Ventral Displacement of Spinal Cord in Spontaneous Intracranial Hypotension: A Sign Easily Be Overlooked

Mao-Hsun Lin, Yung-Chu Hsu

Abstract

Purpose: Spontaneous intracranial hypotension (SIH) is suspected in patients presenting orthostatic headache and needs excluding structural or iatrogenic causes. Image studies are required to confirm the diagnosis and define exact locations of cerebrospinal fluid leakage, but currently there is no single study sensitive enough to make identifications among patients with various symptoms.

Case report: We present a 24-year-old young woman having acute orthostatic headache. She neither had, head trauma, nor recieved neuraxial procedures like lumbar puncture. Brain magnetic resonance image (MRI) with gadolinium enhancement reported normal findings on arrival. She received conservative treatment including analgesics and aggressive intravenous hydration, but her headache improved little. Whole spine MRI with gadolinium enhancement did not demonstrate obvious leakage of cerebrospinal fluid but typical dilated epidural veins with ventral displacement of her thoracic spinal cord. After autologous epidural blood patches therapy, her headache relieved completely.

Conclusion: We review the typical and uncommon findings of spinal MRI in SIH, which is more sensitive than brain MRI in acute stages. Spinal MRI offers the diagnostic value in SIH especially when cranial images do not respond in time.

Keywords: spontaneous intracranial hypotension, ventral displacement of spinal cord, dilated epidural veins, spinal MR image, orthostatic headache

Acta Neurol Taiwan 2020;29:59-63

INTRODUCTION

Orthostatic headache develops and relieves rapidly with postural changes implies causes attributed to alternation of intracranial pressures[1]. Spontaneous intracranial hypotension (SIH) is one common cause of orthostatic headache and diagnosed by clinical history and images findings[2,3]. In some patients with longer disease courses, the typical orthostatic features are lost. Gadolinium enhanced brain and spinal magnetic resonance images (MRI) are thus of importance if typical signs of low intracranial pressures are shown. Findings of

From the Division of Neurology, Department of Internal Medicine, Ditmanson Medical Foundation Chia-Yi Christian Hospital, Taiwan.
Received February 17, 2020. Revised February 25, 2020.
Accepted March 19, 2020.
Correspondence to: Yung-Chu Hsu, MD, Division of Neurology, Department of Internal Medicine, Ditmanson Medical Foundation Chia-Yi Christian Hospital. No. 539, Zhongxiao Rd., East Dist., Chia-Yi City 600, Taiwan.
E-mail: chessergg@gmail.com.tw

intracranial hypotension in brain MRI vary with time and may be absent in acute stages[4,5]. We report a patient with SIH had normal brain MRI but special spinal MRI signs. We also review imaging findings, potential mechanism, and novel MRI series for helping the diagnosis of SIH.

CASE REPORT

A 24-year-old unmarried young woman without previous headache or other medical diseases presented acute orthostatic headache for four days. The characteristic of her headache was explosive and localized in her occiput and bilateral temporal regions, which exacerbated by standing up and relieved rapidly in bed rest. The pain was associated with vomiting. She had difficulties in maintaining her daily activities because of the headache. She reported neither head trauma, nor lumbar puncture before the onset of her headache. She had visited several clinics and a local hospital but got few improvements after taking painkillers prescribed.

During the interview, her consciousness was clear, her neck was supple, and she did not present focal neurological deficits. Physical examination demonstrated neither skin hyper-extensibility nor joint hypermobility. Orthostatic headache and dizziness could be induced easily by postural changes of sitting or standing up. Her vital signs were normal (body temperature 36.9 degrees Celsius, heart rate 97 beats/minute, and blood pressures 122/80 mmHg). We also checked upright blood pressure, which showed 124/82 mmHg with heart rate 86 beats/

