BMC Neurology

CASE REPORT

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Holocord syringomyelia caused by tethered cord syndrome: case report and literature review

Longtao Zheng[1], Zhangzheng Liao[1] and Hongzhou Duan[1*]

Abstract

Background Syringomyelia is a rare disease with diverse etiologies, and the syrinx is typically confined to certain segments of the spinal cord. Case of syringomyelia affecting the whole cord due to tethered cord is extremely rare, and the underlying pathophysiological mechanisms remain poorly understood.

Case presentation We described an 18-year-old male patient who presented with progressive weakness in both lower extremities and bladder dysfunction over the past four years. Magnetic resonance imaging (MRI) of the entire spine revealed a tethered spinal cord with a large syrinx extending from C1 to L5. Common causes of syrinx such as Chiari malformation, intramedullary tumors and spinal cord injury were systematically ruled out, leading to a strong suspicion that the tethered cord was the primary etiology of the extensive syringomyelia. After undergoing un-tethering surgery, the patient experienced significant symptomatic improvement, and the subsequent follow-up MRI examinations demonstrated a remarkable reduction and eventual resolution of the large syrinx.

Conclusions Although rare, tethered cord syndrome can serve as the sole etiology for extensive syringomyelia. For such patients, performing un-tethering surgery can lead to complete resolution of the syrinx and achieve a satisfactory clinical outcome.

Keywords Cerebrospinal fluid circulation, Spinal surgery, Syringomyelia, Tethered cord syndrome

Background

Syringomyelia is a rare disease that refers to the formation of abnormal fluid-filled cavities within the spinal cord. Syringomyelia can be classified into two categories: primary (or idiopathic) and secondary, based on the presence or absence of obstruction in cerebrospinal fluid (CSF) circulation [1]. Numerous etiological factors have been identified as contributors to CSF circulation obstruction, including congenital malformations, with Chiari malformation being the most prevalent, as well as spinal cord injuries, neoplastic lesions, and inflammatory conditions. While syringomyelia can manifest in any segment of the spinal cord, cases involving the entire spinal cord are relatively infrequent, particularly in association with tethered cord syndrome [2]. This article presents a retrospective case study of a patient with a holocord syrinx attributed to tethered cord syndrome treated in the Department of Neurosurgery in Peking University First Hospital in July 2023. Additionally, a review of pertinent literature is conducted to explore the pathogenesis and therapeutic approaches for this condition.

Case presentation

An 18-year-old male patient was admitted to our hospital in July 2023, presenting with progressive weakness in both lower limbs and bladder dysfunction over the past

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four years. Initially, four years prior, the patient sought medical assistance at a local hospital due to weakness predominantly in the right lower limb, which was accompanied by mild varus deformity of the right foot. A lumbar MRI scan was performed, showing a low-lying conus medullaris at the L5-S1 level and a tethered cord, along with a syrinx extending from L1 to L5 (Fig. 1). However, this finding was erroneously interpreted as normal, and no treatment was initiated at that time. Over the past four years, the patient experienced a gradual worsening of weakness in both lower limbs, along with right lower back pain, frequent urination, and incomplete bladder emptying, without any other sphincter control issues or erectile dysfunction. Upon presentation in our hospital, a physical examination revealed an abnormal gait, an elevated arch, and varus deformity of the right foot, although there was no abnormal sensory perception in the lower limbs. Muscle strength was assessed as grade V in the upper limbs and grade IV in the lower limbs. A comprehensive spinal MRI confirmed the low position of the conus medullaris, tethered spinal cord, and a syrinx extending from C1 to L5, without evidence of Chiari malformation, spinal lipoma, or other lesions (Fig. 2). An ultrasound examination of the urinary system indicated the presence of multiple diverticula in the bladder, with a residual urine volume of 380 mL. After ruling out other potential causes, tethered spinal cord was suspected to be the primary etiology of the holocord syrinx, leading to the decision to perform untethering surgery. Based on preoperative spinal positioning, we performed an L5-S1 laminectomy and cut open the dure mater from L5 to S1. A low-lying conus medullaris at the L5-S1 level was confirmed, along with a significantly enlarged cystic filum terminale, approximately 5 mm in diameter, exhibiting increased tension. The filum was found to be adherent to several surrounding cauda equina nerves. After being freed completely and confirmed by an intraoperative neurophysiological monitoring, the filum terminale was cut at the S1 level, revealing a hollow structure with numerous fibrous partitions. A stream of high-tension

