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TETHERED CORD SYNDROME IN ADULTS: EXPERIENCE OF 50 PATIENTS

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Abstract

Background and Aim: Tethered cord syndrome (TCS) encompasses a range of neurological, urological, and orthopedic symptoms and findings resulting from spinal cord malformations. Majority of cases are associated with spinal dysraphism. The clinical presentations vary depending on the individual's age and variation in underlying pathological condition with the most common manifestations encompassing pain, skin-related signs, orthopedic deformities, and neurological deficits. The objective of this study was to investigate the adult patients diagnosed with TCS.

Patients and Methods: This retrospective study was conducted on 50 TCS patients in the Department of Neurosurgery, Mardan Medical Complex, Mardan - Pakistan from March 2020 to April 2023. Individuals aged >18 years diagnosed of tethered cord syndrome were enrolled. The study investigated the occurrence and manifestations of various types of spinal dysraphism, as well as the untreated late-presenting cases' natural progression.

Results: The overall mean age was 32.64±4.8 years with an age range 18 to 70 years. There were 16 (32%) male and 34 (68%) females. Back-Leg pain was the prevalent reason for presentation of tethered cord syndrome in n=30 (60%) cases followed by urological complaint n=10 (20%), skin abnormalities 4 (8.0%), Orthopedic abnormalities 3 (6.0%), Numbness-Contraction 2 (4.0%), and neck-arm pain 1 (2.0%). The incidence of spinal dysraphism associated with TCS such as lipomeningomyelocele, TCS secondary to myelomeningocele, dermal sinus, diastematomyelia, meningocele, and thick filum terminale was 16 (32%), 12 (24%), 8 (16%), 6 (12%), 5 (10%), and 3 (6%) respectively.

Conclusion: The present study found that TCS patients should be promptly shifted for the assessment and treatment shortly after diagnosis. This is crucial because without treatment, they are at risk of experiencing progressive neurological deficits.

Keywords: Tethered cord syndrome, Adults, Spinal dysraphism

INTRODUCTION

Tethered cord syndrome (TCS) is a functional condition affecting the spinal cord resulting from elongation, in which the lower section of the spinal cord is constrained by an unyielding structure [1]. Tethered cord syndrome (TCS) typically becomes evident either at birth or during childhood, characterized by symptoms such as back pain radiating to both buttocks and legs, sensory deficits in the lower extremities, and loss of bladder/bowel control [2, 3]. In contrast, adult tethered cord syndrome (ATCS) is a rare condition, and its diagnosis remains a challenge [4]. The predominant symptom in ATCS is lower back or leg pain [5, 6], and sensory disturbances often exhibit a patchy distribution rather than following a specific dermatomal pattern [7]. Notably, pain in the upper limbs and upper trunk is uncommon. Given that the location of the spinal cord lesion does not align with the presenting symptoms, ATCS with primarily upper body neurological abnormalities may be susceptible to misdiagnosis. The recognized cause of TCS is the traction of the spinal cord, leading to both anatomical and metabolic disruptions that result in clinical symptoms. The tethering of the spinal cord can occur either congenitally (known as primary TCS) or in conjunction with other conditions within the spinal cord or as a result of scarring after surgery (referred to as secondary TCS) [8].

TCS can manifest at any age and its symptoms typically appear in early childhood and remain till their adulthood due to lack of diagnosis. The presentation of TCS can vary depending on the underlying cause, with common symptoms including skin changes, abnormalities in musckeleten system, and urinary issues. The decision-making process regarding the treatment of tethered cord syndrome is intricate. Once the diagnosis is confirmed or when there is a significant probability that tethered cord syndrome is progressing, the neurosurgeon faces the challenging task of selecting from various treatment options. The goal is to enable patients to lead normal lives while also minimizing the risks associated with surgical intervention [9].

Physical examination findings related to tethered cord syndrome (TCS) can vary and may not always present as obvious. They can encompass a combination of abnormalities associated with both lower and upper motor neurons, along with occult spinal dysraphism. The diagnosis of TCS can be challenging because these signs and symptoms can be quite subtle and easily overlooked [10]. In neonates, spinal ultrasound and in older children, magnetic resonance imaging (MRI) are valuable diagnostic tools that typically reveal the underlying cause of TCS. In infants, high-resolution ultrasound examinations can serve as effective screening tests. Radiological abnormalities such as a low-lying conus medullaris, lumbosacral lipoma, filum terminale lipoma, or a thickened filum terminale are key indicators that aid in diagnosing TCS [11-13]. The objective of this study was to showcase the surgical outcomes in a cohort of adult patients who were diagnosed with tethered cord syndrome.

