Management of syringomyelia with Chiari malformation type 1: Case series

By Dodik Tugasworo

WORD COUNT 3580
TIME SUBMITTED 01-DEC-2024 01:17AM
PAPER ID 113249325

Management of syringomyelia with Chiari malformation type 1: Case series

Dodik Tugasworo[1], Retnaningsih[2], Yovita Andhitara[3], Virgi Mayang Maharani[4]

[1]Neurovascular Division, Department of Neurology, Dr. Kariadi General Hospital/Faculty of MedicineDiponegoro University, Semarang, Indonesia
[2]Neurointensive Division, Department of Neurology, Dr. Kariadi General Hospital/Faculty of Medicine Diponegoro University, Semarang, Indonesia
[3]Neurointervention Division, Department of Neurology, Dr. Kariadi General Hospital/Faculty of Medicine Diponegoro University, Semarang, Indonesia
[4]Department of Neurology, Dr. Kariadi General Hospital/Faculty of Medicine Diponegoro University, Semarang, Indonesia

Corresponding author:
Dodik Tugasworo
E-mail: dodiktugasworo2024@gmail.com

ABSTRACT

Introduction: Syringomyelia is a chronic, progressive, degenerative spinal cord condition that manifests as a gap/cavity in the center of the cervical spinal cord. It is frequently associated with congenital anomalies such as Chiari malformation. This report discussed four cases of syringomyelia and the literature review on this topic.

Case presentation: Three female patients with a chief complaint of sensory dissociation in shoulder and hand. The MRI showed the imaging of syringomyelia and Chiari malformation type 1. The management of the disease was pharmacologic treatment, physiotherapy, and decompression surgery.

Conclusion: Syringomyelia and Chiari Malformation are complex neurological disorders. Symptoms may become worse over time. Good outcomes are possible if treatment is not delayed. These conditions can be managed successfully by conservative treatment but if there is a progression of neurological deficit, surgical decompression is a mandatory.

Keywords: syringomyelia, Chiari malformation

INTRODUCTION

Syringomyelia is a chronic, progressive, degenerative spinal cord condition that manifests as a gap/cavity in the center of the cervical spinal cord. This anomaly may extend caudally to the thoracic and lumbar spinal segments or rostrally to the brainstem (syringobulbia). This condition leads to progressive neurological abnormalities, commonly brachial amyotrophy and segmental sensory dissociation.[1–3] Syringomelia is quite rare. It is frequently associated with congenital anomalies such as Chiari malformation.[4,5] As a result, the manifestations of this condition will vary based on the location and size of

the lesion and also the underlying abnormalities. However, this disorder may generally be recognized from typical symptoms such as amyotrophy and dissociated sensory loss.[2,6]

There is still no consensus regarding the pathophysiology of syringomyelia. As a result, the treatment approaches vary considerably. This condition progresses slowly. A person with syringomyelia may remain in the same state for years, if not decades.[7]

Chiari malformations are a set of conditions in which the part of the cerebellum, fourth ventricle, or pons protrudes/herniated downward.[8–10] The pressure exerted on the cerebellum obstructs the passage of cerebrospinal fluid (CSF), resulting in various symptoms, including dizziness, muscle weakness, numbness, vision abnormalities such as nystagmus, headaches, and balance-coordination difficulties. For some individuals, symptoms may vary based on the amount of CNS accumulation and the concurrent pressure of tissues and nerves. Chiari malformations are related with the generation of syrinx (gap/cavity), in the spinal cord, called syringomyelia.[8–11] The incidence of syringomyelia in patient with Chiari malformations is about over 60% in adults and 40% in pediatric patients. Chiari-related syringomyelia made a communication between syrinx cavity and fourth ventricle, called “communicating syringomyelia”.[10] This report discussed four cases of syringomyelia with Chiari malformation type 1

CASE REPORT

Case 1

A 62-year-old female bank employee was presented with a chief complaint of shoulder pain radiating to both hands for three months prior to admission. The pain was described as a burning sensation similar to an electric shock in both hands and resulted in the patient requiring assistance to conduct specific daily tasks. The pain improved considerably during rest. She also felt numbing sensation from the neck to both hands. No history of trauma, malignancy, fever, toothache, heart disease, hypertension, or diabetes mellitus were reported. She also denied any history of a similar condition in the family.