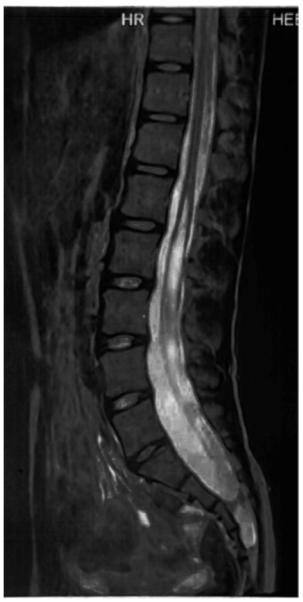

Fig. 1 Four years before admission, the local hospital's lumbar T2 MRI revealed a low-lying conus medullaris positioned at the L5-S1 level, accompanied by a tethered cord and a syrinx extending from L1 to L5

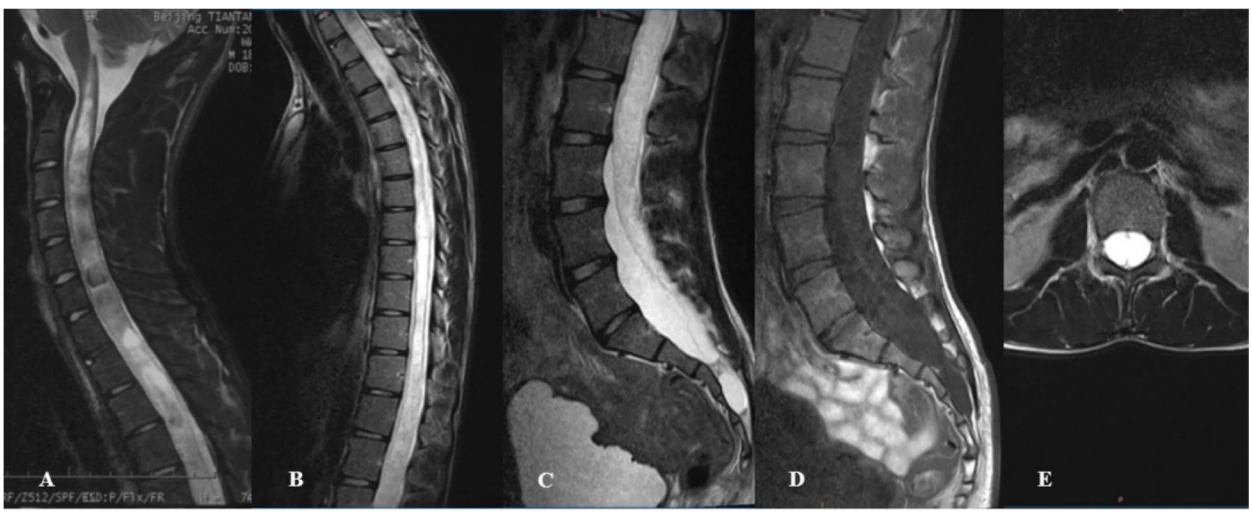

Fig. 2 MRI examinations performed after admission further confirmed the low position of the conus medullaris at L5-S1, the tethered spinal cord, and the presence of a syrinx extending from C1 to L5, with no evidence of Chiari malformation or lumbosacral spinal lipoma. **A** cervical sagittal T2 image; **B** thoracic sagittal T2 image; **C** lumbar sagittal T2 image; **D** lumbar sagittal T1 image; **E** lumbar axial T2 image at L2 segment

fluid gushed out from the incision site immediately, and the conus medullaris shrank and ascended subsequently, suggesting a potential communication between the cystic cavity of the filum and the syrinx. As the filum was cut and the cystic cavity was open, the patient also had an equivalent of a terminal ventriculostomy which was first described by Gandner [3]. Postoperatively, the patient reported significant relief from lower back pain and bladder dysfunction, with an improvement in the strength of both lower limbs. A spinal lumbar MRI conducted five days post-surgery demonstrated a significant reduction in the size of the syrinx (Fig. 3). The patient exhibited satisfactory recovery and attended regular follow-up appointments, during which whole spine MRI examinations at three months and one year (Fig. 4) postoperatively indicated complete resolution of the syrinx. In the most recent one-year follow-up, the patient reported no additional discomfort aside from persistent abnormal gait and varus deformity of the right foot.

