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Original Article

Pediatric Tethered Cord Syndrome: Experience of Tanta University Hospitals

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BACKGROUND: Tethered cord is abnormal fixation of the spinal cord below the first lumbar vertebra. It might be primary congenital or secondary resulting from scarring after intraspinal surgery. This anomaly results in progressive neurological, orthopedic, and urological dysfunctions. Considering the syndrome has a progressive course, surgical treatment cannot be avoided.

OBJECTIVE: Our aim was to analyze the etiology, clinical manifestations and surgical outcome of tethered cord syndrome (TCS) in our center.

PATIENTS AND METHODS: This retrospective study reviewed the data of all patients suffering from TCS in the records of Tanta University Hospitals from January 2019 till October 2021. Detailed clinical, radiological, surgical, intraoperative neurophysiological monitoring and follow up data were reviewed and analyzed.

RESULTS: Nine patients were included in this study. Age ranged from 1-11 years with a mean age of 4.34 years. The causes of tethering included four lipomyelocele (44.4%), two dermal sinus (22.2%), one intradural lipoma (11.1%), one filum terminale lipoma (11.1%) and one case due to adhesions from previous surgery (11.1%). Four cases improved regarding motor and gait disturbance and no case deteriorated. Only two cases were complicated, one with wound infection and one with cerebrospinal fluid (CSF) leak.

CONCLUSION: The TCS could be managed safely in the pediatric population. Efficacy of surgery could result in either clinical improvement or at least preventing further deterioration.

KEYWORDS: Dermal sinus, Filar lipoma, Lipomyelocele, Tethered cord.

INTRODUCTION

Tethered cord is the abnormal situation and fixation of the spinal cord below the first lumbar vertebra. It could be either congenital as with spinal dysraphism or acquired resulting from scarring after intraspinal surgery such as meningomyelocele repair. It results in neurological, urological and orthopedic dysfunction.[1]

The spinal cord forms through primary and secondary neurulation. Failure of primary neurulation can cause an open neural tube defect including myelomeningocele, lipomyelomeningocele, intraspinal lipoma and dermal sinus tract. Failures of secondary neurulation contribute to formation of closed pathologies including lipomas, tight/fatty filum and myelocystocele.[2] Abnormal situation and fixation of the distal spinal cord during embryogenesis leads to stretching of the spinal cord, reduced blood flow, defective oxidative metabolism and abnormal glucose metabolism as well as hypoxia and ischemia of the sacral nerve roots.[3]

Spinal cord tethering incidence is 1 per 4000 births, with slight female gender predominance. The syndrome can manifest itself in children, however milder cases might present in adulthood with differences in the leading symptoms between children and adults. In children, the most common presentations are sensory and motor deficits, visceral manifestations in the form of bladder and/or bowel voiding dysfunction, orthopedic deformities and skin stigmata. However, in adulthood, pain is more common, but motor deficit is quite often. The symptoms are usually progressive. In childhood, symptoms are often worsening during growth spurts; while among adults, trauma or stretching can elicit the complaints.[4]

Considering TCS has a progressive course, surgical treatment cannot be avoided. In childhood, the diagnosis of tethered cord alone should be an indication for surgery for avoiding neurological deterioration.[4,5] Stabilization of neurological situation is achieved by untethering. Early intervention appears to result in a much greater recovery.[6]

We describe here the etiology, clinical manifestations and surgical outcome of tethered cord syndrome in our center.

PATIENTS AND METHODS

Study Design

This retrospective study represents a single institutional experience regarding management of pediatric tethered cord syndrome in our neurosurgical department from January 2019 till October 2021. All patients were children. Patients were subjected to medical history taking and clinical examination including neurological examination (Muscle power was assessed by Medical Research Council Manual Muscle Testing scale), urological assessment, and/or assessment of subjective symptoms, such as pain that was assessed by visual

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analogue scale (VAS). Radiological assessment included lumbosacral spine magnetic resonance imaging (MRI) to diagnose the cause of the tethering in addition to sagittal T2 whole spine exam to rule out associated syringomyelia. Computerized tomography (CT) of the brain was performed to detect hydrocephalus and MRI brain was requested if patients were suspected to have Chiari type II malformation. Whole spine x ray was done to rule out scoliosis. Preoperative urodynamic study and preoperative full laboratory investigations were also performed. All parents/guardians have received all information regarding the outcome and risks with written informed consent under approval of the ethical committee (institutional review board (IRB): 35820/9/22).

