

ADULT TETHERED CORD SYNDROME: TWO CASE REPORTS AND LITERATURE REVIEW

ERİŞKİN GERGİN OMURİLİK SENDROMU: 2 OLGU SUNUMU VE LİTERATÜRÜN GÖZDEN GEÇİRİLMESİ

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#### SUMMARY

Tethered cord syndrome is a childhood disease and rarely seen in adults. In adult patient's pain and bladder dysfunction are essential symptoms in respect of children. While evaluating the low back pain, the diagnosis of tethered cord syndrome must keep in mind at adulthood. Surgery performed by an experienced surgeon is satisfactory at symptomatic patients. Here we presented two cases of adult tethered cord syndrome treated with surgery successfully.

Key words: Surgical treatment, adult, tethered cord syndrome.

Level of evidence: Case report, Level IV.

#### ÖZET

Gergin omurilik sendromu bir çocukluk çağı hastalığıdır ve erişkin yaşta nadiren görülür. Erişkin hastalarda çocuklardan farklı olarak ağrı ve mesane disfonksiyonu esas semptomlardır. Erişkin çağda bel ağrısı ayırıcı tanısında gergin omurilik sendromu mutlaka akılda tutulmalıdır. Semptomatik hastalarda deneyimli bir cerrah tarafından yapılacak cerrahi yüz güldürücüdür. Bu makalede cerrahi ile başarıyla tedavi edilen iki erişkin gergin omurilik sendromu hastası sunulmuştur.

Anahtar Sözcükler: Cerrahi tedavi, erişkin, gergin omurilik sendromu.

Kanıt Düzeyi: Olgu sunumu, Düzey IV.

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#### INTRODUCTION

The symptoms of elongated spinal cord and a thick filum terminale was described by Hoffman and colleagues as tethered cord syndrome (TCS) in 1976 <sup>(4)</sup>. This failure is result of the low lying placement of the spinal cord within the vertebral column during embryogenesis. Ischemia, decrease of the electrophysiological activity and deteriorated oxidative process are the pathophysiological factors of TCS. Lipomas, scar lesions, myelomeningoceles, epidermoid tumors, lipomatous or thick filum terminale are the causes of low lying adherence of the spinal cord <sup>(13-14)</sup>.

TCS is a childhood disease and rarely seen in adults. The real incidence of adult TCS is unknown <sup>(3)</sup>. Surgery for TCS patients at the right time after comprehensive evaluation may prevent neurological deterioration at chosen.

In this manuscript we presented adult TCS cases that was presented with low back pain with the light of the literature.

## CASE-1

Twenty years old woman was admitted to the hospital with the complaint of low back pain for 3 years, numbress and weakness in feet for 4 months. Positive Lhermitte sign, 5/5 dorsal flexion weakness at left foot and 3/5 at left foot, perianal hypoesthesia and hypoactive Achilles reflex were detected at neurological examination. Cystometry showed normal urodynamic signs. Whole spine magnetic resonance imaging and lumbar computed tomography showed evidence of low-lying conus ending at L4-L5 level. Syringomyelia at T1 level, diastomatomyelia at S2-S3 level and thick fatty filum was observed. Surgical detethering of the spinal cord was performed. Fatty filum was resected. In early postoperative period her right foot dorsal flexion paresis improved to 3/5.

# CASE-2

Twenty-three years old man was admitted to hospital with right leg and low back pain after a 10 hours long journey. He had perianal hypoesthesia. Lumbar MRI revealed diplomyelia and tethered cord. Surgical detethering was performed. He was discharged with no additional neurological deficit. At 10 months' follow-up his low back and right leg pain was ameliorated.



Figure-1. Preoperative sagittal MRI of the Case-1 (A) and Case-2 (B).

# DISCUSSION

Spina bifida is the second most common childhood abnormality and most frequent spinal cord disorder in children <sup>(2)</sup>. TCS is part of this entitiy that may cause progressive neurologic deficits. The cord extends to the lower end of the sacrum in normal fetal development. The vertebral column lengthens caudad and the conus ascends the canal to reach the L-3 level at about 30 weeks of gestation. If at this stage the conus is trapped at a low level by a short filum, a lipoma, a sagittal septum, fibrous adhesions, normal ascent would be arrested <sup>(10)</sup>.

The pathophysiology of adult TCS is unclear. Why do some patients with TCS remain asymptomatic till adulthood is still unclear? Some authors postulated that the degree of traction determines the age of symptom onset <sup>(10)</sup>. The spinal cord was ended at L4 and L2 level at our cases; but the roots were placed dorsally at L2 that might be a sign of tethered cord.

