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Technical Notes

Tethered cord syndrome in patients with myelomeningocele: Presentation of 3 cases, technical note on re-anchoring without dural opening

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ABSTRACT

Background: Following myelomeningocele (MMC) repair, 10-30% of patients develop tethered cord syndrome (TCS). Surgical intervention is critical to reverse the stretching of the spinal cord. Here, we describe a technique for spinal cord untethering without dural opening in these patients.

Methods: Three patients underwent spinal cord untethering without dural opening. The surgical technique involved reopening the previous incision and dissecting the scar tissue attached to the dura. A Spongostan sponge was inserted, and lateral sutures were placed between the dural sac and the adjacent muscles. Clinical outcomes, imaging findings, and urodynamic results were evaluated postoperatively.

Results: The technique demonstrated positive outcomes in all three cases. Patients showed symptom improvement, better positioning of the spinal cord on imaging studies, and enhanced bladder function on urodynamic evaluations.

Conclusion: Spinal cord re-untethering without dural opening may be a viable surgical option for selected patients with MMC, offering favorable outcomes with reduced risk.

Keywords: Cord untethering, Myelomeningocele, Tethered cord syndrome

INTRODUCTION

Myelomeningocele (MMC) is a complex congenital defect resulting from incomplete closure of the neural tube requiring surgical repair in the first hours of life.[1] Following MMC repair, approximately 10–30% of the patients will develop tethered cord syndrome (TCS). TCS is caused by stretching in the final portion of the cord due to tissue attachments to the dural sac.[2,11] Signs and symptoms of TCS include lower back pain, urological dysfunction, increased sensitivity to catheterization or changes in urodynamics, scoliosis, and lower-limb deformity.

Surgically reversing the stretching of the spinal cord prevents irreversible damage to the spinal cord.[13] We describe a technique for spinal cord untethering without opening the dura mater in patients with MMC and TCS.

METHODS AND RESULTS

Three cases of patients with MMC and TCS who underwent spinal cord untethering without opening the dura were selected. The technical details of the intervention are described. Signs and symptoms, magnetic resonance imaging (MRI) findings, and pre and postoperative urodynamic studies were evaluated.

Case 1

The patient was a 6-year-old male with a history of MMC functionally classified as grade II.[10] Closure of the defect was performed 24 hours after birth, and the patient required a ventriculoperitoneal shunt (VPS) at 11 days of life. From the age of 3 years, the child developed progressive deformity of the right foot with marked equinus varus, associated with a neurogenic bladder with an abnormal urodynamic study, pain on intermittent catheterization, and presence of syringomyelia at the lumbar level in the MRI [Figure 1]. Surgical extradural spinal cord untethering was performed. Postoperatively, the patient showed significant improvement on urodynamic testing, no pain on catheterization, and no progression of the lower-limb deformity.

Case 2

The patient was a 12-year-old female with a history of MMC grade IV. Closure of the defect was performed at 1 day of life. A VPS was placed at 1 month of life. At the age of 11 years, she started with pain at the lumbar level, and she developed an equinus valgus deformity of the left lower limb. The urodynamic evaluation showed hydronephrosis, increased bladder capacity for age, and detrusor muscle hyperactivity. Based on these findings, the decision was made to proceed with extradural cord untethering [Figures 2 and 3]**.** Postoperatively, at 2 years of follow-up, her symptoms improved, and the urodynamic study showed increased bladder capacity but no detrusor overactivity. The follow-up MRI showed significant improvement compared to the preoperative MRI.

Case 3

The patient was a 10-year-old female with a history of MMC grade IV. Closure of the defect was performed at 6 h of life. VPS was required at 1 month of life. She presented with two distal dysfunctions. She started with pain at the lumbar scar and discomfort during urinary catheterization associated with a progressive gait disturbance. The urodynamic evaluation showed increased bladder capacity with

Figure 1: (a) Preoperative sagittal view of the lumbosacral magnetic resonance imaging (MRI) showing the descent of the spinal cord and tethered cord (red arrow) associated with syringomyelia (yellow arrow) and neurogenic bladder (white arrow). (b) Preoperative axial slice MRI showing evidence of lumbar syringomyelia. (c) The deformity of the lower limbs is observed with marked equinus varus of the right foot. (d-f) Intraoperative images of extradural detachment. The dural sac detached from the subcutaneous plane (white arrow), the dural sac retained at the intracanal level with passing silk sutures (yellow arrows), and the closure of the fascial plane (red arrow) is shown. (g) Lumbosacral MRI postoperative sagittal view reflecting greater space at the lumbar extradural level and reduction of the intracanal cyst as signs of satisfactory unanchoring (red arrow).

