Our Surgical Treatment Results in Adult Tethered Cord Syndrome: An Experience of 9 Cases

Emre Delen 📞 Ahmet Tolgay Akıncı 📞 Osman Şimşek 📞

ABSTRACT

Objective: The aim of this study was to present the results of surgical treatment along with demographic and clinical features in cases with tethered spinal cord syndrome seen in adulthood.

Method: We retrospectively evaluated 73 cases who underwent tethered cord release operations with the diagnosis of tethered cord syndrome between 2004 and 2015 in our clinic. The mean follow-up period was 21.7 (3-63) months. Adult cases consisted of 5 female and 4 male patients aged between 18-55 years. Clinical and radiological characteristics of the patients and follow-up data related to the surgical treatment were recorded. Pain and neurological findings were graded using the scoring system proposed by Klekamp et al.

Results: Nine patients had complaints of back and/or leg pain, two patients had gait ataxia and three patients had bladder dysfunction. Only one patient had a loss of muscle strength. There were cutaneous stigmata in a total of three patients: These were two dermal sinus tracts, one thoracic dermatomal hypertrichosis. Four patients had scoliosis, 4 had syringomyelia and 3 had split cord malformation. The level of conus medullaris was L3 in one patient, L4 in 3 patients, L5 in 2 patients, and S1 in one patient. All patients underwent surgical detethering. Most of the patients reported a decrease in their pain levels. However, sensory dysfunction and sphincter problems remained unresolved.

Conclusion: The main difference between adult cases compared to pediatric cases was related to symptomatology. The pain was the cardinal symptom in almost all cases. While the surgery was effective to diminish pain, it was not effective in resolution of neurological deficits.

Keywords: tethered cord syndrome, adult, surgical treatment

ÖZ

Amaç: Bu çalışmanın amacı erişkinlikte görülen omurilik sendromu oğullarında cerrahi tedavi sonuçlarını demografik ve klinik özelliklerle birlikte sunmaktır.

Yöntem: Klinikimiz 2004-2015 yılları arasında gergin kord sendromu tanısıyla operasyon geçiren 73 olguyu retrospektif olarak değerlendirildik. Ortalama takip süresi 21.7 (3-63) aydı. Erişkin oğullar 18-55 yaş arasına kadar, 4 erkek hasta idi. Hastaların klinik ve radiyolojik özellikleri ve cerrahi tedavi ile ilgili takip verileri alındı. Ağrı ve nörolojik bulgular Klekamp ve arkadaşlarının önerdiği skorlama sistemi kullanılarak derecelendirildi.

Bulgular: Dokuz hastada sırt ve/veya bacak ağrıları yakınıması, iki hastada yürüyüş atakısı ve güç hastada mesane disfonksiyonu vardı. Yalnızca bir hastada kas kuvveti kaybı vardı. Toplam üç hastada cilt bulguları vardı: bunlar iki dermal sinüs trakti ve bir olguda torso dermatom kıllanma artışı idi. Dört hastada sklozya, 4 hastada siringomyelija ve 3 hastada split kord malformationu tespit edildi. Conus medullaris seviyesi bir hastada L3, 3 hastada L4, 2 hastada L5 ve bir hastada S1 idi. Tüm hastalara kord serbestleştirilmesi operasyonu uygulan- di. Hastaların çoğu ağırla seviyelerinde azalma olduğunu bildirdi. Ancak duyusal işlev bozukluğu ve sfinkter disfonksiyonunun işleyişine tespit edildi.

Sonuç: Yetişkin oğullar ile pediatrik oğullar karşılaştırıldığında temel fark semptomatoloji idi. Ağrı hemen hemen tüm vakalarda kardinal semptomdu. Ameliat rejüzyon tedaviminde etkiliyken, nörolojik deficitsin düzeltmesinde etkin değişildi.

