NICeR
Journal of Neurointerventional Case Reports

Syrinx Resolution Following Transvenous Embolization of a Cerebrospinal Fluid–Venous Fistula

CASE REPORT

Nitesh P. Patel [1], Cem Bilgin [2], Ivan Garza [3], and Waleed Brinjikji [2]

[1]Department of Neurosurgery, Mayo Clinic, Rochester, MN, USA
[2]Department of Neurology, Mayo Clinic, Rochester, MN, USA
[3]Department of Radiology, Mayo Clinic, Rochester, MN, USA

CORRESPONDING AUTHOR:
Waleed Brinjikji
Department of Radiology, Mayo Clinic, 200 1st Street SW, Rochester, MN, 55905, E-mail:
Brinjikji.Waleed@mayo.edu

Received: 25 August 2024; Accepted: 26 August 2024; Published Online: 10 October 2024

TO CITE THIS ARTICLE:
Patel NP, Bilgin C, Garza I, Brinjikji W. Syrinx Resolution Following Transvenous Embolization of a Cerebrospinal Fluid–Venous Fistula. Journal of Neurointerventional Case Reports. 2024;1(1):2, 1-3. DOI: https://doi.org/10.70355/NICeR.2

ABSTRACT

Cerebrospinal fluid (CSF)–venous fistulas are a common cause of spontaneous intracranial hypotension (SIH). Transvenous embolization of CSF–venous fistulas has gained increasing recognition among neurointerventionalists as a minimally invasive treatment, with early studies indicating significant symptom improvement or resolution. However, the impact of this treatment on CSF dynamics remains largely unexplored. Herein, we present a case of a 45-year-old female whose syrinx cavity resolved completely following this treatment.

KEYWORDS

spontaneous intracranial hypotension, SIH, transvenous embolization

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is a debilitating condition that can present with a diverse array of symptoms, including orthostatic and cough/Valsalva headaches, tinnitus, hearing loss, visual disturbances, and cognitive impairments.[1] One of the predominant causes of SIH is cerebrospinal fluid (CSF)–venous fistulas, which represent an anomalous connection between the thecal sac or nerve root sleeve and the adjacent foraminal or epidural veins.[1] Transvenous embolization of CSF–venous fistulas has emerged as a promising minimally invasive treatment for these patients. Preliminary studies suggest that this approach yields high rates of symptom improvement or resolution.

CSF–venous fistulas may present with Chiari-like cerebellar tonsillar ectopia due to brain sag. However, the associated syringomyelia is uncommon, with only a few cases reported in the literature.[2] In this case report, we describe a 45-year-old female patient whose syrinx cavity was completely resolved following transvenous embolization.

CASE PRESENTATION

A 45-year-old female presented with a 10-month history of right hand diminished dexterity and paresthesias, 7 months of non-positional Valsalva-induced headaches radiating to the posterior neck, and 2 months of right facial tenderness. Her neurological examination was notable for decreased pinpricked sensation in a C2 distribution, hyperpathia in the right V2 division, and diffuse hyperreflexia without spasticity.

Magnetic resonance imaging (MRI) of the brain (Figure 1A) showed findings consistent with SIH, including cerebellar tonsillar ectopia, pituitary enlargement, and engorgement of the dural venous sinuses. MRI of the cervical spine showed a cervicothoracic syrinx (Figure 1B). MRI of the thoracic and lumbar spine were negative for CSF leak. Right lateral decubitus computed tomography (CT) myelogram showed a right T5 CSF–venous fistula (Figure 2). Left lateral decubitus myelography was negative. She subsequently underwent transvenous embolization of her right T5 CSF–venous fistula.

At 1-year follow-up, she reported complete resolution of her Valsalva-induced headaches and neck pain; however, she reported persistent right facial pain and right-hand paresthesias. MRI of the brain showed complete resolution of her SIH findings (Figure 1C), and MRI of the cervical spine demonstrated dramatic near-complete resolution of her syrinx (Figure 1D).

DISCUSSION

Most commonly, SIH results from one of three types of spinal CSF leaks: a linear dural tear either ventral or posterolateral to the spinal cord, leakage from a meningeal diverticulum, or a spinal CSF–venous fistula. Although orthostatic headache is the classic symptom, the condition can manifest with a broad range of symptoms, including posterior neck pain, nausea, vomiting, hearing loss, tinnitus, gait difficulty, cognitive changes, and personality changes. Typical findings on brain MRI include pachymeningeal enhancement, venous engorgement, pituitary enlargement, brain sagging, and subdural fluid collections.[1]

Chiari-like cerebellar tonsillar ectopia can be seen in SIH as a result of brain sag; however, the associated syringomyelia is rare, and only a few

