

Case Report

Absent sural responses in tethered cord syndrome

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Context: Tethered cord syndrome is a progressive condition usually diagnosed early in life, and leads to a diversity of manifestations including neurologic, urologic, and orthopedic dysfunction.

Findings: We report two cases of tethered cord syndrome associated with a unilaterally absent sural sensory response on nerve conduction studies while other causes of this finding being excluded.

Conclusion/clinical relevance: We believe that this finding is caused by a selective injury of sensory fibers at the level or distal to the dorsal root ganglia.

Keywords: Tethered cord syndrome, Sural response, Gait impairment

Tethered cord syndrome (TCS) is a progressive spinal cord disorder caused by stretch-induced dysfunction of the conus medullaris. Clinical signs and symptoms of TCS may be cutaneous, neurologic, musculoskeletal, and/or genitourinary.¹ Toddlers and adolescents tend to present with gait or running difficulties due to progressive motor dysfunction, sensory deficits, progression of scoliosis, or development of orthopedic foot deformities.² TCS may be associated with myelomeningocele, syringomyelia, diastatomyelia, dermoid sinus and intra or extradural lipoma which may exacerbate the clinical findings.³ Spine MRI demonstrates elongation and caudal descent of the conus medullaris (below L2 vertebral level) and a fatty filum terminale (usually > 3 mm in diameter).⁴ While electrophysiological evaluation for patients with TCS was just limited to intraoperative monitoring, few studies performed a detailed electrophysiological assessment in non-operated patients.^{5,6} We report two pediatric cases with asymmetrically absent sural nerve response prior to their diagnosis of TCS.

Case presentation

Case 1: a 7-years-old girl presented with mild gait impairment and right foot deformity. Physical examination revealed a medical research council (MRC) strength of 4/5 in the right foot dorsiflexion and plantar flexion, absence of knee and Achilles deep-

tendon-reflexes (DTRs) bilaterally, preserved pinprick and vibratory sense with no upper motor neuron signs. Nerve conduction studies (NCS) showed an absent right sural nerve response but normal left sural, bilateral fibular motor, tibial motor responses as well as tibial F-wave responses. Lumbar-spine MRI showed cord tethering at L5-S1. Concentric needle electromyography examination of the tibialis anterior, gastrocnemius and vastus lateralis muscles was normal on the right side.

Case 2: a 13-years-old girl presented with gait impairment. Physical examination showed muscle atrophy over the left calf, weakness in left foot plantar flexion and dorsiflexion with MRC 4/5, but normal lower extremity DTRs, and normal sensory examination. NCS showed absent left sural nerve response and left fibular motor response but normal right sural, right fibular motor and bilateral tibial motor responses including F-waves and H-reflexes. Lumbar-spine MRI cord tethering at L3 level. Concentric needle electromyography showed a chronic neurogenic pattern of recruitment and re-innervation in the left tibialis anterior without any denervation potentials, but was normal in the left vastus lateralis and left gastrocnemius muscles.

Discussion

TCS is characterized by a constellation of neurologic symptoms, occurring as a result of inappropriate tissue adherence between the spinal cord and the spinal canal. In ideal conditions, the cord moves freely with spine flexion and extension. The spinal cord is attached

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to the canal in TCS which puts the cord under tension with flexion/extension spine movements. This induces a recurrent pathologic spinal cord stretch resulting in progressive damage in its structure leading to a variety of neurological manifestations.⁷

Clinical and radiological evaluation remains the gold diagnostic standard in assessing TCS. Neurophysiologic testing including NCS, needle electromyography and evoked potentials (motor and somatosensory) may provide more insight into the extent of neuronal damage in addition to the diagnostic and prognostic value of the intra-operative motor evoked potential and somatosensory evoked potential monitoring during surgical detethering.⁸ Pre-operatively, NCS may show relatively (often asymmetric) abnormal responses in the motor components of peripheral nerves, prolongation in F-waves and H-reflex in the setting of intact sensory nerve responses. Needle electromyography may show evidence of denervation and reinnervation indicating a chronic neuronal injury. These findings suggest that the level of injury is localized to the anterior horn cells or cord roots.⁶

To our knowledge, the electrophysiological finding of unilaterally absent sural responses as in our cases has not been previously reported in the literature. The exclusion of sensory polyneuropathy, sciatic neuropathy, and in setting of no previous history of local trauma to the lower extremity (traumatic isolated sural neuropathy) leads us to believe that a selective nerve fiber injury in the structures of the dorsal root ganglia (DRG) or distally lying structures containing sensory fibers had led to this finding. This can be induced by a repetitive chronic increase in traction of the caudal spinal cord inducing metabolic and electrical dysfunction in the lumbosacral gray matter, with greater stretch in lower segments and little effect on cephalic segments above the lowest pair of dentate ligaments. Various segments of the human lumbosacral cord may elongate slightly at different rates with post-ganglionic stretch effect of the spinal nerves above the threshold of its tolerance. Such differential elongations may be influenced by the duration of the traction effect, presence of arachnoid adhesion and fibrosis, and spinal curvature.⁹

Given the variability of DRG location based on cadaveric studies, the stretch vulnerable points could also

vary. Studies have shown that DRGs at the lumbar levels can be either extraspinal or intraspinal.¹⁰ Levin KH identified 6 subjects with intraspinal canal lesions and reduced superficial peroneal nerve conduction study responses relating the latter finding with an intraspinal localization of DRG.¹¹ In patients where the stretch vulnerable points are proximal to the DRG we expect normal sural responses. We believe that our patients have those points at or distal to the DRG.

Disclaimer statements

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Conflicts of interest Authors have no conflict of interests to declare.

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