İKSSTD 2020;12(1):46-51
doi: 10.5222/iksstd.2020.13007

Araştırma / *Research Article*

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

Emre Delen
Ahmet Tolgay Akıncı
Osman Şimşek

Erişkin Gergin Kord Sendromlu 9 Olguda Cerrahi Tedavi Sonuçlarımız

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 olgularında cerrahi tedavi sonuçlarını demografik ve klinik özelliklerle birlikte sunmaktır.*
Yöntem: *Kliniğimizde 2004-2015 yılları arasında gergin kord sendromu tanısıyla operasyon geçiren 73 olguyu retrospektif olarak değerlendirdik. Ortalama takip süresi 21.7 (3-63) aydı. Erişkin olgular 18-55 yaş arası 5 kadın, 4 erkek hasta idi. Hastaların klinik ve radyolojik 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 ağrısı yakınması, iki hastada yürüyüş ataksisi ve üç hastada mesane disfonksiyonu vardı. Yalnızca bir hastada kas kuvveti kaybı vardı. Toplam üç hastada cilt bulguları vardı: bunlar iki dermal sinüs traktı ve bir olguda torasik dermatomal kıllanma artışı idi. Dört hastada skolyoz, 4 hastada sringomiyeliya ve 3 hastada split kord malformasyonu tespit edildi. Conus medullaris seviyesi bir hastada L3, 3 hastada L4, 2 hastada L5 ve bir hastada S1 idi. Tüm hastalara kord serbesleştirilmesi operasyonu uygulandı. Hastaların çoğu ağrı seviyelerinde bir azalma olduğunu bildirdi. Ancak duyusal işlev bozukluğu ve sfinkter disfonksiyonununda iyileşme tespit edilmedi.*
Sonuç: *Yetişkin olgular ile pediatrik olgular karşılaştırıldığında temel fark semptomatoloji idi. Ağrı hemen hemen tüm vakalarda kardinal semptomdu. Ameliyat ağrıyı azaltmada etkiliyken, nörolojik defisitlerin düzelmesinde etkin değildi.*

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

Received: 18 June 2019
Accepted: 20 January 2020
Publication date: 31 January 2020

Cite as: Delen E, Akıncı AT, Şimşek O. Surgical treatment results in adult tethered cord syndrome: An experience of 9 cases. İKSSTD 2020;12(1):46-51.

Emre Delen
Trakya Üniversitesi Tıp Fakültesi
Beyin ve Sinir Cerrahisi Anabilim Dalı
Edirne - Türkiye
emredelen1979@yahoo.com
ORCİD: 0000-0001-5573-9492

A. T. Akıncı 0000-0002-9937-076X
O. Şimşek 0000-0002-8716-5187
Trakya Üniversitesi Tıp Fakültesi
Beyin ve Sinir Cerrahisi Anabilim Dalı
Edirne - Türkiye

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 (TÜTF-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 c 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	Sphincterfunction
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

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 waekness, n (%)	1 (11.1%)
Sensory disturbance, n (%)	3 (33.3%)
Sphincter disturbance, n (%)	3 (33.3%)
Gait ataxi, n (%)	2 (22.2%)
Skin lesions, n (%)	3 (33.3%)

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 3. The radiological findings of the patients.

Case No	Age (years)	Gender	Concomitant abnormalities	Conus Level
1	28	Female	SCM+S+Sc+Bc	L4
2	55	Female		L5
3	20	Female	SCM+DS+S+Sc+Btv	L4
4	51	Male		L4
5	18	Male	Sc+Hv+Btv	S1
6	27	Female	SCM+Sc+Hv+Btv	L3
7	30	Male	DS	L3
8	44	Female		L3
9	48	Male	M	L5

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

Table 4. The comparison of preoperative and postoperative neurological scores.

