A Presenile Patient with Filar Lipoma Who Developed Tethered Spinal Cord Syndrome Triggered by Lumbar Canal Stenosis

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Abstract

Lumbar canal stenosis (LCS) has been reported as a precipitating factor by which a tethered spinal cord, which is asymptomatic during childhood, develops into tethered cord syndrome (TCS) in adulthood. However, only a few reports on surgical strategies for such cases are available. A 64-year-old woman presented with unbearable pain in the left buttock and dorsal aspect of the thigh approximately 1 year ago. Magnetic resonance imaging showed cord tethering with a filar-type spinal lipoma and LCS due to the thickening of the ligamentum flavum at the L4-5 vertebral level. Five months after the decompressive laminectomy for the treatment of LCS, an untethering surgery was performed at the dural cul-de-sac at the S4 level. The severed end of the filum was elevated rostrally by 7 mm, and the pain subsided postoperatively. This case study shows that surgeries for both lesions should be indicated for adult-onset TCS triggered by LCS.

Keywords: filar lipoma, lumbar canal stenosis, tethered spinal cord syndrome, untethering

Introduction

Tethered spinal cord syndrome (TCS) is a developmental abnormality that impairs the longitudinal movement of the spinal cord and that can be combined with various forms of spinal dysraphism. Although TCS usually occurs in childhood, a tethered spinal cord can be asymptomatic for decades until adulthood, when it may become symptomatic.[1-9] When Pang et al.[7] first described adult-onset TCS, they cited three precipitating factors as the mechanism by which TCS develops in adulthood. These factors include 1) transit stretching of the spine, 2) mechanical constriction/narrowing of the spinal canal, and 3) spinal trauma. The mechanisms causing spinal canal narrowing include lumbar canal stenosis (LCS) due to lumbar spondylosis/spondylolisthesis and intervertebral disc protrusion.[2,3,6-9] When LCS is involved in adult-onset TCS, both an untethering surgery and a decompressive surgery for LCS may be necessary, but only a few studies have reported the appropriate surgical strategy because reports of such cases are rare.[6,7] Herein, we report a case of a presenile patient with a filar lipoma in whom a successful untethering surgery was performed 5 months after decompressive laminectomy for LCS.

Case Report

A 64-year-old woman presented to our hospital approximately a year ago with unbearable pain in the left buttock and dorsal aspect of the thigh. She also complained of lumbago, which had simultaneously developed. Both pains did not worsen with postural change, but she complained of worsening after long walking. Neurologically, she had very mild hypesthesia in the lateral part of the left dorsalis pedis and equivocal weakness in the left anterior tibial, extensor hallucis, and digitorum longus muscles. A neurogenic bladder was ruled out because her pre- and post-voiding echography did not reveal an enlarged bladder or