METHODOLOGY

This retrospective study was conducted on 50 TCS patients in the Department of Neurosurgery, Mardan Medical Complex, Mardan - Pakistan from March 2020 to April 2023. Individual aged >18 years diagnosed of TCS were enrolled. The study investigated the occurrence and manifestations of various types of spinal dysraphism, as well as the untreated late-presenting cases' natural progression. In this study, all patients underwent a comprehensive evaluation of their general and neurological conditions through a combination of clinical assessments, laboratory tests, and imaging examinations. Prior to surgery, every patient received a preoperative spinal MRI. Clinical evaluations were conducted from the time of hospital admission and continued at regular intervals, including assessments at 3, 6, and 12 months following surgery. Urodynamic studies and repeat spinal MRIs were carried out one year after the surgical procedure. The surgical approach involved microsurgery performed under general anesthesia. The primary objective of these procedures was to release the tethering elements and repair the thecal sac. Patients included in this study met the clinical and imaging criteria for a diagnosis of tethered cord syndrome, with specific exclusions: individuals who had previously undergone primary repair of meningiomyelocele, patients with paraplegia, those with hydrocephalus, and individuals under the age of 2 years.

Data collected during both preoperative and postoperative evaluations, as well as investigative studies, were subjected to thorough analysis using statistical software, specifically the Statistical Package for Social Science (SPSS) version 27. The significance level (P value) was set at less than 0.05 to identify statistically significant results.

RESULTS

The overall mean age was 32.64±4.8 years with an age range 18 to 70 years. There were 16 (32%) male and 34 (68%) females. Back-Leg pain was the prevalent reason for presentation of tethered cord syndrome in n=30 (60%) cases followed by urological complaint n=10 (20%), skin abnormalities 4 (8.0%), Orthopedic abnormalities 3 (6.0%), Numbness-Contraction 2 (4.0%), and neck-arm pain 1 (2.0%). The incidence of spinal dysraphism associated with TCS such as diastematomyelia, lipomeningomyelocele, dermal sinus, diastematomyelia, meningocele, TCS secondary to myelomeningocele, and thick filum terminale was 6 (12%), 16 (32%), 8 (16%), 5 (10%), 12 (24%), and 3 (6%) respectively. Demographic details of patients are shown in Table-I. Figure-1 illustrate the common reasons for presentation of tethered cord syndrome. Somatosensory spinal evoked potentials (SSEPS) are shown in Figure-2. Spinal dysraphism refers to a group of congenital developmental disorders affecting the spinal cord and surrounding structures. Tethered cord syndrome (TCS) can be associated with various types of spinal dysraphism as shown in Figure-3. These various forms of spinal dysraphism can contribute to the development of tethered cord syndrome by causing the spinal cord to be abnormally anchored or stretched, leading to a range of neurological symptoms. The specific type of spinal dysraphism can vary among individuals and may require different treatment approaches.

Table-I Demographic details of patients (N=50)

Variables	**Value (Mean ± SD) N (%)**
Age (years)	32.64±4.8
Age Groups (years)	
18-35	42 (84%)
36-50	6 (12%)
51-70	2 (4%)
Gender	
Male	16 (32%)
Female	34 (68%)
Follow-up duration	11 months

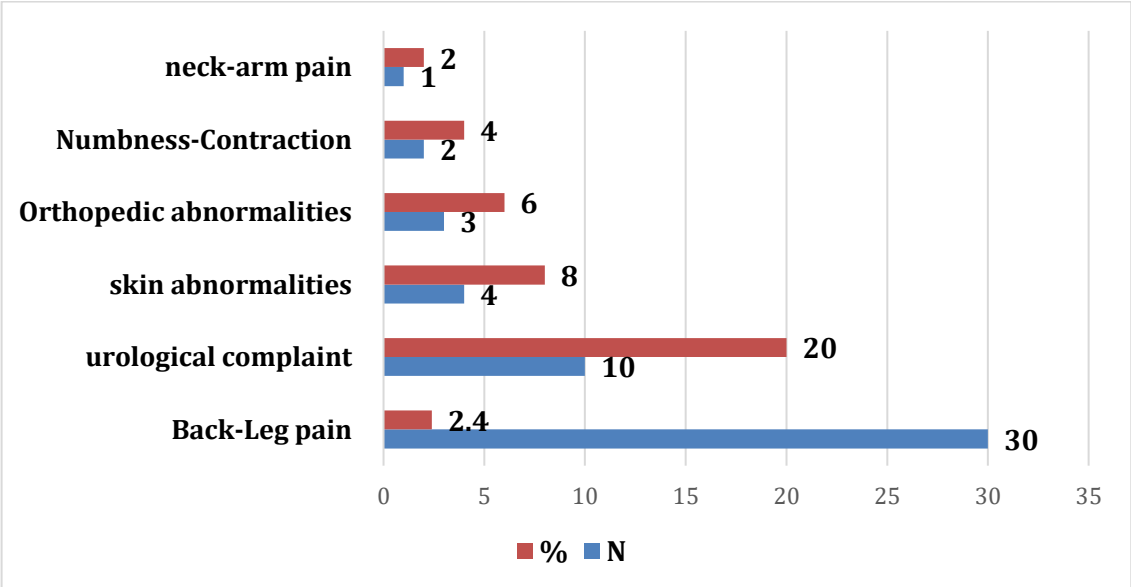

Figure-1 Common presentation of TCS (N=50)

