# Tethered Spinal Cord, Diastematomyelia, and Terminal Syringomyelia in an Adult

## (Report of a case and the review of the literature)

# Erişkin Hastada Gergin Omurilik Sendromu, Ayrık Omurilik Sendromu ve Terminal Siringomiyeli

## (Olgu sunumu ve literatürün gözden geçirilmesi)

ERKAN KAPTANOĞLU, ETEM BEŞKONAKLI, M. ÖZERK OKUTAN, YAMAÇ TAŞKIN

Division of Neurosurgery, Ankara Numune Hospital, Ankara, Turkey

Received: 07.03.2002 ⇨ Accepted: 01.05.2002

**Abstract:** An unusual case of spinal dysraphism in a 74 year old woman is reported. Patient was presented with slight lumbar pain and diagnosed coincidentally. Neurological examination was normal. Magnetic resonance imaging (MRI) revealed tethered spinal cord, diastematomyelia, terminal syringomiyelia, and degenerative changes at lumbar levels. This case is an important example of untreated spinal dysraphism which reached 74 years of age without any complain. The real prevalence of adult asymptomatic spinal dysraphisms is unknown. With the advent of the MRI and long time follow-ups, the real prevalence of spinal dysraphisms may be found.

The authors concluded that, if there is a possibility that tethered cord syndrome and diastometamiyelia may be asymptomatic during wholelife, it should be re-considered whether early surgery in asymptomatic patients is mandatory.

**Key words:** Adult, diastematomyelia, terminal syringomiyelia, tethered cord syndrome

**Running head:** Tethered cord syndrome in adult

**Özet:** Spinal disrafizmi olan 74 yaşındaki bir kadın hasta sunulmaktadır. Hasta hafif bel ağrısı ile başvurmuş ve rastlantısal olarak tanı konulmuştur.

Nörolojik muayene normaldir. Manyetik rezonans görüntülemede gergin omurilik sendromu, ayrık omurilik malformasyonu, terminal siringomiyeli ve lombar vertebralarda dejeneratif değişiklikler saptanmıştır. Bu vaka tedavi edilmemiş spinal disrafizm vakalarına önemli bir örnektir ve 74 yaşına kadar hiçbir yakınımı olmadan ulaşmıştır. Erişkin asemptomatik spinal disrafizm vakalarının gerçek prevalensi bilinmemektedir. Manyetik rezonans görüntüleme ve uzun süreli takipler ile spinal disrafizmlerin gerçek prevalensi bulunabilir. Yazarlar, eğer gergin omurilik sendromu ve ayrık omurilik malformasyonu tüm hayat boyu asemptomatik kalabiliyorsa, asemptomatik hastalarda erken cerrahinin zorunlu olup olmadığının tekrar değerlendirilmesi gerektiği sonucuna varmışlardır.

**Anahtar Sözcükler:** Ayrık omurilik malformasyonu, erişkin, gergin omurilik sendromu, terminal siringomiyeli

## INTRODUCTION

The tethered cord syndrome (TCS) is usually diagnosed in childhood and its onset in adult life is uncommon. Its clinical spectrum comprises low back pain, neurological deficits such as distal motor weakness and trophic and sensory disturbances in legs, urological symptoms and such musculoskeletal signs as scoliosis or foot deformity. The typical patient has a mixed (upper motor neuron and lower motor neuron) sensorimotor deficit involving the lower limbs, usually asymmetrical with pain either in the lower back or the lower limbs. In addition, cutaneous lesions or subcutaneous lipomas in the lumbosacral region may be indirect signs of an intraspinal pathology (17). The late presentation of TCS is possibly related to the degree of tethering and the cumulative effect of microtrauma during flexion/extension (11).

Gupta et al. indicated that there are only two large series of adult TCS in the English literature (11). The patients in these series were diagnosed with myelography and myelo-CT. With the development of MRI, adult TCS series has been increasing in number in recent years (1,3,11,15,16,20,21,23).

