

TETHERED SPINAL CORD: REVIEW OF LITERATURE

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Abstract

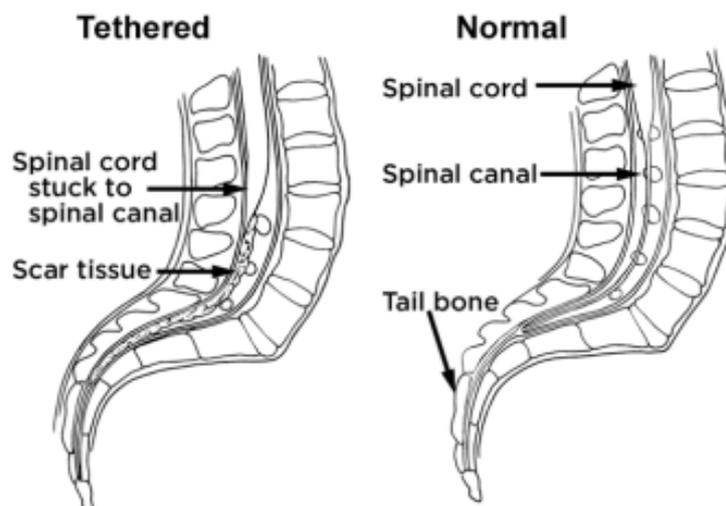
Tethered cord syndrome is a stretch-induced functional disorder associated with the fixation (tethering) effect of inelastic tissue on the caudal spinal cord, limiting its movement. This abnormal attachment is associated with progressive stretching and increased tension of the spinal cord as a child ages, potentially resulting in a variety of neurological and other symptoms. The condition is closely linked to spina bifida, and such presentation in childhood may accompanied with the cutaneous stigmata of dysraphism (hairy patch, dimple, subcutaneous lipoma). There may be associated foot and spinal deformities, leg weakness, low back pain, scoliosis and incontinence. The condition may go undiagnosed until adulthood with the development of sensory and motor problems and loss of bowel and bladder control.

Introduction

Tethered spinal cord syndrome is a neurological disorder caused by tissue attachments that limit the movement of the spinal cord within the spinal column. These attachments cause an abnormal stretching of the spinal cord (Figure 1). Only sporadic cases of tethering along the rest of the neuraxis, including the hindbrain, cervical, and thoracic spinal cord have been documented, always along with some associated congenital

malformations (hydrocephalus, Chiari malformation, myelomeningocele, meningocele, hamartomatous stalk, spina bifida occulta, intramedullary lipoma, intradural fibrous adhesions, the fusion of the sixth and seventh cervical vertebrae, split cord malformation, or low-lying cord)¹⁻¹¹. It is estimated that 20% - 50% of children with spina bifida defects repaired shortly after birth will require surgery at some point to untether the spinal cord.

Figure 1: Normal vs. Tethered Spinal Cord



Discussion

The lower tip of the spinal cord is normally located opposite the disc between the first and second lumbar vertebrae in the upper part of the lower back. In people with spina bifida (myelomeningocele), the spinal cord fails to separate from the skin of the back during development, preventing it from ascending normally, so the spinal cord is low-lying or tethered. In patients with a lipomyelomeningocele, the spinal cord will have fat at the tip and this may connect to the fat which overlies the thecal sac. Although the skin is separated and closed at birth, the spinal cord stays in the same location after the closure. As the child continues to grow, the spinal cord can become stretched, causing damage and interfering with the blood supply to the spinal cord.

Pathology

Tethered cord syndrome can be of a congenital (primary) origin or acquired (secondary or developmental).

Congenital anomalies, particularly spina bifida, are often associated with congenital tethered cord syndrome. The spinal column develops at a greater rate than the spinal cord during fetal development, and abnormal attachments lead to abnormal stretching of the spinal cord. Spina bifida is a birth defect due to incomplete closure of the posterior spinal cord and bony vertebral arch which leaves a portion of the spinal cord protruded through the spinal canal, forming a myelomeningocele. Types of spina bifida associated with tethered cord syndrome include an abnormal connection of inelastic tissue to the caudal spinal cord, dermal sinus tract, a split spinal cord (diastematomyelia), and a benign fatty mass or tumor (lipoma) continuous to the spinal cord. The other fatty anomaly is a lipomyelomeningocele, in which a lipoma extrudes from the spinal canal underneath the meninges, but covered by normal skin.

