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Clinical Study

Tethered cord syndrome: A study of 35 patients

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KEYWORDS

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Abstract *Objectives:* Tethered cord syndrome (TCS) is a stretch-induced functional disorder of the spinal cord in which the caudal part is anchored by an inelastic structure. The majority of cases are related to spinal dysraphism. The presentations differ according to the underlying pathological condition and age, with pain, cutaneous signs, orthopedic deformities and neurological deficits being the commonest. Our aim was to determine the presentations in Saudi patients and to study the natural history of untreated late presenting cases.

Methods: This was a retrospective study of 35 consecutive Saudi patients with TCS seen at a single institution over a period of 7 years. The frequency and presentation of each type of spinal dysraphism and the natural history of untreated, late-presenting cases were studied.

Results: The most frequent spinal dysraphism associated with TCS was lipomeningomyelocele in 12 patients (34.3%), followed by TCS secondary to myelomeningocele in 8 patients (22.8%). Dermal sinus was found in 5 patients, diastematomyelia in 4 patients, meningocele in 3 patients and thick filum terminale in 3 patients. Thirteen patients out of 19 patients over 2 years of age (68.4%), presented with progressive neurological deficits.

Conclusion: Patients suspected of having TCS must be referred and treated by the age of 2 years, or soon after diagnosis, as they are likely to develop progressive neurological deficits if untreated. Normal radiology in the presence of clinical features of cord tethering should not exclude the diagnosis of TCS.

Introduction

Tethered cord syndrome (TCS) is an increasingly recognized clinical disorder, which is usually identified in childhood and is defined as a stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure.[1–3] The disproportionate longitudinal growth between the vertebral column and the tethered spinal cord results in stretching of the conus medullaris and nerve roots. The accepted cause

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of TCS is spinal cord traction, which leads to anatomic and metabolic disorders that are responsible for the clinical presentation.[4] Fixation of the spinal cord can occur congenitally (primary TCS) or in association with other intraspinal pathologies or postoperative scarring (secondary TCS).[5] Most cases are related to spinal dysraphism.

The age at presentation of TCS ranges from early childhood to the 80s.[6] The symptoms related to a congenital tethered cord occur most commonly in childhood, but in many patients, the diagnosis is not established until adulthood.[2] The presentation may also differ according to the underlying pathology, with back and leg pain, cutaneous signs, orthopedic deformities, muscle weakness, urinary dysfunction and neurological deficits being the most common.[7–10] Other manifestations include gait deterioration, foot deformities, progressive scoliosis, and sphincter incontinence.[11] Young children may have skeletal growth deformities, such as a leg-length discrepancy. Adolescent or young adult patients often complain of unexplained long-term back pain.[12–14]

The findings at physical examination may be unimpressive or include a combination of lower and upper motor neuron abnormalities, in addition to one or more of the cutaneous signs associated with occult spinal dysraphism, such as an abnormal tuft of hair in the lumbosacral area ('fawn tail'), fatty masses associated with lipomeningomyelocele or perhaps deep dimples or sinus tracts, or small angiomatous-appearing birthmarks in the mid-lumbar area.[15] The diagnosis can be difficult due to the subtleness of the signs and symptoms, which may be easily overlooked.[8]

Spinal ultrasound in neonates and magnetic resonance imaging (MRI) in older children allow ready diagnosis of TCS and will almost always reveal the cause.[13,15] In infants, a high-resolution ultrasound examination can also be a good screening test. Radiological abnormalities such as low lying conus medullaris, lumbosacral lipoma, filum terminale lipoma, or thick filum terminale are the keys for the diagnosis of TCS.[16]

Surgical untethering should be considered upon appearance of upper motor neuron signs or progression of lower motor neuron signs. In cases of myelomeningocele, low-lying conus is commonly seen without clinical evidence of cord tethering; therefore, radiographic imaging cannot be the sole basis for the diagnosis.[11] At the same time, normal radiology should not exclude TCS. In general, to avoid irreversibility, prompt diagnosis and surgical untethering must be done in patients who have the greatest chance of restoration of neurological function and return to their optimal occupational capacities.[14,17]

The aim of this study was to shed some light on the natural history of TCS, and to determine the common presentation of TCS in Saudi Arabia where many of the patients present later than those seen in industrialized countries. The other aim was to improve awareness, between those who take care of susceptible patients like neurosurgeons, orthopedics, urologist and pediatricians so that patients are referred for the definitive care earlier, for a much better long-term outcome of the treatment.[18]

Materials and methods

We conducted a retrospective study of 35 consecutive Saudi patients with TCS who were seen and managed at King Khalid University Hospital, College of Medicine, King Saud University, Riyadh, Saudi Arabia, over a period of 7 years (1997–2003). Patients with primary myelomeningocele were excluded. The frequency and presentation of each type of spinal dysra-

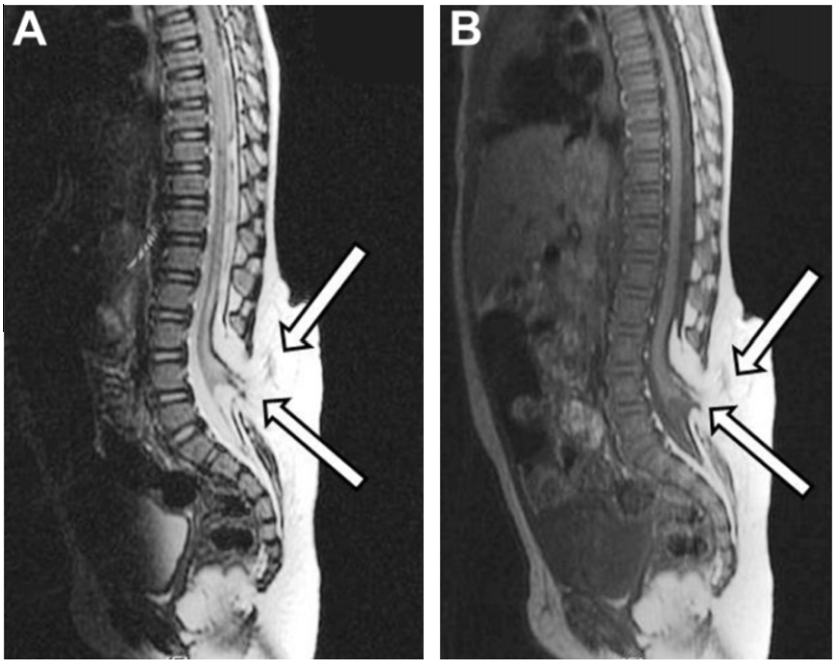

Figure 1: MRI, T2-weighted image (A), and T1-weighted image (B) of a patient with lipomyelomeningocele of the lower lumbar region, showing a low-lying cord at the level of L3-L4 with spina bifida involving lower lumbar and sacral spine. Note intra- and extra-spinal lipoma (arrow).