The present study discusses a case who has tethered spinal cord and free from symptoms over 74 years. We aim not only to call attention to adult TCS, but also emphasise the treatment strategies in adult patients without complaints. We also ask the question "if spinal dysraphisms may be truly occult during lifetime without any complain, should we operate all asymptomatic patients who diagnosed at their childhood or adulthood".

## CASE REPORT

This 74-year old woman presented with slight low back pain. Patient had no complain related to her lumbar pathology during her life until 2 months before the admission. Pain was not radiating to legs. No alleviating or exacerbating factors were reported. Pain did not effect her daily life and social activities and relieved with simple analgesics and bed rest. She has been on antihypertensive treatment for 7 years and has no urinary or other systemic complaints.

*Examination :* The patient's strength was intact in all muscle groups of both lower extremities and no sensory abnormalities were evident. Straight leg raising (SLR) test were negative bilaterally. Deep tendon reflexes, anal tonus and anal reflex were normal. No pathologic reflexes were elicited. Examination and palpation of the lumbar spine revealed no cutaneous markers, and no focal tenderness was noted. No skin lesion was present. There was no urological complain.

*Neuroimaging :* Plain X-ray films showed degenerative changes of the lumbar vertebral bodies. Lumbar magnetic resonance imaging (MRI) showed low conus medullaris at $L_5$ level and extension of the filum terminale to the $S_4$ level, the tethered spinal cord (Figure1). Figure 2 showed that there was a cystic cavity which was iso-intense with cerebro-spinal fluid in the mid-lumbar region, terminal syringomyelia. Axial $T_2$WI revealed dividing of the spinal cord into two hemicords at $L_3$, diastematomyelia, with widening of the vertebral canal (Figure 3). The vertebral bodies are deformed and compressed at almost all visualised levels. Patient was prescribed simple analgesics and recommended bed rest for two weeks. Patient has been pain free for four months.

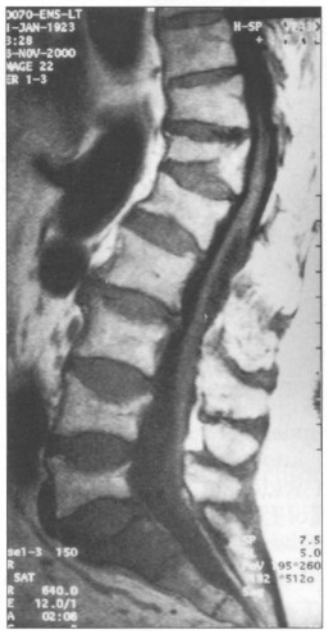

Figure 1: The sagittal $T_1$WI reveals tethered spinal cord extending down to the level of $L_5$. Syrinx cavity is presented at $L_1$-$L_2$ (terminal syringomyelia). The vertebral bodies are deformed and compressed at almost all visualised levels.

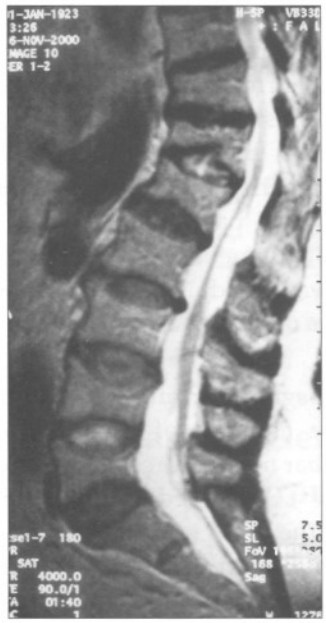

Figure 2: The sagittal T2WI reveals that the conus is as low as $L_5$ and filum terminale extends to the level of $S_4$ (tethered spianal cord). Dilated techal sac is visualised.