Initially, the patient complained of shoulder discomfort, mimicking the common cold; however, the discomfort progressed to a burning sensation, and the patient chose to stay at home rather than visit a doctor. A month after the onset, the condition deteriorated, and the pain started and radiated from the shoulder to both hands. There were no reports of weakness in the limbs, headaches, or dizziness. She finally went to the doctor for a chest x-ray, which was normal; then, she received physiotherapy without progressing symptoms. One week prior to her visit to Dr. Kariadi General Hospital, the patient began to experience weakness in her arm while grasping something and pain radiating from her right shoulder to both hands. There was no fever, loss of consciousness, or visual impairment. The patient then visited Dr. Kariadi General Hospital's neurology clinic. The neurological examination revealed right superior monoparesis (motoric strength 2) and abnormalities in sensory evaluations, including hypesthesia in right cervical(C)5-6 dermatome, while others were within normal limits.

Additionally, a plain cervical MRI was performed on the patient (figure 1), and the lesion was observed to extend spinal intramedullary from vertebrae C2 to thoracic(T)5, indicating syringomyelia. There were two

oval-shaped intramedullary spinal lesions: one at the level of C6-C7 and another at the level of T3-T4 vertebrae, and posterocentral bulgings of the C2-3, C3-4, C4-5, and C6-7 intervertebral discs with compression of the thecal sac, without right or left neural foraminal narrowing. A right posterocentral-posterolateral bulging of the C5-6 intervertebral disc was also found along with compression of the thecal sac and the right neural foraminal narrowing.

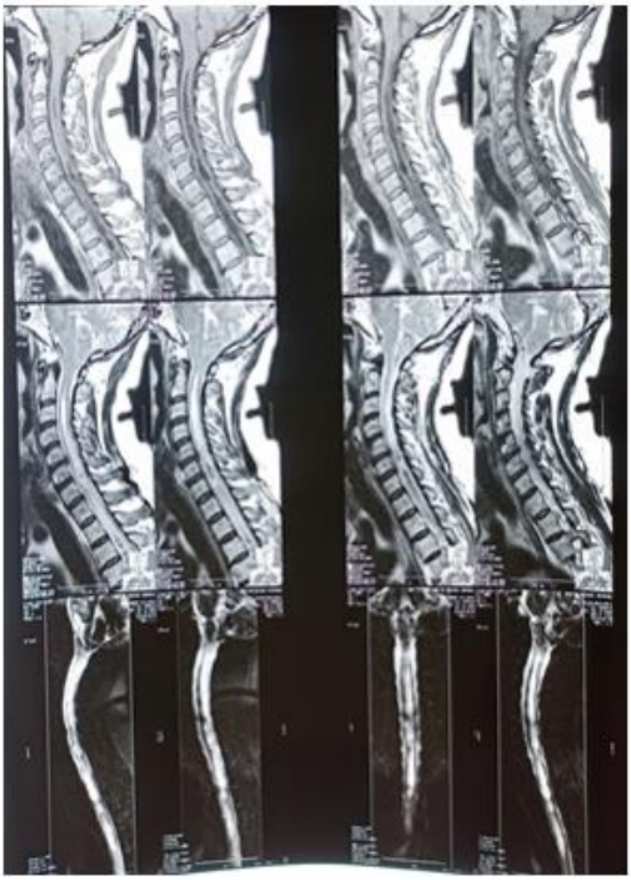

Figure 1. Plain cervical MRI (of case 1) with a spinal intramedullary lesion extending at the level of the C2–T5 vertebrae – tends to show syringomyelia.

The patient was clinically diagnosed with burning shoulder pain, right upper extremity hypesthesia (sensory dissociation), and low motor neuron (LMN) type - right superior monoparesis (with motoric strength 2). The topical diagnosis was cervical spinal cord, while the etiological diagnoses were syringomyelia and cerebellar tonsil herniation (Chiari malformation type 1).