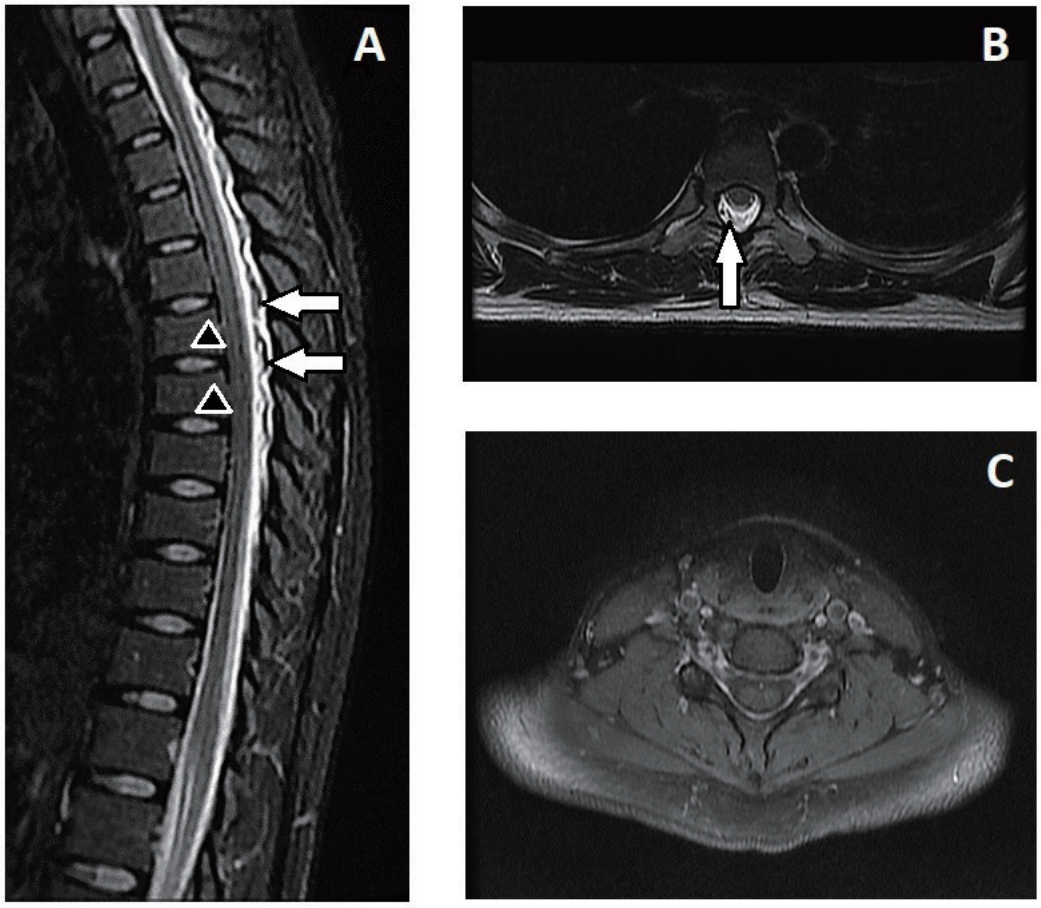

Figure 1. A 24-year-old female had orthostatic headache. A, Sagittal T2-weighted spine MR images showed dilated epidural veins (arrows) and ventral displacement of thoracic spinal cord (arrowheads). B, Dilated epidural veins in axial T2-weighted thoracic spine MR images (arrow). C, Axial T1-weighted spine MR images with gadolinium enhancement showed diffuse dural enhancement.

minute. Routine blood tests reported unremarkable in her blood cell counts, biochemistry, and autoimmune screening (including antinuclear antibodies, complements, and rheumatoid factors).

According to her clinical characteristics, SIH was highly suspected. Brain computed tomography (CT) initially excluded intracranial hemorrhage or other space occupying lesions. Brain MRI and angiography with gadolinium enhancement showed normal findings. Whole spine MRI with gadolinium enhancement did not demonstrate evidence of cerebrospinal fluid leak but dominant dilated epidural veins with ventral displacement of the spinal cord at her thoracic levels (Figure 1).

For symptomatic relief, oral medicines including acetaminophen (1500 mg/day), diphenidol (75 mg/day), and domperidone (30 mg/day) were administered during her hospitalization. Aggressive intravenous hydration (0.9% saline 3000 ml per day) and strict bed rest were also ordered for suspected low cerebrospinal fluid pressure headache. Persistent orthostatic headache was still reported in six days after above treatments. Therefore, autologous epidural blood patch 15 ml at her eighth thoracic vertebral level was done empirically by the anesthesiology specialist. Both her orthostatic headache and dizziness got rapid relief. She was discharged in pain-free states on the next day after epidural blood patches. The headache did not rebound after 3 months follow-up.

