ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 05 | May - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

AN INTERESTING CA(U)SE OF FOOT DEFORMITY IN YOUNG – WHAT NOT TO MISS??

Neurology

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ABSTRACT

Tethered cord syndrome is a clinical phenomenon resulting in anatomic restriction of the normal movement of the spinal cord or vascular compromise leading to hypoxia of its distal structures. The tethered cord syndrome cases mostly appearduring the period of infancy and childhood. Though the adolescent presentation of TCS is well-recognized, it continues to pose significant diagnostic and management controversies. We report a case of tethered cord syndrome with disability in right foot which remained undiagnosed until adolescence.

KEYWORDS

Tethered Cord Syndrome, Conus Medullaris, Filum Terminale (FT)

INTRODUCTION

Tethered cord syndromes (TCS) are a group of rare neurological conditions caused by mechanical tethering or presence of inelastic tissue within the filumterminale, a strand of fibrovascular connective tissue that extends caudally from the conusmedullaris to the coccyx. It has been postulated that the primary functions of the filumterminale are to provide longitudinal support to the spinal cord and to stabilize the conus medullaris during motion that alters the length of the spinal canal (e.g., bending at the waist). When the filumterminale fails to protect the neural elements of the lumbar spine from these changes in length via its interface with the conus, clinical symptoms of TCS can occur. (1)

The spinal cord up to 3rd month of intrauterine life extends throughout the length of the developing vertebral canal and so the length of spinal cord and vertebral canal are equal. The spinal nerves run horizontally from their segment of origin to exit through the corresponding intervertebral foramina. Subsequently, due to the differential growth of spinal cord and vertebral column (the vertebral column becomes much longer than the spinal cord) at birth, the lower end of the cord is at the level of the third lumbar vertebra This process of recession of the spinal cord continues after birth as a result of which, in the adult, the cord usually ends at the level of the lower border of the first lumbar vertebra .The cord and nerve roots are mobile and slide proximally and distally with flexion and extension unless tethered or trapped by severe spinal canal stenosis. (2)

Fixation of the spinal cord can occur congenitally (primary TCS) or in association with other intraspinal pathologies or postoperative scarring (secondary TCS). Most cases of TCS reported are related to spinal dysraphism.(3)

As the spinal column lengthens faster than the cord, tethering lesions cause progressive stretching of the spinal cord. Abnormal traction leads to chronic ischemic changes and neuronal dysfunction.(4)

The most common clinical features of tethered cord include a triad of neurological, urological, and musculoskeletal signs and symptoms. Lumbar lower back and lower extremity pain are the most common symptoms in patients with onset of TCS in adulthood, often with report of pain that is worse with lumbar flexion.(5)

Case Report

14 years old boy who was first born out of non-consanguineous marriage with developmentally normal milestones presented with progressive difficulty in walking. He placed the right foot abnormally on the ground for the past 5 years in durationbut he had no difficulty in gripping slippers, clearing the foot from the ground or tripping of toes.There was no proximal weakness in the lower limb neither truncal or upper limb weakness. There was no flailness or stiffness of the limbs. No sensory, bowel and bladder disturbances. On examination there was wasting in right leg and foot muscles along with hammer toes and callosities on lateral aspect of foot. There is bilateral pescavus deformity (figure 1a and 1b) which wasmore on right comparing with left side. Mild dorsiflexion weakness on right side with absent ankle jerk on right, with flexor plantar on both sides were noted. Rest of neurological examination was normal. There was no cutan eousstigmata, spine deformities and leg length discrepancies. All peripheral pulses were felt. Routine blood workup which included CBC, liver function, renal function, RBS, Sr.electrolytes and TSH were normal .

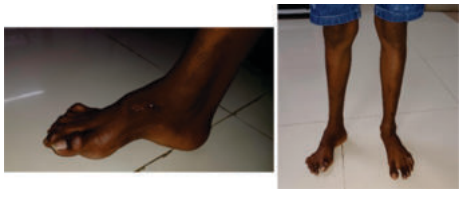

Figure 1a,1b- Showing Hammer Toes, Pescavus Deformity With Distal Wasting Of Both Lower Limbs (RT>Lt)