She received pharmacological and non-pharmacological treatment. The neurosurgery department recommended the patient to have foramen magnum decompression surgery, C1-C2 fusion, and IOM during surgery. The patient continued to have the prior oral treatment regimen following the surgery, with intravenous medications including methylprednisolone 125 mg t.i.d The patient was discharged on the seventh day of treatment. The neurological status of patient was improved in 1 month after treatment (from motoric strength 2 to 4).

Case 2

A 43-year-old female came to the hospital with the primary complaint of numbness in her left hand three years before admission. The left-arm had a sudden numbness following a tingling sensation from the fingertips to the upper arm, and the patient requires family assistance to perform daily chores. The symptoms got worsen when walking for long distances and alleviated after resting. Additional complaints were physical weakness and heaviness – stiffness – numbness of the right hand.

Three years prior to admission, the patient complained of numbness and tingling sensation from the left fingertips to the upper arm, followed by sudden numbness in the left arm. She could not feel pain or distinguish between heat and cold sensation with normal touch and pressure sensation. Subsequently, the patient was referred to a neurologist at private hospital for a head MRI (figure 2). It revealed syringomyelia associated with cerebellar tonsillar herniation consistent with the Chiari malformation type 1, thoracic scoliosis with right-sided convexity, and bulging discs at C4-5 and C5-6 without right or left neural foramen stenosis. She refused a referral plan to a neurosurgeon for a surgical approach. The patient depended solely on alternative treatment and had improved significantly after a few months. There were no complaints of limb weakness; the patient merely noticed that her left hand's fingers frequently move involuntary, as if shaking. There was no bowel or bladder dysfunction. She could resume her daily tasks as a merchant in the market; she walked by herself but felt exhausted after 1 km.

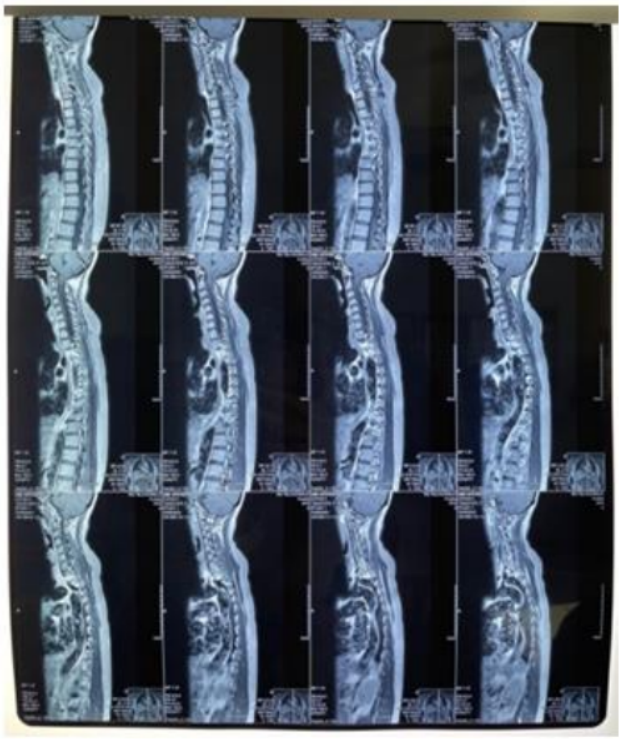

Figure 2. Cervicothoracic MRI with contrast in Case 2. The impression is still visible intramedullary lesions in the central canal at the level of V.C1-2 to Th 11-12 (non-enhanced)