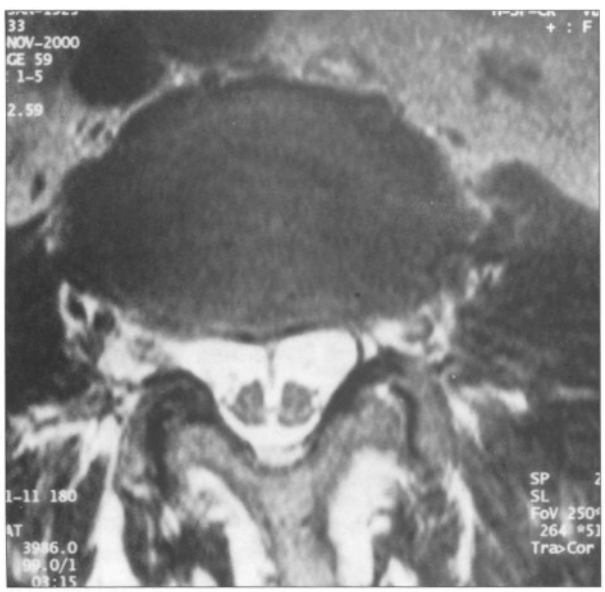

Figure 3: The axial T2WI reveals dividing of the spinal cord into two hemicords (diastematomyelia) with widening of the vertebral canal at the level of $L_{3-4}$.

## DISCUSSION

Tethered cord syndrome in adults is often overlooked. Gupta et al. presented a series of adult TCS and emphasised that there were only two large series of adult TCS in the English literature in 1999 (11). The patients in these series were investigated with myelography and myelo-CT. With the increasing use of MRI, TCS has become an appreciable finding. Numerous reported cases show that the prevalence of adult-onset of tethered cord syndrome may be higher than previously assumed (1,3,4,6,10,11,15,16,20,22,23). However, The real data about asymptomatic patients is not known.

Late neurological deterioration due to the tethering of the spinal cord, termed "tethered spinal cord syndrome," is widely recognised in patients with spinal dysraphism (14). Here, presented case has not deteriorated in her life. She experienced only a slight low back pain which was responsive to bed rest and simple analgesics. It was possible to refer patient's pain to the degenerative changes seen on MRI, because it was relieved by non-steroid anti-inflammatory drugs and bed rest. Hence, we did not name this case as a tethered cord syndrome. Thus, we consider this case as an adult with tethered spinal cord which was diagnosed coincidentally.

**Clinical Features:** In the series, almost all of the patients admitted to the hospital with the complaints of pain, neurological deficits, and urological symptoms (1,3,6,11,15,20,21). The pain was described as severe back and leg pain (23).Akay et al. also presented a series in which all the patients have an additional symptoms to their low back pain such as leg pain or neurological findings (1). Pang et al. emphasised that pain was the most common presenting symptom (20). In his report, low back pain was associated with leg pain, perineal pain, and radicular in character. The straight-leg raising test was positive. We did not consider our patient's pain was a symptom of onset, because it was not associated with a neurologic symptom, or it was not radiating, and there was no accompanying urological symptom.

Gupta et al. reported that cutaneous lesions were present in 11 of 18 patients (11). However, in the series of Pang and Wilberger, this was present in only 40% of adult patients (20). In the presented case, cutaneous lesion was absent. This is the one of the most important findings that brings patient to the hospital even there is no neurologic symptoms.

**Pathogenesis:** An abnormally lowlying conus is described as that situated below the $L_1$-$L_2$ disc space or the inferior aspect of the L2 (11). The degree of traction of the conus is said to determine the age of onset of symptoms (20). In cases of marked tethering and severe stretching of the conus, neurological disturbances appear in infancy or early childhood. A lesser degree of tethering may cause only minor or nonprogressive deficits in childhood. Minimal tethering may remain subclinical in childhood until later aggravated factors reported by Pang et al.(20) But, Gupta et al. reported that there was only one patient presented precipitating symptom in 18 patients in their series (11). Some other authors explained the mechanisms of late onset of TCS in adults. As a person with a tight conus grows older, the cumulative effect of repeated cord traction from years of natural head and neck flexion could ultimatively lead to injury to the conus (12). Dubovitz et al. reported that direct trauma to the back precipitates the symptoms causing deformation of the marginally functioning neuronal elements within the stretched cord (5). Yamada et al. stated that neurological dysfunction in patient with tethered cord correlates with mitochondrial anoxia within the conus (24). Developmental lumbar cord stenosis and this prolapse are also known to precipitate symptoms of TCS (7,8). In our case, conus situated at L5 and extension of the filum terminale is to the level of $S_4$. There is no doubt that cord is tightly fixed to the lower lumbar and sacral area. There is also an additional factor that fixes cord with a septa at $L_3$, a diastematomyelia. Erkan et al. reported that common clinical findings in diastematomyelia were found to be more distinctive in syrinx-associated diastematomyelia cases (6). It is surprising in our case that such a tight fixation and low conus with terminal syringomyelia did not cause any symptoms during her 74 years of age. We accepted this case as asymptomatic and coincidentally diagnosed. Because it is very likely that a patient may have lumbar pain with such degenerated vertebrae. Pain also relieved with simple analgesics and bed rest.