Discussion and conclusions

Syringomyelia is an uncommon disease characterized by the formation of abnormal fluid-filled syrinx within the spinal cord. The association between syringomyelia and tethered cord syndrome (TCS) has been documented extensively in the literature [4, 5]. A retrospective study conducted by Erkan involving 132 patients diagnosed with TCS revealed that 32 patients (24%) exhibited syringomyelia as assessed through MRI [4]. Additionally, in a cohort of 143 patients with occult spinal dysraphism, 24 out of 90 patients (27%) were also found to have syringomyelia [5].

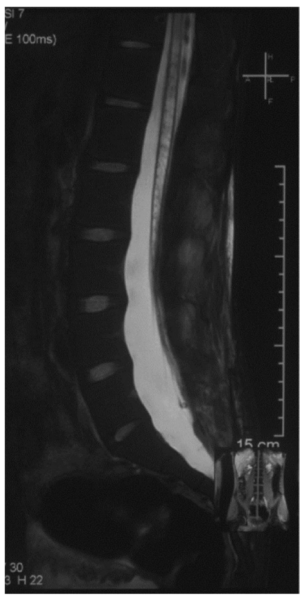

Fig. 3 Five days post-operation, lumbar sagittal T2 MRI demonstrated a significant reduction in the size of the syrinx

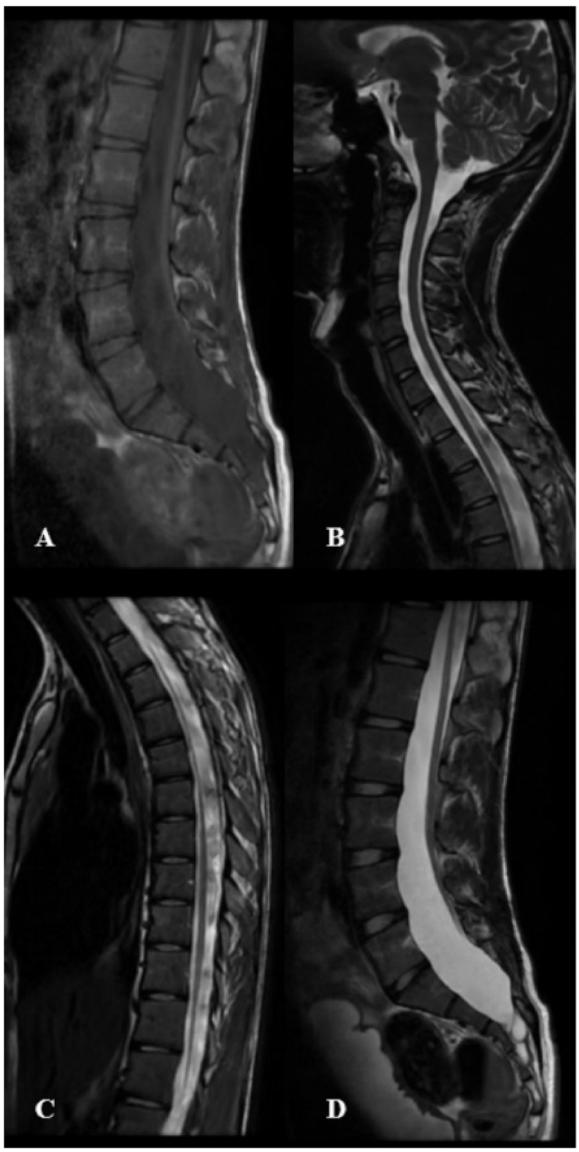

Fig. 4 One year follow-up MRI examinations showed complete resolution of the syrinx. **A** lumbar sagittal T1 image; **B** cervical sagittal T2 image; **C** thoracic sagittal T2 image; **D** lumbar sagittal T2 image

While the correlation between syringomyelia and TCS is well-documented, the underlying pathogenesis linking these two conditions remains incompletely understood. Various hypotheses have been proposed to elucidate the etiology and pathophysiology of syringomyelia. Initial observations indicated that dilation of the central canal and obstruction of cerebrospinal fluid (CSF) circulation occur in the early stages of syringomyelia. One hypothesis posits that atrophy of the spinal cord parenchyma leads to central canal dilation, which compresses the

subarachnoid space and obstructs CSF circulation. This obstruction subsequently prevents CSF from reaching the distal end of the spinal cord, causing it to enter the spinal cord parenchyma, accumulate, and ultimately form a cavity [6]. Other contemporary hypotheses proposed that various factors leading to subarachnoid space obstruction may force CSF into the spinal cord parenchyma and assume that the fluid within the syrinx is indeed CSF [7]. However, these hypotheses have fallen out of favor since it is considered implausible to propel CSF into the spinal cord; rather, it is more likely that CSF would accumulate above the obstructed area and compress the cord, without forming a syrinx within the spinal cord [1, 2].

