Subpial Lumbar Lipoma Associated with Retained Medullary Cord

Takato MORIOKA,[1] Nobuya MURAKAMI,[1] Satoshi O. SUZUKI,[2]
Ryoko NAKAMURA,[3] and Masahiro MIZOGUCHI[4]

[1]Department of Neurosurgery, Fukuoka Children's Hospital, Fukuoka, Fukuoka, Japan
[2]Department of Neuropathology, Graduate School of Medical Sciences, Kyushu University,
Fukuoka, Fukuoka, Japan
[3]Department of Pediatrics, Japan Community Health Care Organization (JCHO), Kyushu
Hospital, Kitakyushu, Fukuoka, