figure Series neuroimages of a 51-year-old man with spontaneous intracranial hypotension

(A) Axial fluid-attenuated inversion recovery (FLAIR) brain MRI sequence from the initial clinical presentation reveals bilateral subdural hematomas. (B) Coronal contrast-enhanced T1-weighted MRI sequence shows pachymeningeal enhancement. (C) Sagittal T1-weighted sequence demonstrates brain sagging, flattening optic pathways, effacement of cisterns, and downward displacement of the cerebellum (arrows). (D) Sequential heavily T2-weighted magnetic resonance myelography reveals a CSF leak in the lower cervical region. (E, F) Cranial CT demonstrates mildly enlarged hematomas and effacement of the cisterns (arrow). (G) Postoperative CT reveals improvement of the cisterns obliteration (arrow). (H, I) Axial diffusion-weighted MRI sequence demonstrates restricted diffusion in pons and bilateral occipital lobes. (J–L) Axial FLAIR and coronal contrast-enhanced T1-weighted MRI sequence and sagittal T1-weighted sequence reveal improvement of subdural hematomas, effacement of the cisterns, and brain sagging. (M) Myelography shows improvement of the cervical CSF leak.

became severe and constant again. Neurologic examinations revealed neck rigidity, without any other positive signs. A repeat head CT demonstrated a mild increase in bilateral SDHs, larger on the right side with increased effacement of the cisterns (figure, E–F). A second EBP was scheduled. The next day, however, the patient exhibited nausea and vomiting, and rapidly

became comatose (Glasgow Coma Scale [GCS] score 5: E1V1M3) with a dilated right pupil, bilaterally sluggish pupillary light reflexes, and urinary incontinence.

Questions for consideration:

1. What is the reason for this patient's coma?
2. What is the treatment at this point?

GO TO SECTION 3

SECTION 3

Decreased level of consciousness and pupillary changes suggest brain herniation. In SIH, SDH enlargement, as well as brain sagging, which results from decreased buoyancy, could give rise to brain herniation. An emergent cranial CT scan was performed, which demonstrated neither obvious hematoma enlargement nor midline shift. The effacement of the cisterns and tight-looking brain on CT illustrated downward herniation of the brain. It is worth noting that coexisting SDHs, even though small, may aggravate brain sagging. Thus in such patients with altered mental status, evacuation of the SDHs is suggested.

Based on this rationale, emergent burr hole drainage of the right SDH was performed. The

patient regained consciousness right after surgery, but he was drowsy and irritated. Neurologic examinations showed dysarthria, blurred vision, and bilateral limited ocular abduction. Bilateral horizontal nystagmus was also noted, while pupil size and pupillary light reflexes were normal. No limb weakness or numbness was found. Postoperative CT revealed broadening of the cisterns, hypodensities in bilateral occipital lobes, and an enlarged left SDH (figure, G).

Questions for consideration:

1. What is the explanation for this patient's new symptoms?
2. What is the next treatment?

GO TO SECTION 4

SECTION 4

The new symptoms indicated brainstem and occipital lobe involvement. These may result from tissue compression during herniation or relate to mechanical stretching or compression of posterior cerebral arteries caused by brain sagging. Cranial MRI showed multiple areas of restricted diffusion in the pons and bilateral occipital lobes (figure, H and I). In view of the special arterial infarct mechanism, we kept this patient lying flat with adequate fluid support, and no antithrombotic therapy.

After drainage of the right SDH, the left hematoma enlarged. We assumed that the CSF leakage continued even after the initial EBP, which could explain the worsening brain sagging and recurrent SDH. Another targeted EBP was performed under general anesthesia, with 11 mL of autologous blood injected into the T2/3 epidural space. Lying flat for the ensuing month, this patient made a good recovery with residual mildly blurred vision. Repeat MRI showed resolving brain sagging and effacement of the cisterns (figure, J–L). Repeat heavily T2-weighted magnetic resonance myelography demonstrated improvement of the cervical CSF leak (figure, M). The patient remained headache-free in the following 2 years of clinic follow-up.

DISCUSSION SIH is not a common disorder, with an incidence estimated at 5 per 100,000 per year.¹ The pathogenesis of SIH is thought to be due to a spontaneous spinal CSF leak through dural defect. Decreased CSF buoyancy causes downward displacement of the brain, leading to traction on pain-sensitive structures and cranial nerves.¹

The prototypical manifestation of SIH is orthostatic headache. However, upon admission to our hospital, this patient experienced headache even while lying flat. We assumed the presence of SDH as the main cause for the change in headache characteristics. SDH is the most frequently reported complication of SIH, with an incidence of about 20%.² It may be caused by the tearing of bridging veins or the rupture of fragile and dilated new dural vessels.¹ With the development of SDH, patients may display a constant headache, a deterioration of their symptoms, or even experience an increased mass effect and midline shift leading to decreased consciousness.³ In our patient, he experienced a sudden change in consciousness. A repeat cranial CT scan revealed no obvious hematoma enlargement but brain descent. As indicated in the literature, downward displacement of the brain and associated distortion of the diencephalon and brainstem may result in central brain herniation.⁴ Hence, we considered central brain herniation secondary to SDH as the immediate cause of coma in this patient with SIH. Repeated neuroimaging is essential to rapidly identify the real reason behind sudden consciousness disturbance in patients with SIH, especially for those accompanied by SDH.

