Tethered Cord Syndrome with Abnormal Gait

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Parents frequently visit their family physicians with concerns of gait abnormality in their children. Minor musculoskeletal anomalies and normal age-related gait variation are the most frequent causes. Tethered cord syndrome is an uncommon pathologic entity that can be initially asymptomatic and subsequently manifest serious neurological sequelae. Prognosis depends on early recognition and prompt surgical correction for optimal outcome. We describe a case of a previously healthy 17-month-old child with abnormal gait who had tethered cord syndrome.

Case Report

A 2-year-old boy was brought to our Family Practice Center with the complaint of abnormal walk for the past 6 months. He had been getting his health maintenance at our center since birth. His great grandmother first expressed concern about the way he walked at age 17 months, but no abnormality was detected at that time. There were no other associated symptoms, including injury, fever, headaches, or weakness.

The prenatal history was remarkable for a maternal second-trimester chlamydial cervicitis that was treated with azithromycin. The child was born at 35 weeks' gestation by spontaneous vaginal delivery to a 21-year-old mother, para 1,1,1,2. The postnatal period was uncomplicated.

His medical history was notable for a second-degree facial burn at 8 months of age. Social services excluded the possibility of child neglect. He was also evaluated for failure to thrive (height and weight less than 10th percentile) at age 1 year despite apparent good nutrition. At that time there were no serious pathologic findings found on his initial medical workup.

His immunizations were up-to-date, his developmental milestones were normal, and there was no family history of gait or feet abnormalities.

He was a thinly built (height and weight remained at less than the 10th percentile), active child with normal temperature and blood. Respiratory and cardiovascular findings were unremarkable. He had lumbar levoscoliosis, overlapping toes, and increased joint laxity caused by possible hypotonia. He had equal length of both extremities, full range of motion, and good ankle dorsiflexion. There were no sacral dimples, tags, nevi, lipomas, or hypertrichosis. He had normal cranial nerves, normal sensory system, normal power (grade 5), normal tone and reflexes, and good coordination. Genitourinary and rectal findings were normal. His gait was characterized by elevation of the left shoulder and left hip with normal stance and balance. A complete blood count and chemistry profile were normal.

Radiographs of the spine and pelvis showed L4 hemivertebra with sharp levoscoliosis. Spinal magnetic resonance imaging showed low-lying conus medullaris at L3, L4 hemivertebra with associated levoscoliosis. There were no intraspinal masses or lipomas. Findings of a renal sonogram and a voiding cystourethrogram were normal.

The pediatric neurologist confirmed the above findings and noted generalized hypotonia. The chromosomal analysis was normal. Operative findings confirmed a low-lying conus medullaris with stretching of the nerve roots. A thickened fatty filum terminale was also found, which was coagulated and cut, along with release of the tethered spinal cord.

The postoperative course was uneventful and the child at a 1-year follow-up examination had a completely normal gait.

Discussion

Tethered cord syndrome is a broadly used term for progressive neurological deterioration localized to lower spinal cord abnormalities (such as fibrous bands or adhesions, thickened filum terminale, di-

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astematomyelia, or intradural lipoma), resulting in traction on the conus medullaris.[1,2]

The family physician can see patients with tethered cord syndrome at any age but most often during childhood. Symptoms and signs vary from being asymptomatic to severe lower extremity neurological deficits with bladder and bowel involvement.[1] Inspection of the back reveals cutaneous stigmata (sacral dimples, hairy nevus, angiomas, or lipomas) in 70% of cases.[3] Children can also have gait abnormalities resulting from scoliosis or asymmetric growth in the foot or leg associated with talipes cavus deformities.[4] A few children complain of diffuse pain in lower extremities or urological symptoms[4] (recurrent urinary tract infections, enuresis, dribbling, delayed toilet training). In contrast, adults usually complain of diffuse bilateral perianal-perineal pain (86% of cases) and urological symptoms (overflow or stress incontinence).

Skin manifestations are found in 50% cases, but feet deformities are rare.[3] The patient might describe a history of urinary tract infections, enuresis, or abnormal micturition, since 20% of the patients will have a neurogenic bladder.[5] Our patient did not have the skin stigmata that would have enabled earlier recognition of tethered cord syndrome. The only attributable clinical feature in our patient was levoscoliosis. Scoliosis can be a primary feature of tethered cord syndrome[5] in 29% of cases.[3] Sensory involvement can manifest as painless trophic skin ulcers in the feet or toes.[6] Our patient had no lower limb skeletal or neurological abnormality except for overlapping toes.