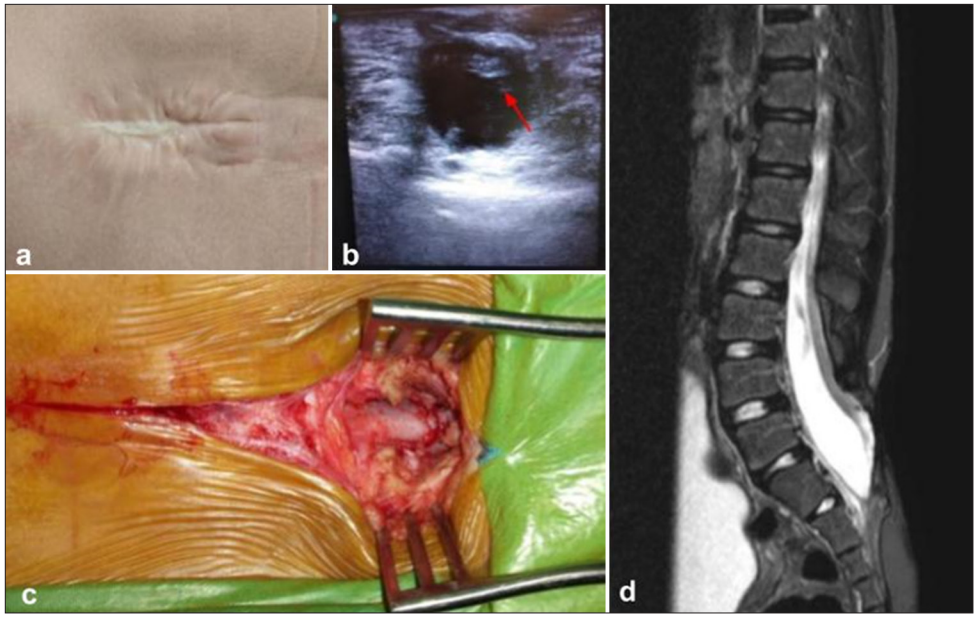

Figure 2: (a) Image of the scar from the patient's first surgical intervention. (b) Ultrasound performed in surgery prior to untethering showed hyperechogenic tissue (marrow) in relation to the overlying scar (red arrow). (c) Intraoperative image of tissue dissection. (d) Preoperative magnetic resonance imaging sagittal cut in T2 sequence showing the dural sac and the spinal cord that protrudes dorsally in close relation to the scar tissue.

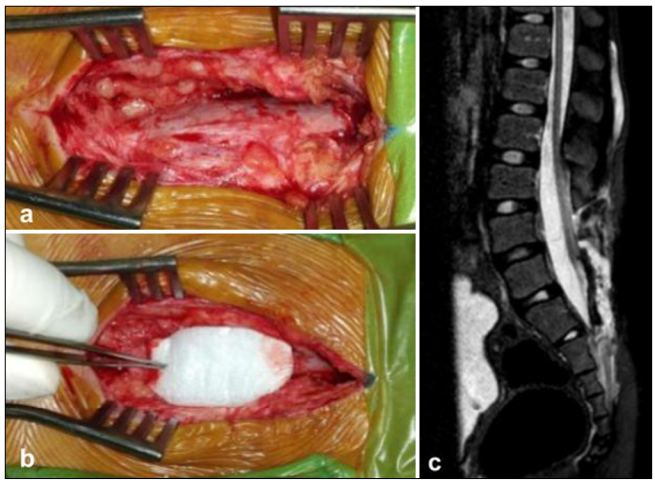

Figure 3: (a) Intraoperative image where the entire dissected dural sac can be seen. (b) Intraoperative image showing the spongostan plate that was placed after the dissection. (c) Magnetic resonance imaging in sagittal section T2 sequence in which improvement of the protrusion of the spinal cord and dural sac is observed in comparison to the preoperative period.

preserved accommodation and areflexia with an elevated leak point pressure requiring catheterization for voiding. Spinal cord MRI showed a tethered cord associated with protrusion of the dural sac posteriorly at the level of L5-S1 [Figure 4]. Extradural cord untethering was performed. Postoperatively, the pain at the lumbar scar diminished, and discomfort during urinary catheterization and gait improved.

For the surgical intervention, the patient was placed in a prone position with pads placed on the pressure points. The previous incision was reopened. The scar tissue adhered to the dura was dissected. To avoid retethering and dorsal displacement of the dural sac, a spongostan plate was inserted, and lateral stitches were placed between the sac and the adjacent muscles. Subsequently, the overlying tissues were closed. Intraoperative neurophysiological monitoring was used in all cases.

The technique of spinal cord untethering without dural opening showed positive results in all three cases. Significant improvement of the symptoms was observed and imaging studies showed an improvement in spinal cord position. In addition, urodynamic evaluation revealed improved bladder function in all cases. The patients had no postoperative complications and were discharged within 72 hours.