Fig. 1

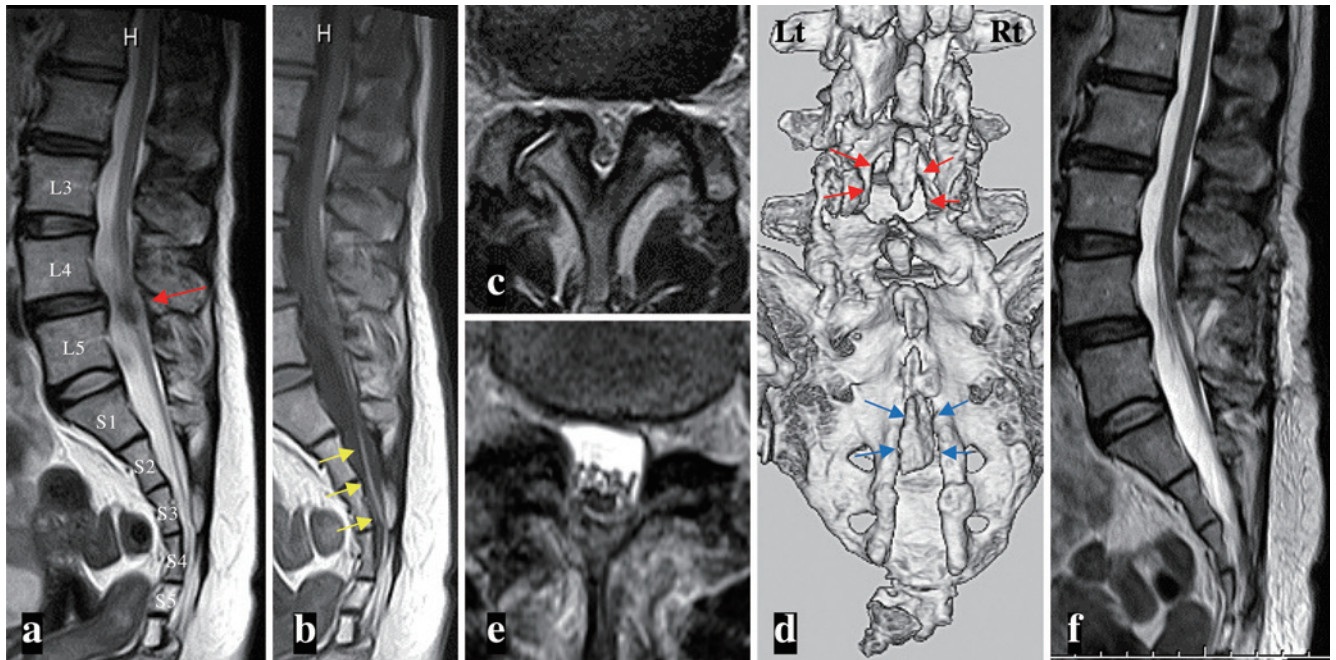**(a-c)** Midsagittal views of T2-weighted (T2WI) (a) and T1-weighted magnetic resonance images (b), and axial view of T2WI (c) at the L4-5 vertebral level (a, red arrow) demonstrate spinal cord tethering with a filar-type spinal lipoma (b, yellow arrows) and lumbar canal stenosis (LCS) due to the thickening of the ligamentum flavum.

(d) Postoperative three-dimensional (3D) reconstruction of a bone-targeted computed tomographic scan shows the extent of the decompressive laminectomy at L4-5 with the preservation of the spinous process (red arrows). An ectopic bone covering the spina bifida at the S3 and upper part of the S4 level is also noted (blue arrows).

(e-f) A month after decompressive surgery for LCS, an axial view of T2WI (e) reveals a successfully enlarged dural sac at the L4-5 level. No change is noted in the degree of spinal cord tethering on a midsagittal view of T2WI (f).

residual urine. Magnetic resonance imaging (MRI) showed cord tethering with a spinal lipoma (Fig. 1a, b). The dural cul-de-sac was located at the S4 vertebral level, where the filar lipoma was terminated. Severe LCS due to the thickening of the ligamentum flavum at the L4-5 vertebral level was also noted (Fig. 1a, c).

Because these pains did not improve with medical treatment for 6 months, a surgery was indicated. First, for the LCS at the L4-5 level, the patient underwent spinous process-splitting laminectomy of the lower two-thirds part of L4 and the upper one-third part of L5 (Fig. 1d). Postoperatively, her lumbago and minute positive neurological findings subsided, but the pain in her left buttock and dorsal thigh persisted. Postoperative MRI confirmed an enlarged dural sac at the L4-5 level with successful decompressive laminectomy (Fig. 1e), but no change was noted in the degree of cord tethering (Fig. 1f).

Five months after the initial surgery, she underwent a second surgery. Following the resection of the ectopic bone covering the spina bifida at the S3 and S4 levels (Fig. 1d), a fatty filum with a diameter of 5 mm, which terminated slightly to the left of the midline of the dural cul-de-sac at the S4 level, was exposed (Fig. 2a). The stimulation of the filum did not evoke a muscle response in the lower limbs

and anus. In addition, we confirmed that nerve roots did not arise from the filum. To untether the cord, the filum was severed at the rostral part of the operative field, and the severed end of the filum was elevated rostrally by 7 mm (Fig. 2b). The caudal part of the filum was also severed at the dural cul-de-sac and resected as a 5-mm long column.

