Clinical Article

Surgical Management and Outcome of Tethered Cord Syndrome in School-Aged Children, Adolescents, and Young Adults

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Objective : The adolescent presentation of tethered cord syndrome (TCS) is well-recognized, but continues to pose significant diagnostic and management controversies. The authors conducted a retrospective study of clinical outcomes after surgical intervention in 24 school-aged children, adolescents, and young adults with TCS.
Methods : All 83 patients with a lipomyelomeningocele (LMMC) underwent untethering surgery for caudal cord tethering between 1987 and 2007. The clinical charts and follow-up data were reviewed. Of these patients, 24 school-aged children, adolescents, and young adults with TCS were studied with respect to the clinical, radiologic, pathologic features, and surgical outcomes.
Results : Untethering procedures were performed in 24 patients (age range, 7-25 years) for TCS of various origins (lipoma, lipomyelomeningocele, and tight filum terminale). Specific circumstances involving additional tugging of the already tight conus, and direct trauma to the back precipitated the onset of symptom in 50% of the patients. Diffuse and non-dermatomal leg pain, often referred to the anorectal region, was the most common presenting symptom. Progressive sensorimotor deficits in the lower extremities, as well as bladder and bowel dysfunction, were also common findings, but progressive foot and spinal deformities were noted less frequently. The most common tethered lesions were intradural lipomas, thickened filum and fibrous band adhesions into the placode sac. The surgical outcome was gratifying in relation to pain and motor weakness, but disappointing with respect to resolution of bowel and bladder dysfunction. Of the 24 patients with TCS, pre-operative deficits improved after surgery in 14 (58.3%), remained stable in 8 (33.4%), and worsened in 2 (8.3%).
Conclusion : The pathologic lesions of tethered cord syndrome in school-aged children, adolescents, and young adults, are mostly intradural lipomas and tight filum. It is suggested that the degree of cord traction results in neurologic dysfunction in late life due to abnormal tension, aggravated by trauma or repeated tugging of the conus during exercise. Early diagnosis and adequate surgical release might be the keys to the successful outcome in school-aged children, adolescents, and young adults with TCS.

KEY WORDS : Adolescent · Intraspinal lipoma · School-aged children · Surgical outcome · Tethered cord syndrome · Young adults.

INTRODUCTION

Tethered cord syndrome (TCS) is a developmental abnormality of the neuroaxis which is usually diagnosed in childhood. Adults with TCS are arguably the most neglected

individuals in the population with a neurosurgical disease. The actual tethering has been attributed to a variety of pathologic entities, including a thickened tight filum terminale[1,8,13], intradural lipomas with or without a connecting extradural component[1,4], intradural fibrous adhesions[1,25], diastematomyelia[5,10], and adherence of the neural placode following previous closure of a myelomeningocele[9,11,14]. However, sufficient differences in the mode of onset, clinical manifestations, and outcome exist between pediatric and adolescent patients with tethered cord to warrant a more detailed analysis of the adult syndrome[19]. The most problematic technical consideration in surgery for the release of the tether-

ed cord is how to preserve functioning neural elements and rebuild the dural sac to maintain normal cerebrospinal fluid (CSF) circulation. The purpose of this paper was to review some unique features in clinical presentation, factors precipitating the onset of symptoms, diagnostic clues, pathologic entities, and surgical outcome in school-aged children, adolescents, and young adults with TCS.

MATERIALS AND METHODS

From 1987 to 2007, 83 patients with lipomyelomeningoceles were treated. Of these patients, 24 school-aged children, adolescents, and young adults with TCS were studied with respect to the clinical, radiologic, pathologic features, and surgical outcomes of this syndrome. The following radiologic investigations were performed on the 24 patients with TCS : metrizamide myelography (initial period in 1987, 1 case), computerized tomography (CT), and magnetic resonance imaging (MRI). In all cases, surgery was performed under continuous intraoperative eletrophysiologic monitoring, including monitoring of bowel and bladder function, with the aim of preserving the functioning neural element during untethering procedures. All tethering was individually treated using an intra-operative monitoring system [somatosensory evoked potential (SEP) and urodynamic study]. The portions adherent to the dura were sharply dissected away, leaving the rest of the mass gliding freely within the dural sac to maintain the functional neural elements. All dural defects were closed with lumbodorsal fascial grafts (9 cases) or Gortex (15 cases). Tethering lesions (lipomas) were maximally debulked and complete untethering of the cord was then performed. At the close of surgery, mega-dural reconstruction (extremely large dura graft for maintaining CSF circulation)[16] was performed.

