Reversal of longstanding neurological deficits after a late release of tethered spinal cord

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The importance of early surgery for tethered cord syndrome in the pediatric population is well established. Optimal treatment and prognosis of tethered cord in adults, on the other hand, is less clear. Some advocate a conservative approach in asymptomatic patients, while others recommend early detethering in all patients. For symptomatic patients, however, there is a consensus in favor of early surgery to prevent progression of neurological deficit. Many studies have reported cessation of neurological decline or reversal of recently acquired neurological deficits in patients with adult tethered cord syndrome. There are limited data in the literature about late surgery for the treatment of tethered spinal cords when the neurological deficits are longstanding. We report on a 37-year-old woman who demonstrated dramatic neurological improvement after surgical release of a tethered spinal cord more than 20 years after the onset of progressive neurological deficits. (DOI: 10.3171/2010.3.FOCUS1078)

KEY WORDS • spinal cord • adult tethered cord • late detethering • neurological improvement

EARLY surgery for TCS in the pediatric population is standard,1,10 optimal treatment and prognosis of TCS in adults is, however, unclear. Some advocate a conservative approach in asymptomatic patients, while others recommend early detethering even in all patients.5,17,19 For symptomatic patients, however, there is a consensus advocating early surgery to prevent progression of neurological deficit.5,13,14,16,17,21,24 Many studies have reported cessation of progressive neurological decline or even reversal of recently acquired neurological deficits in adult patients with TCS.1,4,11,15 There are limited data in the literature about late surgery for the treatment of tethered spinal cords when the neurological deficits are longstanding. We report on a 37-year-old woman who experienced dramatic neurological improvement after surgical release of a tethered spinal cord more than 20 years after the onset of progressive neurological deficits.

Case Report

History and Presentation. This 37-year-old Caucasian woman, in whom a diagnosis of Charcot-Marie-Tooth (CMT) hereditary neuropathy was established, was referred for evaluation of longstanding and progressive neurological deficits including incontinence of urine.

Her early motor development was notable only for a delay in walking until 18 months of age, but subsequent motor development and function appeared normal until high school. At 16 years of age, the patient was referred for orthopedic and podiatric care. She underwent corrective surgery for a toe deformity, but no diagnosis was offered. By 20 years of age, she had bilateral foot drop. She received a diagnosis of CMT hereditary neuropathy and was treated with bilateral ankle foot orthoses. At age 25, she underwent multiple surgical procedures including tendon transfers, Achilles tendon lengthening, and wedge osteotomies.

Urinary incontinence, including nocturnal enuresis, began at age 20 years and progressed in severity despite treatment with anticholinergic medications and a sling procedure. Other problems included progressive numbness over the plantar aspect of both feet, decreased sensation over the left buttock, perianal numbness, and a feeling of being “off balance.” At age 32 years, MR imaging of the brain and entire spine was performed to rule out multiple sclerosis. The only abnormality noted in the report was a tethered cord. The patient was not informed of this finding, and no further evaluation or treatment was advised.

At 36 years of age, nerve conduction studies in the
lower limbs showed evidence of axonal neuropathy, which was suggestive of CMT type 2. In light of her clinical presentation, particularly the urinary incontinence and lack of upper extremity involvement, the diagnosis of CMT type 2 was questioned and she was referred to our institution for management of her progressive neurological deficit and further evaluation.

**Examination.** On physical examination, the patient was 5 feet 6 inches tall and weighed 170 pounds. Examination of the skin demonstrated a 0.7-cm freckle left of the midline in the gluteal cleft. No dermal sinuses or hair tufts were visible. There was a hyperlordotic lumbar curve, and no pain was elicited with back extension maneuvers. Motor examination using the Medical Research Council scale revealed weakness only in the lower limbs: the iliopsoas and quadriceps were 5/5 bilaterally, ankle dorsiflexors 0/5 on the right and 3/5 on the left. The plantar flexors and extensor hallucis longus muscles were 0/5 bilaterally. The patient’s tendon reflexes were hypoactive at the knee and absent at the ankle. Her calf diameters were both 34 cm. In the left S-1 distribution sensory examination was diminished to pinprick and light touch. No deficits were detected in either upper or in the right lower limb. Joint position sense and vibration sensation were decreased on the left as well. The Romberg test was positive. The patient had a “steppage” gait and inability to walk on her toes or heels.

Urodynamic studies with bladder EMG revealed neurogenic bladder dysfunction and a small bladder capacity. There was detrusor overactivity and possible intermittent detrusor sphincter dyssynergia with normal upper tracts. There was significant cough-induced bladder overactivity with evidence of stress incontinence and nonrelaxation with voiding.