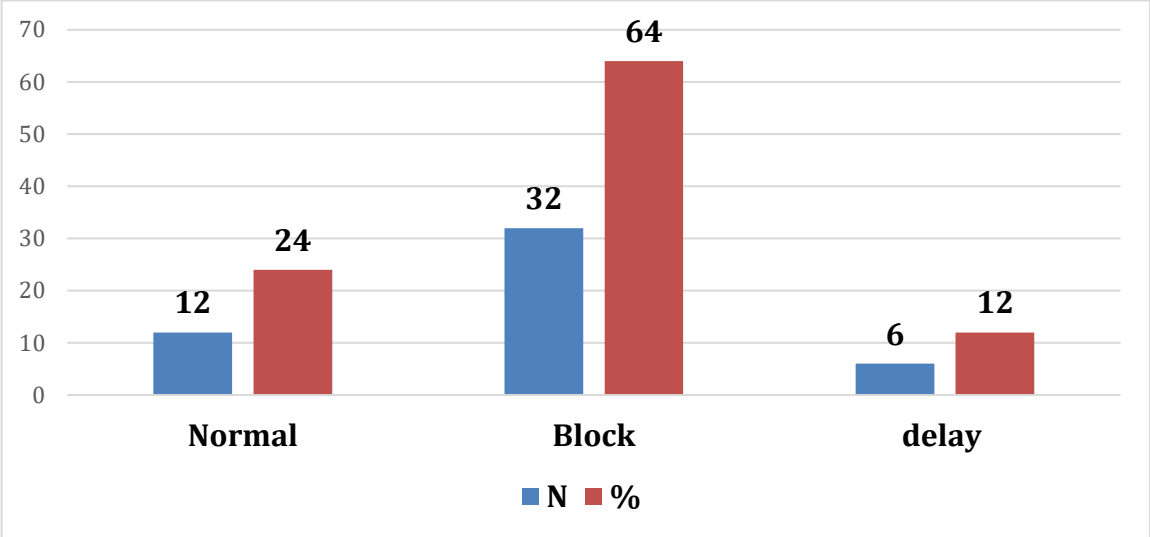

Figure-2 Somatosensory spinal evoked potentials (SSEPS) (N=50)