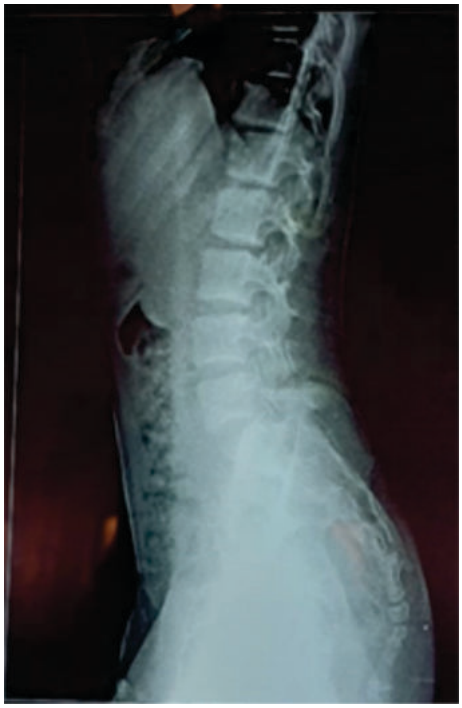

Figure 2: X-ray L-S Spine Shows No Evidence Of Any Bony Deformities

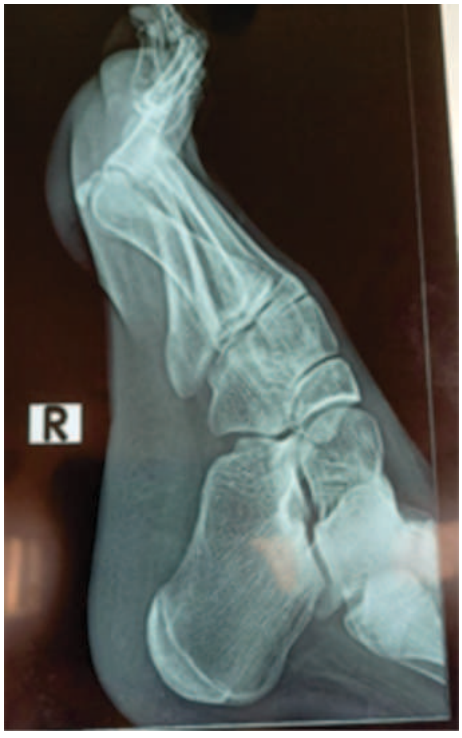

Figure 3:Xray Right Foot Showing Equine Deformity

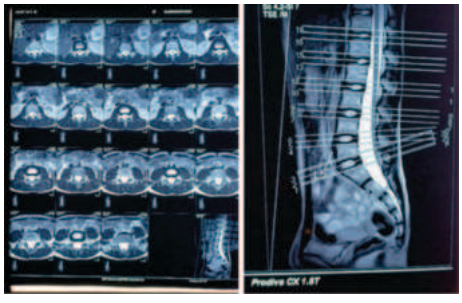

Figure 4 a and 4b,MRI LS Spine Showed Low Lying Conus Medullaris At L5 - S1 With Cord Tethering At L5- S1. Wide Spinal Canal From L3 To Upper Sacral Levels.

- NCS showed normal study.
- Xray LS spine (figure 2) shows no evidence of any bony deformities.
- Xray Right foot(figure 3) showing equine deformity .
- MRI LS spine(figure 4a and 4b) showed low lying conusmedullaris at L5 - S1 with cord tethering at L5- S1. Wide spinal canal from L3 to upper sacral levels.

DISCUSSION

The case presented here highlights the importance of considering tethered cord syndrome in the differential diagnosis of any patients presenting with only deformity in foot without any sensory or bowel and bladder disturbances. Incase report by mishrass et al ., describe two cases of adolescent onset TCS associated with two different etiologies. With first case, an 18-year-old girl who presented due to overflow incontinence in association with TCS was diagnosed to have lumbar meningocele. The second case, a 19-year-girl presenting with perianal anesthesia and bowel and bladder incontinence had lipomyelomeningocele. Both of them underwent untethering surgery.Another study for MR examinations of the spine study done by Raghavan N et al.,among 25 patients with a clinical diagnosis of tethered spinal cord showed that in 21 patients (84%), the level of the tip of the conus was below the mid L2 vertebral body. The causes of the tethering were spinal lipomas (72%), tight filumterminale syndrome (12%), diastematomyelia (8%), and myelomeningocele (8%). In adolescents with radiologically confirmed cord tethering, untethering is an appropriate procedure when the neurologic symptoms and signs are suggestive of TCS. Our case had no associated abnormalities in spinal cord other than tethering. So early diagnosis, adequate surgical releasemight be the keys to successful management in such cases.

CONCLUSION

This is one of treatable cause for the foot deformities in children and adolescent age group which is often not diagnosed early resulting in disability. Soearly recognition and treatment of tethered cord syndrome can significantly improve patient outcomes.

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