Afterward, researchers shifted their focus to the spinal cord itself, and Greitz [8] proposed the widely accepted intramedullary pulse pressure theory. This theory states that obstruction does not directly force CSF into the spinal cord; rather, it alters the pressure distribution within the cord and the subarachnoid space. Under normal circumstances, the pulse pressure in both the spinal cord and subarachnoid spaces is equal, preventing dilation or compression of the cord. However, when the subarachnoid space is blocked, the CSF pulse pressure in the subarachnoid space cannot be transmitted to distally but rather to the spinal cord, resulting in elevated intramedullary pressure compared to the subarachnoid space. This pressure gradient facilitates the movement of fluid from the capillaries into the spinal cord via the extracellular fluid, leading to the formation of a syrinx primarily composed of extracellular fluid derived from the spinal cord's microcirculation, rather than CSF as previously assumed. Plenty of studies have found that patients with syringomyelia often present with obstructions in the subarachnoid space and alterations in CSF dynamics, thereby providing further support to this theory [9–13].

The intramedullary pulse pressure theory provides a framework for understanding various etiologies of syringomyelia, including Chiari malformation, spinal cord injury and neoplastic lesions. However, it presents challenges in elucidating cases of syringomyelia that occur in the absence of clear cerebrospinal fluid circulation obstruction, such as those associated with tethered cord syndrome. Although some studies have observed CSF obstruction in TCS patients, it appears to be a consequence of subarachnoid space compression following syrinx formation, rather than a primary causative factor, since the thickened filum terminale does not directly block CSF circulation in the same manner as congenital malformations or neoplastic lesions [2, 14]. To account for this phenomenon, Milhorat [14] proposed that tethering may directly contribute to syringomyelia by inducing ischemia in the distal spinal cord through mechanical traction and by altering CSF pulse pressure due to decreased compliance of the subarachnoid space. Ng and Seow [15] documented a case of TCS preceding the formation of a syrinx, which subsequently resolved following un-tethering surgery. They posited that tethering serves as a primary cause of syringomyelia by impairing regional spinal blood flow and oxidative metabolism. Erkan [4, 16] also suggested that the severity of tethering, impaired CSF flow, and vascular insufficiency are critical components in the pathophysiology of syringomyelia.

In addition to mechanisms such as ischemia and hypoxia, Bertram [17] has proposed a new hypothesis based on the fluid–structure interaction model. This hypothesis suggests that the tension exerted by the filum terminale on the cord can be separated into two types: axial and radial, with axial tethering being more powerful. The model demonstrates that axial tethering can induce tensile radial stress, which subsequently decreases the fluid static pressure within the cord. This transient reduction in pressure may draw in interstitial fluid, forming a cavity within the previously normal cord tissue. Notably, the closer to the tethered site, the greater the tension. Therefore, the formed cavities are typically longitudinal and situated near the filum terminale, predominantly within the lower third of the spinal cord, a condition referred to as terminal syringomyelia [18–20]. In contrast to Chiari malformation, cases of syringomyelia associated with TCS are less likely to involve the upper segments of the spinal cord, with even fewer documented cases affecting the entire cord. A review of the literature reveals several case reports of holocord syringomyelia in conjunction with TCS, but all of these cases are combined with other associated anomalies (Table 1). For instance, Maheshwari [21] reported a three year old child with a holocord syrinx combined with TCS and diastematomyelia in 2012, but did not provide details regarding subsequent treatment or prognosis. Kemp [22] documented a case of a 14-year-old child with a holocord syrinx, TCS, anterior sacral meningocele and tailgut cyst, noting that postoperative MRI indicated near resolution of the syrinx following the resection of the anterior sacral meningocele and presacral mass. Kapoor [23] described a case presenting with the triad of holocord syringomyelia, tethered cord and Chiari malformation, while Rakip [24] reported a case of syringomyelia with tethered cord associated with the spinal lipoma; in both instances, the syrinx diminished and symptoms improved after untethering surgery.

