Case Report / Olgu Sunusu

Tethered Cord Syndrome

Tethered Cord Sendromu

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ÖZET

Tethered Cord sendomu (TCS) omuriliğin gerilmesiyle nöral doku iskemisinin geliştiği ve sonunda ilerleyici nörolojik kayıpların geliştiği bir hastalıktır. TCS'li çocuklar gece ve gündüz idrar kaçırma, sık idrara çıkma ve sık üriner enfeksiyon geçirmektedirler. Belirti ve bulgular doğumdan itibaren bulunabileceği gibi, çoğunlukla zaman içinde ortaya çıkmaktadır. Korkulan durum nörojenik mesaneye (NM) bağlı renal hasar ve kronik böbrek yetmezliğidir (KBY). Bu makalede, küçüklüğünden beri gece-gündüz idrar kaçıran ve sonra böbrek yetmezliği tablosu ile gelen, TCS'ye bağlı KBY gelişen 14 yaşında bir kız hasta sunulmuştur.

Anahtar Kelimeler: *idrar kaçırma, Tethered Cord sendomu, nörojenik mesane, böbrek yetmezliği*

ABSTRACT

Tethered Cord syndrome (TCS) is a disease in which neural tissue ischemia develops as a result of stretching of the spinal cord and eventually progressive neurological loss develops. Children with TCS have urinary day-night incontinence, frequent urination and frequent urinary infection. Signs and symptoms can be present from birth, but often occur over time. The feared condition is renal damage due to neurogenic bladder (NM) and chronic renal failure (CRF). In this article, we present a 14-year-old female patient who developed CRF due to TCS, who had leaked urine during day and night since her childhood and later presented with renal failure.

Keywords: *urinary incontinence, Tethered Cord syndrome, neurogenic bladder, renal failure*

INTRODUCTION

The conus medullaris, which is at the coccyx level in the 25th week of intrauterine life, rises to the 3rd lumbar vertebra at birth and to the lower end of the 1st lumbar vertebra after the age of 2. (1) During this elevation of the conus medullaris, it adheres to the surrounding tissues and remains below the lumbar 1st vertebra. Afterwards, neurological, urological or orthopedic symptoms occur with stretching of the phylum terminale. (2,3,4) Although the clinical findings vary according to age, it is mostly in the form of abnormalities in urine and stool habits, skin abnormalities in the waist region and foot deformities. 5) Because of the development of NM and CRF due to TCS, early diagnosis and treatment is very important. Here, such a girl case with TCS is presented.

CASE

A 14-year-old girl with normal development presented with day-night urinary incontinence since her childhood. She had urge incontinence. The stool habit was normal. A year ago, he had operated on his right foot pes equinovarus. (Figure 1) The patient had a 1 cm-diameter sacral dimple on the left hip. (Figure 2) There was no neurological abnormality. Her urea was 87.5 mg/dl, creatinine 2.8 mg/dl, hemoglobin level was 9.4 g/dl and she had metabolic acidosis. Renal ultrasonography showed bilateral hydroureteronephrosis, thinning of the renal cortex, increased bladder wall thickness and irregularity. There was no vesicoureteral reflux but the bladder was neurogenic (irregular and reduced capacity) (Figure 3). Urodynamic examination revealed low-capacity high-pressure NM. Lumbosacral magnetic resonance imaging revealed TCS (Figure 4) and she was operated for it. The patient was initiated clean intermittent catheterization at regular intervals and anti-cholinergic treatment. She was followed up with diagnosis of CRF. Bladder augmentation surgery was performed by pediatric urology. Renal functions gradually deteriorated during follow-up. Our patient is now 18.5 years old and is in predialysis period. Although her general condition is stable, creatinine level is 4mg/dl and urea level is 114 mg/dl.

DISCUSSION

TCS is usually a childhood disease. But it can also be seen in adults. TCS is a disease characterized by progressive neurological loss caused by stretching of the lumbosacral spinal cord due to congenital or acquired causes and more common in women. The rapid growth in children aggravates the condition. (1,2) Our case was a girl whose complaints started at an early age.

TCS can be accompanied by fibrous bands, diastometamyelia, meningomyelocele, short thick phylum terminale, meningomyelocele and lipomyelomeningocele. Our patient also had diastometamyelia. Motor loss, urological symptoms, spinal deformities such as scoliosis, foot deformities (such as pes equinovarus), trophic ulcers and skin symptoms are more common in childhood TCS. (1,2) Skin findings are seen in 80-100% of children. These symptoms provide important clues for early diagnosis before neurological loss develops. (6) Our patient also had urological symptoms, NM, vertebral anomaly, right pes equinovarus and sacral dimples on the left hip.

