Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Tethered Cord Syndrome in Association with Chiari Type II Malformation: A Case Report

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DOI: 10.36347/sjmcr.2024.v12i03.024 | **Received:** 02.02.2023 | **Accepted:** 13.03.2024 | **Published:** 27.03.2024

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Abstract **Case Report**

Tethered cord syndrome (TCS) is a rare but potentially debilitating condition that occurs when the spinal cord is abnormally attached to the surrounding tissue. TCS can be associated with Chiari type II malformation (CM-II), a condition where the cerebellar tonsils protrude into the spinal canal. Here we present the case of a 20-year-old female with a history of spinal lipoma and neurogenic bladder who was found to have TCS and CM-II on MRI. The patient underwent surgical untethering with good postoperative outcome.

Keywords: Tethered cord syndrome; Chiari type II malformation; lipomyelomeningocele; spina bifida.

INTRODUCTION

Tethered cord syndrome (TCS) is a neurological disorder caused by abnormal attachment of the spinal cord to the surrounding tissue, resulting in a range of symptoms such as back pain, bladder and bowel dysfunction, and weakness or numbness in the lower extremities. TCS can be congenital or acquired and can be associated with other neurological conditions such as Chiari type II malformation (CM-II) [1].

The aim of this case report is to discuss its clinical presentation, diagnostic evaluation, management, and outcomes.

OBSERVATION

The case report is about a 20-year-old female with a history of spinal lipoma and neurogenic bladder presented with progressive back pain and lower extremity weakness. Upon clinical examination, the patient displayed signs consistent with TCS, including lower limb weakness, sensory disturbances, and urinary dysfunction. Additionally, physical evaluation revealed cerebellar signs, specifically cerebellar ataxia, indicative of the coexisting Chiari type 2 malformation. MRI revealed a low-lying conus medullaris at the level of L5 with associated syringomyelia extending from L2 to L5, as well as a lipomyelomeningocele with intracanalar extension at the level of L5. The patient was also found to have a CM-II with cerebellar tonsillar herniation, and scoliosis with double convexity in the lumbar and dorsal regions without sacro-coccygeal agenesis (Figure 1: images A B C D E F G).