All aforementioned cases involved pediatric or adolescent patients, primarily exhibiting lower extremity weakness and bladder dysfunction. The differentiation of symptoms attributable to syringomyelia versus those resulting from tethered cord syndrome is challenging due to significant symptom overlap. Notably, upper limb

Table 1 Summary of the characteristics of cases with holocord syringomyelia and tethered cord syndrome reported in the literature

Publisher	Publication time	Country	Gender	Age(y)	Syrinx size	Clinical manifestation	Associated anomalies besides TCS
Maheshwari, et al. [21]	2012	India	Male	3	C1-L5	increasing clumsiness and frequent falls	Diastematomyelia
Kemp J, et al. [22]	2014	America	Female	14	C2-L5	progressive bowel and bladder dysfunction and lower extremity weakness	Anterior sacral meningocele and tailgut cyst
Kapoor A, et al. [23]	2014	India	Male	12	C2-L3	progressive weakness and decreased sensation over both lower limbs and hesitancy and slow stream of urine	Chiari malformation
Rakip U, et al. [24]	2022	Turkey	Male	1.5	C7-T10	Not recorded	Spinal lipoma

symptoms or segmental dissociative sensory disorders, which are more commonly associated with syringomyelia, were absent in these cases, as well as in our case presented herein.

In our case, we systematically ruled out potential etiologies such as Chiari malformation, hydrocephalus, and other associated anomalies through comprehensive medical history inquiry and imaging examination. We ultimately hypothesized that the holocord syrinx is predominantly attributable to the tethered cord. The tensile radial stress exerted by the thickened filum terminale on the distal spinal cord may serve as the initiating factor for the formation of the cavity. With the progressive impairment of regional spinal blood flow and oxidative metabolism, the syrinx gradually expands. As the syrinx enlarges and the filum terminale thickens, the circulation of cerebrospinal fluid becomes obstructed, further promoting the development of the cavity and potentially culminating in a holocord syrinx.

Regarding the management of patients with this condition, there has historically been considerable debate among surgeons concerning the appropriateness of performing syrinx shunting or drainage in conjunction with un-tethering surgery [1, 16, 25]. However, an increasing number of clinical studies have shown that un-tethering surgery alone can yield significant therapeutic benefits, with syrinx shunting or drainage being reserved for cases where un-tethering proves ineffective [26–30]. In our case, we performed un-tethering surgery and achieved satisfactory outcomes. Postoperatively, the patient experienced substantial symptom relief, and follow-up MRI indicated the resolution of the syrinx, thereby indirectly supporting our etiological hypothesis. Nonetheless, the inherent limitations of retrospective studies and the relatively brief one-year follow-up period necessitate further multicenter randomized controlled trials to validate the safety and efficacy of un-tethering surgery.

The pathophysiological mechanisms underlying syringomyelia associated with tethered cord syndrome remain incompletely elucidated. Based on this case, we speculate that the tensile radial stress from the filum terminale may lead to a reduction in fluid static pressure within the spinal cord, draw in extracellular fluid and form a syrinx. Ischemia and hypoxia conditions at the end of the spinal cord, as well as impaired cerebrospinal fluid circulation, may further exacerbate the expansion of the syrinx and form a holocord syrinx ultimately. For such patients, it appears that un-tethering surgery may suffice to alleviate symptoms, reduce cavity size, and achieve satisfactory clinical outcomes.

Abbreviations

CSF Cerebrospinal fluid
MRI Magnetic resonance imaging
TCS Tethered cord syndrome

Acknowledgements

Not applicable.

Authors' contributions

ZLT contributed to the collection of medical record data and drafted the manuscript. LZZ contributed to the collection of relevant literature. DHZ contributed to the collection of medical record data and revised the manuscript. All authors read and approved the final manuscript.

Funding

This work was supported by grants from the Interdisciplinary Research Program of Peking University First Hospital (2023IR15).

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient/parent/guardian/ relative of the patient. A copy of the consent form is available for review by the Editor of this journal.

Competing interests
The authors declare no competing interests.

Received: 12 May 2024 Accepted: 6 November 2024
Published online: 11 November 2024

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