It is difficult to evaluate the sphincter dysfunction of the bladder during infantile period. Urodynamic tests should be performed in patients who have day-night urinary incontinence after 4-5 years of age, frequent urination and urinary infection. (7,8,6)

The definitive diagnosis of TCS is made by MRI. The aim of treatment in TCS is to eliminate the pathology leading to stretching of the spinal cord and to prevent damaging of healthy neural structures. Spinal MRI should be performed on patients suspected of TCS. (3,6)

In conclusion, neurological findings in TCS are progressive and diagnosis should not be delayed and surgical treatment should be performed as early as possible. In addition, urinary system functions should be carefully monitored. Even the presence of a small sacral dimple in patients with voiding dysfunction may be a warning sign for primary disease.

References

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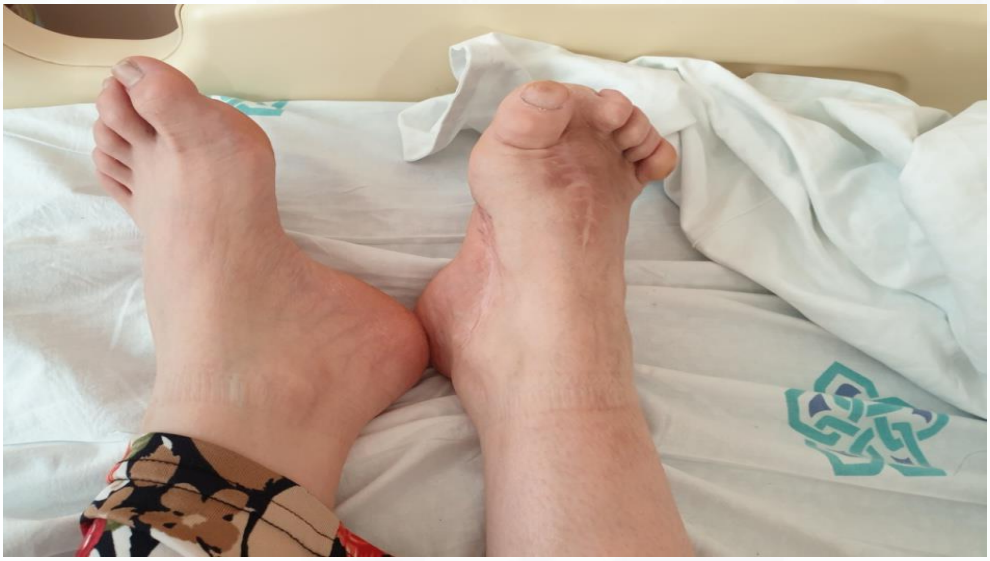

Figure 1- Operated right pes equinovarus

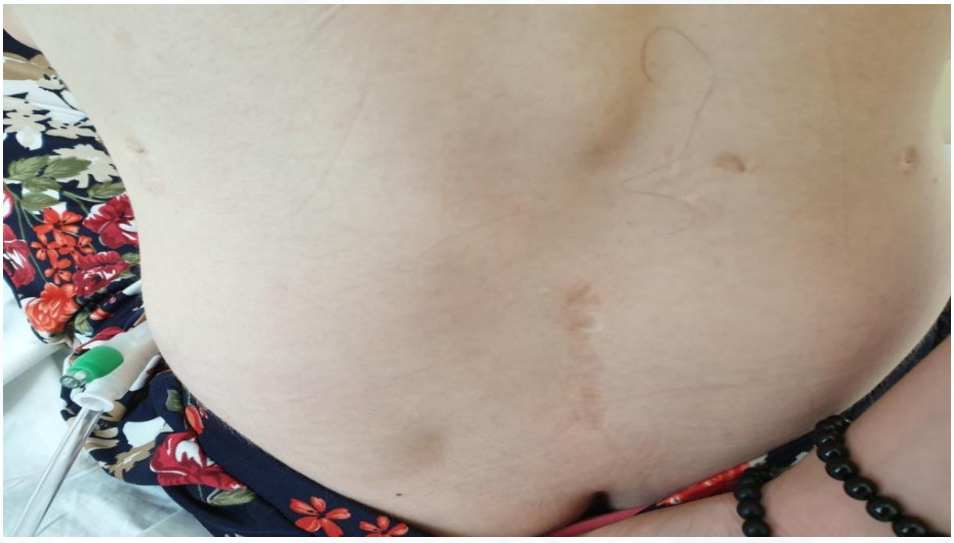

Figure 2 – The TCS operation scar in lumbosacral region and sacral dimple on left hip