Anahtar kelimeler: gergin kord sendromu, yetişkin, cerrahi tedavi
INTRODUCTION

Tethered cord syndrome (TCS) is a developmental anomaly of neuroaxis characterized by spinal cord stretching (1). Due to many accompanying skin manifestations (meningocele, meningomyelocele, hypertrichosis, dermal sinus tract, subcutaneous lipoma, etc.) it is frequently seen and diagnosed in the pediatric age group. Therefore, many studies in the literature have been performed in pediatric cases; data on diagnosis and treatment methods were obtained from pediatric cases. However, adult tethered spinal cord syndrome (ATCS) is rare and has different clinical features than pediatric cases and limited information is available in the literature (2). In recent years, more and more clinical features have been introduced. However, our knowledge of the disease is still insufficient. The role of surgical treatment is often controversial in these patients, especially in the absence of neurological deficits. For all these reasons, new studies are necessary to determine the clinical characteristics and to form a treatment modality for the disease. In this retrospective study, we evaluated clinical and radiological parameters and treatment results of 9 patients who underwent surgery with the diagnosis of ATCS and discussed them in the light of the literature.

MATERIALS and METHODS

The Human Ethics Review Committee of Trakya University approved all of the clinical protocols in this study, and informed consent was obtained from each patient (TÜF-BAEK 2019/261). Amongst 73 patients who underwent a tethered cord release operation between 2004 and 2015 at Trakya University Medical Faculty Hospital, 9 cases who were 18 years or older were included in the study. Demographic characteristics, clinical features and radiological findings of the cases were evaluated. To quantify the clinical and neurological status of the patients before and after surgery we used a scoring system proposed by Klekamp et al. (Table 1) (3,4). Urological tests were performed in 3 patients with bladder dysfunction. Radiological data obtained via magnetic resonance imaging (MRI) were examined for possibly associated spinal cord pathologies.

RESULTS

The mean age of the study population consisting of 5 female, and 4 male patients was 35.6 years (18-55 years). There were no spinal pathologies in medical history in any of these patients. All patients came to the physician because of back and leg pain. Physical examination revealed skin lesions in 3 of 9 patients. These findings were dermal sinus tract in 2 patients and hypertrichosis in the thoracic region in the 3rd patient. In the neurological examinations, motor and sensory loss were detected in one patient, while three of the patients had bladder dysfunction. Two patients had gait ataxia. Demographic and clinical findings of patients are summarized in Table 2.

The radiological findings of the patients were as follows: scoliosis, n=4; hemivertebrae, n=2; butterfly vertebrae, n=2; block vertebrae, n=2; diastomatomyelia, n=3, and 4 syringomyelia, n=4. Level of the conus was at L3 in one, at L4 in 3, at L5 in 2 and at S1 in one patient. Three cases had no spinal cord anomaly (Table 3).

Table 1. The neurological state assessment scale.

| Score | Pain | Sensory disturbance | Motor weakness | Gait ataxia | Sphincter function |
|-------|------|---------------------|----------------|-------------|-------------------|
| 5     | None | Normal              | Full power     | Normal      | Normal            |
| 4     | Slight, no medication | Present, not significant | Movement against resistance | Unsteady, no aid | Slight disturbance, no catheter |
| 3     | Good control w/medication | Significant, function not restricted | Movement against gravity | Mobile w/aid | Residual, no catheter |
| 2     | Insufficient control w/medication | Some restriction of function | Movement w/o gravity | Few steps w/aid | Rarely incontinent |
| 1     | Severe despite medication | Severe restriction of function | Contraction w/o movement | Standing w/aid | Frequent catheter |
| 0     | Incapacitating | Incapacitated function | Paralysis | Paralysis | Premanent catheter |
The mean follow-up period was 21.7 months (3-63 months). Clinical and neurological status of all cases, before and after surgery is summarized in Table 4. In the postoperative period, there was no improvement in the pain complaints of two patients contrary to the other seven patients who reported improvement. None of the patients with sensory or motor loss showed any improvement. In 2 patients with gait ataxia, 1 point improvement was detected. None of the patients with bladder dysfunction improved. Residual urine was not detected in 2 patients without preoperative urinary dysfunction, while it was present in another patient who had bladder dysfunction preoperatively. All patients underwent surgical detethering. Three patients required re-operations. Two patients developed CSF fistula requiring early surgical repair. During the follow-up period, a pseudomeningocele sac was found in the control MRI of a patient who had severe pain after 20 months which was repaired surgically.

