

Spontaneous syrinx resolution in patient with Chiari I malformation: illustrative case

Elizabeth Gallo, PA-C,¹ Gazanfar Rahmathulla, MD,¹ Dinesh Rao, MD,² Kourosh Tavanaiepour, DO,¹ and Daryoush Tavanaiepour, MD¹

¹Department of Neurosurgery, and ²Division of Neuroradiology, University of Florida, College of Medicine, Jacksonville, Florida

BACKGROUND Chiari malformations include a spectrum of congenital hindbrain herniation syndromes. In patients with the most common subtype, Chiari malformation Type I, 50% to 75% develop a syrinx. The pathogenesis of syringomyelia is not well understood, with multiple theories outlined in the literature. Although the presence of a syrinx in a patient with Chiari malformation is generally accepted as an indication for surgical intervention, there are documented cases of spontaneous resolution that support a more conservative approach to management.

OBSERVATIONS The authors reported a case of spontaneous resolution of a cervical syrinx in an adult with an unchanged Chiari malformation.

LESSONS Given the possibility of spontaneous resolution over time, the authors believe a more conservative approach of observation with periodic surveillance, magnetic resonance imaging, and neurological examination should be considered in the management of a patient with a Chiari malformation and associated syringomyelia.

<https://thejns.org/doi/abs/10.3171/CASE21236>

KEYWORDS Chiari malformation; syrinx; syringomyelia; spontaneous resolution

Chiari malformations include a spectrum of congenital hindbrain herniation syndromes that are characterized by the degree of herniation of the cerebellar tonsils through the foramen magnum. The most common subtype, Chiari I malformation, is radiographically defined as displacement of the cerebellar tonsils 5 mm or more below McRae line.¹ In patients with this condition, approximately 50% to 75% develop a syringomyelia.² The development of syringomyelia is not well understood, and multiple theories are outlined in the literature. Although the presence of a syrinx in a patient with Chiari malformation is generally accepted as an indication for surgical intervention with the goal of restoring normal cerebrospinal fluid (CSF) flow,³ documented cases of spontaneous resolution exist to support consideration toward a more conservative approach to management. We report a new case of spontaneous resolution of syringomyelia associated in an adult with an unchanged Chiari malformation.

Illustrative Case

A 46-year-old woman presented to our clinic in February 2019 with a known history of a Chiari malformation. Her condition was diagnosed in 2003 after a motor vehicle accident. She reported being advised of

a syrinx and required evaluation by a neurosurgeon. The patient was offered surgical intervention but declined it at that time. When she presented to our clinic, she had recently been involved in another car accident, which left her experiencing posterior headaches. On examination, the patient did not have any neurological deficits. Previous cervical spine magnetic resonance imaging (MRI), completed in 2009, revealed a syrinx extending from C2 to T1 level with cerebellar tonsillar ectopia (Fig. 1). Follow-up MRI in 2019 demonstrated significant decrease in size of the syrinx extending from C2 to C3 (Fig. 2).

Discussion

Observations

The association between Chiari I malformation and syringomyelia is well documented, but the pathogenesis of syringomyelia remains controversial. Several theories have been presented in the literature over the years to explain the formation of a syrinx. Most early theories referred to how CSF entered the spinal cord from the fourth ventricle via the central canal.⁴ Craniospinal pressure dissociation was

ABBREVIATIONS CSF = cerebrospinal fluid; MRI = magnetic resonance imaging.

INCLUDE WHEN CITING Published June 28, 2021; DOI: 10.3171/CASE21236.

SUBMITTED April 21, 2021. **ACCEPTED** May 5, 2021.

© 2021 The authors, CC BY-NC-ND 4.0 (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



FIG. 1. Sagittal T2-weighted MRI demonstrating a Chiari I malformation and associated syringomyelia in the cervical cord.

proposed by Williams and the theory of CSF pulsations, or the “hydrodynamic theory,” was postulated by Gardner.⁴⁻⁷

Other theories suggested CSF entrance from the subarachnoid space via perivascular spaces.⁴ Ball and Dyan proposed that under the pressure from subarachnoid obstruction, CSF would pass into the



FIG. 2. Sagittal T2-weighted MRI obtained 9 years after the initial scan redemonstrating the Chiari I malformation but with complete resolution of the cervical syringomyelia.