RESULTS

The age distribution and type of lesions of the lipomyelomeningocele (type I, dorsal; type II, transitional; and type III, mixed) are shown in Table 1. The ages ranged from 7-25 years, with a mean of 16.5 years. The mean age of onset was 16 years. Among the 24 patients with TCS, there were 7 with type I, 9 with type II, and 8 with type III. The presenting complaints that led to a neurosurgical consultation in adulthood are listed in Table 2. Pain was the most common presenting symptom, and was a prominent complaint in 22 patients (92%). Eighteen patients (75%) presented with progressive leg weakness or walking difficulties, and most patients had saddle anesthesia or hypesthesia. Fifteen patients (63%) had bladder symptoms, 3 had spastic symptoms,

and 5 had hypotonic symptoms. Ten patients had bowel dysfunction and 14 patients had foot deformities. An intradural mass associated with an extradural mass was demonstrated in 24 patients, a low level of the conus was noted in 21 patients, and a tethering band was identified in 17 patients with radiologic studies.

Twenty-four patients underwent surgery for release of the tethered conus. Four different tethering entities were encountered at the time of surgery. Twenty-four patients had various combinations of two or more types of pathology (Table 3), each of which was thought to contribute to the tethering process. All intradural lipomas were either densely adherent to the thecal wall or were connected with an extradural component through a dural defect, where the conus was in effect anchored. During the surgery, there were no different characteristics comparing infants and young children with TCS.

The surgical outcome for the 24 patients is shown in Table 4. The relief of pain was most gratifying; 20 (95%) of the patients with significant pain became asymptomatic following surgery. Eighteen patients had significant impro-

Table 1. Age distribution and types of lesions in 24 patients with intraspinal lipomas

Age (years)	No. of patients	Types of lesions		
		I	II	III
7-10	8	5	2	1
11-16	9	2	3	4
17-25	7		4	3
	24 (100)	7 (29.2)	9 (37.5)	8 (33.3)

I : dorsal lipoma, II : transitional lipoma, III : mixed lipoma

Table 2. Presenting signs and symptoms in 24 patients

Signs and symptoms	No. of patients (%)
Pain	22 (92)
Sensorimotor deficits	18 (75)
Bladder dysfunction	15 (63)
Spastic	3
Hypotonic	5
Nonspecific	12
Bowel dysfunction	10 (42)
Trophic Ulceration	3 (13)
Deformity of foot	14 (58)
Scoliosis	4 (17)
Back lesions	24 (100)

Table 3. Pathologic entities in 24 patients with tethered cord syndrome

Pathologic entities	No. of patients (%)
Lipomeningocele or tight filum	5 (20.8)
Lipoma or tight filum, placode	12 (50)
Lipoma or adhesion	4 (16.7)
Lipomeningocele or adhesion	3 (12.5)
Total (%)	24 (100)

Table 4. Surgical outcome in 24 patients with intraspinal lipomas with tethered cord syndrome

Preoperative complaints of deficits	No. of cases	Outcome			
		Normal	Improved	Unchanged	Regressed
Pain	22	14	7	1	0
Sensorimotor deficits	18	2	10	6	0
Sphincter dysfunction	15	0	5	9	1
Trophic ulceration	3	0	2	1	0
Scoliosis	4	0	1	3	0
Foot deformity	14	0	1	12	1

vement in sensorimotor deficits. Only 6 patients were unchanged by surgery. The results for sphincter dysfunction were disappointing; only 5 of 15 patients (33.3%) with sphincter dysfunction were helped by surgery.

DISCUSSION

The difference in growth rates between the spinal cord and musculoskeletal elements accounts for the fact that the conus medullaris may terminate anywhere from the T12 to the L2-L3 interspace[3]. A conus below the L2-L3 interspace in an adult is abnormal, and has been termed a "tethered conus"[8]. The syndrome is usually diagnosed in childhood, and although the presentation is variable and may be insidious, a tethered conus should be suspected in patients with an unexpected spastic gait, a neurogenic bladder, bowel dysfunction, lower extremity weakness, scoliosis, or foot deformities[8,17,24]. These symptoms and signs may be related to traction upon the conus and cauda equina by an intradural mass and tethering band, and may grow worse with age owing to accelerated skeletal growth in relation to the neuroaxis during puberty.

After experimental tethering in growing cats, Kang et al.[15] reported a 32% reduction in spinal cord blood flow (SCBF) in the distal spinal cord close to the tethering within 2 weeks. Untethering of the cord 2 weeks after tethering resulted in an increase in SCBF, with a return to the normal level. This study supports the hypothesis that spinal cord dysfunction in the early stage of tethering results from mechanical and vascular damage to a spinal cord segment, which results in widespread ischemic involvement of the cord in a prolonged tethering effect during growth.