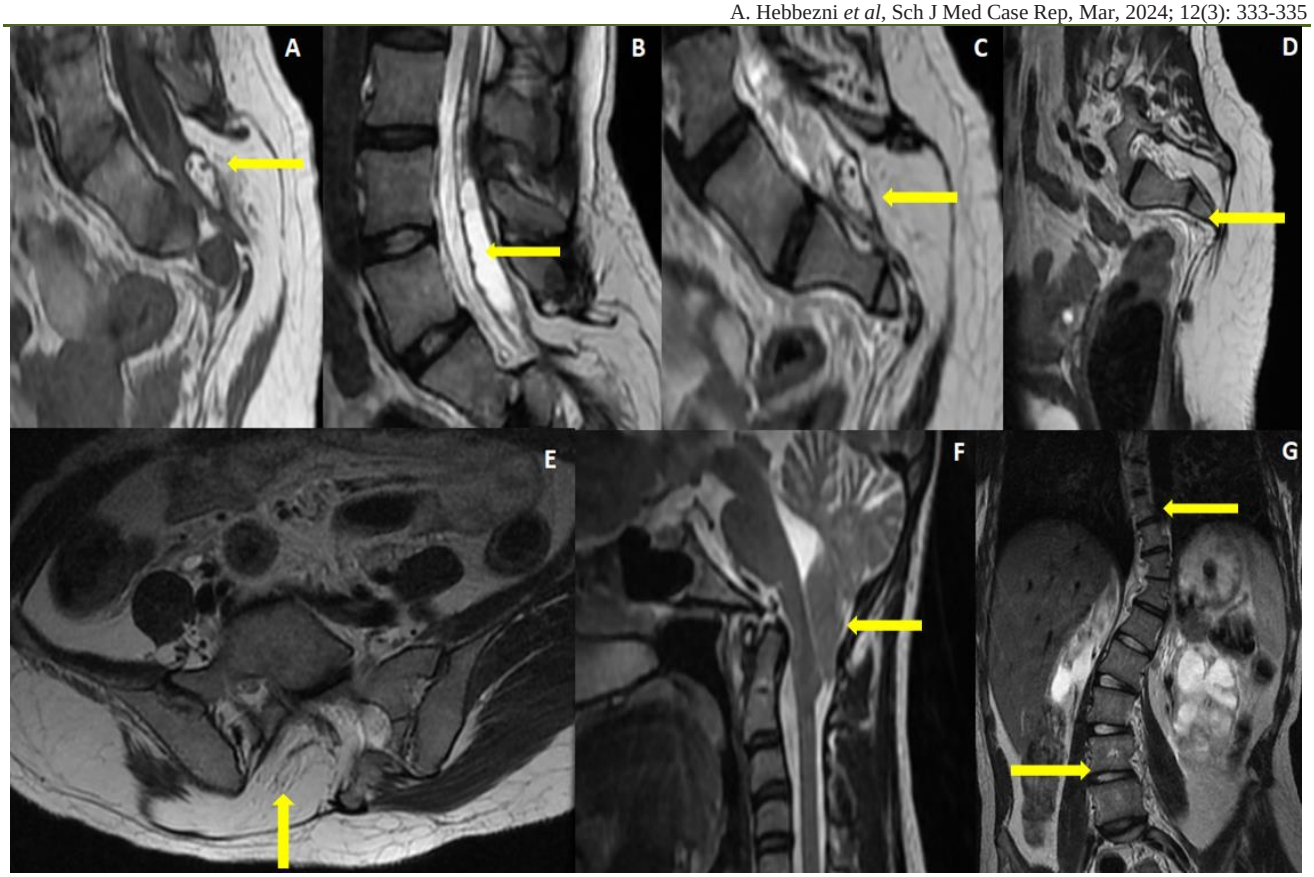

Figure 1: A) Sagittal T1 showing lipomyelomeningocele with intracanalar extension at the level of L5 and low-lying conus medullaris at the level of L5 in T1 (yellow arrow); B) Sagittal T2 showing syringomyelia extending from L2 to L5 in T2 (yellow arrow); C,D,E) Sagittal and axial T2 showing failure of closure of the posterior arch at the level of L5 with visualization of a poorly defined intracanalicular and posterior parietal lesion with fatty signal, encompassing the roots of the cauda equine without sacro-coccygeal agenesis (yellow arrow); F) Sagittal T2 showing Chiari type 2 malformation with cerebellar tonsillar herniation (yellow arrow); G) Coronal T2 showing scoliosis with double convexity in the lumbar and dorsal regions (yellow arrow)

DISCUSSION

Tethered cord syndrome in association with Chiari type II malformation is a rare estimated at 0,72% but well-documented entity that presents unique diagnostic and management challenges. Early diagnosis and surgical intervention are crucial in preventing permanent neurological damage [2].

This case highlights the association between Chiari II malformation and tethered cord syndrome. Chiari II malformation is a congenital disorder of the central nervous system characterized by the downward displacement of the cerebellar tonsils and medulla through the foramen magnum into the cervical spinal canal. It is commonly associated with myelomeningocele, hydrocephalus, and other central nervous system abnormalities [3].

Tethered cord syndrome, on the other hand, refers to a group of neurological disorders caused by an abnormal attachment of the spinal cord to the surrounding tissues. This abnormality can cause spinal cord tension and stretching, leading to a wide range of symptoms such as back pain, leg pain, and muscle weakness [4].

In this case, the patient had a lipomyelomeningocele with tethered cord syndrome in addition to the Chiari II malformation. The lipomyelomeningocele is a form of spina bifida that involves a fatty mass in the spinal canal, often associated with a tethered spinal cord. The combination of these two conditions can cause neurological deficits that range from mild to severe.

Magnetic Resonance Imaging (MRI) plays a pivotal role in the comprehensive evaluation of patients presenting with tethered cord syndrome (TCS) concomitant with Chiari type 2 malformation. This non-invasive imaging modality provides invaluable insights into the intricate anatomical complexities of this dual pathology. Our study demonstrates that MRI not only confirms the presence of TCS through the identification of a low-lying conus medullaris and a thickened filum terminale but also elucidates the extent of cerebellar tonsillar herniation characteristic of Chiari type 2 malformation. The multi-planar capabilities of MRI

allow for precise localization and measurement of spinal cord tethering, aiding in both diagnosis and surgical planning. These findings underscore the crucial role of MRI in the comprehensive assessment of patients with tethered cord syndrome and Chiari type 2 malformation, ultimately contributing to more informed clinical decision-making and improved patient outcomes [1].

The management of TCS associated with CM-II requires a multidisciplinary approach involving neurosurgeons, radiologists, and urologists. Surgery is the primary treatment modality, with the goal of releasing the tethered spinal cord and decompressing the cerebellar tonsils. The timing and extent of surgical intervention depend on several factors, including the severity of symptoms, degree of tethering, and presence of associated anomalies [5, 4].

There is also ongoing debate regarding the role of prophylactic surgery in patients with CM-II without symptoms or signs of TCS. Some studies suggest that prophylactic surgery may prevent the development of TCS and associated neurological deficits, while others caution against unnecessary surgical intervention [2].

Several studies have reported favorable outcomes following surgical untethering in patients with TCS and CM-II. However, the long-term prognosis depends on various factors such as the degree of preoperative neurological deficits, the presence of associated anomalies, and the extent of surgical intervention [6].

CONCLUSION

Tethered cord syndrome in association with Chiari type II malformation is a rare but important entity that requires early diagnosis and surgical intervention to prevent permanent neurological damage. The management of this condition should be individualized based on the patient's symptoms, radiological findings, and associated anomalies.

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