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	4/4	5/5	3/4	5/5	18 months
Case 5	2/5	5/5	5/5	5/5	4/4	26 months
Case 6	3/5	5/5	5/5	4/5	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 [5,14,17,18]. 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 [14,17,19,24]. Electromyography and urodynamic tests can be utilised for decision making [13,25]. Promising results were reported in the recovery of pain and neurological losses [14]. 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 [12]. Nevertheless, 17% of the patients experienced recurrence of the pain after surgery and 5% of the patients had motor deficit again [24]. 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 [1,12,26,27].

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 [27]. 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.

Ethics Committee Approval: Approval was obtained from Trakya University Faculty of Medicine Scientific Research Ethics Committee (01.07.2019 / Decision number: 12/05).
Conflict of Interest: There is no conflict of interest.
Funding: There is no financial support.
Informed Consent: Informed consent was obtained from each patient.

REFERENCES

1. Aufschnaiter K, Fellner F, Wurm G. Surgery in adult onset tethered cord syndrome (ATCS): Review of literature on occasion of an exceptional case. Neurosurg Rev. 2008;31(4):371-83. https://doi.org/10.1007/s10143-008-0140-x
2. Hertzler DA, DePowell JJ, Stevenson CB, Mangano FT. Tethered cord syndrome: a review of the literature from embryology to adult presentation. Neurosurg Focus. 2010;29(1):E1. https://doi.org/10.3171/2010.3.FOCUS1079
3. Klekamp S. Klekamp J, Samii M. Introduction of a score system for the clinical evaluation of patients. Acta Neurochir (Wien). 1993;123(3-4):221-3. Available from:https://europepmc.org/abstract/med/8237513.
4. Klekamp J. Tethered cord syndrome in adults. J Neurosurg Spine. 2011;15(3):258-70. https://doi.org/10.3171/2011.4.SPINE10504
5. Kılıçkesmez Ö, Barut Y, Taşdemiroğlu E. Erişkin gergin omurilik sendromunda MRG bulguları. Tani Girişim Radyol. 2003:9(3): 295-301.
6. Akay KM, Erşahin Y, Cakir Y. Tethered cord syndrome in adults. Acta Neurochir (Wien). 2000;142(10):1111-1115. Available from: https://www.dirjournal.org/eng/makale/815/50/Full-Text https://doi.org/10.1007/s007010070038
7. Yamada S, Lonser RR. Adult tethered cord syndrome. J Spinal Disord. 2000;13(4):319-23. Available from: https://journals.lww.com/jspinaldisorders/Fulltext/2000/08000/Adult_Tethered_Cord_Syndrome.8.aspx https://doi.org/10.1097/00002517-200008000-00008
8. Yamada S, Won DJ, Yamada SM, Hadden A, Siddiqi J. Adult tethered cord syndrome: relative to spinal cord length and filum thickness. Neurol Res. 2004;26(7):732-4. https://doi.org/10.1179/016164104225017929
9. Gupta SK, Khosla VK, Sharma BS, Mathuriya SN, Pathak A, Tewari MK. Tethered cord syndrome in adults. Surg Neurol. 1999;52(4):362-9. Available from: https://www.sciencedirect.com/science/article/pii/S0090301999001214. https://doi.org/10.1016/S0090-3019(99)00121-4
10. Kothbauer K, Seiler RW. Tethered spinal cord syndrome in adults. Nervenarzt. 1997;68(4):285-291. Available from: https://link.springer.com/article/10.1007/s001150050126. https://doi.org/10.1007/s001150050126
11. Garcés-Ambrossi GL, McGirt MJ, Samuels R, et al. Neurological outcome after surgical management of adult tethered cord syndrome. J Neurosurg Spine. 2009;11(3):304-9. https://doi.org/10.3171/2009.4.SPINE08265
12. Lee GYF, Paradiso G, Tator CH, Gentili F, Massicotte EM, Fehlings MG. Surgical management of tethered cord syndrome in adults: indications, techniques, and long-term outcomes in 60 patients. J Neurosurg Spine. 2006;4(2):123-31. https://doi.org/10.3171/spi.2006.4.2.123
13. Vernet O, Farmer JP, Houle AM, Montes JL. Impact of urodynamic studies on the surgical management of spinal cord tethering. J Neurosurg. 1996;85(4):555-9. https://doi.org/10.3171/jns.1996.85.4.0555
14. Düz B, Gocmen S, Secer HI, Basal S, Gönül E. Tethered cord syndrome in adulthood. J Spinal Cord Med. 2008;31(3):272-278. Available from: https://www.tandfonline.com/doi/abs/10.1080/10790268.2008.11760722. https://doi.org/10.1080/10790268.2008.11760722
15. McLone DG. The adult with a tethered cord. Clin Neurosurg. 1996;43:203-209. Available from: https://academic.oup.com/neurosurgery/issue/39/5.
16. Stetler WR, Park P, Sullivan S. Pathophysiology of adult tethered cord syndrome: review of the literature. Neurosurg Focus. 2010;29(1):E2. https://doi.org/10.3171/2010.3.FOCUS1080
17. Hüttmann S, Krauss J, Collmann H, Sörensen N, Roosen K. Surgical management of tethered spinal cord in adults: report of 54 cases. J Neurosurg. 2001;95(2 Suppl):173-8. Available from: https://thejns.org/abstract/journals/j-neurosurg/ 95/2/j-neurosurg.95.issue-2.xml https://doi.org/10.3171/spi.2001.95.2.0173
18. Paradiso G, Lee GYF, Sarjeant R, Hoang L, Massicotte EM, Fehlings MG. Multimodality intraoperative neurophysiologic

