A Case of Maternal Spina Bifida Associated with Tethered Spinal Cord and Mode of Delivery

Bir Maternal Spina Bifida ile İlişkili Gergin Spinal Kord Sendromu Olgusunda Doğum Şekli

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ABSTRACT

Tethered spinal cord syndrome (TCS) or occult spinal dysraphism sequence refers to a group of neurological disorders related to malformations of the spinal cord. Spinal lipomas and cutaneous stigmata are the most common manifestations of the occult spinal dysraphic lesions. The spinal cord normally hangs loose in the spinal canal, free to move up and down with growth, bending and stretching,while a tethered cord is held taut at the end.
A 41 year-old 35 weeks and 4 days pregnant woman with a lumbosacral giant lipoma associated with spina bifida and tethered cord syndrome was admitted to our clinic, an uneventful vaginal birth was achieved, and this rare case is hereby presented with MRI findings. *(Gazi Med J 2012; 23: 164-6)*

Key Words: Open neural tube defects, spina bifida, tethered cord syndrome, parturition

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ÖZET

Gergin spinal kord sendromu (GSK) veya okült spinal disrafizm sekansı , spinal kord malformasyonları ile ilişkili bir grup nörolojik bozukluktur. Okült spinal disrafik lezyonlara en sık eşlik eden bulgular spinal lipomlar ve cilt bulgularıdır. Spinal kord normalde kolumna spinalis içinde serbest bir halde bulunup rahatlıkla büyümeyle birlikte yukarı-aşağı hareket ederek gereğinde bükülüp, gerilebilir. Gergin spinal kord ise uç kısmından sıkıca tutturulmuştur. 41 yaşındaki 35 hafta 4 günlük gebeliği olan hasta lumbosakral dev lipom ve buna eşlik eden spina bifida ve gergin kord sendromu ile başvurmuş ve vajinal yolla doğumu gerçekleştirilmiş olup nadir görülen bu olgu MRI görüntüleri eşliğinde sunulmuştur. *(Gazi Med J 2012; 23: 164-6)*

Anahtar Sözcükler: Açık nöral tüp defektleri, spina bifida, gergin spinal kord sendromu, doğum

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INTRODUCTION

Tethered spinal cord syndrome (TCS) has been defined as a spectrum of congenital anomalies resulting in an abnormally low position of the conus medullaris (1).

Most commonly, TCS is related to spinal dysraphism. The signs and symptoms correlate with the radiological definition in which the conus medullaris is anatomically lower than the L-2 vertebra or below the L1-2 disc space (2).

Magnetic resonance imaging (MRI) is the gold standard in the detection of TCS. Sagittal T1- and T2-weighted images are best for localizing the level of the conus, whereas T1- weighted axial MRI images are better for measuring the diameter of the filum and demonstrating the presence of fat within the filum terminalis. MRI also shows the associated lesions as a cause of tethering (3).

Although mostly diagnosed and detethered in infancy and early childhood period, with manifestations

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of orthopedic deformities in the form of gait disturbances and scoliosis, motor-sensory loss or sphincteric dysfunction with urinary-fecal incontinence, sometimes the pathology might be overlooked due to the lack of manifestations mentioned or the means of detection.

In this case, the patient may continue into adulthood with minor signs and symptoms or only cutaneous manifestations such as hypertrichosis, capillary hemangioma, dermal sinus tract, subcutaneous lipoma or an asymmetric gluteal cleft (4).

Women during their childbearing ages might present with untreated TCS with or without spinal dysraphism and their mode of delivery might present a dilemma to the clinician.

CASE REPORT

A 41 year old gravida 1, para 0, 35-week- 4 days pregnant woman was admitted to our clinic with premature rupture of the membranes, 2 cm cervical dilatation and 30% effacement. The amniotic fluid was clear and there was no sign of fetal distress in the recordings of cardiotocography.

She had hypothyroidism and was on 2x0.1 mg/day levothyroxin treatment.

Due to advanced maternal age, amniocentesis was performed at 18 weeks 5 days, when the normal number and structure of fetal karyotype were found.

Detailed ultrasonography at 22 weeks 5 days of the pregnancy with special attention to the fetal spinal column demonstrated no sonographically visible pathology.

On physical examination , she had a large bulge on her lumbosacral region about 23 cm to 15 cm and a tuft of hair over the skin (Figure 1).

The patient wassent for consultation to the department of neurosurgery and they reported that there was no motor or sensory deficit and the deep tendon reflexes were normal.

We can now retrospectively state that she was a neglected case of spinal dysraphism because there were no previously taken MRI scans to discern whether there was only a giant sacral lipoma or a true spinal defect.

The uterine contractions were sufficient to cause progressive cervical dilatation and she soon entered the active phase of delivery. Until the consultations and arrangements were made, the cervical dilatation and effacement were 5 cm and 60% respectively.

As the patient did not have any motor defect which might have hindered pushing efforts in the second stage of labor, vaginal delivery chance was estimated to be high and the patient was closely monitored .