The patient complained of heaviness, stiffness, and numbness in her right hand in early 2021. The patient could still grasp objects with her right hand but could not hold them for an extended period. Additionally, her left hand had become increasingly weak over the last two years, and the hand's muscles appear to have shrunk. Her left arm continued having the same symptoms and even worsened, and she could not move her arm. She became easily fatigued and could not walk for long distances; after approximately 100 meters, she would be exhausted. She sought alternative treatment and acupuncture (21 regimens) without any improvement; the patient returned to the neurologist and performed another head MRI in early September 2021, which revealed a more severe condition than the previous MRI result. The patient agreed to be referred to the neurosurgery division at Dr. Kariadi General Hospital. When she arrived at Dr. Kariadi General Hospital, the patient reported weakness in all four limbs and could not perform any daily activities. The left hand remained numb, and her right hand was heavy, stiff, and numb. Due to frequent fatigue, the patient could still walk in slow paces for short distances. There was a reduced motor function in all four extremities; the muscle strength examination observed a general weakness of 444/443 444/444. There was muscle atrophy in the right superior and inferior extremities and a sensory dissociation; Meanwhile, autonomic nerve function was found to be within normal limits. The patient had a chest x-ray, revealing normal results for heart and lung, thoracic vertebral scoliosis with right-sided convexity. Lateral and anteroposterior thoracolumbar x-rays showed lumbar spondylosis, normal lower-lumbar thoracic vertebrae alignment, and no listhesis.

The patient received pharmacological and non-pharmacological treatment. The neurosurgery department recommended foramen magnum decompression and C1-C2 fusion (lateral mass) procedure. The neurological status of patient was improved in 1 month after treatment (from motoric strength 2 to 4). The patient was discharged on the 1 weeks of treatment. The neurological status of patient was improved in 1 month after treatment (from motoric strength 4 to 5).

Case 3

A 45-year-old female teacher reported numbness from the neck to right hand’s fingers continuously, felt for 5 years prior to hospital admission. She could do daily activities independently. She also complained neck pain on the right side, headache, sharp and burning sensation. She went to the Private Hospital and underwent routine physiotherapy for 2 years but the complaints did not improve, she was referred to the Dr. Kariadi General Hospital. Cervical MRI showed a mass, surgery was suggested but she refused.

Currently she complained neck pain and numbness radiating to her right arm, sometimes she feels tight on her right eye, low vision at night and stagger when she walks. No history of nausea, vomiting, seizures, syncope, limb weakness. No history of fever, illness, toothache, trauma, hypertension, diabetes mellitus or heart disease was reported. No family members were sharing similar symptoms. She has taken medicine regularly from neuro clinic which consist of diclofenac sodium, thiamine, pyridoxine, folic acid and gabapentin.

The latest MRI (figure 3) revealed herniation of the inferior aspect of the cerebellar tonsils inferior to the foramen magnum resulting in foramen stenosis, narrowing of the foramen Magendi, and suspicion of non-

communicating hydrocephalus Chiari malformation type I. Syringomyelia starts from craniocervical junction level to the upper body T2, accompanied by suspicion of bleeding areas. The size was relatively stable compared to the previous and arachnoid cyst in the posterior fossa in the right and left medullary-cerebellar cistern.

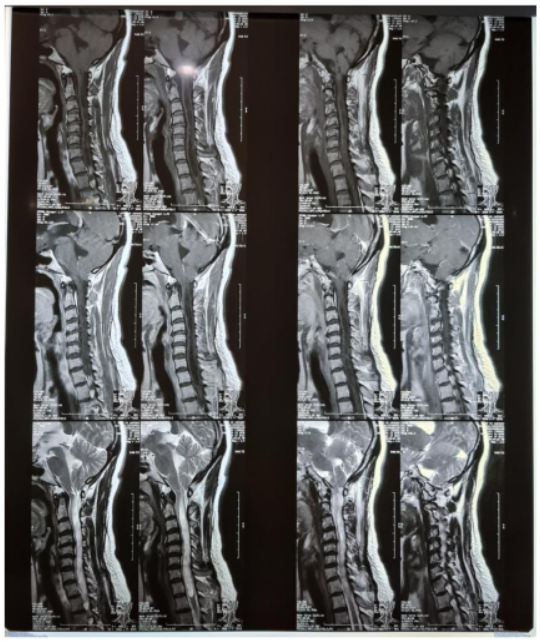

Figure 4. Herniation of the inferior aspect of the cerebellar tonsils inferior to the foramen magnum resulting in foramen magnum stenosis, narrowing of the foramen Magendi, and suspicion of non-communicating hydrocephalus of Chiari malformation type I.

