Child's Nervous System
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CASE REPORT

Occult tethered cord syndrome: a reversible cause of paraparesis not to be missed

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Abstract

A 15-year-old female former gymnast with a history of pectus excavatum was reviewed due to unexplained paraparesis and urinary incontinence since age 10. Symptoms were commenced with intolerable upper back pain and development of a soft mass at the sacrum that remitted spontaneously. Brain and whole spine MRI imaging and blood and CSF testing were normal. The combination of skeletal, neurological, and bladder symptoms with normal lumbar MRI and abnormal urodynamic and neurophysiological studies led to the clinical suspicion of occult tethered cord syndrome (oTCS). Surgical cord "untethering" was performed leading to remarkable postoperative clinical improvement. oTCS is a recently defined functional disorder of the spinal cord due to fixation (tethering) of the conus medullaris by inelastic elements that may lead to severe neurological impairment. High clinical suspicion is required as oTCS is a treatable spinal cord disorder.

Keywords Occult tethered cord syndrome · Paraparesis · Pectus excavatum · Neurogenic bladder

Introduction

Tethered cord syndrome (TCS) refers to a constellation of skeletal, neurological, and bladder or bowel symptoms due to fixation effect of inelastic tissue on the conus medullaris resulting in stretching and limited movement of the cord within the spinal canal. TCS is related to congenital vertebrospinal anomalies, for which the umbrella term spinal dysraphism is used, or acquired conditions, such as scar tissue from prior spinal surgery [1]. Diagnosis is based on imaging findings, such as low position of the conus below the level of the mid L2 vertebral body and/or identification of tethering elements [2]. However, normal MRI does not exclude TCS diagnosis [3]. Thus, in case of TCS symptoms but normal imaging the term "occult TCS" is used [4].

Case presentation

A 15-year-old female former gymnast was reviewed due to unexplained paraparesis since age 10. Apart from mild scoliosis and pectus excavatum that required surgery at age 14, her personal and family history was unremarkable.

Symptoms were commenced abruptly with intolerable chest and upper back pain in T4–T7 distribution that did not respond to painkillers. About 2–3 weeks afterwards she developed moderate lower limb weakness that partially responded to IV dexamethasone administration. Brain, cervical, and thoracic spine MRIs were normal. After 11 months a superficial soft mass appeared suddenly at the sacrum and remained visible for 3–4 weeks. As soon as the mass resolved severe lower limb weakness, inability to stand or walk and urinary incontinence became evident. Lumbar spine MRI; extensive immunologic,

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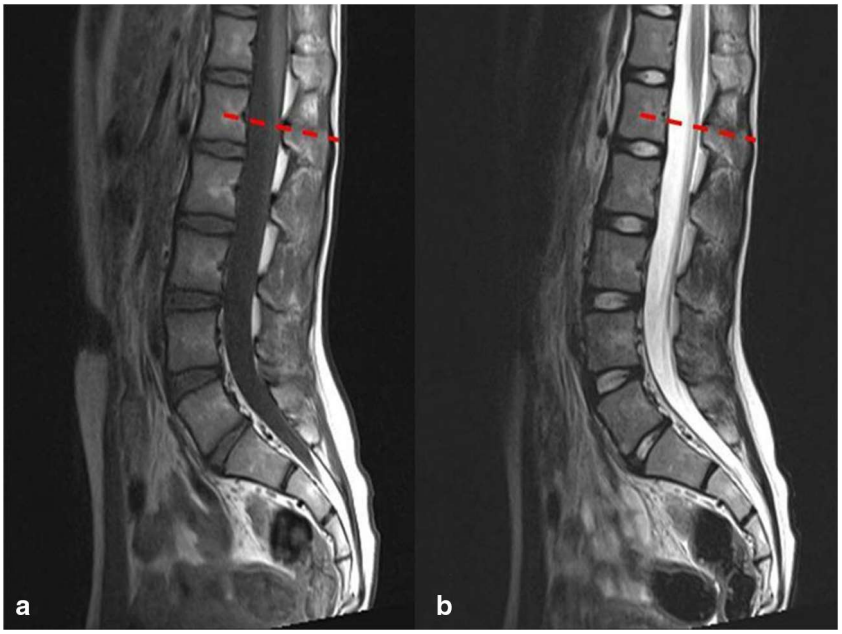

