Case report

Arnold-Chiari 1 malformation type 1 with syringohydromyelia presenting as acute tetraparesis: A case report

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Context: A 19-year-old woman who presented to a community hospital after awakening with tetraparesis, generalized paresthesia, and severe neck pain, and was transferred to an acute care hospital.

Findings: Magnetic resonance imaging of the head and spine was performed and revealed a cystic lesion extending from the C1 level to the C6 level as well as an Arnold-Chiari type 1 malformation. Emergent surgical posterior fossa decompression with duraplasty and C1 laminectomy was undertaken. Most symptoms improved immediately postoperatively. On post-operative day 15, the patient was transferred to our acute rehabilitation hospital for an additional 16 days. With continued aggressive therapy, she demonstrated complete resolution of tetraparesis as well as significant improvement in muscle strength and function in addition to resolution of paresthesia and neck pain. Functional independence measure scores were 69/126 on admission to 110/126 on discharge from the rehabilitation hospital. Her tetraparesis eventually resolved; manual muscle testing scores on follow-up 2 months later were 5/5 in all four extremities.

Clinical relevance: This is the first reported case of Chiari I malformation with syringohydromyelia presenting as acute tetraparesis, generalized paresthesia, and neck pain. Surgical decompression leading to resolution of symptoms made other etiologies extremely unlikely and there was no history of trauma. The different theories on the pathogenesis of syringomyelia are discussed.

Keywords: Tetraparesis, Syringomyelia, Arnold-Chiari malformation, Paresthesia, Neck pain, Rehabilitation, Physical

Introduction

Syringomyelia is a pathologic cystic cavity within the spinal cord containing cerebrospinal fluid (CSF). It is most commonly seen as a complication of an Arnold-Chiari type 1 malformation, which is the caudal herniation of the cerebellar tonsils through the foramen magnum and into the cervical spinal canal.¹ Syringomyelia can also occur as complication of hemorrhage, tumor, meningitis, arachnoiditis, or trauma.² Symptoms most often begin to appear in early and middle life.^{1,3} These symptoms usually consist of pain and weakness that present and progress gradually.²

Case report

A 19-year-old woman with a history of pulmonary embolism and recurrent deep vein thrombosis due to antiphospholipid syndrome with poor compliance on

warfarin therapy presented to an acute care hospital after awakening with tetraparesis, generalized paresthesia, and severe neck pain.

The night before the patient began to feel heaviness and numbness in her lower extremities before going to sleep. The following morning patient awoke with tetraparesis, generalized paresthesias, and neck pain and was taken to her local hospital. The patient was then transferred to our tertiary care hospital for further management. On presentation, initial manual muscle examination revealed 2/5 muscle strength in the upper extremities bilaterally and 0/5 muscle strength in the lower extremities. Further examination demonstrated decreased sensation to light touch at and below the level of T1, 1+ reflexes throughout with the exception of no reflexes at the ankles bilaterally. Cranial nerves II–XII were grossly intact except for CN XI; there was decreased strength with shoulder shrug. The patient also demonstrated ptosis and mydriasis. There was no

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urinary retention on admission. Rectal examination was not performed due to patient refusal. Admission laboratory tests were significant only for a C-reactive protein of 11.7 mg/dl and white blood cell count of 19.9 k/ μ l. Computerized tomography (CT) of the head without contrast was unremarkable; specifically, no hydrocephalus was present (Fig. 1). Magnetic resonance imaging (MRI) of the head and spine with and without contrast revealed a cystic lesion involving the cervical spinal cord from the C1 level extending to the C6 level as well as an Arnold-Chiari type 1 malformation (Fig. 2). Due to the acute onset of symptoms, emergent surgical intervention was felt to be required and the patient underwent posterior fossa decompression with duraplasty and C1 laminectomy. The procedure did not reveal any evidence of intra-syringal hemorrhage or arachnoiditis. Immediately postoperatively, the patient demonstrated 3/5 strength in the bilateral upper extremities and 1/5 strength in the lower extremities bilaterally. Postoperative CT demonstrated successful decompression (Fig. 3).

Three weeks before this she had been evaluated for severe occipital headache that was worse with sitting and associated with nausea and vomiting. She was evaluated at an outside hospital and her workup included a lumbar puncture as part of a meningitis workup. She was placed on intravenous antibiotics because initial CSF white blood cell count was elevated (327 cells/ μ l, 92% neutrophils). CSF cultures did not demonstrate any growth after 72 hours. She was discharged at that time on oral antibiotics for a concomitant right otitis media. During her admission she also received a CT

head without contrast that demonstrated normal appearing anatomy with the exception of moderate sinusitis. The day prior to presenting to our acute care hospital, she presented to the same local community hospital with complaints of unresolved occipital headache, at which point she refused another meningitis workup and was discharged after symptomatic management.