monitoring findings during surgery for adult tethered cord syndrome: analysis of a series of 44 patients with long-term follow-up. Spine (Phila Pa 1976). 2006;31(18):2095-102. https://doi.org/10.1097/01.brs.0000231687.02271.b6
19. Pang D, Wilberger JE. Tethered cord syndrome in adults. J Neurosurg. 1982;57(1):32-47. https://doi.org/10.3171/jns.1982.57.1.0032
20. Bauer DF, Shoja MM, Loukas M, Oakes WJ, Tubbs RS. Study of the effects of flexion on the position of the conus medullaris. Childs Nerv Syst. 2008;24(9):1043-5. https://doi.org/10.1007/s00381-008-0640-6
21. Kesler H, Dias MS, Kalapos P. Termination of the normal conus medullaris in children: a whole-spine magnetic resonance imaging study. Neurosurg Focus. 2007;23(2):E7. https://doi.org/10.3171/FOC-07/08/E7
22. Tubbs RS, Oakes WJ. Can the conus medullaris in normal position be tethered? Neurol Res. 2004;26(7):727-31. https://doi.org/10.1179/016164104225017910
23. Selçuki M, Vatansever S, Inan S, Erdemli E, Bağdatoğlu C, Polat A. Is a filum terminale with a normal appearance really normal? Childs Nerv Syst. 2003;19(1):3-10. https://doi.org/10.1007/s00381-002-0665-1
24. Rajpal S, Tubbs RS, George T, et al. Tethered cord due to spina bifida occulta presenting in adulthood: a tricenter review of 61 patients. J Neurosurg Spine. 2007;6(3):210-5. https://doi.org/10.3171/spi.2007.6.3.210
25. Oz O, Ulas UH, Duz B, Yucel M, Odabasi Z. Electrophysiological findings in patients with adult tethered cord syndrome. Turk Neurosurg. 2010;20(1):16-20. Available from: http://turkishneurosurgery.org.tr/pdf/pdf_JTN_714.pdf
26. Lapsiwala SB, Iskandar BJ. The tether ed cord syndrome in adults with spina bifida occulta. Neurol Res. 2004;26(7):735-40. https://doi.org/10.1179/016164104225017956
27. Selçuki M, Mete M, Barutçuoğlu M, Duransoy YK, Umur AS, Selçuki D. Tethered cord syndrome in adults: Experience of 56 patients. Turk Neurosurg. 2015;25(6):922-9. https://doi.org/10.5137/1019-5149.JTN.11700-14.1