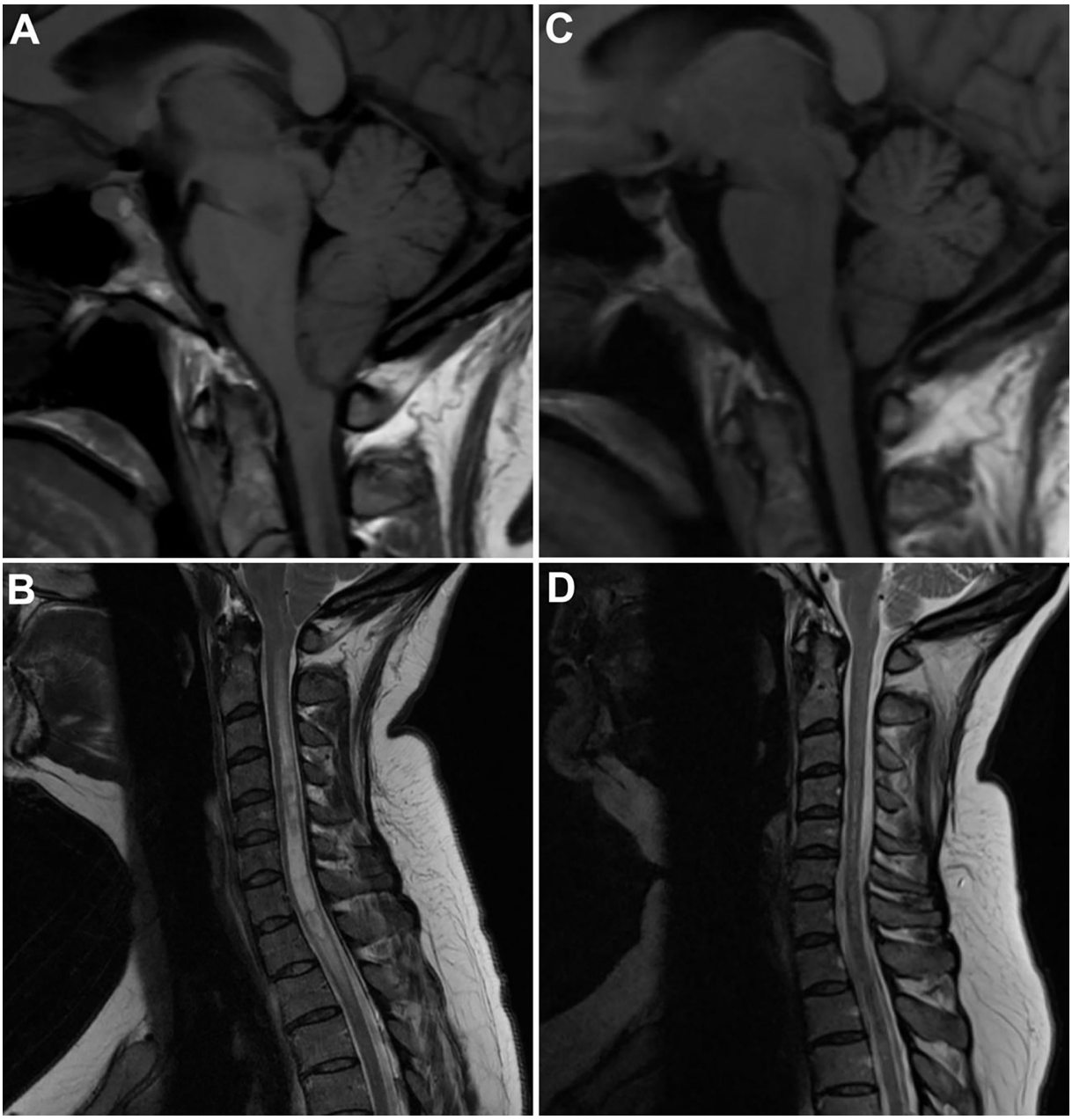

FIGURE 1. Initial imaging findings of spontaneous intracranial hypotension and cervical syringomyelia. (A) T1-weighted sagittal brain magnetic resonance imaging (MRI) showing cerebellar tonsillar ectopia through the foramen magnum, resulting in the compression of the surrounding cervicomedullary space. There is also enlargement of the pituitary gland and engorgement of the straight sinus. (B) T2-weighted sagittal cervical spine MRI showing an expansile, loculated syrinx in the cervicothoracic region. (C) T1-weighted sagittal brain MRI demonstrating complete resolution of the tonsillar ectopia, brainstem compression, pituitary enlargement, and venous engorgement. (D) T2-weighted sagittal cervical spine MRI demonstrating near-complete resolution of the syrinx.

cases have been reported.[2] While these findings in our patient could have been mistaken as a Chiari malformation, the associated pituitary enlargement and venous engorgement raised suspicion for SIH, and further diagnostic evaluation ultimately led to the diagnosis of a CSF–venous fistula, an abnormal connection between a nerve root sleeve and adjacent paraspinal veins.