In the second stage of labor, she was not allowed to push forcefully and a vacuum extractor was held ready for use but not needed. A sufficiently large episiotomy was performed in an effort to protect the perineum and to ease the passage of the fetus.

The delivery was uneventful, 6 hours 15 minutes after the admission, 2300 grams, 46 cm male fetus was delivered vaginally with APGAR scores of 8 and 10 for the first and fifth minutes respectively. Examination of the newborn by the pediatrician was reported to be normal.

Postpartum MRI scans were performed and MRI at T1 and T2 weighted sagittal images demonstrated that there was an open spina bifida at S1 level associated with the conus medullaris at L5 level, the thickened filum terminalis and the peripheral lipomatosis from L3 to L5, indicating a case of TCS (Figure 2, 3).

DISCUSSION

Formation of the neural tube begins by the process of neurulation at 18 to 28 days of gestation. This process progresses in a coordinated manner when the ectoderm over the notochord proliferates to form the neural plate, which later bends into the neural folds. Each fold later fuses in the midline to form the closed neural tube (5).

This process continues in the craniocaudal direction between 22-23 days to 25-27 days of embryologic life . The openings that are formed at the cranial and caudal regions are termed the cranial and caudal neuropores. The cranial neuropore closes approximately on day 24 and the caudal neuropore on day 26. Failure of the caudal (posterior) neuropore closure results in spina bifida.

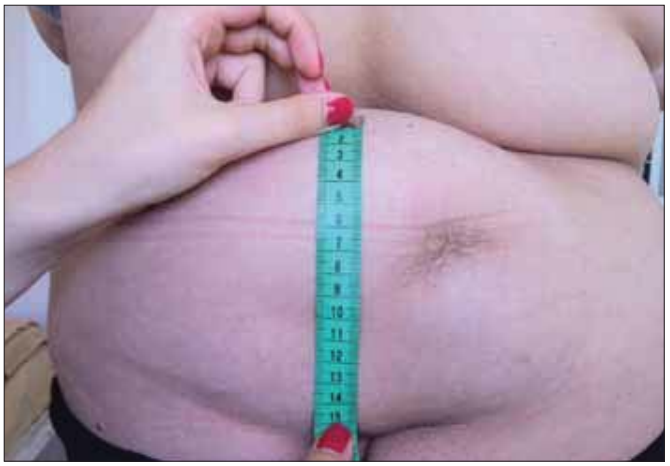

Figure 1. Lumbosacral giant lipoma

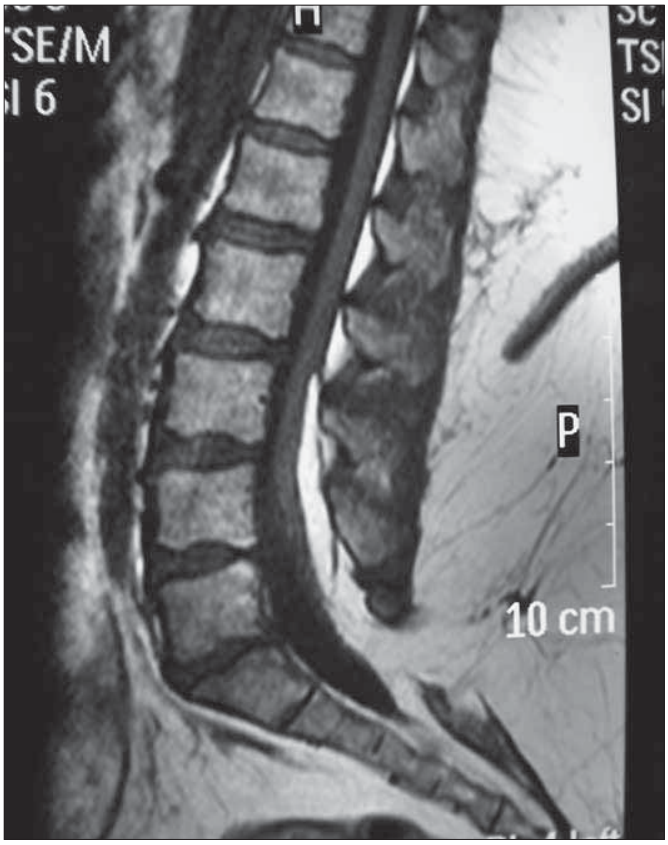

Figure 2. T1 weighted image of maternal lumbosacral spine demonstrating the presence of an open neural tube defect at the level of S1 vertebra , the low lying conus medullaris at L5 and lipomatous accumulation at the end of the thickened filum terminalis