It is difficult to understand the mechanisms how the patient tolerated such severe pathology. This is the only asymptomatic adult case who have tethered spinal cord, diastematomyelia, and terminal syrinx, in the literature. We considered any relation with height of the patient, but no literature was found showing these relation.

**Imaging:** Tethering of the spinal cord can be resulted from intradural lipomas, diastematomyelia, thickened tight filum terminale, dermal sinus tracts, intradural adhesions, and adhesions after myelomeningocele operations (11). MRI is the choice of diagnostic tool to demonstrate all these lesions. In the presented case, MRI demonstrates a combination of low lying conus (tethered spinal cord), diastematomyelia and terminal syringomyelia.

**Treatment:** Hood et al. did not recommend surgery until neurological deterioration (13). Pang et al. also indicated that surgery is essential in tethered cord syndrome, in symptomatic patients (20). The surgical outcome is gratifying in relation to pain and motor weakness but disappointing in the resolution of bowel and bladder dysfunction (2,9,11,20). Gupta et al. recommended that surgery should be offered to all adult patients with TCS, once the diagnosis is established, even if the patient has no neurologic deficit (11).

We should classify adult patients as tethered cord syndrome (TCS) who have symptoms and as tethered spinal cord patients who have no symptoms at all. We recommend early surgery in adult TCS according to Akay et al., Basar et al., Giddens et al., Hood et al., Iskandar et al., and Pang et. al. (1,2,9,13,15,20). We should find out the real prevalence of asymptomatic adult tethered spinal cord patients to conclude weather early surgery is necessary in asymptomatic patients at any age.

**Classification:** Gupta et al. classified patients into two categories (11). Patients with progressive symptoms since childhood and those who had tethering secondary to previous meningocele surgery were excluded. First category of patients those who were symptomatic for the first time in the adult life. Second category included those who were diagnosed in childhood but presenting in adulthood with new or progressive symptoms. Same classification was also made by Pang et al. (20). McLone divided adults with a TCS into three groups (19). The occult group with known cutaneous markers, but neglected. Second group, truly occult cases with no skin lesions who appear to be intact over years and then, as adolescent or young adult begin to develop neurological,

orthopedic, and urological deficits, The third and the largest group is the postrepair group. Although Koyanagi et al. reported that none of the patients with TCS older than 5 years of was asymptomatic, we recommend to add the fourth group to McLone's classification, which has tethered spinal cord with or without accompanying pathologies who never had any symptoms in their entire life (18). We are not agree that the third group is the largest, because we do not know the real prevalence of adult asymptomatic occult spinal dysraphism. There may be more undiagnosed adult patients than we assume. There is no discussion in the literature about asymptomatic coincidentally diagnosed adult tethered spinal cord patients reached 7th or 8th decade.

Here, we would like to emphasise another hypothesis, "If this patient had been diagnosed in her childhood, she would have been operated on". Thus, there would have been a possibility of deterioration because of retethering after operation (14). We now clearly see that this operation would have been unnecessary. Even there is only one case, this patient proves us that there is a possibility of an asymptomatic life with tethered spinal cord and diastematomyelia with a such tight filum terminale. We concluded that asymptomatic patients may be followed-up until neurological deterioration develops, if it would. We should not make our final conclusions on the timing of surgery in asymptomatic patients at any age until we know the real prevalence of asymptomatic adult cases.