The asymptomatic patients became symptomatic due to the situations, that may cause sudden spinal cord traction like the lithotomy position during childbirth, movement occurring during road traffic accidents. There were no history of childbirth or accident at our cases. The symptoms were slightly begun and progressed on a long time at the first case and there was a history of a long journey at the second case.

In adult patients pain and bladder dysfunction are essential symptoms in respect of children<sup>(1)</sup>. Low back and leg pain that may spread to genital and rectal area can be the key symptom at adults. Surgery may be beneficial at the patients who has just low back pain or bladder dysfunction. Early surgical treatment in patients without deficit have been reported better results in postoperative outcomes <sup>(6-7)</sup>.

These facts show that surgery give good results in adult patients. Patients may have motor and sensory deficits, what symptoms are numbness, weakness, muscle's atrophy. Approximately 50 percent of patient have bowel and bladder dysfunction, which are presented as constipation and frequency of urination. Our first patient have low back pain, numbness and weakness in feet and observed Lhermitte sign, weakness in dorsal flexion for 5/5 in left leg, 3/5 in right leg, perianal hypoesthesia and hypoactive Achilles reflex. Her right foot dorsal flexion paresis improved to 3/5 at follow-up visits. The second patient had perianal hypoesthesia.

Adults with TCS may be aware of their significant symptoms and this carelessness come to a conclusion with late diagnose and finally disability <sup>(9,11)</sup>. If TCS is not treated this disease may progress 27 % in first, 40 % in second and 60 % in 5th year <sup>(11)</sup>.

Magnetic resonance imaging is the gold standard for the diagnosis of TCS. Visualization of filum terminale, meningomielocele, Syringomyelia, adhesions are mandatory before the operation. Syrinx cavity could be detected. This syrinx may cause pain, motor deficit, atrophy and rarely headaches. 73 % of patients tortured from back pain, 56 % of patients from leg pain <sup>(8)</sup>. There was syringomyelia at T1 level in our first case and the second case had diplomyelia.

Cranial computed tomography for detecting hydrocephalus may be valuable. Urodynamic tests can detect the changes of bladder function. Somatosensory evoked potentials (SEP) and EMG can give useful information <sup>(12)</sup>. SEP is useful to compare pre and postoperative evidence and shows the benefits of surgery <sup>(5)</sup>.

While evaluating the low back pain, the diagnosis of TCS must keep in mind at adulthood. Surgery performed by an experienced surgeon is satisfactory at symptomatic patients.

## REFERENCES

- 1. Akay KM, Ersahin Y, Cakir Y. Tethered cord syndrome in adults. *Acta Neurochir* (Wien) 2000; 142: 1111–1115.
- Cardoso M, Keating RF. Neurosurgical management of spinal dysraphism and neurogenic scoliosis. *Spine* 2009; 34: 1775-1782.
- 3. Garg K, Tandon V, Kumar R, Sharma BS, Mahapatra AK. Management of adult tethered cord syndrome: Our experience and review of literature. *Neurol India* 2014; 62: 137-143.
- 4. Hoffman HJ, Hendrick EB, Humphreys RP. The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. *Childs Brain* 1976; 2: 145–155.
- Kale SS, Mahapatra AK. The role of somatosensory evoked potentials in spinal dysraphism do they have a prognostic significance? *Childs Nerv Sys* 1998; 14: 328– 332.
- Keating MA, Rink RC, Bauer SB, Krarup C, Dyro FM, Winston KR. Neurological implications of the changing approach in management of occult spinal lesions. *J Urol* 1988; 140: 1299-1301.
- Kondo A, Kato K, Kanai S, Sakakibara T. Bladder dysfunction secondary to tethered cord syndrome in adults: Is it curable? *J Urol* 1986; 135: 313-316.
- Lee GY, 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: 123–131.
- 9. McLone DG. The adult with a tethered cord. *Clin Neurosurg* 1996; 43: 203–209.
- 10. Pang D, Wilberger JE Jr. Tethered cord syndrome in adults. *J Neurosurg* 1982; 57: 32-47.

- 11. Phuong LK, Schoeberl KA, Raffel C. Natural history of tethered cord in patients with meningomyelocele. *Neurosurgery* 2002; 50: 989–995.
- Roy MW, Gilmore R, Walsh JW. Evaluation of children and young adults with tethered spinal cord syndrome. Utility of spinal and scalp recorded somatosensory evoked potentials. *Surg Neurol* 1986; 26: 241–248.
- 13. Yamada S, Won DJ, Yamada SM. Pathophysiology of tethered cord syndrome. Correlation with symptomatology. *Neurosurg Focus* 2004; 16: E6.
- 14. Yamada S, Won DS. What is the true tethered cord syndrome? *Childs Nerv Syst* 2007; 23: 371–375.