Initial treatment for SIH is generally conservative, such as bed rest and hydration.¹ For those who fail conservative management or have severe complications, the EBP is an appropriate treatment option.¹ However, there is no consensus regarding the management of patients with SIH with SDH. With previous experience, an effective EBP treatment with complete closure of the CSF leak could result in spontaneous hematoma regression and good recovery.^{5,6} However, it is worrisome that surgical drainage of SDH in patients with SIH may aggravate brain sagging, exposing patients to higher risk of central brain herniation. Decline in consciousness was reported in some patients with SIH with SDHs after surgical evacuation.⁷ Chen et al.⁸ suggest early surgical evacuation to prevent brain herniation in patients with SDH 10 mm and decreased GCS scores. Based on our experience with this ultimately successful case, surgical evacuation of SDHs may relieve pressure and improve distortion of the diencephalon and brainstem, resulting in the positive outcome of life-saving intervention at the height of a crisis situation.

With proper diagnosis and treatment, the majority of patients with SIH would have benign outcomes. However, as in this case, irrecoverable sequelae secondary to brain herniation or posterior circulation stroke may be encountered.^{4,9} Posterior cerebral arteries may be mechanically stretched or compressed during central brain herniation.⁴

Patients with SIH may develop life-threatening complications, including subdural hematoma, brain herniation, and posterior cerebral stroke. Effective EBP is the mainstay and crucial treatment for patients with SIH. Surgical evacuation of the SDH may help to stabilize the patient and create a limited therapeutic window of opportunity for further EBP treatment.

AUTHOR CONTRIBUTIONS

Dan Zhang evaluated and treated the patient, gathered all relevant clinical information, performed literature research and analysis, and drafted the manuscript. Jin Wang evaluated and treated the patient, designed the teaching file, and revised the manuscript. Feifang He performed the epidural blood patches. Shuxu Yang performed the subdural hematoma drainage. Xiangyang Gong provided an expert neuroradiology opinion on the imaging and provided the neuroimages.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES

1. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. *JAMA* 2006;295:2286–2296.
2. Lai TH, Fuh JL, Lirng JF, Tsai PH, Wang SJ. Subdural haematoma in patients with spontaneous intracranial hypotension. *Cephalalgia* 2007;27:133–138.
3. De Noronha RJ, Sharrack B, Hadjivassiliou M, Romanowski CA. Subdural haematoma: a potentially serious

- consequence of spontaneous intracranial hypotension. *J Neurol Neurosurg Psychiatry* 2003;74:752–755.
4. Schievink WI. Stroke and death due to spontaneous intracranial hypotension. *Neurocrit Care* 2013;18:248–251.
 5. Wang J, Zhang D, Gong XY, Ding MP. Rapid resolution of subdural hematoma after targeted epidural blood patch treatment in patients with spontaneous intracranial hypotension. *Chin Med J* 2014;127:2063–2066.
 6. Chen HH, Huang CI, Hseu SS, Lirng JF. Bilateral subdural hematomas caused by spontaneous intracranial hypotension. *J Chin Med Assoc* 2008;71:147–151.
 7. Dhillon AK, Rabinstein AA, Wijdicks EF. Coma from worsening spontaneous intracranial hypotension after subdural hematoma evacuation. *Neurocrit Care* 2010;12:390–394.
 8. Chen YC, Wang YF, Li JY, Chen SP, Lirng JF. Treatment and prognosis of subdural hematoma in patients with spontaneous intracranial hypotension. *Cephalalgia* 2016;36:225–231.
 9. Chi NF, Wang SJ, Lirng JF, Fuh JL. Transtentorial herniation with cerebral infarction and dural hemorrhage in a patient with spontaneous intracranial hypotension. *Cephalalgia* 2007;27:279–282.

Neurology®

Clinical Reasoning: A benign but potentially life-threatening headache

Dan Zhang, Jin Wang, Feifang He, et al.

Neurology 2016;87:e168-e173

DOI 10.1212/WNL.0000000000003203

This information is current as of October 10, 2016

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/87/15/e168.full
References	This article cites 9 articles, 1 of which you can access for free at: http://n.neurology.org/content/87/15/e168.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Low pressure syndrome http://n.neurology.org/cgi/collection/low_pressure_syndrome Other cerebrovascular disease/ Stroke http://n.neurology.org/cgi/collection/other_cerebrovascular_disease__stroke
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2016 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.