

## Cervical cerebrospinal fluid venous fistula with syringomyelia treated with suboccipital decompression: illustrative case

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**BACKGROUND** Cerebrospinal fluid (CSF) venous fistulas are a recently discovered and underdiagnosed cause of spontaneous spinal CSF leak, which may lead to spontaneous intracranial hypotension. Most cases occur in the thoracic spine, and only 2 cases were reported in the cervical spine. Treatments include the epidural blood patch, fibrin glue injection, and surgical ligation of the fistula.

**OBSERVATIONS** The authors report the treatment of a C6–7 CSF venous fistula, for which direct ligation was not feasible, with suboccipital decompression, leading to the complete resolution of the symptoms. Based on the clinical course and outcome in our patient, the authors summarize the previous theory and propose a hypothesis for the pathophysiology of headache and other symptoms in patients with CSF venous fistulas.

**LESSONS** The symptoms of CSF venous fistulas may be linked not only to intracranial hypotension but also to the altered CSF dynamics induced by tonsillar herniation. Suboccipital decompression should be considered as a potential treatment option, especially in patients with Valsalva-induced headache who show a poor response to surgical ligation, patients in whom surgical ligation is not feasible, and patients with foramen magnum obstruction. Further investigation of the pathophysiology of CSF venous fistulas is warranted and should be performed in the future.

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**KEYWORDS** cerebrospinal fluid venous fistula; spontaneous intracranial hypotension; spontaneous spinal cerebrospinal fluid leak; syringomyelia; tonsillar herniation; suboccipital decompression

Cerebrospinal fluid (CSF) venous fistulas are a recently discovered, underdiagnosed source of spontaneous spinal CSF leakage that might cause spontaneous intracranial hypotension (SIH).<sup>1,2</sup> Established treatment includes an epidural blood patch, fibrin glue injection, and direct ligation of the fistula—considered the optimal treatment. The CSF venous fistula is predominantly thoracic, with only 2 cases reported in the cervical spine (C7/T1). Here, we report treatment of a C6–7 CSF venous fistula, in a patient for whom direct ligation was not feasible, with suboccipital decompression, leading to the complete resolution of the symptoms.

### Illustrative Case

A 30-year-old woman with a history of well-controlled Evans syndrome presented with orthostatic headache accompanied by dizziness, nausea, and a dragging sensation in the occipital region for

four months. The pressure headache usually lasted for ~15–20 minutes and subsided spontaneously. Associated nuchal rigidity, shoulder soreness, otalgia, and tinnitus developed later. She denied any trauma history. Magnetic resonance imaging (MRI) showed typical findings of SIH (brain sagging, pachymeningeal enhancement, and venous distention) and a linear hyperintense signal at the C2–3 level on T2-weighted imaging, which raised the suspicion of intracranial hypotension due to a ventral spinal CSF leak (Fig. 1). In the following 6 months, she received a radiologically targeted epidural blood patch and multilevel fibrin glue injection, but without success. Furthermore, the headache was no longer restricted to the occipital region and radiated to the frontal region. A Valsalva-induced headache was also reported.

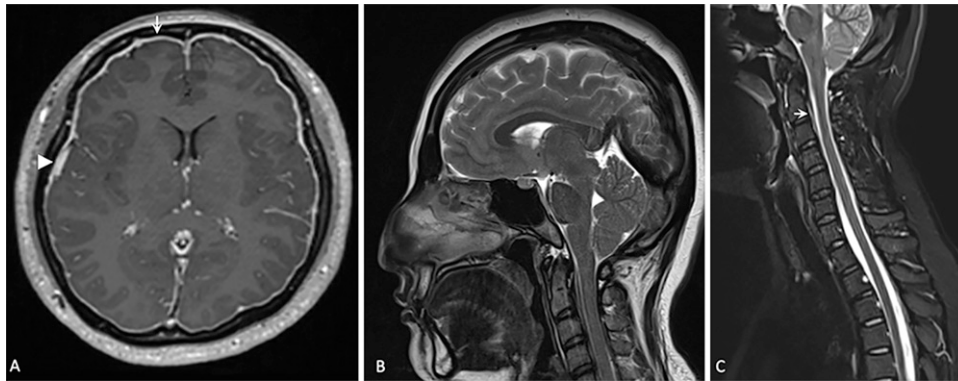
Sequential examinations were arranged (Fig. 2). MRI showed aggravated tonsillar herniation and newly formed syringomyelia at

**ABBREVIATIONS** CSF = cerebrospinal fluid; CT = computed tomography; DSM = digital subtraction myelography; MRI = magnetic resonance imaging; SIH = spontaneous intracranial hypotension.