Neurological examinations revealed a decrease of strength on the superior right arm, and hyperesthesia from right hand fingertip according to the C3 dermatome. She underwent neurosurgery for decompression The neurological status of patient was improved in 2 months after treatment (from motoric strength 3 to 4).

DISCUSSION

Chiari malformation was initially documented more than a hundred years ago, involving abnormalities in the posterior fossa, typically characterized by the descent of the cerebellar tonsils via the foramen magnum. The early comprehension of this condition was primarily derived from postmortem examinations, and has been significantly augmented by the introduction of MRI technology.[1] The surgical treatment of Chiari abnormalities has also developed in a similar manner. While the specific surgical approach may differ across surgeons, the primary objectives of the procedure remain consistent. These objectives include alleviating pressure on the brainstem and distortion of cranial nerves, restoring the normal circulation of cerebrospinal fluid through the foramen magnum, and minimizing the size of any accompanying syrinx cavity. Syrinx cavities are typically linked to Chiari abnormalities, however primary spinal syringomyelia (PSS) can also be

induced by many factors such as trauma, infection, degeneration, and other causes that result in a partial obstruction of cerebrospinal fluid (CSF) flow in the spinal subarachnoid space.[2] Similar to syringomyelia linked to Chiari abnormalities, the primary objective of PSS surgery is to restore cerebrospinal fluid (CSF) circulation through the obstructed region. Aside from MRI, myelography with CT can be highly beneficial in assessing and treating these individuals by finding specific areas of cerebrospinal fluid (CSF) blockage that may be suitable for surgical intervention. [3]

Chiari malformation type I is the predominant cause of non-communicating syringomyelia. The imaging of patients with this illness shows the presence of cerebellar tonsillar ectopia, which can extend beyond the foramen magnum.[10-11] Additionally, it is proposed that disturbed absorption of extracellular fluid through intramedullary venous channels in the spinal cord microcirculation may result in the accumulation of extracellular fluid. Due to its sensitivity to pressure, symptoms may exacerbate with the Valsalva maneuver or other activities that can impact cerebrospinal fluid (CSF) pressure.[11] Symptoms related to the ear and nervous system include vertigo, ringing in the ears, sensation of pressure in the ears, reduced hearing, and visual disturbances characterized by the perception of objects moving back and forth. Cerebellar abnormalities manifest as tremors, dysmetria, ataxia, and impairments in walking and balance. Syncope refers to a temporary loss of consciousness or fainting. The patient can have sleep disruptions such as snoring, sleep apnea, and palpitations due to compression of the brainstem.[12,13] Progressive scoliosis occurs due to damage to the anterior horn cells that control the paraspinal axial muscles. Horner syndrome can occur in individuals with cervical or upper thoracic trauma.[14]

Physical examination of first patient revealed right superior monoparesis (motoric strength 2) and abnormalities in sensory evaluations, including hypesthesia in right cervical(C)5-6 dermatome, while others were within normal limits. Physical examination of second patient revealed reduced motor function in all four extremities; the muscle strength examination observed a general weakness of 444/443 444/444. There was muscle atrophy in the right superior and inferior extremities and a sensory dissociation. Physical examination of third patient showed decrease in the ability to move, strength, and sensory in the right superior extremity in the form of hypesthesia, decrease in pain and temperature sensation in the right superior extremity. Neurological examinations of third patient revealed a decrease of strength on the superior right arm, and hyperesthesia from right hand fingertip according to the C3 dermatome.[14,15]

Magnetic Resonance Imaging (MRI) is the preferred diagnostic procedure, which can be performed with or without the use of contrast agents.[16] It clearly defines the important anatomical features and enables precise imaging of the syrinx in both sagittal and axial orientations. MRI readily identifies the precise position, dimensions, and scope of the syrinx cavity, as well as the extent of cerebellar tonsillar ectopia.

