

Low cerebrospinal fluid pressure may not be necessary in the diagnosis of spontaneous intracranial hypotension: A report of four cases

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Abstract

We presented four patients with sub-acute onset orthostatic headache in occipital and sub-occipital regions and neck pain. No loss of consciousness, neurologic deficit, trauma or cranial/spinal surgery history was noted. They had normal cerebrospinal fluid (CSF) opening pressure, normal laboratory studies, and diffuse pachymeningeal enhancement or sagging of brain on gadolinium-enhanced magnetic resonance imaging (MRI). Their symptoms resolved with intravenous large isotonic fluid or epidural blood patch. The diagnosis of spontaneous intracranial hypotension requires history of orthostatic headache, demonstration of lower CSF pressure, and abnormal findings on MRI. But these patients may have normal CSF opening pressure. CSF hypovolemia rather than CSF hypotension has been proposed as the underlying cause. Therefore, the CSF pressure may not be necessary for diagnosis in such patients with typical radiographic features. Thus, in the presence of convincing clinical symptoms and imaging abnormalities, a normal CSF pressure should not discourage the clinician from searching for a source of CSF leak.

Keywords: spontaneous intracranial hypotension (SIH), low cerebrospinal fluid (CSF) pressure, orthostatic headache

INTRODUCTION

The diagnosis of spontaneous intracranial hypotension (SIH) requires detailed history taking, demonstration of low cerebrospinal fluid (CSF) pressure, and abnormal findings on magnetic resonance imaging (MRI). However, SIH remains underdiagnosed due to low index of suspicion.¹ The current International Headache Society criteria tend to make the diagnosis of SIH-associated headache ambiguous since many patients have clinical and imaging features of SIH in the absence of low CSF pressure. The identification of low CSF pressure may not be necessary for the diagnosis of SIH.

CASE REPORTS

We present here four patients with subacute onset moderate headache in occipital and sub-occipital regions, with pressure-like tightness sensation and neck pain. The headaches were precipitated by standing or walking, and were relieved with recumbent position. There was no history of loss of consciousness, neurologic

deficit, trauma, or cranial/spinal surgery. The laboratory studies showed normal results. The gadolinium-enhanced MRI revealed the presence of diffuse pachymeningeal enhancement (Figure 1) and sagging of brain (Figure 2). The CSF protein level was elevated in three of the four cases, but the opening pressure was within the normal range in all of them. (Table 1) Extradural CSF leakage was found in three of the four cases with the whole spine MRI (Figure 3). All of the four patients were successfully treated with intravenous isotonic fluid (Patient 2, 3 and 4) or epidural blood patch (Patient 1).

DISCUSSION

The CSF pressure is usually lower than 60 mm CSF in patients with SIH, but our four cases have consistently normal CSF opening pressure. Chung *et al.* reported normal CSF pressure in 18% of the patients in their case series.² CSF hypovolemia rather than CSF hypotension per se has also been proposed as the underlying cause of the headache in patients with normal CSF pressure who

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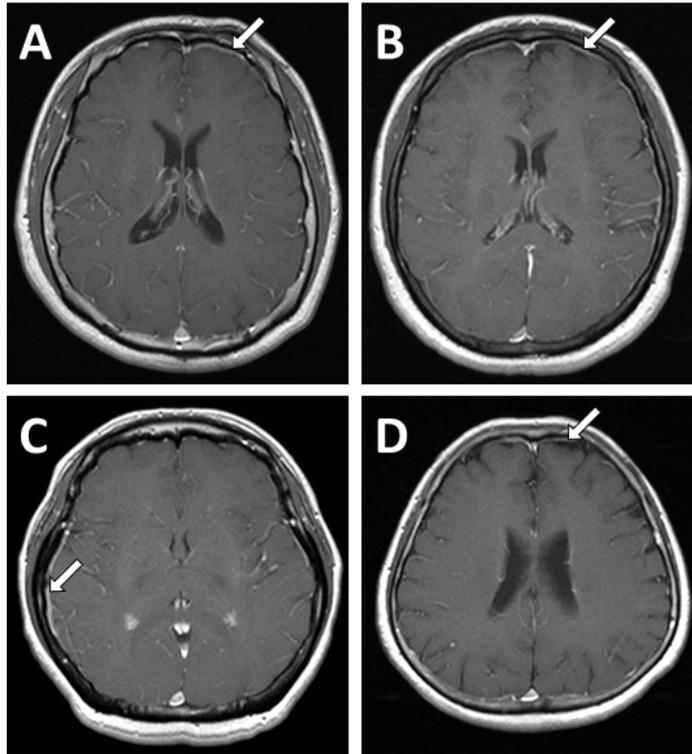


Figure 1. Axial view of the gadolinium-enhanced T1-weighted brain MRI of the four patients, showing pachymeningeal enhancement. (arrows) A. Patient 1; B. Patient 2; C. Patient 3; D. Patient 4.

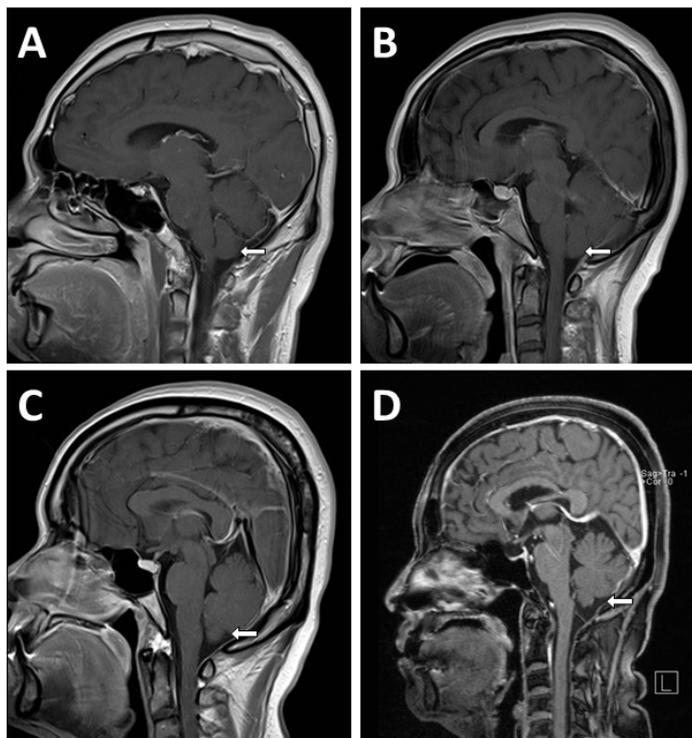


Figure 2. Sagittal view of the gadolinium-enhanced T1-weighted brain MRI of the four patients, showing sagging of brain. (arrows) A. Patient 1; B. Patient 2; C. Patient 3; D. Patient 4.

Table 1: The cerebrospinal fluid (CSF) data of the four patients

CSF data	Gender	Age	Open pressure	White cell count	Protein
Patient 1	Male	38 years old	170 mm CSF	6 count/ul	64 mg/dl
Patient 2	Female	41 years old	150 mm CSF	6 count/ul	100.8 mg/dl
Patient 3	Female	40 years old	90 mm CSF	3 count/ul	77 mg/dl
Patient 4	Female	69 years old	140 mm CSF	0 count/ul	36.6 mg/dl

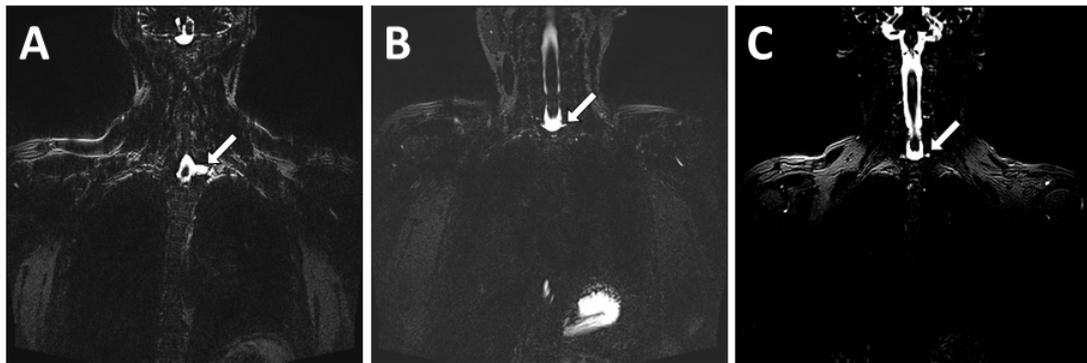


Figure 3. Coronal view of the T2-weighted whole spine MRI performed in three of the four patients, showing extradural CSF leakage. A. Patient 1, CSF leakage at left T1-2 level; B. Patient 3, CSF leakage at bilateral C-T junction; C. Patient 4, CSF leakage at bilateral C-T junction.

have clinical and radiographic features that are otherwise typical orthostatic headache.³ Altered distribution of cerebrospinal elasticity due to loss of CSF has been postulated as another plausible mechanism for headache.⁴

SIH is characterized by low CSF pressure in the cranial cavity in the absence of head injury or lumbar puncture. The estimated annual incidence of SIH is 5/100,000.¹ The peak incidence is around age 40, with male to female ratio of 1:2.

Headache attributed to low CSF pressure and headache attributed to SIH have been defined by the International Headache Society (IHS) as shown in the Table 2.⁵ Nevertheless, the criteria may be so stringent since the actual CSF pressure is not necessarily lower than 60 mm CSF and the evidence of CSF leakage on imaging may not be found, sometimes just sagging of brain is noted. Chung *et al.* considered the presence of at least two of the following three criteria to diagnose

Table 2: Headache attributed to low cerebrospinal fluid pressure (CSF) and headache attributed to spontaneous intracranial hypotension (SIH) defined by the International Headache Society (IHS) as shown in The International Classification of Headache Disorders, 3rd edition (ICHD-3)

ICHD-3: 7.2 Headache attributed to low cerebrospinal fluid pressure

- A. Any headache fulfilling criterion C
- B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

ICHD-3: 7.2.3 Headache attributed to spontaneous intracranial hypotension

- A. Headache fulfilling criteria for 7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure, and criterion C below
- B. Absence of a procedure or trauma known to be able to cause CSF leakage
- C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

SIH: orthostatic headache, CSF opening pressure < 60 mm CSF, and abnormal imaging findings, including diffuse pachymeningeal enhancement or sagging of brain on gadolinium-enhanced MRI as adequate for diagnosis of SIH.² All of our four cases meet the criteria. Besides, in the presence of convincing clinical features and imaging abnormalities, a normal CSF pressure should not discourage the clinician from searching for a source of CSF leak as the syndrome of orthostatic headaches caused by CSF leak can be seen with persistently normal CSF pressures.

In conclusion, this study suggests that identification of low CSF pressure may not be necessary for the diagnosis of SIH. A high index of clinical suspicion or imaging evidence without heavily relying on stringent diagnostic criteria or CSF pressure is key to making a timely diagnosis of SIH. Furthermore, a lumbar puncture may be omitted if intended only for CSF pressure measurement. This will avoid possible complications, as well as diagnostic confusion in case the CSF opening pressure may be normal.

DISCLOSURE

Conflict of interest: None

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