Tethered cord refers to the abnormal fixation of the spinal cord observed in patients with dysraphism.[2] Prolonged impaired mobility of the spinal cord and nerve roots stretches the spinal cord longitudinally, which potentially compromises the blood supply and causes ischemic damage to the neural tissues.[5] Yamada et al[7] described two groups of tethered cord patients, one with primary elongation of spinal cord producing excessive tension as described by Hoffman et al,[8] and the other with structural lesions (tumors,[9–12] myelomeningocele,[13] scar formation,[14] or a bony spicule[15]) that result in overstretching or restricting the spinal cord movement. Our patient had the primary tethered cord syndrome as described by Sarwar et al[16] in that he was asymptomatic with no associated lower cord anomalies. No urodynamic evaluation was performed for our patient, but when it is performed, it can show detrusor hyperreflexia and bladder-sphincter dysergy.[2,17] Our patient had no clinical evidence of bladder or bowel involvement. Loss of bladder or bowel control should trigger a thorough search for detecting spinal cord abnormality.

High incidence of long-tract signs has been observed in patients with diastematomyelia because the septum is located in the mid to high lumbar or thoracic cord.[6] Symptoms usually develop gradually, except when initiated by a sudden blow to the back or fall on the buttocks.[6] Hyperreflexia associated with motor dysfunction or the Babinski sign can be found in 10% to 17% of patients with tethered cord syndrome.[18] Exacerbation of the neurological deficits or deterioration during growth spurts with relative increased cord tethering is typical of this disorder.[17]

An embryologic developmental defect has long been considered to play a role in spinal cord tethering.[19] Barson[20] has shown that the conus lies at the level of L1–2 disk by 2 months of age. More rapid growth of the vertebral canal than that of the spinal cord results in the ascension of the spinal cord. This process starts in the 8th to 9th week of fetal age and ceases at the postnatal age of 3 years.[21] The cervical cord moves 1.8 to 2.8 cm with the neck flexion and extension. A very active patient with a phase of rapid growth might thus become more symptomatic at an earlier stage.[19]

Biochemically tethered cord syndrome can be regarded as a metabolic dysfunction of the spinal cord. The steady traction of the neurons causes impairment of the oxidative metabolism that has been shown to improve after surgical untethering.[22]

Tethered cord syndrome should be considered in the differential diagnosis of patients who have any lower extremity deformities or subtle neurological abnormalities. Any cutaneous sign of occult spinal dysraphism, even in a neurologically intact child, justifies a thorough neurological assessment to facilitate early detection of any upper motor neuron signs. Patients with imperforate anus should be considered for screening for spinal dysraphism, because both conditions can coexist[23]

The initial evaluation of potential tethered cord should include a thorough neurological examination and appropriate imaging studies. Spina bifida occulta or sacral abnormality is found in 90 of the patients with tethered cord syndrome,[24,25] whereas

only a few children with incidental radiologic findings of spina bifida occulta have cord tethering.[19] Radiographs might show associated vertebral anomalies, such as bifid vertebrae, laminar defects, sacral agenesis, and hemivertebra, in approximately 90% of cases.[26] Hemivertebra in our patient was an strong indicator to look for associated spinal anomalies. Spinal dysraphism is the commonest anomaly associated with hemivertebrae and is reported to occur in 15% to 25% of cases.[27]

Sonography might be helpful in young children for evaluating spinal dysraphism in the absence of spina bifida.[28] Magnetic resonance imaging has excellent specificity and sensitivity.[29] Urodynamic studies are recommended in most cases of spinal dysraphism.[18] The most common imaging and operative finding of tethered cord syndrome are known to be an elongated spinal cord below the L2 level and thick filum terminale (>2 mm).[19,28]

Management of patients with tethered cord syndrome is controversial. Studies have shown that the potential for reversal of upper motor neuron symptoms can be poor once neurological signs or orthopedics deformities are detected,[30] and prophylactic untethering is advocated.[7,30] A low-lying conus on imaging studies does not translate into symptoms of tethered cord syndrome, and not all patients with tethered cord actually develop tethered cord syndrome.[31] Many neurosurgeons advocate careful follow-up with close surveillance for the development of upper motor neuron signs to determine the need and timing of surgical untethering.[2]

Surgical intervention in our patient resulted in recovery of his gait, and his neurological status remained completely normal.

Implications

Tethered cord syndrome should be considered in patients with gait disturbance. Its manifestations are subtle and consequences are severe, yet it is eminently treatable. Although most patients evaluated by a family physician have minor musculoskeletal problems, searching for a treatable cause is prudent and rewarding.

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