

Chiari type I malformation with occult tethered cord syndrome in a child

A case report

Yuan Zhou, MD, Lin Zhu, MD, Yixing Lin, MD, Huilin Cheng, MD, PhD*

Abstract

Rationale: Chiari type I malformation (CM1) and occult tethered cord syndrome (OTCS) are considered to be malformations associated with subtle structural abnormalities of the terminal filum. Few studies have reported patients with CM1 and OTCS. Treatment strategy for patients of CM1 associated with OTCS is controversial.

Patient concerns: A 14-year-old child was admitted with intermittent pain and numbness in the right upper limb. And he had urinary frequency, neck pain, back pain, and numbness simultaneously. The imaging examinations showed CM1, syringomyelia, small fat in the filum at the level of the L2 vertebral body but the conus medullaris at the aspect of the L1 vertebral body.

Diagnoses: The child was diagnosed with CM1 associated with OTCS.

Interventions: Patient underwent sectioning of filum terminale (SFT) under electrophysiological monitoring during the first hospital and posterior fossa decompression (PFD) during the second hospital.

Outcomes: After first discharge pain of the right upper limb was relieved, but he still felt numbness. And his numbness was relieved after second discharge. The imaging examinations also showed corresponding improvement during the 2-year follow-up period.

Lessons: For pediatric patients with CM1 and TCS, treatment trouble is not only to choose the staging operation or simultaneous operation but also staging procedures for treatment of 2 lesions. Detailed preoperative evaluation is essential for development of individualized surgical plan. Staging operation of firstly minimally invasive SFT and later PFD may be helpful for such cases owing to its positive effect on both the symptoms and imaging findings.

Abbreviations: CM = Chiari malformation, CM1 = Chiari type I malformation, OTCS = occult tethered cord syndrome, PFD = posterior fossa decompression, SFT = sectioning of filum terminale, TCS = tethered cord syndrome.

Keywords: Chiari malformation, filum terminale, occult tethered cord syndrome, pediatric, posterior fossa

1. Introduction

Chiari malformation (CM) is a kind of malformation characterized by cerebellar tonsil herniation into the foramen magnum, of which Chiari type 1 malformation (CM1) is the most common, and is often associated with syringomyelia.^[1] Occult tethered

cord syndrome (OTCS) refers to a clinical syndrome related to tethering of the spinal cord by the filum, but the conus is in normal position.^[2] However, few studies have reported patients with CM1 and OTCS and treatment strategy of CM1 with OTCS is still inconclusive. We report a case of 1 child with the above 2 diseases and review the relevant literature.

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Department of Neurosurgery, Jinling Hospital, Jinling School of Clinical Medicine, Nanjing Medical University, Jiangsu, China.

* Correspondence: Huilin Cheng, Department of Neurosurgery, Jinling Hospital, Jinling School of Clinical Medicine, Nanjing Medical University, 305 East Zhongshan Road, Nanjing, Jiangsu 210002, China (e-mail: huilin_cheng@yahoo.com).

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2. Case report

A 14-year-old child was admitted to our hospital with a more than 1-year history of intermittent pain and numbness in the right upper limb. One year ago, he had an onset of intermittent pain and numbness in the right upper limb without apparent inducement. And he had urinary frequency, neck pain, back pain, and numbness simultaneously. His symptoms were refractory to medical management. He had no lower limb numbness, pain, weakness, muscle atrophy, perineal numbness, or stool dysfunction. His physical examination indicated that the tactile, pain, and temperature sense of the right torso decreased. The deep sense was normal yet. Right Babinski sign was positive. Double Hoffman and left Babinski signs were negative. Cervical and thoracic magnetic resonance imaging (MRI) (Fig. 1A) showed CM1 and syringomyelia. Lumbar MRI (Fig. 1B, C Fig. 1) in our hospital pointed out small fat in the filum at the level of the L2 vertebral body but the conus medullaris at the aspect of the L1 vertebral body. Urodynamic testing indicated neurogenic bladder. Patient underwent sectioning of filum terminale (SFT)