Procedure and Monitoring

We did untethering of the cord under intra-operative electrophysiological monitoring device ISIS (Inomed, Emmindengen, Germany) in form of somatosensory evoked potential (SSEP), motor evoked potential (MEP) and direct nerve root stimulation of muscles of the anal sphincter; while dealing with each pathological cause of tethering. In cases with dermal sinus and infected epidermoid **(Fig. 1)**, ellipse incision was done around the sinus and the track was followed till the dural defect. The bone was widened and the dural opening was extended below and above the dural defect exposing the dermoid cyst that was carefully removed utilizing usual microsurgical techniques with identification and division of the filum. In cases with filar lipoma **(Fig. 2),** exposure was done through routine hemilaminectomy with division of the thickened yellow filum. In cases with lipomyelocele **(Fig. 3)**, linear skin incision was performed with exposure of the subcutaneous lipoma followed by exposure of the bony posterior element defect and the dural defect which was widened. Safe debulking of the intradural lipoma was done and the filum was identified and sectioned. In cases with intradural lipoma, exposure was done through routine laminectomy with safe debulking of the lipoma and identification and division of the filum. In cases with recurrent tethered cord, untethering was done by exposing the dural scar and widely opening the dura around the scar then identification and microsurgical separation of the placode from the dorsal dural scar.

One month postoperatively, MRI lumbosacral spine and CT brain was repeated for all patients. Urodynamic studies was repeated for the patients who had preoperative visceral abnormality to be compared with the preoperative data. All patients were reexamined monthly for 9-12 months postoperatively.

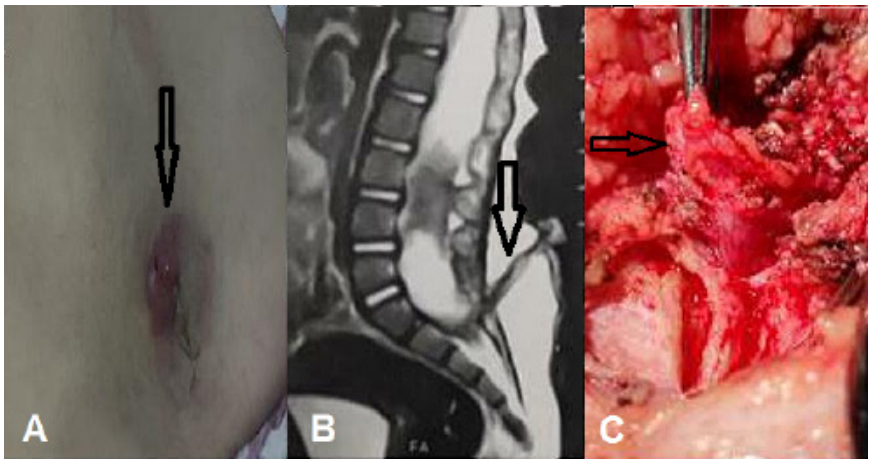

Fig 1: (A) Photography shows the dermal sinus. (B) Preoperative sagittal T2-weighted MRI shows hyperintense mass attached to an expanding cord causing cord tethering with associated dermal tract and sinus. (C) Intra-operative photography shows dissection of the adherent dermoid cyst from the cord.