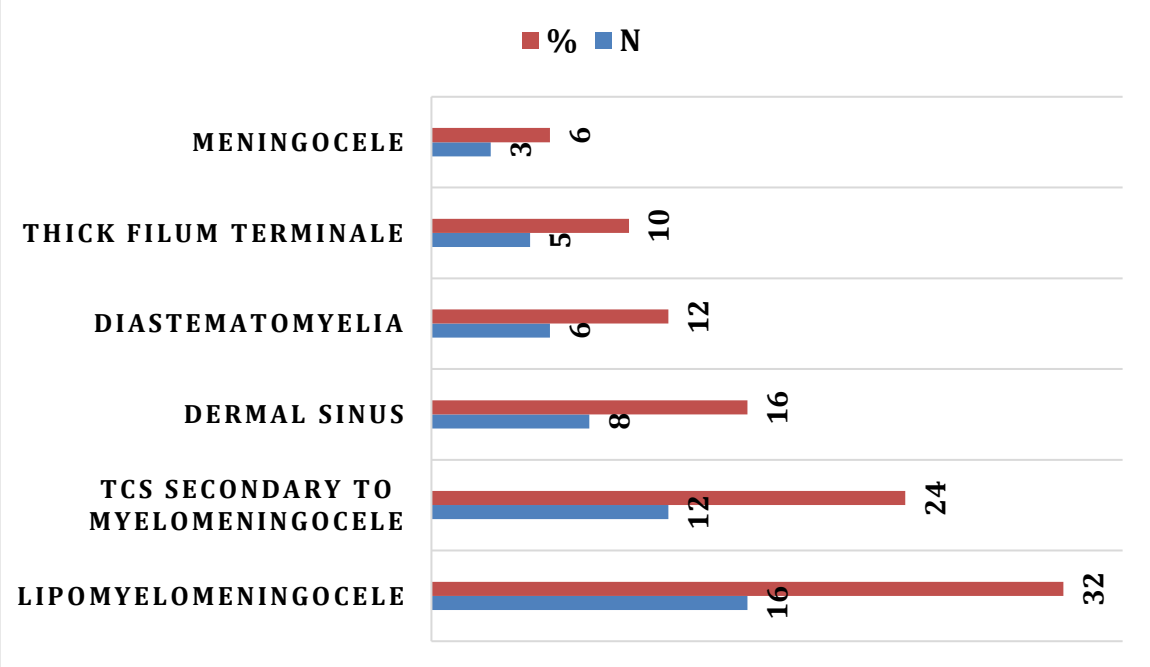

Figure-3 incidence of spinal dysraphism associated with TCS

DISCUSSION

The present investigation mainly focused on the experience of 50 patients of tethered cord syndrome in tertiary care hospital and found that Back-Leg pain was the prevalent reason for presentation of tethered cord syndrome and lipomeningomyelocele was the spinal dysraphism associated with TCS. Furthermore, it is advisable to promptly refer individuals suspected of having TCS for assessment and treatment by the age of 2 years or shortly after receiving a diagnosis. This is crucial because without treatment, they are at risk of experiencing progressive neurological deficits. Tethered cord syndrome (TCS) is a clinical condition that arises due to congenital anomalies and can lead to the craniocaudal stretching of the spinal cord [14-16]. This stretching exerts pressure on the spinal cord, restricting blood flow to this region. Consequently, it induces local tissue hypoxia, which eventually results in impaired mitochondrial oxidative metabolism. This metabolic disruption can, in turn, cause cellular dysfunction and ultimately lead to the death of affected cells [17, 18].

The present study observed that majority of TCS patients were hospitalized due to variety of primary reasons with array of symptoms such as visible skin abnormalities, back-leg pain, musculoskeletal anomalies, and disruptions in anal sphincter function. Similar findings were

reported in earlier studies [19, 20]. Likewise, Sun et al. [21] reported that 79% cases had sensory symptoms and back-leg pain as a major complaints. Furthermore, bladder dysfunction was found in 53% patients. Similar study conducted by Costa et al [22] on 27 TCS patients concluded that prior to surgical intervention, the radicular pain, urinary issues, lower back pain, and motor deficits was found in 54%, 19%, 49%, and 51% respectively.

Tethered cord syndrome (TCS) has been observed in association with various pathological conditions, including tumors, dermal sinuses, myelomeningocele, bone or fibrous spicules, lipomyelomeningocele, and fatty filum terminale [23]. Choi et al [24] reported that the incidence of tight filum terminale and combination of lipoma and tight filum terminale was 28.3% (17/60) and 18.3% (11/60) respectively.

In a study conducted by Day and colleagues, which involved 61 patients, findings included lipomyelomeningocele in 25 patients, tight filum terminale in 22 patients, and split cord malformations in 15 patients. These coexisting conditions can contribute to the development or exacerbation of TCS symptoms and often require specialized evaluation and treatment [25]. Following surgery, Vepakomma and colleagues et al [26] observed notable improvements in various aspects of tethered cord syndrome (TCS) in their study. Specifically: 1. Back pain showed improvement in 78% of patients. 2. Leg pain demonstrated improvement in 83% of patients. 3. Motor deficits exhibited improvement in 64% of patients. 4. Urological dysfunction saw improvement in 50% of patients. These findings highlight the positive impact of surgical intervention in relieving TCS-related symptoms, including pain, motor issues, and urological problems.

In this particular study, the most commonly observed spinal dysraphism associated with tethered cord syndrome (TCS) was lipomeningomyelocele, accounting for 32% of cases. Following that, TCS secondary to myelomeningocele constituted 24% of cases. Notably, the prevalence of TCS secondary to myelomeningocele was significantly higher than typically testified range 3-15% [27, 28].

Li et al. [29] reported recommended that infants exhibiting mid-line lumbar cutaneous abnormalities, such as hemangiomas, lipomas, hair patches, or dimples, should undergo evaluation to rule out the possibility of tethering of the spinal cord. This highlights the importance of early assessment and diagnosis of potential TCS-related issues in infants with such cutaneous signs. The slow rate of clinical deterioration often plays a significant role in the late diagnosis of TCS. Moreover, prior neurological deficiencies can make it challenging to detect subtle neurological changes.

Due to the intricate nature of TCS and the necessity for thorough care, it is advisable for patients with myelomeningocele to undergo continuous follow-up by a multidisciplinary team. It's important to recognize that myelomeningocele is not a static condition, and problems can persist beyond childhood. Therefore, an understanding of the potential complications associated with TCS and the importance of early detection and intervention justify the need for lifelong follow-up protocols in susceptible patients to prevent additional handicaps [30].

CONCLUSION

The present study found that TCS patients should be promptly shifted for the assessment and treatment shortly after diagnosis. This is crucial because without treatment, they are at risk of experiencing progressive neurological deficits.

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