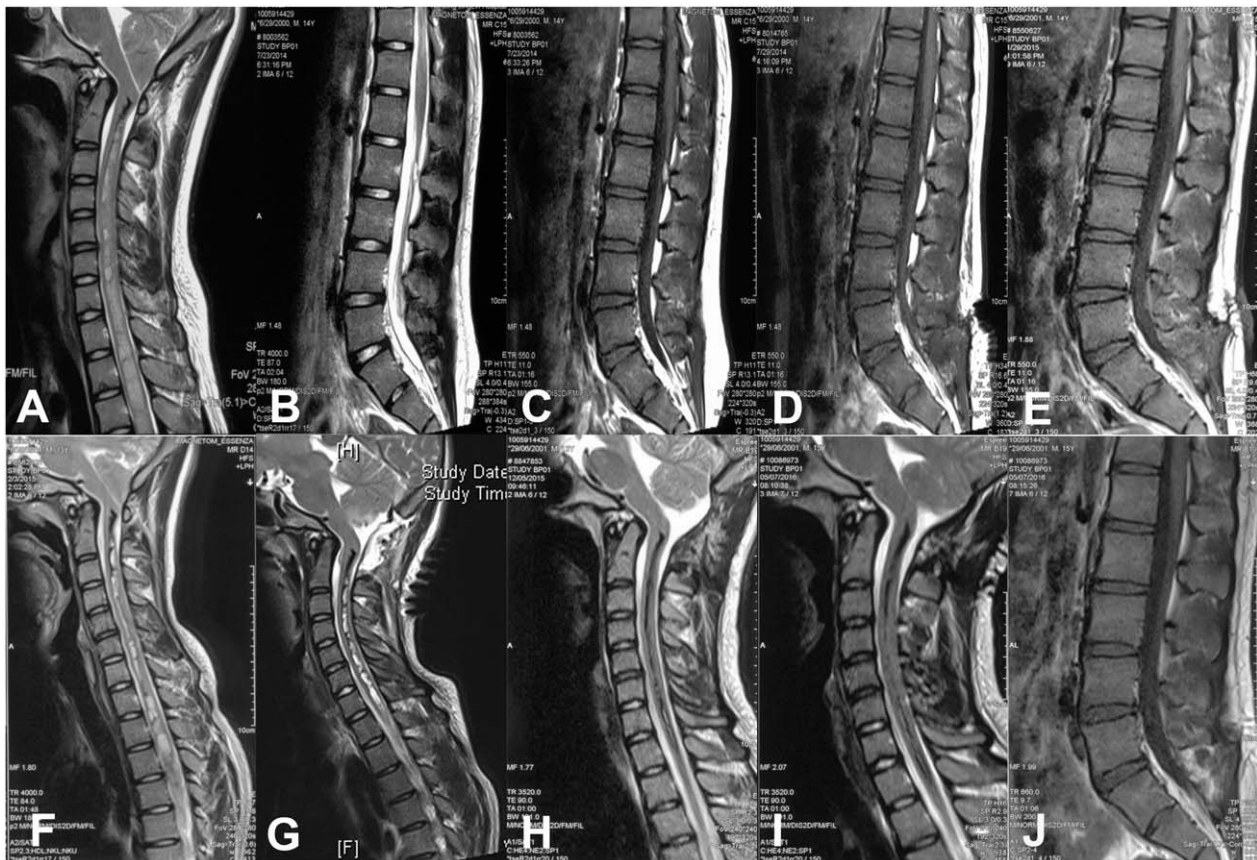


Figure 1. MRI demonstrating the evolution and resolution of Chiari type 1 malformation (CM1), syringomyelia, and the fat in the filum, (A) Scan of cervical and thoracic vertebrae performed on first admission demonstrating CM1 and syringomyelia, (B) T2 imaging of lumbar vertebrae performed on first admission demonstrating fat in the filum at the level of the L2 vertebral body but the conus medullaris at the aspect of the L1 vertebral body, (C) T1 imaging performed on first admission demonstrating fat in the filum at the level of the L2 vertebral body, (D) Four days post first operation demonstrating neither fat nor the position of conus medullaris had changed, (E) Half a year post first discharge demonstrating no fat but the position of conus medullaris was unchanged, (F) Cervical and thoracic MRI performed on second admission demonstrating unchanged CM1 and less syringomyelia than previous scan, (G) Four days post second operation demonstrating a good retraction of cerebellar tonsil, no significant change in syringomyelia, (H) Half a year post second discharge demonstrating no CM1 and significantly reduced syringomyelia, (I) Eighteen months post second discharge demonstrating no CM1 and further reduction of syringomyelia, (J) Eighteen months post second discharge demonstrating no fat and conus medullaris in normal position.

under electrophysiological monitoring within 4 days after initial admission. During operation, a thickened fatty filum was detected and cut. Pathology showed hyperplastic fibers and adipose tissue. After symptomatic treatment, pain of the right side of the neck, shoulder, and back was relieved, while both urinary dysfunction and urodynamic testing had improved. Four days after operation, MRI (Fig. 1D) showed that neither fat nor the position of conus medullaris had changed. Half a year after discharge, review of MRI (Fig. 1E) showed the fat in the filum radiographically vanished, but the position of conus medullaris was unchanged.

