

Case Report

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Tethered Cord Syndrome Causing Chronic Kidney Disease in a Child: A rare event Case Reports

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The fixing of the spinal cord, regardless of the underlying cause of the fixation, is called a tethered cord. Tethered cord syndrome is a group of diseases that develops as a result of the regression of the spinal cord due to congenital or acquired causes and is characterized by evaluative neurologic losses. In stretched tethered cord syndrome which is observed during childhood usually manifested by skin symptoms, motor losses, urologic symptoms, and evaluative spinal cord deformations such as scoliosis are more frequently observed while perineal and primal pain, urologic symptoms, and motor losses are more frequent in adults. Tethered cord complicating a neurogenic bladder in childhood is a rare disease. So, this case report is presented.

Keywords: Tethered cord; Child; Chronic Kidney Disease; Neurogenic Bladder.

Running Title: Tethered Cord Syndrome Causing Chronic Kidney Disease

Introduction

The spinal cord is free to move up and down within the spinal canal. The fixing of the spinal cord, regardless of the underlying cause of the fixation, is called a tethered cord [1]. When there is severe pain or neurologic deterioration in response to fixation, it is called end cord syndrome called the tether [2]. Beyond infancy, the spinal cord in humans ends in the conus medullaris at about the level of L 1. The position of the conus below L2 is consistent with a congenital tethered spinal cord [3,4]. The attachment to the spinal cord usually causes the spinal cord to end lower in the lumbar or sacral spinal canal. The bottom of the spinal cord is tethered (tied down) and cannot move freely. As the child grows, the spinal cord becomes stretched. When stretched, the spinal cord cannot carry messages between the brain and body properly, causing symptoms of the disease [5]. Spinal cord tethering may be either primary or secondary.

The primary form is typically a form of occult spinal dysraphism (OSD) while the secondary type usually occurs following a meningomyelocele [6,7].

Tethered cord syndrome is a group of diseases that develops as a result of the regression of the spinal cord due to congenital or acquired causes and is characterized by evaluative neurologic losses [8]. In the majority of the cases, tethered cord syndrome is a congenital disease of childhood and the patient is born with it; it may also be present in adults. The most frequent causes are isolated spinal cord syndrome, short and thick filum terminale, intradural lipoma, and adhesions that develop after lipomyelomeningocele and meningomyelocele surgery. In stretched tethered cord syndrome which is observed during childhood usually manifested by skin symptoms, motor losses, urologic symptoms, and evaluative spinal cord deformations such as scoliosis are more frequently observed while perineal and primal

pain, urologic symptoms, and motor losses are more frequent in adults [9,10].

Clinical features of the tethered cord include variable degrees of back pain and leg pain, sensory and motor deficits, orthopedic deformities, bladder and or bowel dysfunction, and loss of spinal curvature [11,12].

The earliest features of subclinical neurovesical dysfunction are depressor hyper - reflexes and depressor - sphincter dye synergic resulting in visico- urethral reflux and hydronephrosis [13].

Tethered cord may cause neurogenic bladder in which there is loss of bladder innervation, resulting in failure to understand the fullness of bladder, incontinence, recurrent urinary tract infection, and increased risk of renal damage [13, 14]. The aim of the treatment is elimination of the pathology leading to regression and protection of healthy neural structures. Prognosis depends on early recognition and prompt surgical correction (detethering) for an optimal outcome [15].

Tethered cord complicating a neurogenic bladder in childhood is a rare disease. So, this case report is presented.

Case Report

A 12-year-old boy was admitted to the Department of Pediatric Nephrology, Bangabandhu Sheikh Muja Medical University (BSMMU), Dhaka, Bangladesh on April 2, 2015 with the complains of urine dribbling since birth. Dribbling was continuous in nature and was never dry. He also complained of localized flank pain associated with low grade intermittent fever for 5 days. He gave a history of repeated attacks of similar pain and fever for which he was treated by a registered physician. He also gave a history of fecal incontinence since infancy. On query, his brother stated that he had a swelling over his lower back at birth not covered by skin which resolved spontaneously. He had no weakness in his lower limbs. He underwent per-urethral surgical intervention twice without significant improvement. On Examination, the boy was well alert, mildly pale, and febrile and had normal vital signs except blood pressure (BP) which was raised (140/100 mmHg, both systolic and diastolic BP were above the 99th centile for age, sex and height). His height for age was on the 50th centile (149 cm) and weight for age was on the 10th centile (32 kg). There was a hypo-pigmented area over his lower back (figure 1). On nervous system examination, both knee and ankle jerk were diminished and anal grasp (tone) was also



Figure 1. Hypopigmented area over back

decreased. Examination of the loco-motor system revealed bilateral pes cavus and cocking of toes. Genitourinary system examination revealed a palpable bladder and a tender renal angle without palpable or ballot-table kidneys. Other systemic examinations revealed normal findings (figure 2).