**DISCUSSION**

TCS is described as the clinical syndrome associated with lower extremity motor and sensory loss as well as urinary and intestinal dysfunction due to the longitudinal traction of conus medullaris (5). It has a wide spectrum of clinical presentations according to patients age (2). Pediatric cases are frequently diagnosed with the presence of other congenital anomalies of the spine, such as myelomeningocele, meningocele, and lipomyelomeningocele. In pediatric cases, congenital anomalies of other systems are common (2,6,7). In this respect, these patients are easy to recognize. For this group of patients, the dominant opinion in the literature is to treat this condition surgically. Even though similar mechanisms are

| Table 2. The clinical and the demographical features of the patients. |
| --- |
| **Number of patients** | n:9 |
| **Age (average±SD)** | 35.6±14.4 (18-55 years) |
| **Female/Male, n (%)** | 5 (55.5%) / 4 (44.5%) |
| **Back and leg pain, n (%)** | 9 (100%) |
| **Motor weakness, n (%)** | 1 (11.1%) |
| **Sensory disturbance, n (%)** | 3 (33.3%) |
| **Sphincter disturbance, n (%)** | 3 (33.3%) |
| **Gait ataxia, n (%)** | 2 (22.2%) |
| **Skin lesions, n (%)** | 3 (33.3%) |

| Table 3. The radiological findings of the patients. |
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| **Case No** | **Age (years)** | **Gender** | **Concomitant abnormalities** | **Conus Level** |
| 1 | 28 | Female | SCM+S+Sc+Bc | L4 |
| 2 | 55 | Female | SCM+DS+Sc+Hv+Btv | L5 |
| 3 | 20 | Female | SCM+S+Sc+Hv+Btv | L4 |
| 4 | 51 | Male | | L4 |
| 5 | 18 | Male | Sc+Hv+Btv | S1 |
| 6 | 27 | Female | SCM+S+Sc+Hv+Btv | L3 |
| 7 | 30 | Male | DS | L3 |
| 8 | 44 | Female | | L3 |
| 9 | 48 | Male | | L5 |

**DS**: dermal sinus, SCM: Split cord malformation, diastomatomyely, S: siringomyeli, M: Meningocele, Sc: Scoliosis/ Hv: Hemivertebrae, Btv: “butterfly” vertebra, Bc: Bloc vertebrae

| Table 4. The comparison of preoperative and postoperative neurological scores. |
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| **Case No** | **Pain** | **Motor weakness** | **Sensory Disturbance** | **Gait ataxia** | **Sphincter Function** | **Follow-up Time (month)** |
| **preoperative/postoperative neurological scores** |
| Case 1 | 2/5 | 5/5 | 5/5 | 5/5 | 5/5 | 5 months |
| Case 2 | 2/5 | 5/5 | 4/4 | 5/5 | 4/4 | 52 months |
| Case 3 | 4/4 | 5/5 | 5/5 | 5/5 | 5/5 | 63 months |
| Case 4 | 4/4 | 5/5 | 3/4 | 5/5 | 4/4 | 18 months |
| Case 5 | 2/5 | 5/5 | 5/5 | 5/5 | 5/5 | 26 months |
| Case 6 | 3/5 | 5/5 | 4/5 | 5/5 | 5/5 | 3 months |
| Case 7 | 1/3 | 5/5 | 4/4 | 5/5 | 5/5 | 6 months |
| Case 8 | 0/2 | 5/5 | 2/2 | 5/5 | 5/5 | 20 months |
| Case 9 | 4/5 | 5/5 | 5/5 | 5/5 | 3/3 | 3 months |
involved, ATCS has differences in clinical features and in patient management. Patients, who have been asymptomatic for many years, may consult a physician later in their life, most usually with back and leg pain and without neurological deficits. In this case, the place of surgical treatment is controversial, and even the diagnosis might be overlooked.\(^{1,2}\)

It is controversial why cases are symptomatic in adult years. Moderate cord tension can be tolerated by patients until adulthood and the disease may be asymptomatic. Yamada et al. explained the reason why patients become symptomatic over time as follows: 1) Loss of elasticity as a result of fibrosis in the filum terminale, 2) A sudden growth attack, 3) An increase in physical activity during the young adult period.\(^7,8\) The main focus is on repetitive microtrauma and on the degenerative process of the spine.\(^1,8\) Besides, conditions causing laxity in lumbosacral ligaments such as giving birth in the lithotomy position, herniated disc and fracture, leg exercises, prolonged sitting, physical exercise, heavy lifting, traffic accidents, which may cause a reduction in the dimensions of the spinal canal are the facilitating factors.\(^1,2,6,9,10\)

