

# A Rare Case of Chiari Type-1 Malformation Accompanied by Symptomatic Cerebrospinal Fluid Hypovolemia: Comparison of Congenital Chiari Type-1 Malformation and Acquired Chiari Malformation Secondary to Cerebrospinal Fluid Hypovolemia: Case Report

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## Abstract

A 23-year-old woman was injured in a rear-end collision. She had general malaise and posterior neck pain, which were more severe when she was in an upright position. Magnetic resonance imaging (MRI) revealed the presence of cerebellar tonsil descensus and syringomyelia in the spinal cord. Radioisotope (RI) cisternography showed signs of an early accumulation of RI in the bladder, and a delayed accumulation of RI in the cerebral fornix. We considered the possibilities of cerebrospinal fluid (CSF) hypovolemia and congenital Chiari type-1 malformation as being responsible for her headache. To obtain a definitive diagnosis, we performed gadolinium (Gd)-enhanced MR cisternography and found evidence of CSF leakage. We performed an epidural blood patch (EBP), and her symptoms resolved. In 2 years since the episode, her symptoms have not recurred, and additional treatment has not been required. In addition, MRI performed 2 years after the EBP did not reveal any changes. There seems no previous report which described successful differentiation of pre-existing congenital Chiari type-1 malformation from the acquired one caused by symptomatic CSF hypovolemia. Because treatment protocols differ between these two conditions, the establishment of a correct diagnosis is important.

Key words: congenital Chiari type-1 malformation, cerebrospinal fluid hypovolemia, gadolinium-enhanced magnetic resonance cisternography, syringomyelia, epidural blood patch

## Introduction

Cerebrospinal fluid (CSF) hypovolemia is caused by a persistent or intermittent leakage of CSF. According to Mokri, who reported in 1991 that patients with spontaneous intracranial hypotension (SIH) showed pachymeningeal enhancement on gadolinium (Gd) contrast magnetic resonance imaging (MRI), the three definitive symptoms of SIH are: (1) orthostatic headaches, (2) pachymeningeal enhancement, and (3) low opening pressure on lumbar puncture.<sup>1</sup> The diagnostic criteria of SIH (or low-CSF-volume headache) described in the International Classification of Headache Disorders, 2nd Edition (ICHD-II),

include orthostatic headache that may be associated with one or more symptoms, such as images suggestive of low CSF pressure or CSF leaks, low CSF pressure, or a rapid improvement of the symptoms after epidural blood patch (EBP).<sup>2</sup>

However, recent case reports have shown that the clinical features of CSF hypovolemia are diverse, i.e., there are some atypical cases that do not meet the diagnostic criteria of SIH mentioned above.<sup>1</sup> In particular, some cases of CSF hypovolemia without orthostatic headache, pachymeningeal enhancement on brain MRI, or decreased CSF pressure have been reported. For this reason, some authors feel that the existing diagnostic criteria are insufficient.<sup>3</sup> *The 2007 Guideline for Treatment of Cerebrospinal Fluid Hypovolemia* was released

by a study group in Japan and covers the wide variety of symptoms observed in this disorder.<sup>4)</sup>

Here, we describe our experience with a very rare case of Chiari type-1 malformation accompanied by symptomatic CSF hypovolemia with a good outcome after careful diagnosis and treatment.

### Case Report

The patient was a 23-year-old woman who was injured in a rear-end motor-vehicle collision. She presented with malaise and posterior neck pain, and a nearby clinic established a diagnosis of traumatic cervical syndrome and prescribed conservative treatment with drugs and a cervical collar. However, her symptoms did not improve after 2 months of treatment, and she was referred to our department for examination. Before the accident, she had not experienced headache. Therefore, she had never received a radiological examination of the head. At the time of the hospital visit, 2 months after the accident, she complained of intense general malaise and posterior neck pain, which were more severe when she was in an upright position. She reported that very intense malaise often confined her to bed. There were no accessory symptoms related to headache or any other neurological deficits.

