



Case Report

Late-Onset Tethered Cord Syndrome in a Patient with Spina Bifida: A Case Report

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Abstract

Tethered cord is a clinical syndrome associated with short and thick filum terminale. It can occur because of congenital or acquired reasons and can lead to progressive neurological deficits. A thorough medical history, physical examination, imaging, and electrophysiological tests are helpful in its diagnosis.

An 11-year-old patient with a prior diagnosis of spina bifida was operated for pes equinovarus at the age of 1.5 years. The patient visited our physical medicine and rehabilitation polyclinic with complaints of pain and weakness in the left ankle and region around the knee. She had been suffering from walking disruption, thinning of leg muscles, and pain for past 1 year. After imaging, a diagnosis of tethered cord syndrome was made.

Follow-up of patients diagnosed with spina bifida during growth period is important to prevent complications such as syringomyelia and tethered cord syndrome.

Keywords: Spina bifida; syringomyelia; tethered cord syndrome.

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Tethered cord syndrome is a clinical condition that manifests itself with traction of the spinal cord. Myelomeningocele, lipomyelomeningocele, and diastematomyelitis may result in a tethered/short filum terminale; furthermore, a dermal sinus, tumor, hemangioma, or scar tissue may induce tethered cord syndrome.^[1,2] This syndrome is clinically symptomatic and manifests itself with progressive neurological findings. It may become apparent after birth and in early childhood and may also be seen in advanced ages. In conditions manifesting themselves during adulthood, urological symptoms are more prominent.^[3]

Syringomyelia is a chronic disease whose pathogenesis is not completely known; it is characterized by longitudinal cavitations in the spinal cord.^[4] Although clinically, it manifests itself more frequently with sensory symptoms such as pain and change in the perception of heat, in many

patients it may be detected incidentally.^[5] In this paper, we present the case of a patient who was referred to our clinic with lower extremity weakness and was diagnosed with tethered cord syndrome along with the presence of syringomyelia; furthermore, we present an overview of the available literature.

Case Report

A 10-year-old female patient consulted our outpatient clinic of physical medicine and rehabilitation with the complaints of pain and weakness in the left ankle and around the knee region. She had been diagnosed with spina bifida and had undergone surgery at the age of 1.5 years because of pes equinovarus with an uneventful postoperative period. Motor developmental stages of the patient had a normal course. Since the last year, she had started limping

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while walking, her leg muscles had got thinner, and she had been suffering from pain. The patient did not report any history of trauma. On physical examination, the range of joint motion at plantar flexion and dorsiflexion was found to be 10°, whereas on motor examination, the following grades of muscle strength were detected: m. external hallucis longus (2/5), m.tibialis anterior (however, this muscle could not be evaluated fully because of contracture) (3/5), and m.gastrocnemius (3/5). Tibialis anterior ve gastrocnemius muscles were atrophic. Sensory examina-

tion was unremarkable, and the patient was found to have pes cavus deformity. Magnetic resonance imaging (MRI) of the lumbar spine revealed the following: spinal cord terminated at the level of L3 vertebral inferior endplate (tethered cord) (Fig. 1); fusion of vertebral bodies; lateral posterior elements at L4-L5 level (Fig. 2); and syringohydromyelic cavitations at the distal spinal cord (Fig. 3). EMG findings consistent with anterior horn involvement at L5-S1-S2 levels on the left side, which also affected the posterior root of the spinal ganglion, were detected. Urodynamic, MRI, and somatosensory evoked potential (SEP) examinations did not reveal any pathology. The patient was referred to the neurosurgery department for consultation, and surgery was scheduled after making a diagnosis of tethered cord syndrome.



Figure 1. Spinal cord terminates at the level of inferior endplate of L3 vertebra (low-lying conus medullaris-arrow).



Figure 2. Fusion of vertebral bodies and their posterior elements at L4 and L5 vertebral levels.



Figure 3. A longitudinally oriented syrinx cavity is seen (arrow).

Discussion

Tethered cord syndrome develops as a result of congenital or acquired stretching of the spinal cord. The evolving imaging modalities have shown that the manifestations of tethered cord may become evident not only in association with occult type dysraphism but also they may be related to tumor(s), trauma, arachnoiditis, postsurgical meningocele, and meningomyelocele.^[6] In pediatric patients with tethered cord syndrome skin manifestations, motor functional loss, urological symptoms, and progressive spinal cord deformities are more frequently seen, whereas in adults perineal and perianal pain, urological symptoms, and motor functional loss are more frequently observed.^[7] We assume that in our patient, progressive muscle weakness, pain, and muscular atrophies developed as clinical manifestations of tethered cord syndrome. Further sophisticated tests were requested to finalize the diagnosis and rule out a peripheral pathology.

Diverse electrophysiological abnormalities may be seen in patients with tethered cord syndrome. During SEP examination, latencies of cortical responses may be observed. According to one report, fine-needle electromyography (EMG) findings were consistent with chronic neurological involvement, and in nerve conduction studies, decrease in motor unit action potentials and abnormal H reflexes were detected.^[8]

Syringomyelia is not a disease, but it is a clinical entity that manifests as a result of the development of longitudinally oriented cystic cavities in the spinal cord and related spinal cord compression with ensuing neurological symptoms.^[9] The cases of syringomyelia not related to and accompanied by trauma, spinal tumor, and craniocervical or intracerebral pathologies are termed as idiopathic syringomyelia.^[4]

It is frequently accompanied by Chiari malformation. For

radiological diagnosis, methods such as intravenous and intrathecal contrast-enhanced tomography and myelography may be used; however, MRI is the gold-standard diagnostic method. As is the case with tethered cord syndrome, syringomyelia may yield symptoms such as muscle weakness, paresthesia, hyperesthesia, dysesthesia, non-radicular segmental pain, analgesia, or anesthesia.^[10] In our patient, complaints of segmental pain around the left ankle and knee along with muscle weakness suggest that both syringomyelia and tethered cord syndrome may contribute to clinical manifestations. Electrophysiological findings of syringomyelia are nonspecific, and affected muscles may display polyphasic MUPs, fibrillation, positive sharp waves. Sensory nerve action potentials remain intact.^[11] In our patient, tibial and peroneal nerve motor conduction studies did not yield any abnormality; furthermore, left peroneal nerve sensory action potential could not be elicited. Left sural nerve action potential exhibited a low amplitude. Sensory conduction velocities of the other examined nerves were within normal limits. Fine-needle EMG of muscles innervated by left L5-S1-S2 nerve roots revealed normal, slightly dispersed, sustained, polyphasic motor unit potentials without any acute denervation potentials. EMG results of other muscles were within physiological limits. These findings were consistent with anterior horn involvement at L5-S1-S2 levels on the left side, which also affected the posterior root of the spinal ganglion. Besides, on MRI, we did not encounter any associated pathology apart from the tethered cord and syringomyelia in this patient with a history of surgery for spina bifida; thus, surgery was planned for this patient.

Conclusion

It should always be remembered that a clinically occult period may be present in patients with the diagnosis of spina bifida, and during chronic phase, additional pathologies of medulla spinalis may contribute adversely to the clinical course of the disease. As a critically important issue, particularly adolescent patients with the diagnosis of spina bifida should be closely followed regarding tethered cord syndrome and syringomyelia.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Conflict of Interest: None declared.

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