Treatment options for CSF–venous fistula treatment include epidural blood patching, microsurgical ligation nerve root ligation, and transvenous embolization.[1] Our patient underwent transvenous embolization of her right T5 CSF–venous fistula in which the epidural venous plexus, paraspinal vein, and intercostal vein at the level of the fistula were embolized with Onyx liquid embolic agent to completely seal drainage of CSF into the venous system. This novel endovascular procedure has shown encouraging clinical results and is emerging as an important treatment option.[3,4]

To our knowledge, this is the first reported case of syrinx resolution following transvenous embolization of a CSF–venous fistula and demonstrates the importance of considering SIH as

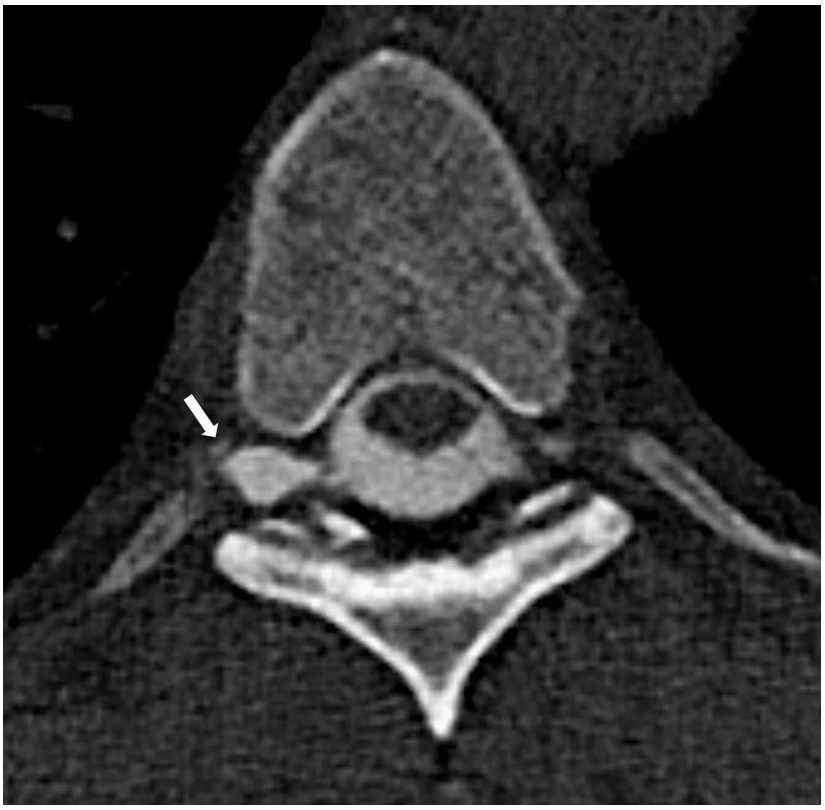

FIGURE 2. Evidence of cerebrospinal fluid (CSF)–venous fistula on computed tomography (CT) myelogram. Venous opacification (white arrow) along the right T5 nerve root sleeve diverticulum on the right lateral decubitus CT myelogram indicated the presence of a right T5 CSF–venous fistula.

a potential cause of a syrinx associated with tonsillar ectopia, especially when other findings of SIH are apparent on brain MRI. Accurate diagnosis of SIH, instead of Chiari malformation, is imperative in avoiding an unnecessary suboccipital decompressive surgery.[2]

Patient consent

Written consent for the publication of patient images and associated medical information was obtained using the institutional form.

Contributors

All authors contributed to drafting the manuscript and approved the final version for submission.

Funding

None

Acknowledgements

None

REFERENCES

1. Schievink WI. Spontaneous intracranial hypotension. The New England Journal of Medicine. 2 Dec 2021; 385(23): 2173–2178. DOI: https://doi.org/10.1056/NEJMra2101561.
2. Middlebrooks EH, Okromelidze L, Vilanilam GK, et al. Syrinx secondary to Chiari-like Tonsillar herniation in spontaneous intracranial hypotension. World Neurosurgery. Nov 2020; 143: e268–e274. DOI: https://doi.org/10.1016/j.wneu.2020.07.108.
3. Brinjikji W, Garza I, Whealy M, et al. Clinical and imaging outcomes of cerebrospinal fluid-venous fistula embolization. Journal of Neurointerventional Surgery. Oct 2022; 14(10): 953–956. DOI: https://doi.org/10.1136/neurintsurg-2021-018466.
4. Brinjikji W, Savastano LE, Atkinson JLD, et al. A novel endovascular therapy for CSF hypotension secondary to CSF-venous fistulas. AJNR American Journal of Neuroradiology. May 2021; 42(5): 882–887. DOI: https://doi.org/10.3174/ajnr.A7014.