Spinal MRI revealed that her cerebellar tonsil was descended by 8 mm from the foramen magnum, with syringomyelia formation from C3 to Th7 (Fig. 1A). Brain MRI did not reveal pachymeningeal enhancement. There were neither traumatic changes nor brain sagging or subdural fluid collection (Fig. 1B, C). Radioisotope (RI) cisternography demonstrated signs of early RI accumulation in the bladder, and delayed accumulation of RI in the cerebral fornix. However, there was no evidence of

CSF leakage (Fig. 2). CSF pressure measured at the time of the performance of lumbar puncture for RI cisternography was 10 cmH<sub>2</sub>O. MR myelography yielded images that were suggestive of a diffuse CSF leak in the bilateral paravertebral region; however, it did not identify the source of the leak.

Although MRI findings suggested that these manifestations could be due to a congenital Chiari type-1 malformation, CSF hypovolemia was strongly suspected on the basis of the findings of the RI cisternography and MR myelography, and the symptom of orthostatic pain. To obtain a definitive diagnosis of CSF hypovolemia and provide appropriate treatment, we performed additional Gd-enhanced MR cisternography. Because intrathecal administration of Gd is contraindicated in Japan, we obtained an authorization to perform this procedure from our hospital's internal ethics committee, and thoroughly explained about informed consent to the patient and her family before performing Gd-enhanced MR cisternography.

Our examination was conducted according to the method established by Albayram et al. as follows:<sup>5)</sup> (1) a solution of Gd-contrast medium (0.5 mL Gd-contrast medium + 4 mL saline) was injected into the subarachnoid space from the lumbar spine; (2) the patient stayed in an elbow-to-knee position for 15 min, and then in a supine position for another hour before; and (3) whole-spine MRI was performed.<sup>5)</sup> Gd-enhanced MR cisternography revealed that the contrast medium was leaking from the left side of the L4–5 area (Fig. 3). This evidence allowed us to establish a diagnosis of CSF hypovolemia. We performed EBP on the leak site, and her symptoms resolved 3 days later. In 2 years since the procedure, she had no recurrence of symptoms and had not required additional treatment. Moreover, she gave birth to a healthy infant 1 year after treatment. MRI performed 2 years after EBP showed no

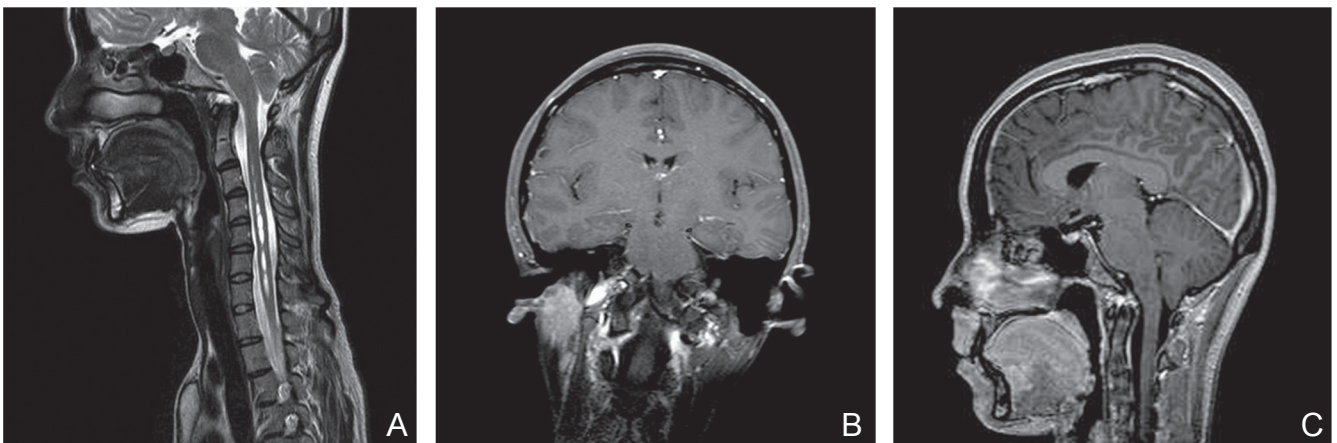


Fig. 1 Spinal T<sub>2</sub>-weighted magnetic resonance (MR) imaging on admission revealed the presence of cerebellar tonsil descensus (8 mm below the foramen magnum) and syringomyelia in the spinal cord, from C3 to Th7 (A). Brain coronal and sagittal T<sub>1</sub>-weighted MR images with gadolinium did not reveal pachymeningeal enhancement, brain sagging, or subdural fluid collection (B, C).

change in the degree of cerebellar tonsil descensus or syringomyelia (Fig. 4).