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**FIG. 1.** Initial imaging showed pachymeningeal enhancement (*arrow*) and venous distention (*arrowhead*) on postcontrast T1-weighted MRI scan (**A**) and brain sagging with tonsillar herniation (**B**) and linear hyperintense signal (*arrowhead*) at the C2–3 level (**C**) on T2-weighted MRI scans, which raised the suspicion of intracranial hypotension resulting from ventral spinal CSF leakage.

the C2–T2 level. Conventional computed tomography (CT) myelography revealed a hyperdense paraspinous vein sign at the C6–7 level. Digital subtraction myelography (DSM) confirmed a CSF venous fistula at the right C7 nerve root sleeve. She received a radiologically targeted epidural blood patch again, which gave her transient relief from her headache. However, the symptoms relapsed, leading to a more severe headache, nuchal rigidity, intrascapular pain, generalized hyperreflexia, and right arm numbness 1 week later. Follow-up DSM showed a persistent CSF venous fistula at the right C7 nerve root sleeve. Given the rapid deterioration of symptoms and worsened myelopathy, which could be due to reasons other than intracranial hypotension, she underwent suboccipital craniectomy and dural augmentation for decompression.

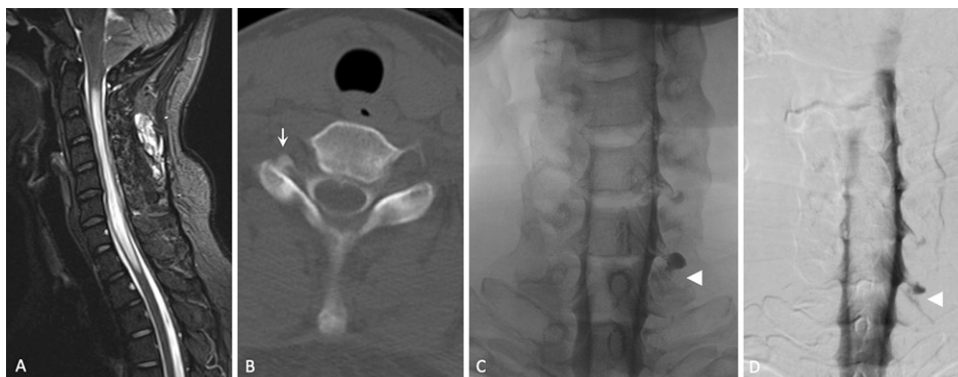
The patient tolerated the surgery well, and her perioperative course was smooth without any complications. After overnight observation in the intensive care unit, she was transferred back to the general ward the next morning. She was discharged on the sixth postoperative day with resolved symptoms. No recurrence of symptoms was reported during follow-up at the outpatient clinic. Postoperative MRI at 8 months showed completely resolved syringomyelia (Fig. 3).

## Discussion

### Observations

CSF venous fistula was discovered in 2014 and has increasingly been recognized as a cause of spontaneous spinal CSF leakage that may lead to SIH.<sup>1</sup> It is an abnormal connection between the subarachnoid space and the epidural venous plexus.<sup>2,3</sup> A recent literature review described a right-sided predominance of CSF venous fistulas with a single level of involvement. Most cases (95.2%) occur in the thoracic spine;<sup>2</sup> the cervical or lumbar<sup>2,4–6</sup> spine is an unusual location for CSF venous fistulas. To our knowledge, only 2 cases were reported in the cervical spine, and both of them were at the C7–T1 level.<sup>7</sup>