A CT scan demonstrated a Grade I isthmic spondylolisthesis at the L5–S1 level. Magnetic resonance imaging showed the spinal cord to be tethered at S1–2; it terminated in a lipoma at the S-2 level (Fig. 1). Comparison with the previously obtained spine MR images showed no appreciable change. Electromyography of the lower extremities showed evidence of chronic bilateral lumbosacral radiculopathy at L5–S1. Nerve conduction velocity was abnormal only in the lower limbs, with markedly diminished amplitude of compound muscle action potentials and no response to stimulation of the left tibial nerve. Left sural amplitude was diminished as well.

The clinical, electrophysiological, and radiological features were consistent with and attributable to a diagnosis of symptomatic tethered spinal cord.

**Operation and Postoperative Course.** Surgical treatment involved S1–2 laminectomies, intradural exploration with untethering of the spinal cord, sectioning of the fatty filum terminale with intraoperative EMG monitoring, and subtotal resection of the intradural lipoma (Fig. 2). The extent of the lipoma resection was dictated by intraoperative EMG findings. Pathological examination showed adipose tissue consistent with lipoma encasing a small portion of the filum terminale. The patient had an uneventful postoperative recovery. Postoperative MR imaging showed untethering of the spinal cord, resection of the fatty filum, and subtotal resection of the lipoma (Fig. 3).

Three months postoperatively, the patient reported an increase in sensation over the posterior thighs and perineum. Nocturnal incontinence had resolved completely. Postoperative motor examination was noteworthy for 5/5 power in bilateral proximal lower extremities and in the left distal lower extremity (dorsiflexion and plantar flexion). Right lower extremity power and extensor hallucis longus strength remained 0/5. Sensory examination improved throughout the left lower extremity and perineum but was not normal as compared with the opposite side.

Additionally, the patient reported marked improvement in sacral sensation, menstrual awareness, and sexual feeling.

**Discussion**

Tethered cord syndrome is a congenital syndrome resulting in mechanical rostrocaudal traction on the spinal cord and variable neurological deficits. It is a part of the constellation of disorders related to a defective retrogressive differentiation of the caudal neural tube.

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**Fig. 1.** Left: Preoperative axial T2-weighted MR image at the L-5 pedicle level demonstrating a bony defect dorsally with enlargement of the thecal sac and dorsal location of the spinal cord. Right: Preoperative sagittal T1-weighted MR image showing a low-lying spinal cord terminating in a lipoma at the S1–2 level (arrow).
Approximately 75% of pediatric patients with TCS have progressive neurological deficits. These may include radicular pain, weakness, asymmetrical hyporeflexia, spasticity, sensory changes, and sphincter dysfunction. The upper motor neuron symptoms are believed to result from mechanical rostrocaudal traction and ischemic damage to the cord. The goal of a detethering surgical procedure is to release the conus from the abnormal filum terminale, which anchors it distally. If the procedure is performed early, excellent neurological recovery is expected in sphincter function, pain, motor deficits, and sensation. Preferably, surgery is performed before the onset of fixed or irreversible neurological deficits. Objective improvements in blood flow and nerve conduction velocities have been reported after early surgical untethering of the spinal cord. The surgery itself is an effective procedure associated with low morbidity and prevents long-term morbidity resulting from the natural progression of the disease process.

In adult TCS, the late onset of symptoms is attributed to repeated microtrauma during flexion and extension of a mechanically tethered spinal cord. A traumatic event that acutely stretches the conus may cause acute deterioration or may even precipitate initial presentation in previously undiagnosed adults. Studies have shown that detethering in an adult population may relieve pain and improve sphincter function.

Several features in our patient’s history and findings on physical examination were not compatible with her diagnosis of an axonal form of CMT, including sparing of the hands, bulky calf muscles, saddle hypesthesia, neurogenic bladder, and asymmetrical deficit in the lower extremities.

Several studies of adult TCS have shown that the potential for reversal of upper motor neuron dysfunction can be poor once neurological symptoms are detected. Lee et al. reviewed several large studies looking at the natural history and long-term outcome. They found that detethering was clearly beneficial when symptoms were of recent onset. Similarly, Aufschnaiter et al. published a case report of an excellent outcome in a patient with symptoms and objective MR imaging diagnosis of tethered cord 1 year prior to surgery. After a literature review of more than 450 patients, these authors concluded that excellent results are generally achieved in adult patients with symptoms 1 year or less in duration.

The neurological deficits that reversed in our patient...
were present for more than 20 years, and there was MR imaging evidence of a tethered cord 5 years prior to surgery. This report challenges the usual presumption that late definitive surgery may not be as beneficial in such cases. The fact that the symptoms remained progressive may be the clue to reversibility. Given this experience and that described in other anecdotal reports, we propose that in chronic cases, detethering more than 5 years after symptom onset may result in significant improvement, particularly when progression has continued.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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