The patient's hospital course at our acute care hospital was complicated by a thrombus in the right basilic vein for which a vena cava filter was placed. The patient was placed on enoxaparin injections and bridged to warfarin. Further complications included urinary tract infection as well as mild hemoptysis which resolved with temporary discontinuation of anticoagulation. The patient also experienced urinary retention and blurry vision due to exposure keratopathy.

After an acute inpatient hospital stay of 15 days, she was transferred to our acute rehabilitation facility secondary to continued muscle weakness, poor balance, neurogenic bowel and bladder, and generalized functional decline. At that time, manual muscle testing revealed 5/5 strength in the right upper extremity, 4/5 strength in the left upper extremity, 3/5 strength in the hip flexors bilaterally, 4/5 strength in the rest of the right lower extremity, and 5/5 strength in the rest of the left lower extremity. Once again, she declined rectal examination. Functionally, the patient improved significantly during her 16-day rehabilitation stay as

Figure 1 Head CT (axial view, without contrast) at time of presentation was unremarkable.

Figure 2 MRI of the head and spine (T2 weighted sagittal view) at time of presentation revealed a cystic lesion involving the cervical spinal cord from the C1 level extending to the C6 level as well as an Arnold-Chiari type 1 malformation.

Figure 3 Postoperative Head CT (axial view, without contrast) demonstrated successful decompression.

evidenced by her total functional independence measure score increasing from 69/126 to 110/126.

The patient was seen for follow-up 2 months after discharge and her tetraparesis had resolved completely with manual muscle testing revealing 5/5 strength throughout. She was ambulating independently and was independent to modified independent for all activities of daily living. Since this initial follow-up visit, the patient has not maintained her follow up visits. While this is not optimal and does not allow for physical examination, the patient was contacted via telephone 23 months postoperatively and at that time was doing well without further symptoms or new concerns; this is highly suggestive that the syrinx has not recurred, though without imaging this cannot be confirmed.

Discussion

The unusual feature of this case is the acute onset of tetraparesis as a result of syringohydromyelia. Differential diagnosis of acute tetraparesis included traumatic spinal cord injury, multiple sclerosis, transverse myelitis, neoplasm, anterior cord infarction, vitamin B₁₂ deficiency, and infection (bacterial, viral, fungal, parasitic). There was no history or evidence of trauma. Multiple imaging studies did not demonstrate any evidence of neoplasm, cord infarction, or plaques within the brain or cervical spine. The other possibilities were also ruled out by the immediate improvement of symptoms after surgical decompression.

The initial CT most likely did not reveal the syringomyelia because of the surrounding bone causing distortion.⁴ CT myelography is better at revealing syrinx formation, but still misses 10–50% of syrinxes and as

such, MRI is the best diagnostic imaging tool with respect to syringomyelia.^{5,6}

We could not account for the CSF pleocytosis that was found three weeks prior to admission. There are no reports of syringomyelia or Chiari malformation causing CSF pleocytosis independently. One possibility was that initially she was suffering from bacterial meningitis despite there being no growth on culture and that she subsequently improved with treatment, even though it was not of duration that is considered standard of care. A viral etiology is not likely considering the predominance of neutrophils demonstrated in her CSF studies. Another possibility, considering the abnormal presentation of the syringomyelia, is that the pleocytosis may be another unique element in this case. We are confident that symptoms of tetraparesis and generalized paresthesia were not due to an infectious or rheumatologic process considering the immediate improvement of symptoms after decompression in addition to the fact that her symptoms did not return.

Chiari type 1 malformation is the caudal herniation of the cerebellar tonsils through the foramen magnum that exceeds 5 mm. This malformation is typically associated with syringomyelia, with 40–75% of type I malformations having a concomitant syringomyelia, as seen in this patient.⁷ Acute paraparesis has been described as a consequence of Arnold-Chiari type 1 malformation, but only in the presence of acute neck hyper-flexion injuries.⁸ Notably, this patient did not report any history of trauma or neck injury.