Typical clinical symptoms of CSF venous fistulas are associated with SIH, including headache (especially orthostatic or Valsalva-induced headache), dizziness, and nausea/vomiting. Rarely, associated symptoms include nuchal stiffness, photophobia, phonophobia, cognitive disturbance, personality changes, tinnitus, reduced hearing, imbalance, movement disorders (tremor, choreiform movements, parkinsonism, and ataxia), vision alteration (blurred vision, spots, and diplopia), numbness, paresthesia, and hypersomnolence.<sup>2,7,8</sup> The clinical course is insidious, and the duration from symptom onset to diagnosis may be as long as several months to years.<sup>2,4,6,9</sup>



**FIG. 2.** Preoperative T2-weighted MRI scan showed aggravated tonsillar herniation and newly formed syringomyelia at the C2–T2 level (**A**); conventional CT myelography revealed a hyperdense paraspinous vein sign (*arrow*) at the C6–7 level (**B**); and DSM confirmed a CSF venous fistula (*arrowhead*) at the right C7 nerve root sleeve before (**C**) and after (**D**) the radiologically targeted epidural blood patch.



**FIG. 3.** Postoperative MRI scan obtained at 8 months, showing completely resolved syringomyelia and no foramen magnum obstruction.

Pathophysiologically, CSF venous fistula is thought to originate from communication between the subarachnoid space and the epidural venous plexus due to dural weakness surrounding the nerve root sleeves, meningeal diverticula, or rupture of the arachnoid granulations distributed along with the nerve roots.<sup>2,3,8,10</sup> The CSF flow appears to be unidirectional into the venous system.<sup>3</sup> The insidious headache in the early stage of the disease may result from SIH caused by the drainage of CSF from the CSF venous fistula. Postural changes may result in insidious headache and progress gradually.<sup>2,7,11,12</sup> Because the previously reported opening pressures were not as low as expected (84% were 7–20 cmH<sub>2</sub>O),<sup>7</sup> we postulate that there may be an unknown compensatory mechanism that helps overcome the insidious progression of SIH. Brain sagging progresses if this compensation is overwhelmed. The aggravation of downward herniation and tonsillar herniation may cause crowding of the posterior fossa, brainstem distortion and compression, and traction of the cerebral hemispheres and cranial nerves.<sup>2</sup> These changes may result in some uncommon symptoms, including cognitive impairment, cochleovestibular dysfunction, cerebellar signs, and movement disorders.

Diagnosis is based on symptoms and signs of intracranial hypotension and the presence of CSF venous fistula using radiological imaging. Because concurrent epidural CSF leak was not seen in most cases (91%),<sup>5</sup> patients with SIH with no identifiable epidural CSF collection on spine imaging should be investigated for CSF venous fistula.<sup>3</sup> Besides, at least one sign of SIH on contrast-enhanced brain MRI was present in most cases (>91%), including venous distention, pachymeningeal enhancement, brain sagging, and subdural collections.<sup>5,10,13</sup> Several imaging modalities could be used to identify the CSF venous fistula, but the optimal methods are still being explored.<sup>2,3,5</sup> DSM is thought to be the best diagnostic tool, especially with the patient in the lateral decubitus position, which may help increase the rate of fistula detection.<sup>2,3,10,14</sup> Drainage of contrast material into the paravertebral venous system confirms the diagnosis of CSF venous fistula. When DSM is not available, dynamic CT myelography can be an alternative choice,

followed by a combination of conventional CT myelography and MR myelography.<sup>2,3</sup> Performing conventional CT myelography or MR myelography alone often misses the diagnosis of CSF venous fistulas.<sup>2</sup> The presence of the dense paraspinous vein sign with a minimum threshold of 70 Hounsfield units in conventional CT myelography is highly suggestive of a CSF venous fistula.<sup>2,5</sup>

Current treatments for CSF venous fistulas include an epidural blood patch, percutaneous fibrin glue injection, and surgical fistula ligation. Recent case series also reported favorable results regarding the novel use of transvenous embolization of the paraspinous vein with Onyx in 5 patients with thoracic CSF venous fistulas. Almost all patients had conditions refractory to an epidural blood patch or fibrin glue injection. Their symptoms might be relieved transiently after application of an epidural blood patch or fibrin glue injection but recurred later. Surgical ligation of CSF venous fistulas accompanied by nerve root ligation provides better outcomes.<sup>2,7,9,10</sup> However, ~17–24.4% of patients reported no or little (<50%) improvement in headache postoperatively.<sup>7,9</sup> This indicates that the pathophysiology underlying the clinical symptoms of CSF venous fistulas is much more complicated than what is indicated by the above-mentioned known evidence.