Postoperatively, the pain in her left buttock and dorsal thigh subsided over a month. Postoperative MRI confirmed a successful untethering surgery (Fig. 2c). The histopathological examination of the resected filum showed glial fibrillary acidic protein (GFAP)-positive neuroglial tissue in the center of the fibroadipose tissue. However, no central canal-like structures were observed (Fig. 2d, e).

Discussion

Considering the clinical course of the present case, cord tethering by a filar lipoma, which had been asymptomatic until the 60s, developed into TCS and presented as unbearable left buttock and dorsal thigh pain. The TCS was triggered by the mechanical constriction/narrowing of the spinal canal with LCS, as initially advocated by Pang et al.[7] The painful buttock and dorsal thigh on the left side cor-
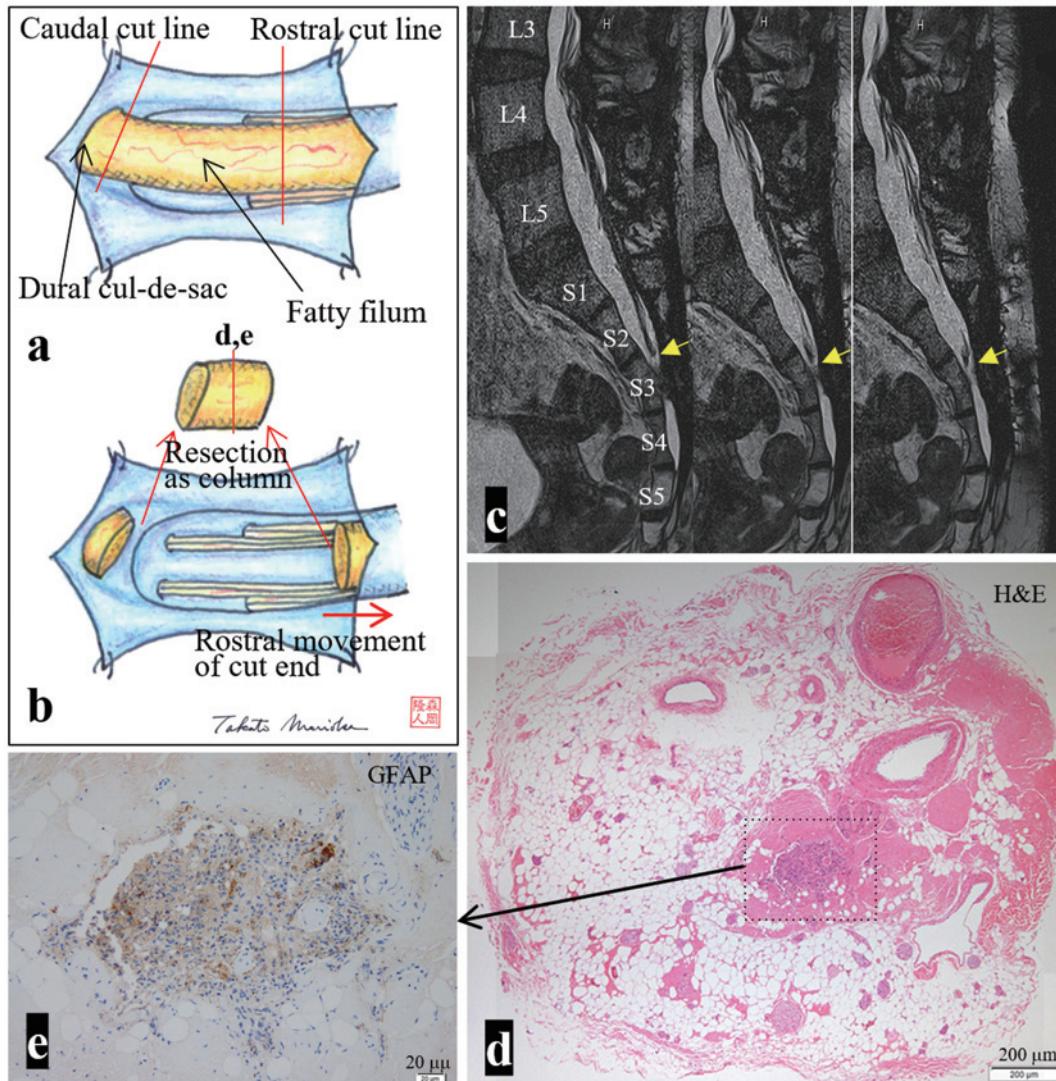
Fig. 2
(a, b) Schematic drawing of the operative findings. (a) Dural opening at the S3 and S4 vertebral levels reveals a fatty filum termi-nated slightly to the left of the midline of the dural cul-de-sac. (b) The filum is severed at the rostral part of the operative field, and the severed end of the filum is elevated rostrally. The caudal part of the filum is also severed at the dural cul-de-sac and resected as a column. (c) Four months after the second surgery, serial sagittal views of 3D heavily T2WI (slice thickness, 1 mm) show a suc-cessful untethering surgery. The severed end is located at the upper part of S3 vertebral level (yellow arrows). (d, e) On the axial section of the resected filum, histopathologically, glial fibrillary acidic protein (GFAP)-positive neuroglial tissue is observed in the center of the fibroadipose tissue. The location of the section is indicated as a red line in (b). The area of the enlarged view is indi-cated as a dotted square in (d). Hematoxylin and eosin (H&E).