## Discussion

According to the diagnostic criteria for SIH (or low CSF-volume headache) specified by the ICHD-II, its major symptom is orthostatic headache associated with neck stiffness, tinnitus, hypacusia, photophobia, or nausea. SIH can be diagnosed when at least one of the following criteria are met: (1) evidence of low CSF pressure on MRI (e.g., pachymeningeal enhancement, brain sagging, or subdural fluid collections); (2) evidence of CSF leakage on conventional myelography, CT myelography, or cisternography; or (3) CSF opening pressure < 60 mmH<sub>2</sub>O in a seated position. Headache resolves soon after EBP is performed.<sup>2)</sup>

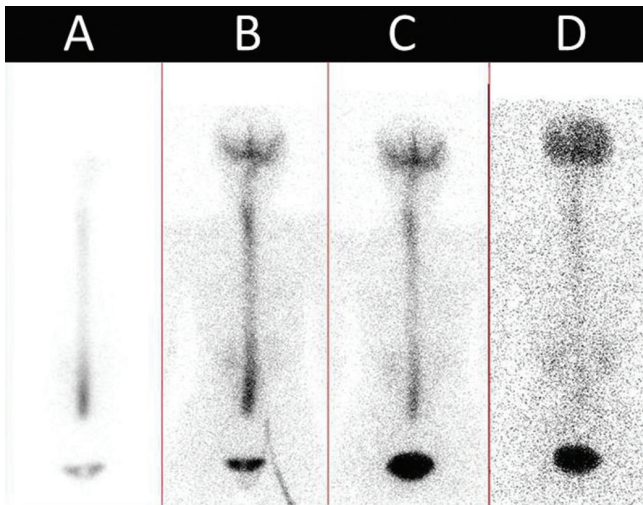


Fig. 2 Sequential images of radioisotope (RI) cisternography were acquired at 1 h (A), 4 h (B), 6 h (C), and 24 h (D) after injection. Early accumulation of RI was observed in the bladder, and a delayed accumulation of RI was observed in the cerebral fornix.

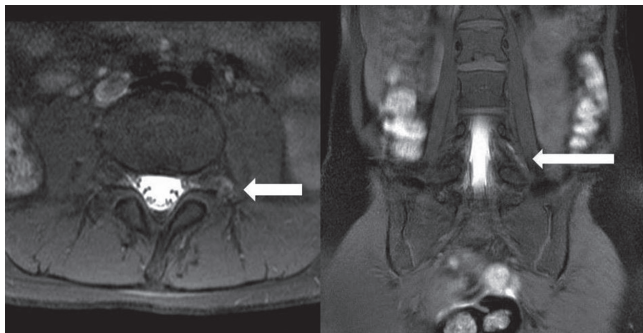


Fig. 3 Gd-enhanced magnetic resonance (MR) cisternography revealed that the contrast medium was leaking from the left side of the L4-5 area (arrow).

However, it is well established that the clinical and radiological findings of CSF hypovolemia are highly variable.

Our patient presented with malaise and posterior neck pain that were more severe when she was in an upright position. Because imaging results were consistent with CSF hypovolemia, and the symptoms remitted after the EBP, our patient was ultimately diagnosed with CSF hypovolemia, despite an incomplete match with the ICHD-II SIH criteria.

Cerebellar tonsil herniation and syringomyelia are characteristic of Chiari type-1 malformation. However, hindbrain herniation is not always secondary to congenital Chiari type-1 malformation. Acquired tonsillar descent can be secondary to a variety of conditions. This is often seen after procedures such as lumbar drainage and lumboperitoneal shunting, as well as chronic leaks of spinal CSF.<sup>6)</sup> This is called acquired Chiari malformation.

A headache caused by acquired Chiari malformation secondary to CSF hypovolemia is an orthostatic headache, and often has a shorter duration than a headache caused by congenital Chiari type-1 malformation. In addition, it is rare that acquired Chiari malformation secondary to CSF hypovolemia is complicated by syringomyelia.<sup>7-11)</sup> Thus, there are some differences between congenital Chiari type-1 malformation and acquired Chiari malformation secondary to CSF hypovolemia (Table 1).<sup>6)</sup> However, SIH cases that were first considered to be congenital Chiari type-1 malformation accompanied by headache have been reported.<sup>12)</sup> Puget et al. reported a case of SIH in which the symptoms and related images improved after EBP,



Fig. 4 T<sub>2</sub>-weighted magnetic resonance imaging performed 2 years after epidural blood patch revealed the presence of cerebellar tonsil descensus and syringomyelia in the spinal cord.