spinal cord by way of Virchow-Robin spaces.^{4,8,9} A blockage of the upward flow of CSF at the upper end of the central canal has been suggested to cause CSF to enter the spinal cord via perivascular spaces as well.¹⁰ It has also been hypothesized that when there is obstruction of CSF at the foramen magnum, the cerebellar tonsils are forced to act as a “piston” on the partially isolated spinal CSF space, imparting a systolic pressure wave on the surface of the spinal cord. As a result, CSF is believed to be forced from the subarachnoid space and into the spinal cord through the perivascular or interstitial spaces.^{9,11} Recent studies have suggested the syrinx fluid is extracellular from the spinal cord microcirculation, not from the CSF in the fourth ventricle or subarachnoid space.⁴ One proposed mechanism describes mechanical distension of the spinal cord, which fills with extracellular fluid.¹² Another proposed mechanism suggests that the dilatation of intramedullary vessels below the subarachnoid block at the foramen magnum partially disrupts the blood–spinal cord barrier, which creates the syrinx with the accumulation of extracellular fluid from the intramedullary microcirculation.^{4,13} Based on the varying hypotheses in the literature, it is evident that altered CSF dynamics play a role in the development of syringomyelia.

For patients with Chiari malformation with syringomyelia, it is generally recommended that they receive surgical intervention to prevent progression and worsening neurological problems. The standard surgical procedure for Chiari I malformation is a posterior fossa decompression with the goal of restoring normal CSF flow across the craniocervical junction.^{3,14,15} Within the procedure of a suboccipital decompression, the amount of bony removal can vary to include a C1 laminectomy. Treatment of the dura itself varies because it can be left intact, scored, or opened. When dura is opened, the arachnoid may be left intact or lysis of arachnoid adhesions may be performed with an intradural dissection.^{2,14,16,17}

Lessons

Although surgical intervention is the widely accepted treatment modality, documented cases of spontaneous resolution of a syrinx associated with a Chiari malformation make the current treatment strategy controversial.¹⁸⁻²⁶ Some authors have demonstrated improvement or resolution of the Chiari I malformation itself, which leads to syringomyelia improvement.^{20,21,23} To our knowledge, few documented cases exist of patients with spontaneous resolution of a syrinx associated with an unchanged Chiari malformation.^{18,19,22,25,26}

One explanation for spontaneous resolution in adults with an unchanged Chiari malformation involves the formation of a communication between the syringomyelia and the subarachnoid space through a tear caused by increased pressure from Valsalva maneuvers.^{19,21,25,27} These types of communications have been demonstrated by MRI and neuropathological examinations.^{22,28} Another explanation in the literature describes spontaneous rupture of arachnoid scarring or thickening at the foramen magnum or foramen of Magendie, which improves the flow of CSF at the foramen magnum and leads to resolution of the syrinx.^{17,20,27} In our case, it could be speculated that our patient’s syringomyelia resolved through either of these mechanisms as a result of the recent motor vehicle accident impact.

Given the possibility of spontaneous resolution over time, close observation with periodic surveillance MRI and neurological examination should be considered in the management of a patient with a Chiari malformation and associated syringomyelia. The addition of our case report to previous descriptions of spontaneous syrinx resolution further supports the concept of a more conservative approach to these patients.

