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Holocord Syringomyelia due to Spinal Dysraphism without Chiari Malformation - A Case Report

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ABSTRACT
Syringomyelia is a cystic enlargement of the spinal cord that causes myelopathy. The commonly used classification is one based on anatomical features or pathogen other than pathophysiological mechanisms. Among the many pathophysiological theories, the most widely accepted is that disturbances in cerebrospinal fluid (CSF) flow in the lower arachnoid spaces disrupt flow velocity leading to syrinx. Although it may appear idiopathically, Type 1 and 2 Chiari, the Dandy-Walker syndrome cerebellar ectopia basilar impression basilar arachnoiditis, posterior fossa tumors and arachnoid cysts, medulla spinalis tumors, spinal trauma and infections are among the most common causes. Syringomyelia is mostly seen with type 1 Chiari malformation in patients. Based on this information, we present this rare patient with tethered cord and holocord syringomyelia without type 1 Chiari malformation. We aimed to contribute to the literature by providing information about the treatment process of the patient.
Keywords: Malformation Arnold-Chiari, Tethered Cord Syndrome, Syringomyelia.

Chiari Malformasyonu Olmayan Spinal Disrafiye Bağlı Holokord Siringomiyeli: Vaka Sunumu

ÖZ
Siringomiyeli, miyelopatiye neden olan omuriliğin kistik genişlemesidir. Yaygın olarak kullanılan sınıflandırma, patofizyolojik mekanizmalar dışında anatomik özelliklere veya patojene dayanan bir sınıflandırmadır. Birçok patofizyolojik teori arasında en yaygın kabul göreni, alt araknoid boşluklardaki beyin omurilik sıvısı (BOS) akışındaki bozuklukların akış hızını bozarak syrinkse yol açmasıdır. İdiyopatik olarak ortaya çıkabilmesine rağmen, Tip 1 ve 2 Chiari, Dandy-Walker sendromu serebellar ektopi baziler izlenim baziler araknoidit, posterior fossa tümörleri ve araknoid kistler, medulla spinalis tümörleri, spinal travma ve enfeksiyonlar en sık nedenler arasındadır. Siringomiyeli hastalarda en sık tip 1 Chiari malformasyonu ile birlikte görülür. Bu bilgiler ışığında, tip 1 Chiari malformasyonu olmaksızın nadir görülen gergin kord ve holokord siringomiyeli olgusunu sunuyoruz. Hastanın tedavi süreci hakkında bilgi vererek literatüre katkı sağlamayı amaçladık.
Anahtar Kelimeler: Malformasyon Arnold-Chiari, Gergin Kord Sendromu, Siringomyeli.

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INTRODUCTION

Syringomyelia is a rare spinal cord pathology caused by the enlargement of the central canal in the spinal cord due to compression of the cerebrospinal fluid (Leclerc, Matveeff, & Emery, 2021). Studies in literature report incidence rates of 8.4 per 100,000 and 0.9 per 10,000 (Brickell et al., 2006; Weier et al., 2008). Symptoms frequently appear between the ages of 20 and 50. Pathophysiological mechanisms have been overlooked in the classification of syringomyelia. The classification is divided into two groups based on anatomy and underlying pathology: foraminal and non-foraminal (Leclerc et al., 2021). Foraminal syringomyelia is frequently associated with craniocervical junction pathologies (Type 1 and Type 2 Chiari, Dandy-Walker, etc.). Non-foraminal syringomyelia, on the other hand, is mostly associated with pathologies arising from arachnoiditis (trauma, infection, etc.) (Leclerc et al., 2021; Tucer, Yilmaz, Ekici, Menku, & Koc, 2014). In addition to the symptoms of the primary pathology causing syringomyelia, cape-type sensory loss and loss of heat and touch sensation are characteristic findings (Greitz, 2006; Roser, Ebner, Sixt, Hagen, & Tatagiba, 2010). According to some studies in the literature, Chiari malformation is the primary pathology accounting for 90% of syringomyelia cases (Fernández et al., 2009). Only 12% of syringomyelia cases without Chiari malformation have spinal dysraphism (Lee, Kim, & Wang, 2020). Although the relationship between tethered cord and syringomyelia is well known, its pathophysiology is unclear (Bruzek et al., 2019). The following case study, which involves spinal dysraphism (tethered cord) and associated holocord syringomyelia, without Chiari malformation, will be examined and discussed. A concise overview of the extant literature pertaining to the management of syringomyelia in conjunction with spinal dysraphism will be presented.