In many individuals, tethered cord syndrome is caused mechanically by an inelastic often-thickened filum terminale. This structure, which is composed of glial tissue and covered by pia mater, is a delicate strand of fibrous tissue, bridging the spinal cord tip and the sacrum. Due to its high viscoelasticity, the filum allows movement of the spinal cord. If abnormal fibrous tissue grows into the filum and replaces glial tissue, the filum loses its elasticity and abnormally fixes the spinal cord, and becomes the mechanical cause of tethered cord syndrome. The inelastic filum is commonly thickened in children, but found less frequently in adolescents and adults.

Genetic factors are involved in development of anomalous caudal spine and spinal cord, e.g. myelomeningocele, and in some cases of lipomyelomeningocele. An important gene associated with tethered spinal cord syndrome is COX8C (Cytochrome C Oxidase Subunit 8C)¹².

Secondary causes include tumors, infection or the development of fibrosis connected to the spinal cord, complication of spinal surgery.

Pathophysiology

The neuronal dysfunction in tethered cord syndrome is caused by the impaired oxidative metabolism, partly due to cellular ischemia, and partly to ion channel dysfunction directly related neuronal membrane stretching. During gestation, the spinal cord is continuous to the brain and runs in the spinal canal to the tailbone area. In general, the spinal cord is protected from external insult by two mechanisms; 1: encased in the spinal column, that is, a rigid structure, 2: floating free in the spinal fluid space of the spinal canal. In addition, the spinal cord is continuous to the filum terminale, which is extremely extensible because of its high visco-elasticity. If the spinal cord

is tethered at its caudal end, and if the spinal cord is unable to grow as fast as the vertebral column in childhood, the spinal cord is stretched beyond its physiological tolerance causing metabolic abnormalities and, ultimately, the various neurological symptoms of this syndrome.

Normally, the spinal cord ascends in the spinal canal as the spinal column starts to grow faster than the spinal cord at 9th weeks of gestation. Consequently, the spinal cord is pulled upwards due to this growth difference. By three months of age, the tip of the spinal cord reaches the normal level between T12 and L2 vertebrae. An elastic, extremely extensible filum allows for the ascension of the less elastic spinal cord. If the filum becomes inelastic in an embryo, then the spinal cord tip is anchored and ceases to ascend. Compensatory to the stretching force, the lower spinal cord naturally grows more than seen in normal subjects, and becomes elongated. Associated with tethered cord syndrome, the elongated cord is often noted in children, but less often in adults¹³⁻¹⁵.

Symptoms in children

The specific symptoms, severity and progression of tethered cord syndrome vary from one individual to another. Due to the variation of the growth rate of the spinal cord and the spinal column, the progression of neurological signs and symptoms is highly variable. Some patients present with congenital tethered cord syndrome at birth, while others develop the symptoms in infancy or early childhood. Others may not develop any noticeable symptoms until adulthood. A high percentage of pediatric patients, with tethered cord syndrome show cutaneous tufts of hair, skin tags, dimples, benign fatty tumors, skin discoloration or hemangiomas. Other symptoms include lower back pain that worsens with activity and improves with rest, leg pain or numbness, gait disturbances, foot and spinal deformities (scoliosis or lordosis),

high-arched feet and hammertoes, and less commonly difference in leg strength. Tethered cord syndrome can also cause difficulties with bladder and bowel control. Affected children may experience involuntary urination or defecation (incontinence) and repeated urinary tract infections.

Symptoms in adults

Back pain from tethered cord is often aggravated by bending slightly forward, by sitting upright with crossed legs, or by holding a moderate weight (such as a baby or a stack of books) at waist level. This pattern of pain is sometimes called the “3-B sign” for bending, Buddha-sitting with legs crossed and baby-holding at the waist level¹⁶.