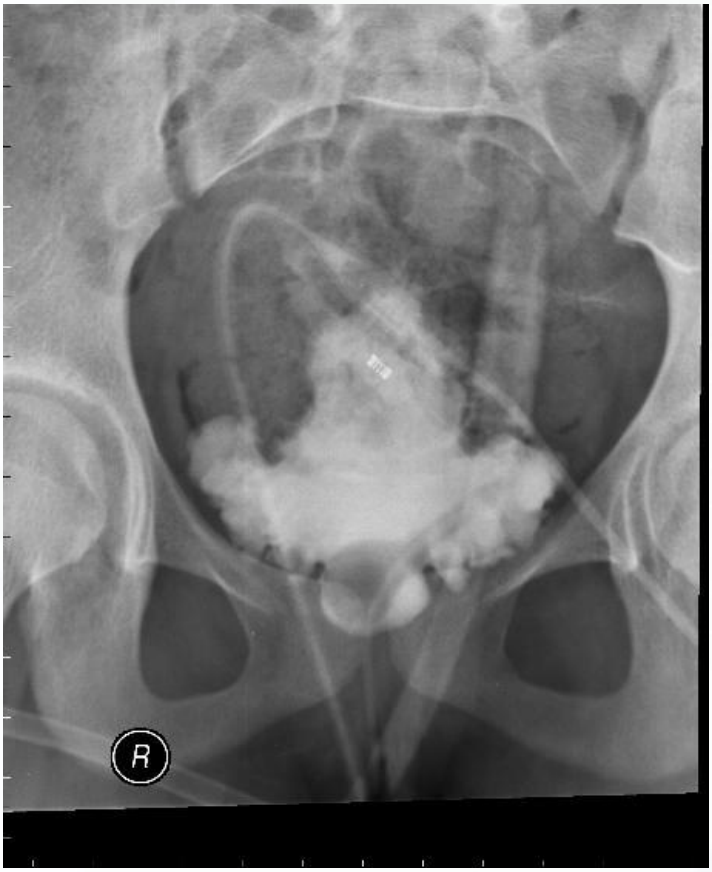

Figure 3 - Irregular neurogenic bladder with reduced capacity

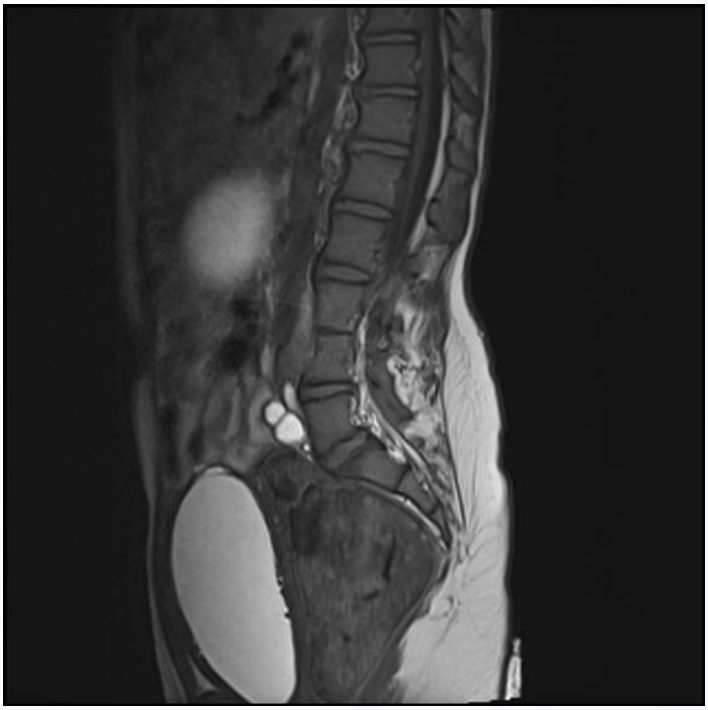

Figure 4 - Tethered cord (Conus medullaris terminated at L4 vertebrae and adherent to the posterior), the arcus fusion defect in L3-4-5 vertebrae and sacral bones, diastometamyelik appearance in the L1-4 vertebrae