

Case Report

Tethered Cord Syndrome: An Unusual Cause of Neurobladder in Adult (Case Report)

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BACKGROUND: Tethered cord syndrome is a progressive functional disorder due to constant or intermittent axial traction on the terminal part of the spinal cord fixed in an abnormal caudal position. It is a frequent complication of spinal dysraphism. The low fixation of the spinal cord prevents normal cranial migration within the vertebral canal. The result being neurological dysfunction. Although quite common and diagnosed early in childhood, this condition may be less severe and remain asymptomatic into adulthood. Diagnosis is essentially based on spinal cord magnetic resonance imaging (MRI) and treatment remains surgical in patients with the symptomatic form.

CASE PRESENTATION: We report the case of a 26-year-old man suffering from tethered spinal cord syndrome, discovered following an initial urinary and then motor symptomatology. We performed spinal cord untethering with lysis of adhesions by detachment of the terminal filum, followed by dural closure and placement of a dural patch. Motor and bladder re-education sessions led to progressive and complete regression of the disorders 16 months after surgery.

CONCLUSION: Low attached spinal cord syndrome is rarely diagnosed in adulthood. Treatment involves several medical and surgical specialties. Delayed diagnosis and management can affect the chances of recovery.

KEY WORDS: Adult, Neurological bladder, Spinal cord, Tethered cord syndrome.

INTRODUCTION

Tethered cord syndrome (TCS) is a progressive functional disorder caused by abnormal pathological attachment of the terminal part of the spinal column. It belongs to the group of occult spinal dysraphism.^{1,2} The fixation of the spinal cord poses a risk of neurological progression due to the growth and ischemic phenomena on the conus and the last nerve roots secondary to stretching. This results in bladder, bowel and orthopedic disorders that may worsen.³

The incidence of TCS is estimated to be between 0.05 and 0.25 cases per 1000 births,⁴ with a widely reported female predominance.^{5,6} It may be due to a primary cause; related to a too short and too thick filum terminale, an intra-spinal sacral lipoma, an occulta spina bifida or diastomatomyelia. There are also secondary causes related to connective tissue adhesions after postnatal surgical closure of a meningocele.⁷ If the diagnosis is made early in childhood, the clinical signs found are generally related to an underlying disease, but for patients in whom no

early dysraphism is found in childhood, the diagnosis is delayed and might be made only in adulthood.⁸ The diagnosis of tethered cord relies on imaging, and MRI is currently the key element in this, showing the location of the conus in a low position (Below L2), and usually revealing the cause of the tethering.⁹ The treatment of TCS is essentially surgical and consists of neurosurgical release of the terminal spinal cord through a posterior approach.

We report the case of a 26-year-old man suffering from TCS discovered following urinary symptomatology that was followed by motor symptoms, for which surgical intervention allowed progressive and complete improvement of the sensory-motor and sphincteric bladder disorders at 16 months post-operatively.

CASE PRESENTATION

A 26 year old man, having benefited from surgery for spina bifida of meningocele type at the age of 8 months without any immediate after-effects, presented to our department. He has been followed up at the urology and andrology department of the university hospital of Conakry for 4 months for dysuria, urinary urgency that has been evolving for 14 days, complicated by urinary incontinence with complete paraplegia that has been evolving for 72 hours. In view of this symptomatology,

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the results of the renal-vesical ultrasound which showed no specific lesion apart from a post-micturition residue estimated at 50cc, and the normality of the biological work-up, the patient was referred to us for exploration and management. His neurological examination on admission revealed tactile and algesic hypoesthesia of the lower limbs with saddle anesthesia. Patellar tendon reflexes were decreased and symmetrical in the lower

limbs. There was a complete motor deficit in the lower limbs (Overall muscle strength: 0/5).

The spinal cord MRI performed 4 months after the onset of the first symptoms and on day 4 of the motor deficit, allowed us to detect a low attached spinal cord with the conus inserted opposite L5-S1 and a spina bifida adjacent to S3-S4 (Fig. 1).