Figure 2. Pes-cavus presentation of the case

Relevant investigations were done. Urine microscopic examination showed plenty of pus cells but no growth was observed on urine culture. Complete blood count revealed a hemoglobin level of 9.2 gm/dl and a total white blood cell count of 11000/cmm (differential count of white blood cells: 55% neutrophils, 40% lymphocytes), and platelet count of 350000/cmm, which were all within normal limits.

Serum Calcium was low (7.5 mg/dl, normal value: 8.5 to 10.5 mg/dl). Serum inorganic phosphate was elevated (6.3 mg/dl, normal value: 3 to 4.5 mg/dl). The level of alkaline phosphatase (490 U/L, normal value: 35 to 136 U/L), serum

parathyroid hormone (512 pg/ml, normal value: 10 to 67 pg/ml), serum creatinine (7.83 mg/dl, normal value 0.3 to 1.1 mg/dl), and blood urea (158 mg/dl, normal value: 15 to 45 mg/dl) was elevated while serum electrolytes were normal. We also noticed a low creatinine clearance rate (10.47 ml/min/1.73 m²).

Ultrasonography findings of the kidney and urinary bladder showed bilateral hydronephrosis with cystitis. Retrograde urethrogram with micturating cystourethrogram (RGU with MCU) showed grade III vesico-ureteric reflux (VUR) with no urethral valve or any other lesion in the urethra.

No significant post voidal residue was present. X-ray of the lumbosacral spine in both views was unremarkable.

MRI of the lumbosacral spine showed spinal cord ending at the level of L3-4 that was thicker than usual, consistent with a diagnosis of tethered spinal cord syndrome (Figure 3).



Figure 3. MRI showing tethered cord

Considering the history, clinical findings, and laboratory investigations, the boy was diagnosed as a case of chronic kidney disease with urinary tract infection due to neurogenic bladder (secondary to tethered cord).

Initially he was managed with continuous catheterization of the bladder, conservative management of chronic kidney disease, intravenous antibiotics, antihypertensive drugs, and counseling of the patient and parents. Finally, renal replacement therapy was scheduled.

Discussion

Tethered cord syndrome is a group of diseases that develops as a result of fixation and stretching of the spinal cord due to various forms of occult dysraphism such as lipomyelomeningocele, myelocystocele, and diastematomyelia [1-3].

These conditions are associated with cutaneous manifestations such as midline lipomas, dermal sinus hair patch, and hemangioma, often with asymmetry of the gluteal fold. In the present case, the patient had meningocele, which is a common presentation of the tethered cord syndrome associated with attachment of the spinal cord. Tethered cord syndrome can also develop in patients who undergo surgical procedures that disrupt the pial surface of the spinal cord [9]. Clinical features of tethered cord include variable degrees of back and leg pain, sensory-motor deficits, orthopedic deformities like pectus excavatus and curving of the spine, and bladder and bowel dysfunction [10]. The reported patient had back pain and pectus excavatus, diminished ankle jerk, and bladder and bowel dysfunction [11, 12]. The earliest features of subclinical neurovesical dysfunction are depressor hyper-reflexes and detrusor sphincter dys-synergic resulting in vesico-urethral reflux and hydronephrosis [12, 13].

Our patient had hydronephrosis and grade III vesico-ureteric reflux. Tethered cord may cause neurogenic bladder in which there is absence or loss of bladder innervation, resulting in failure to understand the fullness of the bladder, incontinence, recurrent urinary tract infection, and increased risk of renal damage [13]. Unfortunately the patient had all these features and, as the operations was not done early, he developed chronic kidney disease manifested by anemia, hypertension, and decreased creatinine clearance rate, a rare presentation of tethered cord.

Direct vertebral graphs, spinal computer assisted tomography, myelography, and MRI are used for evaluation of patients with tethered cord syndrome. In direct vertebra graphs, it is possible to observe spina bifida and bone spicules leading to separate spinal cord.

If there is an associated separate spinal cord, spinal computer - assisted tomography may be used for the evaluation of bone spicules. A final diagnosis of tethered cord syndrome is established using magnetic resonance imaging (MRI) [15].

MRI helps to detect conus medullaris, thick filum terminal, and other native anomalies (hydromyelia) [15, 16].

The aim of treatment is elimination of the pathology leading to the regression of the spinal cord backwards and protection of healthy neural structures [17].

Considering that evolutive neurological observation neurogenic bladder that rarely recovers, heals after its development, the diagnosis should not be surgical intervention should be performed as soon as possible. If surgical intervention is the choice, microsurgical dissection with releasing the spinal cord attachment to the overlying dura is the goal of treatment [17, 18].

Conflict of Interest

None declared

Financial Support

None declared

Conclusion

Our aim was to emphasize the importance of early detection and management in tethered cord syndrome. The management of tethered cord syndrome in neonates and children with occult spinal dysraphism requires a multi - disciplinary comprehensive approach with antenatal counseling, postnatal evaluation and vigilant follow-up, careful investigation and their interpretation followed by timely selective surgical management in trained hands to facilitate neural preservation.

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