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A Case of Tethered Cord Syndrome with Pes Cavus Clinically Suspected with Charcot-Marie-Tooth

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BACKGROUND

Tethered cord syndrome (TCS) is a constellation of signs or symptoms which are caused by abnormal attachment of the spinal cord to surrounding tissue or structure. TCS is associated with tension applied to the spinal cord due to low-lying conus medullaris.

In most cases, symptoms often occur at an early age, while others are asymptomatic until adulthood. A typical symptom is that neurologic symptoms may occur due to ischemic damage caused by spinal cord tension. Motor weakness, sensory dysfunction or abnormal upper motor neuron signs can appear in TCS. Pain is uncommon in childhood, however adults with TCS usually complain of pain. Urologic dysfunctions and deformity of lower extremity or spine such as pes cavus can be accompanied in TCS. However, pes cavus is rare in TCS. In TCS, the goal of surgery is to remove or release tissue that apply tension to spinal cord. Early surgical intervention is associated with better outcomes. In this case report, we describe a first case that was late-diagnosed with TCS at the age of 60s as far as we know.

CASE REPORT

A 38-years-old man first visited our clinic in 1998 for registration of persons with disabilities about pes cavus. The patient was taking medicine for hypertension and did not have familial history associated with neurological disorders. The patient had both feet deformities and walked independently but unstable. Until the age of 38, he had never visited the hospital for above symptoms. Charcot-Marie-Tooth (CMT) disease was suspected only by neurologic examinations including the manual muscle strength test and the shape of foot deformity. The patient had intermittent chronic low back pain and radiating pain in both lower extremities. Magnetic resonance imaging (MRI) examination and genetic test for CMT were initially recommended, but it was not performed due to cost. In the initial manual motor strength test, poor to fair grades in both lower extremities were confirmed. For 24 years, only conservative managements including injection treatment and medications with painkillers were performed.

Since 2019, weakness in both lower extremities gradually deteriorated and the patient started to walk with cane. In 2022, the patient began to have difficulty in urinary management. An electrophysiologic study was performed to evaluate proceeding weakness in both legs and long-lasting back pain with pain radiating along the posterior leg. The result showed small amplitude of compound motor action potential only in left side tibial nerve and cannot obtain late response (F-wave and H reflex). This result is not corresponding with typical electrophysiological findings of CMT disease. At this point, motor power was checked poor grade in both legs (table 1). While subjectively complained of weakening lower extremity muscle strength, objective manual muscle strength tests did not show significant aggravation for 20 years in the progress records.

Table 1. Changes of manual muscle strength grade

	1998~2010		2022	
	Right	Left	Right	Left
Hip flexion	3 (Fair)	3 (Fair)	2 (Poor)	2 (Poor)
Knee extension	3 (Fair)	3 (Fair)	2 (Poor)	2 (Poor)
Ankle dorsiflexion	2 (Poor)	2 (Poor)	2 (Poor)	2 (Poor)
Big toe dorsiflexion	2 (Poor)	2 (Poor)	2 (Poor)	2 (Poor)
Anlke plantarflexion	2 (Poor)	2 (Poor)	2 (Poor)	2 (Poor)

Both knee reflexes were hyperactive and upper motor neuron signs were identified. In response, brain and spine MRI were recommended again to differentiate the upper motor neuron lesion, and brain MRI performed was normal. In spine MRI, arteriovenous malformation, leipomyelomeningocele at the sacral level with sacral defect and accompanying low level conus medullaris below L2 level were identified (figure 1) and were finally diagnosed with TCS. The patient has been referred to another department for surgical treatment, but since the symptoms are not severe, the patient is receiving conservative treatments.

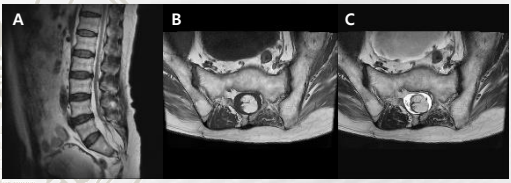

Figure 1. Spine MRI : Sagittal T2 weighted image (A) showing leipomyelomeningocele at sacrum level with low-lying conus; Axial T1 image (B) and T2 image (C) at sacral level of figure 1A with leipomyelomeningocele

DISCUSSION

Pes cavus is a broad spectrum of foot deformity with high medial longitudinal arch that does not flatten on weight bearing. Hereditary Motor-Sensory Neuropathy (HMSN) is the most common cause of pes cavus and it is not easy to think of TCS when pes cavus is the main problem. In our case, CMT was suspected due to a deformity of the foot, and the final diagnosis was difficult because no additional examination was conducted for several years. TCS is mostly associated with spina bifida as a problem with the embryonic development. With the development of prenatal test technology, it is unusual to be diagnosed as TCS for the first time as an adult as in this case.

In conclusion, the patient had symptoms from childhood that could be caused by TCS, such as waddling gait and foot deformity, but did not visit the hospital or perform additional tests. The patient has not been treated for TCS in addition to preservative managements such as trigger point injection or medications for more than 20 years. Therefore, it was possible to verify the natural course of TCS when the causative treatment of TCS was not performed. For about 20 years after first visiting the outpatient clinic in 1998, the patient's lower extremity muscle weakness did not get any worse and there was no rapid aggravation enough to show a clear difference in the manual muscle strength test until 2019. This is a case report in which it was found that the progression of TCS in adults has no significant changes.