Plain cervical MRI of first patient showed lesion extended spinal intramedullary from vertebrae C2 to thoracic(T)5, indicating syringomyelia , and posterocentral bulgings of the C2-3, C3-4, C4-5, and C6-7 intervertebral discs with compression of the thecal sac, without right or left neural foraminal narrowing. Lateral and anteroposterior thoracolumbar x-rays of second patient showed lumbar spondylosis, normal lower-lumbar thoracic vertebrae alignment, and no listhesis. MRI of second patient revealed syringomyelia

associated with cerebellar tonsillar herniation consistent with the Chiari malformation type 1, thoracic scoliosis with right-sided convexity, and bulging discs at C4-5 and C5-6 without right or left neural foramen stenosis. The latest MRI of third patient revealed herniation of the inferior aspect of the cerebellar tonsils inferior to the foramen magnum resulting in foramen stenosis, narrowing of the foramen Magendi, and suspicion of non-communicating hydrocephalus Chiari malformation type I. Syringomyelia starts from craniocervical junction level to the upper body T2, accompanied by suspicion of bleeding areas. Typically, asymptomatic incidental lesions are monitored with periodic imaging in a clinical setting. However, in some places, preventive surgery may be carried out.[19] The occurrence of recurrent or persistent syringomyelia after Chiari decompression in adults is observed in approximately 6.7% of cases on average. When there are large holocord syringes, spinal cord injury can result in long-lasting symptoms or disability, even if the syrinx size is adequately reduced with decompression. [20] Research has indicated that a reduced length of time for the symptoms before surgery leads to more favorable results. Performing surgery at an early stage helps to reduce the negative effects or impairments. [21]

The first patient was diagnosed with syringomyelia and cerebellar tonsil herniation (Chiari malformation type 1) as the etiology of burning shoulder pain, right upper extremity hypesthesia (sensory dissociation), and low motor neuron (LMN) type - right superior monoparesis (with motoric strength 2). The second patient was diagnosed with The patient was diagnosed with syringomyelia at the level of C2-T1 with Chiari malformation type 1. The first patient was given oral medications, physiotherapy, and exercise. The neurosurgery department recommended the patient to have foramen magnum decompression surgery, C1-C2 fusion, and IOM during surgery. The patient continued to have the prior oral treatment regimen following the surgery, with intravenous medications including methylprednisolone 125 mg t.i.d and omeprazole 40 mg b.i.d. The second patient was given oral medications, physiotherapy, and exercise. The neurosurgery department recommended foramen magnum decompression and C1-C2 fusion (lateral mass). The third was recommended some pharmacological treatment and underwent surgery for decompression. However, if the syrinx diameter exceeds 5 mm and is accompanied by edema, it indicates a rapid decline in health. The infrequency of the illness, unpredictable progression, and limited duration of monitoring provide challenges in evaluating therapy outcomes. Nevertheless, performing surgery at an early stage reduces impairments and leads to superior results.[22] Postoperative neurological problems encompass cerebrospinal fluid (CSF) leakage, infection, bleeding, and syrinx recurrence.[23]

CONCLUSION

Syringomyelia and Chiari malformation are complex neurological disorders. Symptoms may become worse over time. Good outcomes are possible if treatment is not delayed. These conditions can be managed successfully by conservative treatment but if there is a progression of neurological deficit, surgical decompression is a mandatory.