After first discharge, he still felt numbness of right upper limb and right back, while occasionally waist pain. He was thus readmitted 192 days after SFT. Cervical and thoracic MRI (Fig. 1F) showed unchanged CM1 and less syringomyelia than previous scan. Patient underwent posterior fossa decompression (PFD) within 2 days after second admission. During operation, we chose subpial cerebellar tonsil resection combined with dural expand repair. We loosened the arachnoid adhesion, dredged the fourth ventricle, the cisterna magna, and the cisterna magna. Four days after operation, MRI (Fig. 1G) showed a good retraction of cerebellar tonsil, no significant changes in

syringomyelia. Half a year after second discharge, numbness was relieved and a review of MRI (Fig. 1H) showed significantly reduced syringomyelia. Eighteen months after second discharge, review of MRI (Fig. 1I, J Fig. 1) showed further reduction of syringomyelia, no fat and conus in normal position. All procedures performed in studies involving human participant were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from the patient included in the study.

3. Discussion

In a narrow sense, CM is ectopia or displacement of the cerebellar tonsil, while in a broad sense, it is a series of cranial cervical junction abnormalities including cerebellar tonsillar herniation.^[1] About 50% to 70% of CM1, the most common of the CMs, were accompanied by syringomyelia.^[3,4] CM1 is typically picked up in later childhood and even in adulthood, and can occasionally radiographically change over time. Milhorat et al^[5] reported that about 14% of CM1 patients were associated with

TCS, and spinal cord traction was considered to be one of the pathological mechanisms of CM1. TCS, defined as a clinical constellation of tethered cord symptoms, is also not typically congenital, with or without associated radiographic findings of tethered cord, but rather typically develops during childhood growth spurts.^[2] OTCS is a recently defined entity, diagnosis of which is dependent on clinical examination supplemented by urodynamic or functional studies.^[2] Neurogenic bladder is the most common symptom of OTCS.

Management of OTCS is still controversial. Surgical treatment such as SFT is preferred for patients presented with urologic dysfunction by some studies and the rate of improvement for urinary symptoms has varied from 60% to 97%.^[6,7] However, recent randomized, controlled pilot study seemed to demonstrate no objective difference in urological outcome between medical management plus or minus filum section for patients with OTCS.^[8] PFD is currently the most important operation for CM1. However, as far as CM1 pediatric patients with OTCS are concerned, as a result of 2 lesions need surgery, treatment trouble is not only to choose the staging operation or simultaneous operation, but also staging procedures for treatment of 2 lesions.^[9] Some studies demonstrated that it was feasible to perform both PFD and SFT procedures safely during the same operation.^[9] But it was necessary to assess the operation opportunity. Other studies have reported that there was clinical improvement in patients with CM1 after SFT.^[10] They concluded that a tethered spinal cord affects coordinated spinal column growth, resulting in compensatory growth mechanisms that lead to CM1 and advocated SFT as the first choice for the treatment of CM1 patients with TCS. And some studies found that OTCS may be involved in the formation of CM1 and children with a fatty or thickened filum terminale demonstrated a greater amount of tonsillar displacement.^[10]

The patient's presenting symptoms were representative of both CM1 and OTCS. He was young and growth was not complete. It was so difficult to determine the operation mode. On the basis of above conditions and opinions of his family, we first chose the minimally invasive surgical approach—SFT to treat OTCS. Fortunately, the symptoms of urinary dysfunction and pain were relieved after SFT earlier, but numbness still existed. Nearly 192 days post first operation, PFD was performed. The facts proved that this staging operation for the child was available due to its effect. In addition, we found a strange phenomenon that the small fat in the filum disappeared by itself after SFT. It is easy to understand how a thickened filum may exert tension on the conus. We conclude that after SFT abnormal tension on the conus

by the filum would be relieved, then small adipose tissue in the filum might be dispersive and shift toward the periphery followed by terminal displacement. The real cause is still unknown.

4. Conclusion

Our case shows the difficulty in determining the causal relationship between radiographic findings and clinical symptoms about CM1 and OTCS. Staging operation may be a good choice, although first SFT and later PFD are available for children with CM1 and OTCS. Of course, detailed preoperative evaluation, which is very important for treatment of children with CM1 and OTCS, includes whether there are brainstem symptoms caused by CM1 and spinal cord symptoms associated with OTCS, whether preoperative MRI has abnormal performance. Similarly, patient's physical status, family status, and family opinions are especially important. Only in this way can individualized surgical plan be developed.

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