Many different presentations of syringomyelia have been described. Most often, progressive weakness in the arms and legs is seen, in addition to stiffness in the back, shoulders, arms, or legs. Chronic and sometimes

severe pain is also seen.² Rare reported presentations of syringomyelia include progressive vocal fold paralysis⁹ and acute respiratory failure.¹⁰ One case report describes syringomyelia that presented as paraparesis coupled with urinary incontinence that slowly progressed over a 3-month period and was associated with an intramedullary lesion from T10 to the sacral spinal canal.¹¹ The lesion was found to be a syringohydromyelia intraoperatively.¹¹ While the location of the syringomyelia in the aforementioned case accounts for the patient only experiencing symptoms in his lower extremities, this current case is still unique in the acute onset of paresis. Paraparesis and even tetraparesis in the setting of a cervical syrinx have also been reported in the setting of a malfunctioning CSF shunts.^{12–14} This was not a possible explanation in our case though, given that our patient did not have a CSF shunt. Gower's intrasyringal hemorrhage associated with hematomyelia linked to syringomyelia has been described to cause acute muscle weakness.¹⁵ However, there was no evidence of such hemorrhage in this case. Other cases of acute paraparesis associated with hematomyelia caused by spinal cord arteriovenous malformation have also been reported.¹⁶ Once again, it is worth noting that our patient had neither hematomyelia nor a spinal cord arteriovenous malformation. Certainly, there have been other reports of paresis associated with syringohydromyelia, but we believe this to be the first reported case of syringomyelia associated with tetraparesis that presented in such an acute manner in the absence of other pathology or risk factors, specifically in the absence of a spinal cord arteriovenous malformation or a malfunctioning CSF shunt.

We also considered the possibility that the previous lumbar puncture 3 weeks prior to presenting to our hospital with acute tetraparesis was the inciting factor for syrinx or Chiari formation. However, there are no reported cases of lumbar puncture causing syringomyelia and only one case report of an acquired Chiari 1 malformation, without cervical syrinx, which occurred after multiple traumatic lumbar punctures.¹⁷ Considering that this patient only received a single lumbar puncture and that she demonstrated a syrinx in addition to the Chiari malformation, it seems unlikely that the lumbar puncture caused her Chiari malformation with syringohydromyelia. The possibility also exists that the previous lumbar puncture exacerbated an already present Chiari malformation that was not seen on the initial CT 3 weeks prior. Chiari malformation usually goes undetected on CT examination if only axial cuts are performed.¹⁸ Nonetheless, even if it was visualized, possible meningitis is a strong justification for lumbar

puncture even when a Chiari malformation is known and as such her management would likely not have differed. Moreover, tetraparesis did not develop until 3 weeks after lumbar puncture and developed acutely, not chronically, which makes this scenario less likely. We could not identify any literature reports of syrinx formation in the setting of single uncomplicated lumbar puncture in a patient with Chiari malformation.

Many theories exist that attempt to explain the pathogenesis of syringomyelia. Some theories suggest that CSF from the fourth ventricle creates a syrinx in the spinal cord due to either arterial pulsation or a pressure dissociation between the subarachnoid spaces of the cranium and spinal cord.^{19–23} Other theories suggest that CSF coming directly from the subarachnoid space creates a syrinx as a direct result of the anatomical defect of the cerebellar tonsils.^{24–26} Still, other theories propose that the fluid in the syrinx originates from accumulated extracellular fluid.^{27–30} Another theory suggests that decreased venous compliance within the subarachnoid space disrupts CSF flow.³¹ None of these theories can fully explain or predict the unusual presentation of acute tetraparesis as evidenced in this case.

Conclusion

We present a unique case of syringomyelia and associated Chiari I malformation presenting as acute tetraparesis. Treatment in the acute setting remains surgical decompression, followed by acute inpatient rehabilitation as with any other patient with tetraparesis. Recovery with significant functional improvement and resolution of tetraparesis was demonstrated.