DISCUSSION

We presented a young female with orthostatic headache which presented a typical course of SIH. There were no predisposing traumatic or iatrogenic insults before her headache. Connective tissue diseases associated with SIH such as Marfan syndrome, Ehlers-Danlos syndrome, retinal detachment at a young age, or spontaneous arterial dissection were not favored in her physical features and family history. She received brain and whole spine MRI with gadolinium enhancement shortly after the onset of her headache. Brain MRI on the fourth day was normal without findings of SEEPS (subdural fluid collections, enhancement of the pachymeninges, engorgement of venous structures, pituitary hyperemia, and sagging of the brain) as reported by Schievink, but there were epidural venous dilatations and ventral displacement of the spinal cord in her spinal MRI. Although no obvious cerebrospinal fluid leak site was seen in her image studies, her headache completely relieved after autologous epidural blood patches.

Brain MRI with gadolinium enhancement has been proposed to be a useful image tool in helping confirm the diagnosis of SIH[6,7]. However, about 25% patients of SIH presented none of SEEPS in their brain MRI[8]. Spinal images especially CT or MR myelography are also commonly used in patients of suspected SIH for defining the exact location of CSF leakage. And some special findings of spinal images in SIH were found, such as spinal fluid collection, dural enhancement, dilated epidural veins, a thickened or enlarged ventral lateral epidural venous plexus, high T2 signal intensity between the C1 and C2 spinous processes (C1–C2 sign), and active contrast extravasation[9], to add the particular value in making diagnosis. The prevalence of each sign ranged 31-77%. Spinal epidural CSF collection, dural enhancement, dilated epidural veins, and C1-C2 signs were seen in more than half patients. Spinal epidural venous dilatation may also appear in other various etiologies such as arteriovenous malformation, thrombosis or occlusion of the inferior vena cava, or abdominal mass lesions[10], but it is a specific sign pointing intracranial hypotension when accompanied by other typical findings in spine MRI.

The presence of signs in brain MRI like diffuse pachymeningeal enhancement (DPE) was associated with the timing of examinations[4,5]. As reported by Fuh et al, 24.5% (13/53) patients of SIH did not have DPE in their initial brain MRI with gadolinium enhancement conducted in shorter durations after their headache onset. Eight of the thirteen patients with negative DPE received follow-up brain MRI, and six of them (75%) developed DPE later. The sign would also disappear with time. The earliest disappearance of DPE was 25 days from headache onset, whereas the longest duration of DPE was 90 days. The outcome did not differ between patients having positive or negative DPE in their initial brain MRI.

In an original study of Watanabe et al, eighteen patients of SIH were enrolled and received both brain and spinal MRI with gadolinium enhancement. The sensitivity of SIH was 83% (15/18) for brain MRI and 94% (17/18) for spinal MRI[11], which means spinal MRI is more sensitive than brain MRI for diagnosis in acute stages of

SIH. Two of the three patients having normal brain MRI findings presented either spinal epidural fluid collection or epidural vein distension, and both patients received MRI study within two day after their headache onset. The most prevalent abnormal findings of spinal MRI were epidural fluid collection (89%) and distension of epidural veins (78%). The possible pathology of distended epidural veins was proposed that decreased thecal CSF leads to collapse of spinal subarachnoid spaces and results in enlargement of epidural spaces[12], and reduced CSF volume rather than intracranial hypotension plays a major role[13]. Ventral displacement of the spinal cord is another result of above pathologic changes.

Gadolinium enhanced brain MRI without the requirement of spine MRI was recommended in the initial studies of the diagnostic algorithm made by Mokri[7], and heavily T2-weighted MR myelography is also useful to exactly localize leakage of CSF besides CT myelography or digital subtraction myelography[14]. Brain and spine MRI combined with digital subtraction myelography were recommended by Farb et al for CSF leak localization and indications of epidural blood patches[15]. Special findings in spinal images of SIH patients are also has high diagnostic values. And it is proposed by Medina et al that combined cranial and spinal MRI with gadolinium enhancement should be arranged simultaneously after failed response of conservative treatments for confirming clinical suspicions of headache attributed to intracranial hypotension.

The limitation of our case is that she did not completely fulfill the ICHD-3 criteria of SIH. Lumbar puncture was recommended for confirming low cerebrospinal pressures after her image studies failed to localize CSF leakage, but the patient and her parents refused lumbar puncture because of worries about possible adverse effects of postdural puncture headache.