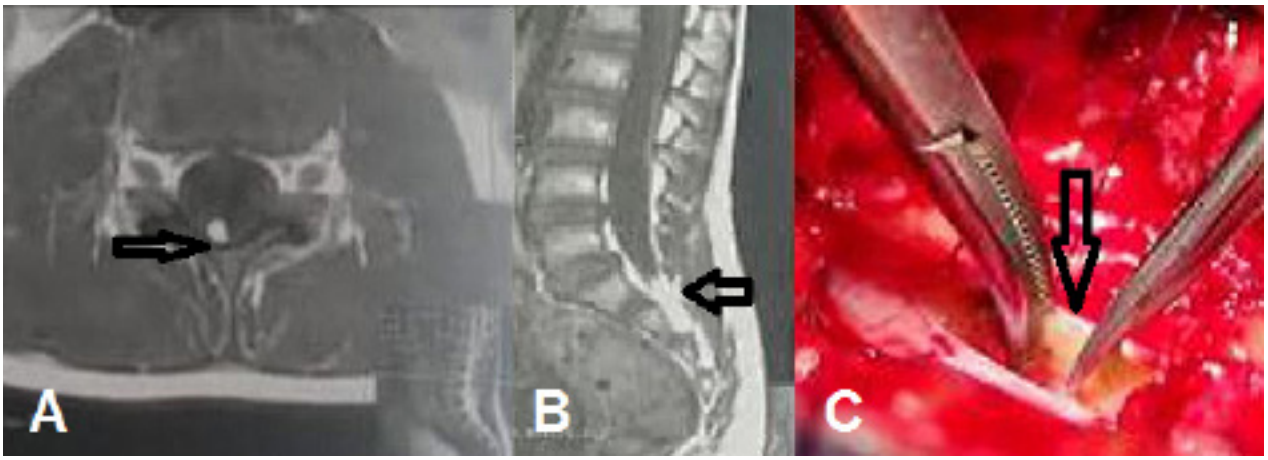

Fig 2: (A & B) Preoperative axial and sagittal T1-weighted images show hyperintense filar lipoma. (C) Intraoperative photography shows division of the thickened fatty filum.