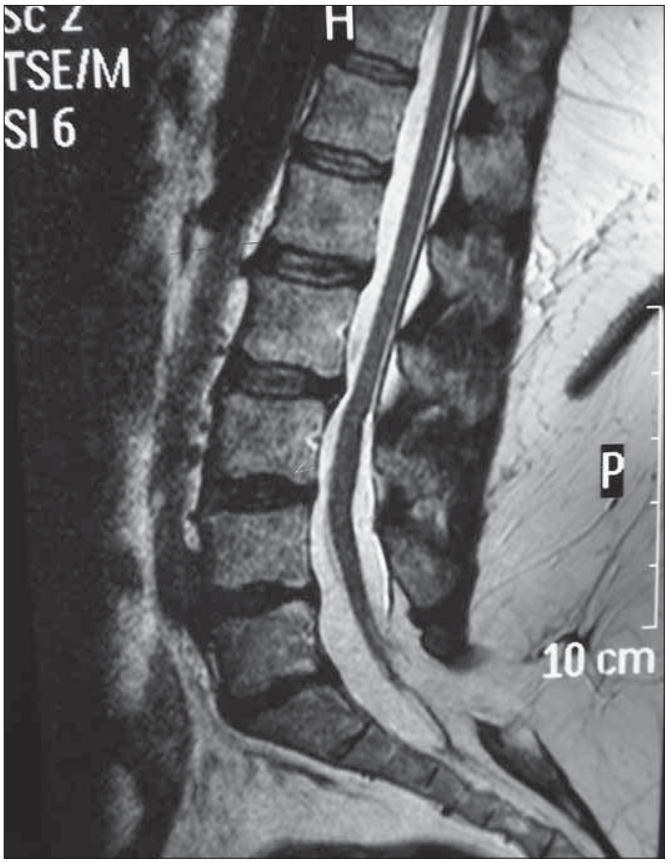

Figure 3. T2 weighted image of maternal lumbosacral spine demonstrating that the spinal cord is held taut at the end, namely, ''tethering"

Between 43 to 48 days, the ventriculus terminalis is formed at the caudal end of the neural tube and is the site for the conus medullaris to develop (6).

Due to the disproportionate growth rate of the vertebral column to the spinal cord, the neural tissue distal to the ventriculus terminalis lenghtens and forms the filum terminale. Hence the conus terminalis continues to ascend in the postnatal period , reaching the adult level of L1-2 by about 3 months of age.

Anomalies located in the dorsal aspect of the lumbosacral spinal cord, such as myelomingoceles, lipomyelomeningoceles, lipomas, and meningocele manqué, may cause a combination of TCS and local effects on the spinal cord.

An elongated cord and a thick terminal filum may be related to an increased tension that occurred in the embryonic stage, and these features support the diagnosis of TCS if the signs and symptoms localize the lesion to the lumbosacral cord.

Due to the anchoring effect of caudal lesions hindering the spinal cord from freely moving, the oxidative metabolism in the taut spinal cord is impaired by intermittent stretching (7).

Symptoms related to a congenital tethered cord occur most commonly in childhood, so it was initially regarded as a pediatric problem; but sometimes the neurological dysfunction may not be seen until adulthood.

Signs and symptoms related to TCS depend on the presence of associated spinal dysraphism and the age of presentation. In infancy, cutaneous manifestations, orthopedic deformities, scoliosis, spasticity and absence of deep tendon reflexes might be seen.

Children and toddlers may present with both motor and sensory deficits, arrest in the developmental stages, loss of bladder control or anal incontinence and in adulthood, the exacerbation of already present pain, aggravation of sphincteric dysfuntions, and sometimes new onset diagnosis of TCS might be suspected by a history of sexual dysfunction. In the absence of spina bifida, pain is the most common presentation (6, 8).

Pang and Wilberger stressed the fact that the onset of symptoms might be related to the degree of cord traction rather than the level of tethering or origin of lesions. Natural head and neck flexion as minor trauma over time has been indentified as a contributor to the onset of symptoms in a tethered spinal cord (9).

The effect of pregnancy and childbirth as a cause of trauma to TCS remains to be determined. It is not elucidated whether pregnancy related stretching of the spinal cord contributes to the aggravation of already present symptoms or causes TCS to become symptomatic.

In a study to describe the antenatal complications, mode of delivery and outcome of pregnancy in women with spina bifida, the duration of mean gestation was 36.6 weeks, with 12 cesarean and 11 vaginal deliveries. Wheelchair dependent patients were more prone to have cesarean delivery with a 1 in 5 vaginal delivery ratio. However, vaginal delivery was possible in 10 out of 18 pregnancies in independently mobile women (10).

Under continuous monitorization and close observation of the mother and the fetus, we chose vaginal delivery after consulting with the neurology and neurosurgery departments, as earlier reports indicate that the vaginal route of delivery in pregnancies with maternal spina bifida without neurologic deficits is possible in more than half of cases.

From here we can deduce that pregnancies complicated by spina bifida are likely to have preterm delivery and, as the level of neurologic deficit decreases, vaginal delivery is more likely to occur and the authors concluded that women with spina bifida who become pregnant generally have positive outcomes.

Conflict of Interest

The authors have no conflict of interest to declare.

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