DISCUSSION

MMC is a complex congenital defect characterized by an incomplete closure of the neural tube during fetal development. The surgical repair of the defect consists of neurulation of the placode, dural closure, and subsequent musculoaponeurotic system and skin closure. Adhesions that form between the repaired dural sac and the overlying tissue cause stretching of the cord in some patients. TCS is a progressive disorder that results in orthopedic, urologic, and neurological dysfunction.[5,4,11]

Patel *et al*.[11] were the first to investigate in cadavers whether an extradural section of the filum terminale was possible. In four of the five specimens studied, there was minimal

movement of the intradural filum terminale, and, therefore, the authors concluded that the technique was unlikely to have a significant effect.[7] The main limitation of this study is that the analysis was conducted on cadavers. While this provides valuable insights, it does not account for the potential clinical impact, as even minimal spinal cord movement in living patients could lead to symptom improvement.

In recent years, there has been increasing interest in less invasive surgical procedures to avoid the postoperative complications associated with the traditional technique.[3,6] Based on this concept, we propose a surgical approach to spinal cord untethering without dural opening in MMC patients [Figure 5]. The technique was performed in three selected cases in which the MRI showed that the dorsal side of the sac with the attached conus was adhered to the outer part of the canal generating stretching and microtrauma to the conus and epiconus. In all cases, there was improvement in symptoms, MRI findings, and urodynamic pattern.

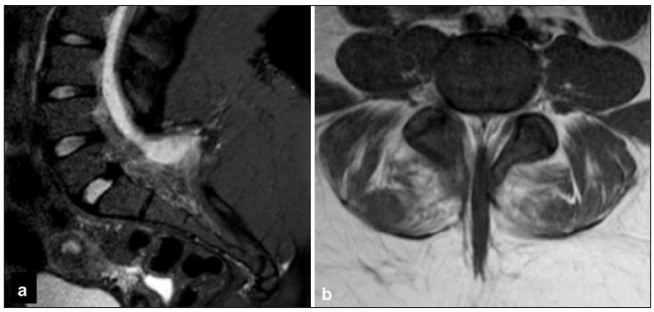

Figure 4: Preoperative magnetic resonance imaging of the lumbar spine. (a) In the T2 sequence, sagittal cut, the tethered cord is observed associated with the emergence of the dural sac projected posteriorly at the level of L5-S1. (b) In the T2 sequence, the axial cut shows protrusion of the dural sac in a dorsal direction and adherence to the subcutaneous cell plane.

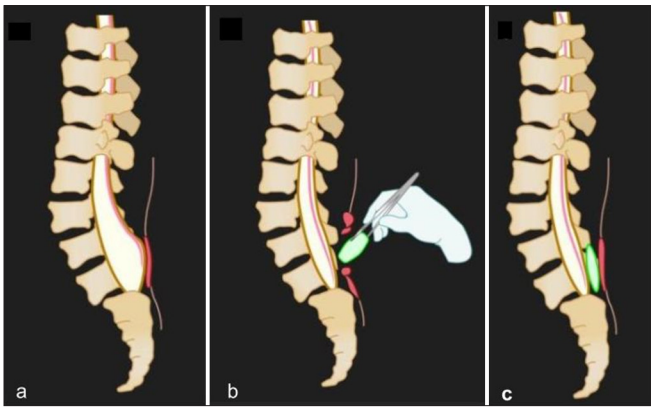

Figure 5: Surgical technique schematic. (a) Preoperative: The dural sac is protruding and tethered to scar tissue, leading to dorsal elongation of the spinal cord. (b) The dural sac is seen freed from the scar tissue (marked in red). By detaching it from the scar tissue, the dura mater is no longer pulled posteriorly, reducing tension on the spinal cord. The placement of the spongostan sheet is shown in green. (c) Final Surgical Image: The dural sac is clearly separated from the scar tissue.

Pre and postoperative urodynamic studies are mandatory, as secondary release surgery has been shown to improve urological outcomes significantly.[14,15] Intraoperative neurophysiological monitoring was used during all procedures since it has been associated with improved long-term outcomes; however, in the future, this modality may no longer be necessary.[4]

The mechanism by which we consider that this technique could be appropriate in these certain patients is because the nervous tissue adheres to the dural sac. We propose that this technique may be appropriate for certain patients because the nervous tissue is adhered to the dural sac, which in turn is attached to the scar tissue from the MMC closure. By dissecting the scar tissue from the dura, dorsal traction on the spinal cord is reduced, potentially alleviating postoperative symptoms.

Advantages of the extradural spinal cord untethering technique include a shorter operative time compared to the traditional technique in which duroplasty is often necessary.[8] In addition, there is a lower risk of complications that could delay hospital discharge, such as central nervous system infection, fistula formation, or cerebrospinal fluid collection.[2,6,9,12] After surgery involving dural opening, progressive decubitus changes are recommended, which may be challenging in pediatric patients. Using the above-described technique, patients are free to move already in the immediate postoperative period. This technique may provide significant clinical improvements in patients with MMC and TSC, alleviating the associated neurological and urological symptoms.

CONCLUSION

Extradural spinal cord untethering appears to be an effective and safe technique for the management of TCS due to cord retethering in patients with MMC. The findings of this study support the use of this technique, showing clinical and functional improvements in the treated patients. Nevertheless, further studies and long-term follow-up are needed to validate the technique and compare it with traditional approaches fully.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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