Fig. 1 Lumbar MRI: T1 (**a**) and T2 (**b**) sequences exhibiting normal conus medullaris termination at mid L1 vertebral body level (red dotted lines) and absence of fatty or tight filum terminale, defined as > 2 mm in diameter

hematologic, and infectious blood; and CSF testing were normal. Since then the patient has remained papaparetic, only experiencing minor transient improvement after steroid administration (5-day course of 1 g Methyprednisolone/day).

Motor examination revealed mild upper (MRC grade 4/5) and severe lower limb (1/5) weakness. Apart from mild distal lower limb muscle atrophy muscle bulk was generally well preserved. Although upper and lower limb tendon reflexes were brisk (+ 3), muscle tone was normal and plantar reflexes were downgoing. Higher cortical and cerebellar testing, cranial nerves, and sensory examination including light touch, pinprick, vibration, and proprioception, abdominal, and anal reflexes were intact.

For the past 5 years this case had remained a mystery as all tests, including brain and whole spine MRI imaging and blood and CSF sampling, were normal. Additional tests were ordered. Urodynamic testing revealed

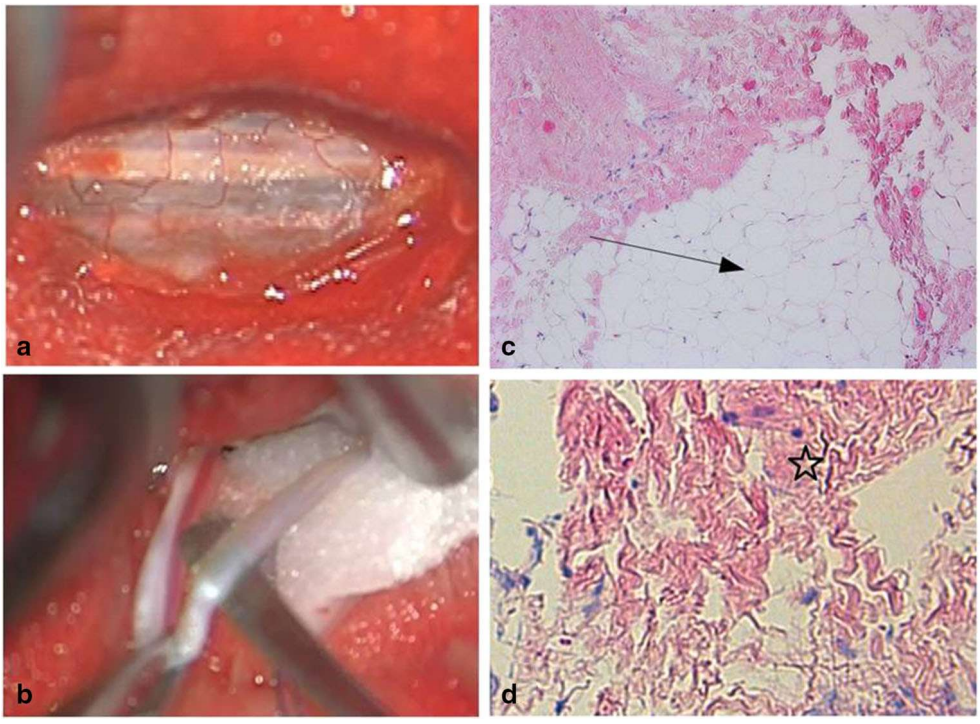

Fig. 2 Perioperative and histological findings. The filum was visible prior to opening the dura (**a**) because of its dorsal location and overstretching (**b**). Histological examination revealed invasion of adipose tissue (**c**, arrow) and degeneration of elastin fibers (**d**, star)

neurogenic flaccid bladder with increased cystometric capacity and underactive detrusor without significant post-void residual volume, as in case of conus medullaris/cauda equina syndrome. Neurophysiological testing was also performed. Motor and sensory nerve conduction studies were normal. Denervating features (fibrillations 1+) from proximal and distal lower limb muscles were EMGraphically recorded. However, the respective motor unit action potential (MUAP) morphology was normal. Interestingly, abnormal MUAP activation from upper and lower limb muscles was evident. Thus, findings were consistent with combined upper and lower motor neuronopathy.