In adults, the tethered cord syndrome includes constant, often severe back and leg pain, which may extend to the rectum and genital area in some cases. Progressive sensory and motor deficits may affect the legs potentially resulting in numbness, weakness or muscle wasting (atrophy) in the affected areas. More than 50% of the affected individuals experience bladder and bowel dysfunction, manifested by increased frequency or urgency of urination or constipation.

Diagnosis

A diagnosis of tethered cord syndrome is made based upon identification of characteristic signs and symptoms that can neurologically locate the lesion to be above the attachment of the anomalies to the spinal cord. A detailed patient history and a thorough clinical evaluation, plus imaging exams must be carried out.

Ultrasound may be a useful in the pediatric population due to the lack of ossification of the posterior arch of the spine in normal infants and the presence of a bony defect in patients with spina bifida. Ultrasound obtains images of the spinal cord moving in the thecal sac.

Magnetic resonance imaging (MRI) is useful in visualizing the conus medullaris, assessing the thickness of the filum terminale, identifying traction lesions, and evaluating associated bony dysraphisms. In one study of 25 patients in 84% the tip of the conus was below the level of the mid-L2 vertebral body¹⁷. Prone imaging may be useful in patients who have undergone tethered cord surgery or in those in whom clinical suspicion is high, while supine MRI imaging demonstrated no abnormalities. Prone MRI imaging is however of little value when supine MRI has demonstrated the defect. Imaging features are of a low conus medullaris (below L2) and thickened filum terminale (>2 mm)^{18,19}.

Myelogram can show pressure on the spinal cord or nerves due to tethered spinal cord.

The CT or CAT scan show how the dye flows around the spinal cord and nerves. In some cases, electromyography (EMG) and nerve conduction studies may be used to assess nerve function.

Treatment

Untreated, tethered cord syndrome has a progressive course. Surgical release, in selected patients, can dramatically improve function.

Untethering is generally performed only if there are clinical signs or symptoms of deterioration. The surgery involves opening the scar from the prior closure down to the covering (dura) over the myelomeningocele. Sometimes a small portion of the bony vertebrae (the laminae) are removed to obtain better exposure or to decompress the spinal cord. The dura is then opened, and the spinal cord and myelomeningocele are gently dissected away from the scarred attachments to the surrounding dura. Once the myelomeningocele is freed from all its scarred attachments, the dura and the wound are closed. In individuals with severe arachnoiditis (adhesion of the meninges to the spinal cord) found by

MRI or CT scan, careful evaluation of pain and neurological condition is required to find if surgical treatment is warranted. At surgery, release of arachnoid adhesion must be performed with meticulous technique. Or re-adhesion or extensive scar formation might follow the surgery. To circumvent this problem, two special surgical procedures have been advocated: 1: transection of the spinal cord to relieve severe back and leg pain, and 2: shortening of the spinal column by resection of one or two vertebrae to relieve spinal cord tension.

The child usually can resume normal activities within a few weeks. Recovery of lost muscle and bladder function depends upon the degree and length of preoperative implications. The combined complication rate of this surgery is usually only 1 to 2 percent. Complications include infection, bleeding, damage to the spinal cord or myelomeningocele, which may result in decreased muscle strength or bladder or bowel function. Many children require only one untethering procedure. However, since symptoms of tethering can occur during periods of growth, 10 to 20 percent require repeated surgery.

Bladder impairment is much less likely to resolve after surgery because of the vulnerability of the nerves supplying the bladder to permanent damage from progressive tethering. In some patients, the best result from surgery is cessation of neurological deterioration, but in over half of patients, there is the hope of sustained symptomatic improvement, even after many years of having the syndrome as an adult.

Early referral of patients with this condition will help surgeons to optimize the timing of detethering²⁰.

The surgical prognosis varies depending upon the presenting symptoms and tethering-producing anomalies.

Conclusion

With treatment, patients with tethered spinal cord syndrome have a normal life

expectancy. However, some neurological and motor impairments may not be fully correctable. Prompt surgical intervention results in reversal, or at least stabilization, of symptoms in many cases and can prevent further functional decline.

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