Ethical considerations

The study was conducted retrospectively and does not require ethics committee approval as it uses anonymized data. This article has been prepared in accordance with the principles of the Declaration of Helsinki and does not require ethics committee approval.

CASE PRESENTATION

A 3-year-old female patient presented to the paediatric clinic with complaints of delayed walking, urinary incontinence and spinal curvature. Imaging results revealed the presence of tethered cords and syringomyelia, thus necessitating the admission of the patient to our clinic. The patient was incapable of standing or walking without support. The presence of pes equinovarus deformity was observed in both feet. Radiographic imaging revealed the presence of scoliosis. The patient's imaging revealed that the spinal cord terminated at the level of the fourth lumbar vertebra, and that the terminal portion appeared thicker than normal, indicative of a tethered cord. The presence of an image consistent with syringomyelia (holocord syringomyelia) was identified in the central canal, extending from the brainstem and spinal cord, and continuing distally. No pathology was detected at the patient's cervicomedullary junction (see Figure 1).

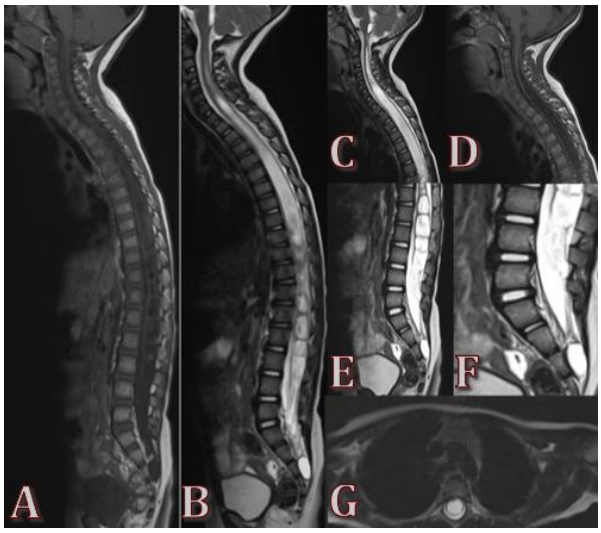


Figure 1. Sagittal T1 and T2 magnetic resonance imaging (A and B) show a finding consistent with syringomyelia (holochord syringomyelia) in the central canal, starting from the brainstem and spinal cord and continuing distally.

No pathology is observed at the cervicomedullary junction (C and D). The patient's images reveal that the spinal cord terminates at the ‘lumbar four vertebrae’ level and that the terminal is thicker than normal (tethered cord) (E and F). T2 axial magnetic resonance imaging (G) shows a syringomyelia covering almost the entire spinal canal.

The patient exhibited severe neurological symptoms associated with syringomyelia; therefore, in addition to tethered cord release surgery, a syringo-subarachnoid shunt was planned. A syringo-subarachnoid shunt was placed at the levels of the filum terminale and thoracic vertebrae 9. Subsequent postoperative follow-up revealed no further complications. Postoperative imaging revealed that the syringomyelia had regressed (see Figure 2).

In the course of the outpatient follow-up examination, it was noted that the patient had commenced ambulation without the use of assistive devices, and that the issue of urinary incontinence had shown signs of improvement.

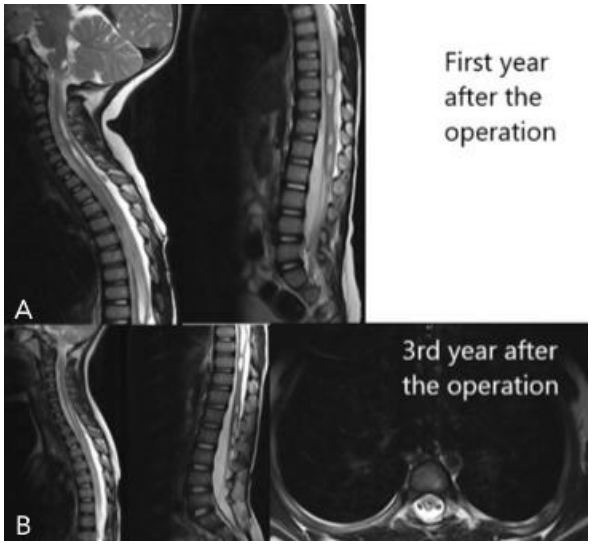


Figure 2. A, T2 sagittal magnetic resonance image taken in the first year after surgery. B, images taken in the third year after surgery.

