CASE REPORT

Tethered Cord Syndrome Accompanied by Unilateral Muscle Atrophy in the Calf Muscle

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A 40-year-old man developed slowly progressive muscle atrophy in his calf muscle. The patient became aware of asymmetry of his right big toe during in his teens; muscle atrophy was revealed at the age of 25 years. Multi-image examinations revealed “tethered cord syndrome” with spina bifida occulta. Spinal magnetic resonance image revealed a pathogenetic mechanism of asymmetry of muscle atrophy in the calf muscle. This suggested that spinal MRI imaging is one of the most useful diagnostic methods for this disease.

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Key words: MRI, CT, spina bifida occulta, spinal cord malformation, spinal dysraphism

Introduction

In the early embryonal period, the spinal cord terminal exists at the level of terminal sacrum, then it ascends to the L4 level at week 17 in the embryonal period. After that, the spinal cord terminal gradually ascends to the L1 or L2 levels (1). Tethered cord syndrome (TCS) has come to be recognized as a longitudinal traction of the conus, due to several mechanisms (2).

Recently, diagnostic procedures of TCS have been remarkably improved by the employment of magnetic resonance imaging (MRI) (3–5) and other diagnostic imaging equipments. We report a case of TCS associated with spina bifida occulta together with unilateral muscle atrophy in the calf muscle; the asymmetry was confirmed by MRI.

Case Report

A 40-year-old man was admitted to our hospital because of muscle atrophy in his right calf muscle. He was born as a full-term, normal delivery baby. He was a good runner until 12 years of age; in junior high school, he could not run any faster than at a young age. At that point, he became aware that his right big toe was smaller than that of the left side. At the age of 15 years, he became aware of the asymmetric size of his feet, because his left shoe felt more tight than his right shoe. At the age of 25 years, his wife noticed muscle atrophy in his right calf muscle. At the age of 35 years, he noticed difficulty in flexing the right big toe, as well as hypesthesia of the right sole. At age 39, he noticed slight difficulty of urination and constipation. His past and family histories were not significant. On admission at age 40, the physical examination revealed no abnormal findings except for muscle atrophy in the right calf muscle (Fig. 1), scoliosis and pes cavus. Neurological examination revealed muscle strength of the right flexor digitorum longus was 1/5, while the strength of the right extensor hallucis longus, extensor digitorum longus, tibialis anterior and gastrocnemius was slightly decreased. Deep tendon reflexes were symmetrically decreased. Plantar responses were flexor bilaterally. Sensory examination revealed hypesthesia to pain, touch and vibration in the segmental area L5 and S1–4 of the right lower extremity.

Blood and chemical examinations were all within normal ranges. Lumbosacral x-ray showed spina bifida occulta at the L5 and sacral levels (Fig. 2). On MRI, the T1-weighted sagittal image revealed that the spinal cord was displaced to the posterior portion of the spinal canal and was tethered between L5 and S1, and there was an oval-shaped, low-intensity tubular lesion at the level of the conus medullaris (Fig. 3a). T2-weighted sagittal image showed widening of the subarachnoidal space, and indicated that the tubular lesion was a high intensity lesion. No abnormal shadow of fatty tissue in the L5 spina bifida area was observed (Fig. 3b). T1-weighted sagittal image showed widening of the subarachnoidal space, and indicated that the tubular lesion was a high intensity lesion. No abnormal shadow of fatty tissue in the L5 spina bifida area was observed (Fig. 3b). T1-weighted axial image after gadolinium injection showed no enhancement, the spinal cord adhered to the right side at the L5 level; a low intensity round lesion was also seen (Fig. 3c). Postmyelographic axial CT finding revealed that the spinal cord adhered to the right side at the L5 level (Fig. 4a) and metrizamide enhancement was seen in the syrinx-like formation (Fig. 4b), which exhibited a low-intensity tubular lesion on the T1-weighted image.

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The cerebrospinal fluid, taken by inferior occipital puncture, showed 87 mg/dl protein, normal cells, sugar and chloride. Cystometry showed hypotonic bladder. The intracystic pressure was elevated to over 100 cm H₂O during voiding and residual volume was 200 to 300 ml. On electromyography, delay of motor conduction velocity, below knee joint both peroneal and tibial nerve, was observed predominant in the right lower extremity. Needle EMG revealed that a neurogenic pattern was predominant in the right lower extremity.

An untethering operation was not performed, because it was thought that no surgical improvement can be anticipated in the early stage of the syndrome (6, 7).

Discussion

The present case has suffered from deformities of the foot since in his teenage year, and developed slowly progressive muscle atrophy in his right calf muscle. Multi-imaging examinations revealed TCS with spina bifida occulta.

This syndrome is more common in the fields of neurosurgery, orthopedics and pediatrics, whereas reports are rare in the field of internal medicine.

The symptoms of TCS have been reported to include pain in the back and leg, motor-sensory disturbance, bladder dysfunction, constipation, trophic ulcerations, as well as cutaneous manifestations such as lipoma, hypertrichosis and nevus (6, 8–10). TCS is divided into two categories according to the age of onset: child-onset type and adult-onset type. Child-onset type is more common, and the majority of cases have deformities of the lower extremities or vertebra accompanied by cutaneous manifestations of the lumbosacral portion (8, 9). However, adult-onset type is less common, and usually includes symptoms such as pain in the back or leg, sensory-motor dysfunction and bladder dysfunction. Most cases develop symptoms after a trauma, and diagnoses of narrowing of the spinal canal or herniated disc were made at an earlier stage (6, 10). Moreover, TCS can be classified into 2 categories by the location of traction: low-placed conus medullaris (LPCM) and tightfilm terminalis (8, 11). It has been reported that most LPCM cases have difficulty in walking and deformity of the feet more frequently than pain.

In the present case, deformities were seen in his childhood, including asymmetry of the big toe and feet; however, cutaneous manifestations were not seen. In the classification of TCS, LPCM seemed most likely. Moreover, right calf muscle atrophy resulted from adhesion or tethering of the spinal cord to the right side, according to CT scan and MRI. Muscle atrophy in the calf muscle is rarely seen in adult-onset TCS; however, in this case it was thought that the child-onset TCS developed into muscle atrophy, due to the lack of an early untethering operation (6, 7). We do not know why dysfunction is more predominant in motor than in sensation, although this was thought to be due to the fact that sensory nerves are less affected than motor nerves of long-standing tethering as reported in large case studies (9, 12). The pathogenetic mechanism of TCS has been indicated to have a relationship to lipoma, diastematomyelia, fibrous adhesions, occult myelomeningocele, postmyelomeningocele repairment and dermal sinus tract (6, 8, 9). However, here no fatty tissue was detected, and also no enhancement was seen in the gadolinium enhanced images. In the present case, fibrous adhesion was considered to be the most probable cause (9, 13). A tubular formation in the region of the conus medullaris seen on the T1-weighted image was thought to be
neoplastic syringomyelia, resulting from circulatory disturbance of CSF with long-term tethering of the spinal cord, because that tubular lesion was shown as a high-intensity lesion on the T2-weighted MRI and metrizamide enhancement was seen in postmyelographic axial CT.

These results suggested that MRI was the most useful
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diagnostic method for this syndrome. An untethering operation is recommended in the earlier stages of this syndrome, because long-standing TCS may develop into numerous neurological and other complications.

References


Fig. 4. a) Post myelographic axial CT reveals the spinal cord adhered to the right side of spinal canal at L5 level. b) Post myelographic delayed CT scan reveals that metrizamide enhancement is seen in the syrinx-like formation, the area which exhibited a low intensity tubular lesion in MRI image.