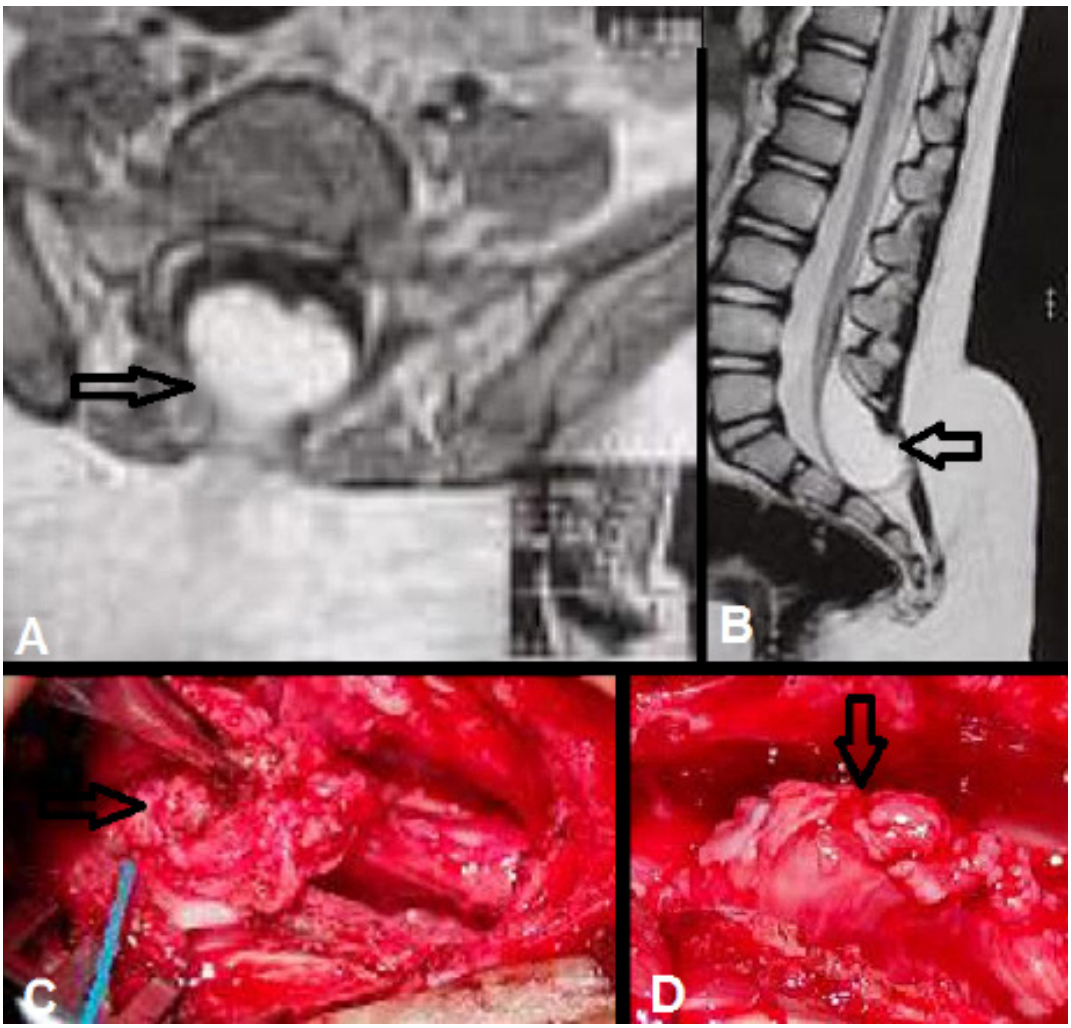

Fig 3: (A & B) Preoperative axial T1 and sagittal T2-weighted images show lipomyelocele with tethered cord. (C & D) Intraoperative photography shows dissection of the adherent lipoma from the cord and reconstruction of the cord after near total lipoma excision.

Statistical Analysis

The statistical packages for the social sciences (SPSS) software version 21 was used to analysis the data. Descriptive data included mean ± standard deviation for parametric data and number and percentage (%) for frequency.

RESULTS

Nine patients were included in the current study **(Table 1)**; 5 males (55.6%) and four females (44.4%). Our patients' ages ranged from 1-11 years with a mean age of 4.34 ± 3.29 years. As regards the cause of tethering, there were four lipomyelocele (44.4%), two dermal sinuses (22.2%), one intradural lipoma (11.1%), one filum terminale lipoma (11.1%) and one case due to adhesions from previous surgery (11.1%). Only seven children had skin stigmata (77.8%); four children showed subcutaneous lipoma (44.4%), two patients had dermal sinus (22.2%) and one case showed an old scar of repaired myelomeningocele (11.1%). Delay between age at onset of the disease and age at time of surgery ranged from 3 to 12 months. Five patients had motor deficit (55.6%) but only three of them had progressive gait deterioration (33.3%). Only four cases improved regarding motor deficit with improvement of the associated gait disturbance. One case did not improve, but no case deteriorated. Six patients had bowel and bladder dysfunction (66.7%), 3 of these cases showed improved bladder functions as confirmed by the postoperative urodynamic study. Three patients had preoperative back pain which completely resolved within the first month postoperatively Three patients had sensory deficits (33.3%) that showed no change postoperatively.

One child developed hydrocephalus (11.1%) and was treated by ventriculoperitoneal shunt. None of our cases showed scoliosis or Chiari type II malformation. We had only two complicated cases; one of them had a wound infection which was treated conservatively by daily dressing until complete cure. The other case had CSF leak which was managed by surgical repair with dural patch and daily dressing until complete cure. None of our cases has developed retethering manifestations till the end of the follow up period.

Table 1: Demographic, clinical and follow up data of studied patients

Parameter	Patients (Total = 9)
Age (years):	
Minimum-maximum	1-11
Mean ± standard deviation	4.34± 3.29
Gender:	
Male	5 (55.6%)
Female	4 (44.4%)
Causes of tethering:	
Lipomyelocele	4 (44.4%)
Dermal sinus	2 (22.2%)
Adhesions	1 (11.1%)
Intrdural lipoma	1 (11.1%)
Fatty filum	1 (11.1%)
Complaints:	
1. Visceral manifestations	6 (66.7%)
2. Motor deficits	5 (55.6%)
3. Gait disturbance	3 (33.3%)
4. Back pain	3 (33.3%)
5. Sensory deficits	3 (33.3%)
Dermal stigmata:	
▪ Subcutaneous lipoma	4 (44.4%)
▪ Dermal sinus	2 (22.2%)
▪ Scar	1 (11.1%)
▪ No skin stigmata	2 (22.2%)
Postoperative hydrocephalus:	1 (11.1%)
Functional outcome:	
1. Visceral manifestations	3 improved (50%)
2. Motor deficits	4 improved (80%)
3. Gait disturbance	3 improved (100%)
4. Back pain	3 improved (100%)
5. Sensory deficits	No improvement
Complications:	
No complications	7 (77.8%)
Wound infection	1 (11.1%)
Cerebrospinal fluid leak	1 (11.1%)