There are several reported cases of adult presentations of tethered conus, and it is unclear why these patients should have been symptom-free and the diagnosis overlooked for so long. Sostrin et al.[23] described three patients with acquired spinal stenosis and a tethered conus, and suggested that the stenosis made the primary tethered conus symptomatic. Pool[21] theorized that the tethered conus eventually became symptomatic in one patient because local stretching and ischemia occurred from the repetitive and insidious low

back trauma associated with the patient's occupation. The description of spinal vasculature in patients with scoliosis given by Hilal and Keim[12] supports this hypothesis, since they demonstrated that tension on the conus caused arterial stretching and ischemia, with resultant cord damage.

Experimental studies[2] also indicate that the damage from local cord stretching is absorbed within a short distance above or below the level of the insult, and this may explain the causal relationship of these patients' symptoms. These clinical and experimental reports, indicating a delicate balance between skeletal maturation and traction upon the tethered conus, may explain why some patients remain symptom-free until adulthood.

Pain is an uncommon presenting complaint in children with TCS[1,13] as opposed to about 90% incidence in our group of school-aged, adolescents, and young adults. In a large number of our cases, the pain, when present, is located in the lumbosacral region, with variable radiation down both legs. The anal and perineal pain described by 12 of our patients was reported rare in children[1,13]. Progressive foot deformities are one of the most common presenting features in children with TCS[1,22]. None of our patients without a pre-existing foot deformity developed this problem with the adult onset of neurologic dysfunction. This suggests that the insult to the adult conus causes flank sensorimotor deficits or even atrophy, but does not result in any type of muscle imbalance. Progressive scoliosis was noted in 33% of a series of children with TCS[13], but progressive spinal deformities have been rarely described in reports of adult TCS[17,19]. The incidence of urologic symptoms appears to be similar in childhood and adult syndromes[1,4,7]. Unlike adults, however, our cases rarely showed urgency, incomplete voiding, or stress incontinence.

In the management of adult patient with TCS, three questions should be considered. Should it be treated aggressively? Should it be studied and operated on as soon as possible? Would it be reasonable to prophylactically operate on patients who are neurologically intact? In general, we do not agree with aggressive treatment of adults with TCS because they have relatively advanced neurologic deficits, and the functional zone they have retained is very narrow, which means that it is necessary to maintain the balance between skeletal maturation and conus traction upon the TC. If aggressive treatment is tried, it could result in a worsening of their neurologic deficits. Pre-operative studies are necessary to plan the treatment in order to avoid operative morbidity or complications. However, if adult patients

with TCS have impending neurologic deficits and/or progression of their existing neurologic deficits, it is reasonable and prudent to operate as soon as possible.

There are various controversial opinions about operating on patients who are neurologically intact or for prophylactic purposes[6]. However, if tethering pathology is confirmed through a radiologic investigation, it might be recommended that the patient is limited in exercise or work, and performed an operation for prophylactic spinal cord release. The operative results for pain are most gratifying[20,23]. Most of our patients had no pain after surgery, and sensorimotor deficits also responded favorably with surgical release of the conus. Many authors with experience in childhood TCS have commented on the poor operative results in terms of urinary dysfunction[7,17]. In the childhood TCS patients, sensory deficits, musculoskeletal atrophy, and voiding difficulty were improved after early untethering procedures[18]. However, pain relief and sensorimotor deficits were improved after untethering in adults TCS patients. Our results for bladder dysfunction in school-aged, adolescents, and young adult TCS patients are equally disappointing. Of the 24 patients with TCS, the pre-operative sensorimotor deficits improved after surgery in 12 patients (66.6%), remained stable in 6 (33.3%), and worsened in 1 (6.7%) patient with bladder dysfunction and a foot deformity. Early diagnosis, adequate release of the tethered cord, and reforming the dural sac might be the keys to successful management. In the school-aged children, adolescents, and young adults with TCS, untethering is an appropriate procedure when there are neurologic symptoms and signs and a confirmed tethering effect based on neuroimaging.

CONCLUSION

Symptomatic onset or aggravation of preexisting patient's complaints could be anticipated for the majority of TCS in school-aged children, adolescents, and young adults. Pain is the most common presenting feature; pain is usually bilateral and often involves the anorectal region. The pathologic lesions causing tethering are intradural lipomas and a tight filum. The surgical outcome is excellent for the resolution of pain and sensorimotor deficits, but disappointing for bladder dysfunction. Early diagnosis and adequate surgical release might be the keys to a successful outcome in school-aged children, adolescents, and young adults with TCS.

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