**Correspondence:** Erkan Kaptanoğlu
Division of Neurosurgery,
Ankara Numune Education and
Research Hospital, Ankara, Turkey
Phone : +90-532-435 1057
E-mail : ekaptanoglu@hotmail.com
Fax : +90-312-312 6876

## REFERENCES

1. Akay KM, Erşahin Y, Çakır Y: Tethered cord syndrome in adults. Acta Neurochir (Wien) 142:1111-1115, 2000
2. Başar H, Aydoğanlı L, Yüksel M, Başar M, Akdemir G, Kaptanoğlu E: The outcome of urological findings in operated tethered cord patients. Int Urol Nephrol 29(2):167-171, 1997
3. Begeer JH, Staal-Schreinemachers AL: The benefits of team treatment and control of adult patients with spinal dysraphism. Eur J Pediatr Surg 6, Supp 1:15-16, 1996
4. Caruso R, Cervoni L, Fiorenza F, Vitale AM, Salvati M. Occult dysraphism in adulthood: a series of 24 cases. J Neurosurg Sci 40:221-225, 1996
5. Dubowitz V, Lorber J, Zachary RB: Lipoma of the cauda equina. Arch Dis Child 40:207-13, 1965
6. Erkan K, Unal F, Kiris T: Terminal sryngomyelia in association with tethered cord syndrome. Neurosurgery 45(6):1351-1360, 1999
7. Freeman LW: Late symptoms from diastematomyelia. J Neurosurg 18:538-41, 1961
8. Garcia FA, Kranzler LI, Signeria EB: Diastematomyelia in an adult. Surg Neurol 14:93-4,1980
9. Giddens JL, Radomski SB, Hirshberg ED, Hassouna M, Fehlings M: Urodynamic findings in adults with the tethered cord syndrome. J Urol 161:1249-1254, 1999
10. Gokay H, Barlas O, Hepgul KT: Tethered cord in the adult mimicking the lumbar disc syndrome: report of two cases. Surg Neurol 39:440-442,1993
11. Gupta SK, Khosla VK, Sharma BS, Mathuriya SN, Pathak A: Tethered cord syndrome in adults. Surgical Neurol 52(4):362-370, 1999
12. Guthkelch AN: Diastematomyelia with median septum. Brain 97:729-42, 1974
13. Hood RW, Riseborough EJ, Hehome AM: Diastematomyelia and structural spinal deformities. J Bone Joint Surgery 62A:520-8, 1980
14. Inoue HK, Kobayashi S, Ohbayashi K, Kohga H, Nakamura M: Treatment and prevention of tethered and retethered spinal cord using a Gore-Tex surgical membrane. J Neurosur 89:689-693, 1994
15. Iskandar BJ, Fulmer BB, Hadley MN, Oakes WJ: Congenital tethered spinal cord syndrome in adults. J Neurosurgery 88:958-961, 1998
16. Kaplan JO, Quencer RM: The occult tethered cord syndrome in the adult. Radiology 137:387-91, 1980
17. Kothbauer K, Seiler RW: Tethered spinal cord syndrome in adults. Nervenarzt 68(4):285-91, 1997
18. Koyanagi I, Iwasaki Y, Hida K: Surgical treatment supposed natural history of the tethered cord with occult spinal dysraphism. Childs Nerv Syst 13:268-274, 1997
19. McLone DG: The adult with a tethered cord. Clin Neurosurg 43:203-209, 1996
20. Pang D, Willberger JE. Tethered cord syndrome in adults. J Neurosurg 57:32-42, 1982
21. Ratliff J, Mahoney PS, Kline D: Tethered cord syndrome in adults. South Med J 92(12):1199-203, 1999
22. Satar N, Bauer SB, Shefner J: The effects of delayed diagnosis and treatment in patients with an occult spinal dysraphism. J Urol 154:754-758, 1995
23. Yamada S, Lonser LL. Adult tethered cord syndrome. J Spinal Disord 13(4):319-23, 2000
24. Yamada S, Zinke DE, Sanders D: Pathophysiology of "tethered cord syndrome" J Neurosurg 54:494-503, 1981