The clinical triad of skeletal deformities, voiding dysfunction, and lower limb weakness is typical of TCS [1]. Urodymanic and neurophysiological studies were also consistent with TCS as the whole spinal cord and, especially, its caudal end seemed affected. Due to the fact that lumbar MRI was normal (Fig. 1), "occult TCS" was suspected and surgical "untethering" was arranged. Intraoperatively, the stigmas of spina bifida, a small subcutaneous lumbar lipoma and spinous process deformity, were revealed. Minimally invasive surgical approach was performed involving intersection of the interspinous ligament, excision of ligamenta flava, and expansion of L5-S1 interlaminar space by the interspinous distractor. As evidenced in Fig. 2 a and b, a tight filum, not seen on MRI, located dorsally and causing thickening of the dura was revealed and sectioned. Histological examination exhibited invasion of adipose tissue and degeneration of elastin fibers (Fig. 2 c and d). Six months postoperatively remarkable clinical improvement was recorded as lower limb muscle strength reached 4−/5, the patient could stand and walk with assistance and bladder symptoms improved considerably.

Discussion

"Occult TCS" (oTCS) is a recently defined entity [4] whose diagnosis is based on the typical TCS clinical triad of skeletal, neurological, and bladder or bowel symptoms [1] supplemented by abnormal urodynamic [5] and neurophysiological studies [6]. Standard lumbar MRI is, by definition, normal.

Our case fulfilled oTCS diagnostic criteria as mild scoliosis and pectus excavatum accompanied by lower limb weakness and neurogenic bladder were evident. Typical symptom onset in childhood was also present. TCS symptoms usually develop during childhood growth spurts due to the discrepancy between the rapidly growing musculoskeletal elements of the spinal column in relation to neuroaxis that causes excessive stretching of the anchored spinal cord [4]. Furthermore, clinical findings were accompanied by normal lumbar MRI imaging (Fig. 1), as conus medullaris terminated in mid L1 vertebral body level and no tethering elements, such as fatty or tight filum, defined as > 2 mm in diameter, were evident [3]. Finally, diagnosis was assisted by abnormal neurophysiological and urodynamic findings. oTCS diagnosis led to the decision to proceed with surgical "untethering." During surgery a tight filum terminale, not seen on MRI, located dorsally and causing a thickening of the dura—intraoperative diagnostic criterion of oTCS [7]—was revealed (Fig. 2) and sectioned resulting in remarkable postoperative improvement.

Our case exhibits several intriguing clinical features as TCS symptom onset with severe chest and upper back pain [8] and after resorption of a soft mass, most probably a meningocele that had suddenly developed at the sacrum, is rarely mentioned [9]. Moreover, our clinical (profound lower limb weakness accompanied by brisk reflexes and minimum muscle atrophy) and neurophysiological findings (fibrillations plus abnormal MUAP activation) are in keeping with the proposed stretching-induced vascular compromise of the spinal cord [1, 10] and combined upper and lower motor neuronopathy [11] in case TCS, as the same phenomenon is encountered in chronic spinal cord ischemia [12, 13]. Furthermore, the mentioned transient improvement after steroid administration is in keeping with the therapeutic effect of steroids in case of spinal cord ischemia and edema [14].

Conclusion

oTCS is an underdiagnosed entity that may lead to severe neurological impairment. Prompt diagnosis is crucial as it is a treatable functional disorder of the spinal cord.

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Data availability Data and material are available upon request.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

Informed consent Patient's consent to participate and for publication are available upon request.

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