DISCUSSION

The TCS has a wide variety of clinical presentations caused by excessive stretching on the spinal cord. The majority of cases are related to spinal dysraphism. The syndrome presents clinically at any age with wide spectrum of clinical presentations including pain, neurological deficits and deformities. Surgical untethering is recommended in patients with progressive or new onset presentation. Early intervention appears to result in a much greater recovery with better results and outcomes.[6] The benefits of surgery are debated in asymptomatic patients.[1,7] The results of the study of Bui et al. demonstrated that in childhood the diagnosis of tethered cord alone should be an indication for surgery for avoiding neurological deterioration.[8] All our cases were symptomatic at the time of presentation, so we operated without any further delay.

The cutaneous manifestations of TCS include lipomas, hypertrichosis, hemangiomas, cigarette burns, dermal si¬nus, and meningocele.[8] In our study 77.8% of the patients had skin stigmata; 44.4% subcutaneous lipoma, 22.2% dermal sinus and 11.1% old scar of repaired myelomeningocele.

The neurological manifestations with TCS are highly variable including motor weakness, gait abnormality and sensory dysfunction.[8] In a study by Stavrino et al. almost all patients presented by some degree of motor deficit with postoperative improvement rate of 73%.[9] Herman et al. reported a 79% improvement in motor function

in 153 myelomeningocele and lipomyelomeningocele patients.[10] A study of Bowman et al. found that 70% of myelomeningocele patients gained muscle strength in the lower extremities as compared to preoperatively.[11] In our study group, 55.6% of the patients had motor deficits with 80% improvement rate postoperatively, while 33.3% had sensory deficits that showed no change postoperatively; none of our patients deteriorated postoperatively.

In our series, pain had the best response to surgical treatment with 100% improvement. It seems that pain improves dramatically secondary to untethering, regardless of the underlying cause and pathological condition, with almost high majority of children showing improvement.[12] Additionally, 75% improvement rate has been reported with repeated untethering procedures.[13]

In children presenting with tethered cord, the urological dysfunction and abnormalities include large varieties with only less than half of the patients actually having no symptoms.[14] Urodynamic dysfunction will not be detected in toddlers until their toilet-training. In the literature, the results are generally satisfactory even with repeated procedures,[15] with improvement rates between 50% and 100% (Well above 70%), with recommendations given for early untethering.[9,14,16,17] On other hand, an improvement rates of 30–60 % has been reported by other studies,[10] while others did not find significant improvement.[18] In our series, 66.7% of the patients had bladder dysfunction with 50% improvement rate as confirmed by the postoperative urodynamic study.

We used electrophysiological monitoring in all cases. Electrophysiology could significantly reduce the deficit risk postoperatively from 9.4 to 2.9%.[4] However, certain authors have stated earlier that intraoperative monitoring during the surgical release of tethered cord has no significant benefit.[19]

Regarding the effect of surgical untethering in myelomeningocele patients who had scoliosis, many studies in the literature demonstrated that surgery will improve the curvature in some cases or at least will arrest curve progression.[20]

The common surgical complications after untethering surgery include CSF leakage, wound infection, meningitis, bladder voiding dysfunction, and neuro¬logical deficit and deterioration.[21] We had only two complicated cases; one of them had wound infection which was managed conservatively by daily dressing until complete cure. The other case had CSF leak which was treated by surgical repair with dural patch and daily dressing until complete cure.

Retethering resulting in symptoms is a frequent unfavorable situation following untethering surgery. While variable methods have been reported to prevent retethering, none has been proven in a prospective manner.[22] Current series did not show retethering during the follow up period.

Limitations of the current study

The retrospective nature of the study, the small number of included cases (As we are not a pediatric center) and the relatively short follow-up period are the main study limitations.

CONCLUSION

Tethered cord syndrome could be managed safely in the pediatric population. Efficacy of surgery could result in either clinical improvement or at least preventing further deterioration.

List of Abbreviations

CSF: Cerebrospinal fluid.
CT: Computerized tomography.
IRB: Institutional review board.
MEP: Motor evoked potential.
MRI: Magnetic resonance imaging.
SPSS: Statistical packages for the social sciences.
SSEP: Somatosensory evoked potential.
TCS: Tethered cord syndrome.
VAS: Visual analogue scale.

Disclosure

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

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