In general, the first reason for ATCS patients to apply to a physician, are waist, back, and leg pains. These complaints of patients may not be tolerated despite medical treatment. While in a large series of 62 patients, 59 patients reported pain; in other studies, this rate has been reported between 45% to 68.1%\(^11-13\). All of the patients in our series had complaints of pain. In addition, 6 out of the 9 patients with pain had at least 3 points lesser scores according to the neurological scale we used. The pain had definitely a negative effect on their daily living activities. A significant decrease was observed in the postoperative pain of the patients (Table 4). From this point of view, it can be said that the best benefit obtained from the surgical treatment is pain relief, and the pain that cannot be cured with medical treatment is a good reason to decide on the surgical treatment.

In neurological examination performed at the time of diagnosis, neurological deficits are reported in a significant percentage of cases (59% -72%)\(^15,16\). In our series of 9 patients, only 1 patient had a motor deficit and there was no amelioration during the postoperative period. Our study results were not fully compatible with the literature in terms of these data. The reason might be the small sample size which is the main limitation of this study. However, the fact that our study included ATCS patients without neurological deficits made us think that the data could contribute to the literature in the decision-making process in such patients.

The findings of bladder dysfunction, most commonly hyperactive bladder, frequently accompany TCS cases.\(^2\) In previous studies, it has been reported that bladder dysfunction findings in TCC vary between 36.3% and 71%\(^11,12,14\). These findings may be present at the time of diagnosis or may be an early predictor of neurological deterioration.\(^2,13\) In this regard, it may be beneficial to support the diagnosis in patients diagnosed with MRI and by performing urological tests. Bladder dysfunction was detected in three of our patients. The postoperative evaluation revealed no improvement in these complaints (Table 4).

When TCS is clinically suspected, the next step in diagnosis is to perform an MRI. Radiological imaging provides benefit in diagnosis and directs surgical treatment by revealing the accompanying spinal cord anomalies. The presence of diastematomyelia was reported to be present in between 9-35% of patients with ATCS\(^9,14,17,18\). In our series, diastematomyelia was found in 3 out of 9 cases which was consistent with the literature. For these cases, it has been reported that segmentation anomalies of a vertebral corpus such as hemivertebra, butterfly vertebra, and block vertebra are threshold conditions.\(^19\). Therefore, it is important to determine these anomalies in the planning of surgical treatment. The accompanying spinal anomalies are summarized in Table 3. As classical information, the conus medullaris is located below the L2 level, and its downward position is correlated with the symptoms and the complaints of the patients.\(^19-21\). In our series, conus medullaris ends distal to L2 in all cases. However, in recent years it has been reported that TCS may develop with the conus medullaris at the normal position.\(^22\). The data obtained from the samples of filum terminale excised during the operation have been evaluated which suggests that these patients could benefit from surgical treatment.\(^23\). The fact that many series includes patients with conus medullaris located at L2 level or more distally has suggested that ATCS cases with conus medullaris at the normal position might have been skipped.
An important point in management of these cases is to determine the necessity and the timing of the surgical treatment. This question becomes even more important when patients have no neurological deficits and have moderate clinical complaints. Although a consensus is not present in the literature, the prominent opinion is that early surgical treatment could provide good patient management, similar to the pediatric cases. Electromyography and urodynamic tests can be utilized for decision making. Promising results were reported in the recovery of pain and neurological losses. Although recovery of 60% is reported in the long-term results of bladder dysfunction, patients suffering from pain and motor-sensory loss gain major benefits from surgical treatment. Nevertheless, 17% of the patients experienced recurrence of the pain after surgery and 5% of the patients had motor deficit again. In the light of the other studies in the literature and this study, it can be suggested that surgical procedure is an effective method for preventing the progression of the disease and the neurological deterioration.

As a result; ATCS patients, unlike pediatric cases, may consult a physician with an asymptomatic clinical picture. Even if an MRI is performed, the diagnosis may be overlooked. Considering that most of our patients are cases without neurological deficits; we can conclude that in the absence of neurological deficits, surgical treatment can be considered as a good alternative, especially for the amelioration of pain and motor-sensory deficits. Further studies with a larger sample size might lead us to much more precise information for effective patient management.

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