In our patient, the CSF venous fistula was located at the C6–7 level, which considerably increased the surgical risk with respect to ligation of the C7 nerve root and injury of the vertebral artery. Suboccipital decompression was performed to arrest the rapid deterioration, and the symptoms resolved dramatically after the operation. We believe that the clinical symptoms are not merely related to the intracranial hypotension but are also associated with the altered CSF dynamics induced by tonsillar herniation.

As the brain descends gradually, the subarachnoid space and CSF flow at the foramen magnum become partially obstructed, resulting in a CSF flow jet at the foramen magnum and interruption of the transmission of systolic pulse pressure to the distal CSF, which causes a Venturi effect on the spinal cord. According to the intramedullary pulse pressure theory, the uncoupling of the pulse pressure in the CSF and cord (medullary–subarachnoid pressure dissociation) creates a pressure gradient from the center of the cord outward, causing distention of the spinal cord and the central canal. Syringomyelia occurs after repetitive mechanical distention of the spinal cord.<sup>15,16</sup> In addition, distention of the spinal cord may gradually encroach into the surrounding subarachnoid space and expel the CSF through the CSF venous fistula, thereby aggravating the brain sagging and forming a vicious circle. Suboccipital decompression disrupts the vicious circle to restore the balance by eliminating the Venturi effect on the spinal cord and diminishing the medullary–subarachnoid pressure dissociation.

Foramen magnum obstruction can also explain the Valsalva-induced headache presented by most patients. The Valsalva maneuver may induce a transient pressure dissociation between the head and spine, which is associated with impaired CSF flow that is secondary to significant foramen magnum obstruction. During the Valsalva maneuver, the initial spinal pressure elevation shifts CSF to the head. However, unlike what occurs in healthy persons, the shifted CSF cannot return to the spinal canal immediately at the end of the Valsalva maneuver due to foramen magnum obstruction, resulting in increased intracranial pressure. The theory was first proposed in Chiari malformation patients<sup>17,18</sup> and then extrapolated to CSF venous fistula patients by Duvall and colleagues.<sup>7</sup> This theory

was confirmed in our patient because the Valsalva-induced severe headache resolved completely after suboccipital decompression.

Nevertheless, because most case reports or case series do not provide adequate information about the sequence of symptoms and images of brain sagging, we could not corroborate our findings with those of previous case reports. Further investigation should be performed in the future.

## Lessons

CSF venous fistula is an abnormal connection between the subarachnoid space and the epidural venous plexus. Patients with SIH who have no identifiable epidural CSF collection should be investigated for CSF venous fistula, and the DSM in the lateral decubitus position is thought to be the best diagnostic tool currently. Established treatments for CSF venous fistulas include an epidural blood patch, fibrin glue injection, and surgical ligation of the fistula. Epidural blood patches and fibrin glue injection are not as effective as surgical ligation of the CSF venous fistula along with nerve root ligation. As observed in our patient, the pathophysiology of the clinical symptoms of CSF venous fistulas may be due not to intracranial hypotension alone but also to the altered CSF dynamics induced by tonsillar herniation. Therefore, in addition to the direct treatment of the CSF venous fistula, suboccipital decompression should also be considered, especially in patients with Valsalva-induced headache, who have a poor response to surgical ligation or in whom surgical ligation is not feasible and in those with foramen magnum obstruction.

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## Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

## Author Contributions

Conception and design: all authors. Acquisition of data: Hsieh. Analysis and interpretation of data: Hsieh, Lai. Drafting the article: Huang, Hsieh. Critically revising the article: Huang, Hsieh, Lai. Reviewed submitted version of manuscript: Hsieh, Kuo. Approved the final version of the manuscript on behalf of all authors: Huang. Administrative/technical/material support: Lai. Study supervision: Huang.

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