DISCUSSION

Syringomyelia CSF is a rare spinal cord pathology resulting from circulatory impairment. This circulatory impairment causes the central canal of the spinal cord to expand into a fluid-filled cavity (Greitz, 2006). The prevalence of syringomyelia is a subject that has received scant attention in epidemiological literature. A survey of extant studies reveals that this is an exceedingly rare pathology (Brickell et al., 2006; Weier et al., 2008). In addition to classification based on anatomical and pathological features, syringomyelia can be divided into two categories based on aetiologia: idiopathic and symptomatic syringomyelia. In this distinction, the causes of secondary syringomyelia include craniocervical junction pathologies, posterior fossa tumours, infections, cerebellar ectopia, basilar invagination, and related pathologies (Huang et al., 2011; Kim & Kim, 2009; Takeuchi et al., 2003; Tucer et al., 2014). Milhorat et al. evaluated 105 postmortem cases in autopsy series and classified syringomyelia into three groups based on their findings. They identified central canal enlargement associated with the fourth ventricle (communicating) in 47 patients, central canal enlargement not associated with the fourth ventricle (isolated, non-communicating) in 23 patients, and extra canalicular syringomyelia within the parenchyma in 35 cases. In communicating syringomyelia, the cavity is partially or completely lined with ependyma. Non-communicating syringomyelia was associated with pathologies affecting CSF dynamics, such as spinal arachnoiditis, cervical spinal stenosis, basilar compression, and type 1 Chiari malformation (Kim & Kim, 2009 ; T. H. Milhorat, Capocelli, Anzil, Kotzen, & Milhorat, 1995). The extra-anal type has been associated with spinal cord injuries (hemorrhage, infarction, etc.).

In addition to the present study, syringomyelia has been classified in various ways in numerous studies. In the syringomyelia literature, researchers have developed various theories about the pathophysiology of syringomyelia (Leclerc et al., 2021). Gardner and Angel's theory is the 'hydrodynamic theory'. Due to posterior brain herniation, CSF circulation is affected and, as a result, they propose the 'water hammer' effect, which results in CSF being directed towards the central canal with each pulse (GARDNER & ANGEL, 1958). Williams proposes the theory that in patients with posterior brain herniation, pressure in the foramen magnum reaches 100 mmHg during Valsalva maneuvers, and that cerebrospinal fluid (CSF) exerts a suction force pulling it into the syringomyelia cavity as a result of the cranial pressure difference (Williams, 1972). Ball and Dayan state that in cases of foramen magnum obstruction, increased CSF pressure (coughing, sneezing) causes it to progress through the perivascular Virchow-Robin spaces (Ball & Dayan, 1972). However, there is no comprehensive study that could classify these three theories as positive or negative (Edward H. Oldfield, 2017). In addition to these, one approach suggests that herniated cerebellar tonsils create a 'piston-like' effect, causing CSF to accumulate in the central canal (E. H. Oldfield, Muraszko, Shawker, & Patronas, 1994). According to another approach, the rapid movement of CSF over the spinal cord causes a low-pressure area in the spinal cord, which consequently promotes the accumulation of extracellular fluid and the formation of syringomyelia (Greitz, 2006 ; Honey et al., 2017 ; Koyanagi & Houkin, 2010). Our research on the symptoms of syringomyelia indicates that this topic must be addressed within a very broad spectrum. Due to the symptoms caused by primary pathology, they are mostly seen as a secondary result of certain pathologies. The most common sensory symptom is paresthesia/hyperesthesia; non-radicular segmental pain is observed (Thomas H. Milhorat et al., 1999). While the sense of touch and vibration is preserved, the sensation of pain and temperature is lost. There is a 'cloak-like' hypoesthesia in the upper extremities. Due to problems with pain perception, patients may suffer serious burns or injuries. Fine motor skills are affected as a result of weakness in the hand muscles. Progressive syringomyelia may cause spasticity in the lower extremities. Progressive scoliosis may be seen due to involvement of the anterior horn and, as a result, involvement of the paraspinal muscles (Batzdorf, Khoo, & McArthur, 2007). The differential diagnosis of syringomyelia includes tumours, cysts and myelomalacia. From a radiological imaging perspective, contrast-enhanced and non-contrast-enhanced MRI of the spinal cord should be performed to confirm the diagnosis of syringomyelia. Consequently, this approach facilitates the diagnosis of spinal dysraphism and numerous analogous spinal cord pathologies.