References

1. Mukherjee S, Kalra N, Warren D, et al. Chiari I malformation and altered cerebrospinal fluid dynamics—the highs and the lows. *Childs Nerv Syst.* 2019;35(10):1711–1717.
2. Hersh DS, Groves ML, Boop FA. Management of Chiari malformations: opinions from different centers—a review. *Childs Nerv Syst.* 2019;35(10):1869–1873.
3. De Vlieger J, Dejaegher J, Van Calenbergh F. Posterior fossa decompression for Chiari malformation type I: clinical and radiological presentation, outcome and complications in a retrospective series of 105 procedures. *Acta Neurol Belg.* 2019;119(2):245–252.
4. Koyanagi I, Houkin K. Pathogenesis of syringomyelia associated with Chiari type 1 malformation: review of evidences and proposal of a new hypothesis. *Neurosurg Rev.* 2010;33(3):271–285.
5. Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocoele. *J Neurol Neurosurg Psychiatry.* 1965;28(3):247–259.
6. Gardner WJ, Angel J. The mechanism of syringomyelia and its surgical correction. *Clin Neurosurg.* 1958;6:131–140.
7. Williams B. On the pathogenesis of syringomyelia: a review. *J R Soc Med.* 1980;73(11):798–806.
8. Ball MJ, Dayan AD. Pathogenesis of syringomyelia. *Lancet.* 1972;2(7781):799–801.
9. Meadows J, Kraut M, Guarnieri M, et al. Asymptomatic Chiari Type I malformations identified on magnetic resonance imaging. *J Neurosurg.* 2000;92(6):920–926.
10. Milhorat TH, Miller JI, Johnson WD, et al. Anatomical basis of syringomyelia occurring with hindbrain lesions. *Neurosurgery.* 1993;32(5):748–754.
11. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. *J Neurosurg.* 1994;80(1):3–15.
12. Greitz D. Unraveling the riddle of syringomyelia. *Neurosurg Rev.* 2006;29(4):251–264.
13. Levine DN. The pathogenesis of syringomyelia associated with lesions at the foramen magnum: a critical review of existing theories and proposal of a new hypothesis. *J Neurol Sci.* 2004;220(1-2):3–21.
14. Batzdorf U, McArthur DL, Bentson JR. Surgical treatment of Chiari malformation with and without syringomyelia: experience with 177 adult patients. *J Neurosurg.* 2013;118(2):232–242.
15. Schijman E, Steinbok P. International survey on the management of Chiari I malformation and syringomyelia. *Childs Nerv Syst.* 2004;20(5):341–348.
16. Durham SR, Fjeld-Olenec K. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. *J Neurosurg Pediatr.* 2008;2(1):42–49.
17. Klekamp J, Batzdorf U, Samii M, Bothe HW. The surgical treatment of Chiari I malformation. *Acta Neurochir (Wien).* 1996;138(7):788–801.
18. Deniz FE, Oksüz E. Spontaneous syringomyelia resolution at an adult Chiari Type 1 malformation. *Turk Neurosurg.* 2009;19(1):96–98.
19. Jack CR Jr, Kokmen E, Onofrio BM. Spontaneous decompression of syringomyelia: magnetic resonance imaging findings. Case report. *J Neurosurg.* 1991;74(2):283–286.
20. Klekamp J, Iaconetta G, Samii M. Spontaneous resolution of Chiari I malformation and syringomyelia: case report and review of the literature. *Neurosurgery.* 2001;48(3):664–667.
21. Kyoshima K, Bogdanov EI. Spontaneous resolution of syringomyelia: report of two cases and review of the literature. *Neurosurgery.* 2003;53(3):762–769.
22. Santoro A, Delfini R, Innocenzi G, et al. Spontaneous drainage of syringomyelia. Report of two cases. *J Neurosurg.* 1993;79(1):132–134.
23. Sun JC, Steinbok P, Cochrane DD. Spontaneous resolution and recurrence of a Chiari I malformation and associated syringomyelia. Case report. *J Neurosurg.* 2000;92(suppl 2):207–210.
24. Sung W-S, Chen Y-Y, Dubey A, Hunn A. Spontaneous regression of syringomyelia: review of the current aetiological theories and implications for surgery. *J Clin Neurosci.* 2008;15(10):1185–1188.
25. Vaquero J, Ferreira E, Parajón A. Spontaneous resolution of syrinx: report of two cases in adults with Chiari malformation. *Neurol Sci.* 2012;33(2):339–341.
26. Yuan C, Yao Q, Zhang C, Jian F. Spontaneous resolution of syringomyelia with a 16-year serial magnetic resonance imaging follow-up: a case report and literature review. *World Neurosurg.* 2019;130:432–438.
27. Ozisik PA, Hazer B, Ziyal IM, Ozcan OE. Spontaneous resolution of syringomyelia without Chiari malformation. *Neurol Med Chir (Tokyo).* 2006;46(10):512–517.
28. Milhorat TH, Capocelli AL Jr, Anzil AP, et al. Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. *J Neurosurg.* 1995;82(5):802–812.

Disclosures

Dr. K. Tavanaiepour reported personal fees from Astura Medical outside the submitted work.

Author Contributions

Conception and design: D Tavanaiepour, Rao, K Tavanaiepour. Acquisition of data: D Tavanaiepour, Rao. Analysis and interpretation of data: D Tavanaiepour, Rahmathulla. Drafting the article: D Tavanaiepour, Gallo, Rao. Critically revising the article: D Tavanaiepour, Gallo, Rahmathulla, Rao. Reviewed submitted version of manuscript: D Tavanaiepour, Gallo, Rahmathulla. Approved the final version of the manuscript on behalf of all authors: D Tavanaiepour. Study supervision: D Tavanaiepour.

Correspondence

Daryoush Tavanaiepour: University of Florida, College of Medicine, Jacksonville, FL. daryoush.tavanaiepour@jax.ufl.edu.