Other diseases with autonomic dysfunction, like postural orthostatic tachycardia syndrome (POTS), may present orthostatic headache mimicking SIH[16]. The diagnostic criteria of POTS include heart rate increment of not less than 30 beats per minute or above 120 beats within 10 minutes of active standing or head-up tilt[17], both of which did not appear in our patient. The pathophysiology of orthostatic intolerance in POTS attributed to decreases in spinal venous pressure and volume of cerebrospinal fluid due to an absolute or orthostatic hypovolemia[18]. As reported, the common headache related to POTS is migraine like instead of orthostatic headache in SIH. In this article, they reported 4 cases with longer courses (by POTS criteria, > 6 months) of orthostatic headache. There was a four cases report[16] about orthostatic headaches without CSF leak in postural tachycardia syndrome. These four patients presented normal open CSF pressure, and unlike our patient, case 1 and 4 did not respond to epidural blood patch therapy. Case 2 and 3 were diagnosed as POTS by tilting table test so did not undergo epidural blood patch.

In conclusion, we review the typical and uncommon findings of spinal MRI in SIH, which is more sensitive than brain MRI in acute stages. Spinal MRI offers the diagnostic value in SIH especially cranial images do not respond in time.

REFERENCE

1. Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders, 3rd edition. Cephalalgia 2018;38(1): 1-211
2. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. JAMA 2006;295:2286-2296
3. Amrhein TJ, Kranz PG. Spontaneous intracranial hypotension: imaging in diagnosis and treatment. Radiol Clin N Am 2019;57:439–451
4. Mokri B, Atkinson JLD, Dodick DW, Miller GM, Piepgras DG. Absent pachymeningeal gadolinium enhancement on cranial MRI despite symptomatic CSF leak. Neurology 1999;53:402–404
5. Fuh JL, Wang SJ, Lai TH, Hseu SS. The timing of MRI determines the presence or absence of diffuse pachymeningeal enhancement in patients with spontaneous intracranial hypotension. Cephalalgia 2008;28(4):318-322
6. Friedman DI. Headaches due to low and high intracranial pressure. Continuum (Minneap Minn) 2018;24(4):1066–1091
7. Mokri B. Spontaneous intracranial hypotension. Continuum (Minneap Minn) 2015;21(4):1086–1108
8. Schoffer KL, Benstead TJ, Grant I. Spontaneous intracranial hypotension in the absence of magnetic

resonance imaging abnormalities. Can J Neurol Sci 2002;29:253-257
9. Medina JH, Abrams K, Falcone S, Bhatia RG. Spinal imaging findings in spontaneous intracranial hypotension. AJR 2010;195:459–464
10. Lee JH, Song WJ, Kang KC. Myelopathy-mimicking symptoms of epidural venous engorgement and syringomyelia due to inferior vena cava stenosis at the thoracolumbar junction in a patient with Budd-Chiari syndrome. J Neurosurg Spine. 2015;23(4):467-470
11. Watanabe A, Horikoshi T, Uchida M, Koizumi H, Yagishita T, Kinouchi H. Diagnostic value of spinal MR Imaging in spontaneous intracranial hypotension syndrome. AJNR 2009;30:147–151
12. Kranz PG, Gray L, Malinzak MD, Amrhein TJ . Spontaneous intracranial hypotension: pathogenesis, diagnosis, and treatment. Neuroimag Clin N Am 2019;29:581–594
13. Kranz PG, Malinzak MD, Amrhein TJ, Gray L. Update on the diagnosis and treatment of spontaneous intracranial hypotension. Curr Pain Headache Rep 2017;21(8):37
14. Wang YF, Lirng JF, Fuh JL, Hseu SS, Wang SJ. Heavily T2-weighted MR myelography vs CT myelography in spontaneous intracranial hypotension. Neurology 2009;73:1892–1898
15. Farb RI, Nicholson PJ, Peng PW, Massicotte EM, Lay C, Krings T, terBrugge KG. Spontaneous intracranial hypotension: a systematic imaging approach for CSF leak localization and management based on MRI and digital subtraction myelography. AJNR 2019;40:745–753
16. Mokri B, Low PA. Orthostatic headaches without CSF leak in postural tachycardia syndrome. Neurology. 2003;61:980–982
17. Fedorowski A. Postural orthostatic tachycardia syndrome: clinical presentation, aetiology and management. J Intern Med. 2019;285(4):352-366
18. Garland EM, Celedonio JE, Raj SR. Postural Tachycardia Syndrome: Beyond Orthostatic Intolerance. Curr Neurol Neurosci Rep. 2015;15(9):60