Furthermore, cranial imaging and craniocervical junction imaging should be performed in cases of Chiari malformation and cranial pathologies. Furthermore, cine MRI obtained at the level of the foramen magnum can be used to demonstrate obstruction in the cerebrospinal fluid circulation pathway. While it is not a diagnostic tool for syringomyelia, electromyography (EMG) can be utilized to exclude the presence of neuropathies.

If syringomyelia accompanies another pathology, treatment and management initially involve treating the primary pathology. Syringomyelia tends to regress with improved cerebrospinal fluid circulation, which often occurs when the primary pathology is corrected (Roy, Slimack, & Ganju, 2011). Shunts are employed in cases of idiopathic syringomyelia or in instances where syringomyelia persists following treatment of the underlying primary pathology. Specifically, the utilization of Syring subarachnoid, Syring pleural and Syring peritoneal shunts may be indicated. Syringomyelia caused by spinal dysraphism without craniovertebraljunction anomalies is reported in some sources in the literature at a rate of 12%. Although the relationship between tethered cord and syringomyelia is well known, the direct pathophysiological relationship is unknown (Bruzek et al., 2019). In a study conducted on paediatric patients with tethered cord syndrome who did not have significant imaging abnormalities such as lipomyelocele, lipoma, or thickened/fatty filum terminale, no change in the size of syringomyelia was observed in most patients after surgery (Kulwin et al., 2013). In 2020, a cohort study involving 25 patients reported that there was no significant improvement in syringomyelia despite tethered cord surgery (Bruzek et al., 2019). In another series of 16 patients, only 25% of patients experienced a reduction in syrinx size following tethered cord surgery (Kulwin et al., 2013). Syrinx drainage is also recommended in cases of spinal dysraphism and large syringomyelia. Good results have been observed in these cases (Erkan, Unal, & Kiris, 1999 ; Iskandar, Oakes, McLaughlin, Osumi, & Tien, 1994). In the case presented, due to the presence of tethered cord and holocord syringomyelia, which resulted in severe symptoms, both tethered cord release and Syring subarachnoid shunt surgery were performed during the same surgical session. The patient's inability to be managed with spinal dysraphism alone, as is possible with milder cases of syringomyelia, led us to adopt this approach. As demonstrated in pertinent literature, a number of studies involving a limited number of cases have indicated that positive outcomes in relation to syringomyelia have been elusive in instances where tethered cord release has been the sole procedure performed (Bruzek et al., 2019; Kulwin et al., 2013). The combination of direct syringomyelia drainage with untethering has been demonstrated to yield superior clinical outcomes and significant improvement in syringomyelia compared to untethering alone. Specifically, the terminal ventriculostomy procedure for the management of terminal syringomyelia has been demonstrated to yield a 91% improvement in motor and sensory neurological disorders (Mosleh & Sohn, 2024).

CONCLUSION

The present study is an account of the treatment of a patient with holocord syringomyelia due to spinal dysraphism without craniocervical junction pathology. The study is of significance as it contributes to the limited literature on syringomyelia.

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Conflict of Interest

The author declare no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Author Contributions

Planning, design: AST, EC; **Materials, methods and data collection:** AST, MA, MOB; **Data analysis and interpretation:** AST, MA; **Writing and editing:** MOB, EC.

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Ethical Approval

The study was conducted retrospectively and does not require ethics committee approval as it uses anonymised data. This article has been prepared in accordance with the principles of the Declaration of Helsinki and does not require ethics committee approval.

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