Conflict of Interest: None

REFERENCES

1. Hart DJ. Syringomyelia. Encycl Neurol Sci. 2014;3:378–81.
2. Sternberg ML, Gunter ML. Syringomyelia. J Emerg Med. 2017;53(2):e31–2. doi: 10.1016/j.jemermed.2017.04.029.
3. Yudoyono F, Widhiatmo AO, Wirjomartani BA. Progressive Neurological Deficit in Adult Idiopathic Syringomyelia (IS): Case Report and Literature Review. J Med Heal. 2016;1(3):258–63. doi: 10.28932/jmh.v1i3.521.
4. Di Rocco C, Pang D, Rutka JT. Textbook of Pediatric Neurosurgery. Cham: Springer Cham; 2020. 243–275 p.
5. Killory BD, Preul MC, Spetzler RF. Chiari Malformations. Encycl Neurol Sci. 2014;1:770–2.
6. Hankey GJ, Wardlaw JM. Clinical Neurology. 1st ed. Florida: CRC Press; 2008.
7. Heiss JD, Snyder K, Peterson MM, Patronas NJ, Butman JA, Smith RK, et al. Pathophysiology of primary spinal syringomyelia. J Neurosurg Spine. 2012;17(1):367--380. doi: 10.3171/2012.8.SPINE111059.
8. Urbizu A, Toma C, Poca MA, Sahuquillo J, Cuenca-León E, Cormand B, et al. Chiari Malformation Type I: A Case-Control Association Study of 58 Developmental Genes. PLoS One. 2013;8(2):1–10. doi: 10.1371/journal.pone.0057241.
9. McClugage SG, Jerry Oakes W. The Chiari I malformation. J Neurosurg Pediatr. 2019;24(3):217–26. doi: 10.3171/2019.5.PEDS18382.
10. Holly LT, Batzdorf U. Chiari malformation and syringomyelia. J Neurosurg Spine. 2019;31(November):619–28. doi: 10.3171/2019.7.SPINE181139.
11. Klekamp J. How Should Syringomyelia be Defined and Diagnosed? World Neurosurg. 2018;111:e729–45.
12. Chang HS, Nakagawa H. Hypothesis on the pathophysiology of syringomyelia based on simulation of cerebrospinal fluid dynamics. J Neurol Neurosurg Psychiatry. 2003;74(3):344–7. doi: 10.1136/jnnp.74.3.344 .
13. Hidalgo J, Tork C, Varacallo M. Arnold Chiari Malformation. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022.
14. Labuda R, Simplice B, Nwotchouang T, Ibrahimy A, Allen PA, Oshinski JN, et al. A new hypothesis for the pathophysiology of symptomatic adult Chiari malformation Type I. Med Hypotheses. 2022;158:1–8. doi: 10.1016/j.mehy.2021.110740.
15. Goetz L, De JO, McAvoy S. Posttraumatic Syringomyelia. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022.
16. Walid M. Syringomyelia: A Complication of an Underlying Pathology. J Clin Med Res. 2010;2(2):102–4. doi: 10.4021/jocmr2010.04.291w.
17. Sheony V, Sampath R. Syringomyelia. StatPearls [Internet]. Treasure Island (FL): StatPearls

Publishing; 2022.

18. Flint G. Syringomyelia: diagnosis and management. Pract Neurol. 2021;21(5):403–11. doi: 10.1136/practneurol-2021-002994.
19. Leclerc A, Matveeff L, Emery E. Syringomyelia and hydromyelia: Current understanding and neurosurgical management. Rev Neurol (Paris). 2021;177(5):498–507. doi: 10.1016/j.neurol.2020.07.004.
20. Lee JS, Song GS, Son DW. Surgical Management of Syringomyelia Associated with Spinal Adhesive Arachnoiditis, a Late Complication of Tuberculous Meningitis: A Case Report. Korean J Neurotrauma. 2017;13(1):34–8. doi: 10.13004/kjnt.2017.13.1.34.
21. Greenberg JK, Yarbrough CK, Radmanesh A, et al. The Chiari Severity Index: a preoperative grading system for Chiari malformation type 1. *Neurosurgery*. 2015;76(3):279-285. doi:10.1227/NEU.0000000000000608.
22. Mora J.R., Rison R.A., Beydoun S.R. Chiari Malformation Type I with Cervicothoracic Syringomyelia Masquerading as Bibrachial Amyotrophy : A Case Report. *Journal of Medical Case Reports*. 2015; 9 (11)) : 1-6.
23. Sivakumaran P, Ashwood N, Kamal M, Jayakumar N. The Pathogenesis of Chiari Malformation and Syringomyelia: A Case Report and Systematic Review of Current Theories. *Cureus*. 2023;15(10):e47301. Published 2023 Oct 19. doi:10.7759/cureus.47301