References

- Banerji NK, Millar JHD. Chiari malformation presenting in adult life. Its relationship with syringomyelia. *Brain* 1974;97(1):157–68.
- Syringomyelia Fact Sheet. Bethesda, MD: National Institute of Neurological Disorders and Stroke; 2011. NIH Publication No. 10–3780.
- Van Gijn J, Giesbergen PCLM, Hoogengraad TU. Attacks of pain in the leg from classic syringomyelia. *J Neurol Neurosurg Psychiatry* 1997;63(3):410.
- Klekamp J, Samii M. Syringomyelia diagnosis and treatment. Chapter 4, Syringomyelia associated with diseases of the spinal canal; pp. 111–93. Heidelberg: Springer-Verlag Berlin; 2002.
- Aubin ML, Vignaud J, Jardin C, Bar D. Computed tomography in 75 clinical cases of syringomyelia. *Am J Neuroradiol* 1981;2(3):199–204.
- Broadbelt AR, Stoodley MA. Post-traumatic syringomyelia: a review. *J Clin Neurosci* 2003;10(4):401–8.
- Fernandez A, et al. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). *BMC Musculoskeletal Disorders* [Internet]. 2009 December 17 [cited 2010 August 29]; 10 (S1). Available from: <http://www.biomedcentral.com/1471-2474/10/S1/S1>.
- Vicek BW, Ito B. Acute paraparesis secondary to Arnold-Chiari type I malformation and neck hyperflexion. *Ann Neurol* 1987; 21(1):100–1.
- Viswanatha B. Syringomyelia with syringobulbia presenting as vocal fold paralysis. *Ear Nose Throat J* 2009;88(7):E20.

- 10 Al Bashapshe A, Bhatia H, Aziz S. Acute respiratory failure as a first manifestation of syringomyelia. *Lung India* 2010;27(2):93–5.
- 11 Abuzayed B, Dashti R, Erdinciler P, Ozlen F, Kafadar AM, Ciplak N. Terminal syringomyelia mimicking an intramedullary tumor: case report. *Turk Neurosurg* 2010;20(2):247–50.
- 12 Martinez-Lage JF, Alarcon F, Lopez-Guerrero AL, Murcia MF, Vilar ARE, Almagro MJ. Syringomyelia with quadriparesis in CSF malfunction: a case illustration. *Childs Nerv Syst* 2010;26(9):1229–31.
- 13 Beswick E, St George EJ, Solanki GA. Paraparesis secondary to expansion of cervical syrinx: an unusual presentation of ventriculo-peritoneal shunt malfunction. *Periatr Neurosurg* 2005;41(6):334–6.
- 14 Milhorat TH, Bolognese PA, Black KS, Woldenberg RF. Acute syringomyelia: case report. *Neurosurgery* 2003;53(5):1220–2.
- 15 Hamlat A, Adn M, Yahia MB, Morandi X, Brassier G, Guegan Y. Gowers intrasyringal hemorrhage case report and review of the literature. *J Neurosurg Spine* 2005;3(6):477–81.
- 16 Srivatanakul K, Songsaeng D, Ozanne A, Toulgoat F, Lasjaunia P. Spinal arteriovenous malformation associated with syringomyelia. *J Neurosurg Spine* 2009;10(5):436–42.
- 17 Sathi S, Stieg PE. “Acquired” Chiari I malformation after multiple lumbar punctures: case report. *Neurosurgery* 1993;32(2):306–9.
- 18 Erbay SH, O’Callaghan MG, Bhadelia R. Is lumbar puncture contraindicated in patients with Chiari I malformation? *Am J Neuroradiol* 2005;26(4):985.
- 19 Gardner WJ, Goodall RJ. The surgical treatment of mechanism and importance of encephalography in diagnosis. *J Neurosurg* 1950;7(3):199–206.
- 20 Gardner WJ, Ange J. The mechanism of syringomyelia and its surgical correction. *Clin Neurosurg* 1958;6:131–40.
- 21 Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocoele. *J Neurol Neurosurg Psychiatry* 1965;28(3):247–59.
- 22 Williams B. The distending force in the production of “communicating syringomyelia.” *Lancet* 1969;294(7613):89–193.
- 23 Williams B. On the pathogenesis of syringomyelia: a review. *J R Soc Med* 1980;73(11):798–806.
- 24 Ball MJ, Dayan AD. Pathogenesis of syringomyelia. *Lancet* 1972;2(7781):799–801.
- 25 Milhorat TH, Miller JI, Johnson WD, Adler DE, Heger IM. Anatomical basis of syringomyelia occurring with hindbrain lesions. *Neurosurgery* 1993;32(5):748–54.
- 26 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.
- 27 Greitz D, Ericson K, Flodmark O. Pathogenesis and mechanics of spinal cysts. A new hypothesis based on magnetic resonance studies of cerebrospinal fluid dynamics. *Int J Neuroradiol* 1999;5(2):61–78.
- 28 Greitz D. Unraveling the riddle of syringomyelia. *Neurosurg Rev* 2006;29(4):251–64.
- 29 Klekamp J. The pathophysiology of syringomyelia – historical overview and current concept. *Acta Neurochir (Wien)* 2002;144(7):649–64.
- 30 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.
- 31 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):271–85.