responded to the left S2-3 region on the dermatome. Therefore, this pain could not be explained by the pres-ence of LCS at the L4-5 level as a result of the first sur-gery. The fact that the symptoms improved for the first time after the untethering surgery also supports this the-ory.

In terms of the appropriate surgical strategy in such cases, recent authors[10-12] have demonstrated improvement in symptoms only after decompressive surgery for the treatment of LCS. However, the symptoms in these cases

were due to compression myelopathy with LCS instead of TCS. In cases with TCS, similar to the present case, a sur-gery for both lesions is considered necessary.[6] When both surgical sites coincided, it is possible to perform both sur-geries in one stage.[6] However, in this case, we believed that the least-invasive untethering surgery could be per-formed at the S3-4 level near the dural cul-de-sac, and we planned a two-staged surgery. However, no studies have re-ported which operation should be first performed. LCS forms later than congenital cord tethering, and the me-

chanical constriction of the spinal canal due to LCS is severe, suggesting that decompressive surgery alone may improve symptoms. Therefore, in this case, priority was given to decompressive surgery for LCS, which can be performed by only epidural manipulation. However, TCS symptoms did not improve, and an untethering surgery was performed after 5 months. As the severed end of the filum was elevated rostrally by 7 mm after the untethering and the symptoms improved, we believe that performing the decompressive surgery first was useful.

The reason that TCS manifested by LCS does not improve with only successful decompression is not clear and remains within the realm of speculation. It is generally believed that the pathophysiology of TCS is related most of all to chronic ischemia of the cord.[3,4,8,9,13] We speculate that, also in this case, chronic ischemia, which has existed subclinically for over 60 years, was aggravated by LCS and manifested the symptoms. This chronic ischemia persisted even after the successful decompression for the LCS, presumably necessitating the untethering surgery.

The retained medullary cord (RMC) was first proposed by Pang et al.,[14] in which a cord-like structure (C-LS) is continuous from the conus medullaris and extends to the dural cul-de-sac, resulting in cord tethering. In this decade, several additional reports regarding RMC have been published.[15-19] RMC is believed to originate from the late arrest of secondary neurulation.[14] The characteristic histopathological findings of C-LS include the presence of a central canal-like ependyma-lined canal with surrounding GFAP-positive neuroglial tissues in a fibrocollagenous tissue, corroborating a remnant of the cavitary medullary cord, which is typically destined to regress.[14] Recently, some studies reported that this histopathological finding was present frequently in filar and caudal lipomas, which are also believed to originate from the secondary neurulation failure.[18,19] Because central canal-like structures are not present from end to end of RMC tissue,[15-18] central canal-like structures were missing in this case whereas GFAP-positive neuroglial tissues were present. The present findings provided further evidence for the idea that RMC and filar lipomas can be considered consequences of the continuum of regression failure during secondary neurulation.[14,15,18,19]

Conclusion

The present case suggests that surgery for both lesions should be indicated for adult-onset TCS triggered by LCS.

Acknowledgments

Conflicts of Interest Disclosure

The authors have no conflict of interest to declare.

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