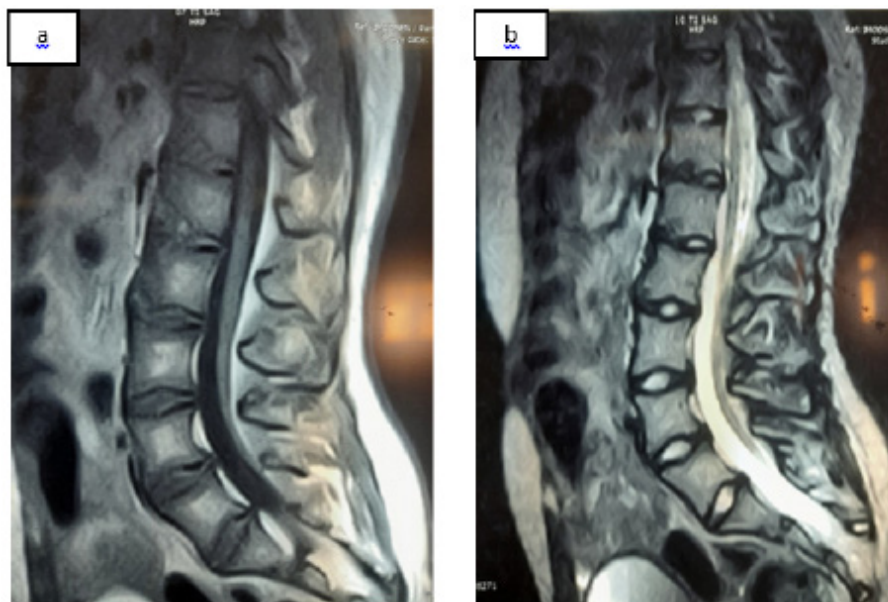


Fig 1: (A) Sagittal MRI T1-weighted images and (B) T2-weighted images showing tethering of the low-lying spinal cord.

The indication for surgical untethering of the low-lying cord was clear. After positioning of the patient, we made a linear posteromedial lumbosacral incision 8 cm long, from L3 to S2. After bilateral musculo-aponeurotic disinsertion exposing the posterior vertebral architecture of the canal, we proceeded to a spino-laminectomy with total durotomy from L4 to S2 to expose the lower attached cord. We performed lysis of adhesions by untethering of the filum terminale, then dural closure and placement of a dural patch. The rest of the surgery went well. There was no improvement in the immediate postoperative period and we had one complication of cerebrospinal fluid (CSF) leak, which was successfully repaired.

The removal of the bladder catheter for a trial period was unsuccessful. The patient therefore opted for intermittent self-catheterization. At 65 days postoperatively, there was a significant improvement in the motor deficit (3/5 in the lower limbs) and sensory modalities. Several sessions of repetitive and regular motor and vesico-sphincter physiotherapy were carried out. We witnessed complete recovery of the sensory- motor disorders 7 months after the operation. The vesico-sphincter disorders persisted for 14 months postoperatively. However, the patient showed bladder autonomy (no need for a urinary catheter) at 16 months postoperatively.

The study was approved by the institutional review board of National teaching hospital, Conakry/ Guinea. Informed consent was obtained from the patient.

DISCUSSION

TCS is a rare condition that may or may not occur as part of a spinal dysraphism in children or young adults in their late teens. We report the case of a 26 year old man who was diagnosed with TCS 25 years after an uncomplicated spina bifida operation. We found in the literature some data according to which the authors reported an average duration of 8 years between the development of the symptoms and the diagnosis.^{10,11} In our case it took 4 months to search for the diagnosis in the urology and andrology departments so that in the end, the diagnosis of certainty was established in the neurosurgery department. In fact, the rarity of this pathology and the subtlety of the symptoms which can mimic other urological and andrological pathologies, as illustrated by the case of this patient, are among the reasons for the delay in diagnosis.

In contrast to our case, motor disorders often precede sphincter bladder disorders.^{10,12} This can of course delay the diagnosis but it should be borne in mind that if the motor deficit is the first clinical sign, symptoms of bladder and bowel dysfunction should be systematically sought during the interview. The patient, too preoccupied with his motor deficit, may not voluntarily give this information, which is nevertheless crucial. Urodynamic examinations have a place in the diagnosis and follow-up of fixed cord syndromes. MRI is the examination of choice for confirming the diagnosis and establishing a complete and accurate assessment.

Treatment is mainly surgical in the symptomatic patient. An early surgical approach is recommended.^{11,13} In 90% of patients, surgery has a good outcome. Significant improvement in pain and motor deficit occurs in 80% and 60% of patients respectively. However, sensory and urinary symptoms are less likely to improve and remain unchanged in about 50% of the patients.¹² As found in our patient, the most frequent surgical complication is CSF leakage. We did not observe any postoperative infection and the evolution was favourable with total regression of the clinical manifestations after 16 months of vesico-sphincter and motor rehabilitation, with an unremarkable urodynamic assessment. We think that it is crucial for clinicians to be aware of adult tethered cord syndrome and its presenting symptoms in order to achieve early diagnosis and subsequent management.

CONCLUSION

Tethered cord syndrome remains a worrying subject in neuro-urology. It involves us at the stage of diagnosis, neurosurgical indication and symptomatic treatment. The diagnosis must be early and quickly suspected in patients with a history of spinal dysraphism.

List of abbreviations

CSF: Cerebrospinal fluid.

MRI: Magnetic resonance imaging.

TCS: Tethered cord syndrome.

Disclosure

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