**Table 1 Comparison of congenital Chiari type-1 malformation and acquired Chiari malformation secondary to CSF hypovolemia**

	Congenital Chiari type-1 malformation	Acquired Chiari malformation secondary to CSF hypovolemia
Headache	Cough headache and neck pain	Orthostatic headache
Duration	Long duration	Short duration
Syringomyelia	30–70%	Rare
Therapy	Posterior fossa reconstruction	EBP, Surgical repair

CSF: cerebrospinal fluid, EBP: epidural blood patch.

but not after foramen magnum decompression (FMD).<sup>10)</sup> Schievink also reported two cases of SIH that were first misdiagnosed as congenital Chiari type-1 malformation and who were then treated by FMD.<sup>13)</sup> Therefore, it is extremely important to differentiate these two distinct diseases via diagnostic testing.

In our patient, we observed cerebellar tonsil descensus and syringomyelia from C3 to Th7 on MRI. Because her imaging findings and CSF opening pressure did not fulfill the ICHD-II criteria exactly, we considered the possibilities of both CSF hypovolemia and congenital Chiari type-1 malformation; thus, we needed to differentiate between these two conditions to establish an appropriate treatment plan. For this reason, we performed Gd-enhanced MR cisternography, which identified the leakage site clearly. Eventually, we were able to establish a definitive diagnosis of CSF hypovolemia based on her symptoms, as well as provide her with a good outcome after a single EBP using the clear identification of the leakage site.

Since the first report describing its use in 2002, Gd-enhanced MR cisternography has become a useful tool for diagnosing CSF rhinorrhea and otorrhea.<sup>14,15)</sup> Since 2008, more than 100 reports have demonstrated the usefulness of Gd-enhanced MR cisternography in diagnosing CSF hypovolemia without causing any serious adverse events.<sup>3,5,16)</sup> Aydin et al. reported follow-up results in patients who had undergone Gd-enhanced MR cisternography for CSF rhinorrhea (during a mean follow-up period of 4.12 years); those authors did not experience any serious adverse events or other lingering problems.<sup>17)</sup> Schick et al. reported their results demonstrating that Gd-enhanced MR cisternography was the best method to identify CSF leakage sites. They also showed that EBP improved symptoms with a low incidence of adverse drug reactions.<sup>16)</sup> Albayram et al. pointed out that Gd-enhanced MR cisternography offers higher spatial resolution than does RI cisternography in terms of identifying the leakage site, and that the Gd-contrast medium is less viscous than the contrast medium used for CT myelography; thus, Gd-enhanced MR cisternography is expected to identify leakage sites more effectively.<sup>5)</sup> However, intrathecal administration of

Gd is contraindicated in Japan. To obtain an authorization from ethics committee an informed consent is necessary to perform Gd-enhanced MR cisternography.

It has been speculated that acquired Chiari malformation may appear as a complication of SIH.<sup>12,18)</sup> Although it is rare, there are reported cases of SIH appearing with complications of cerebellar tonsil descensus and syringomyelia.<sup>7–11)</sup> However, syringomyelia in all of these cases improved within 4 months after treatment.

Based on these reports, our patient was thought to be a very rare case of asymptomatic congenital Chiari type-1 malformation appearing with a complication of CSF hypovolemia; this diagnosis was supported by the fact that no changes were observed in the MRI findings collected 2 years after the EBP. We consider that it is necessary to continue long-term imaging follow-up in this patient. There seems no previous report which described successful differentiation of pre-existing congenital Chiari type-1 malformation from the acquired one caused by symptomatic CSF hypovolemia.

## Conclusion

We report our experience with a patient with headache in whom it was difficult to differentiate between CSF hypovolemia and congenital Chiari type-1 malformation. We have compared these two conditions. Because the treatment protocols differ between these two conditions, it is important to establish a correct diagnosis.

## Conflicts of Interest Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices in the article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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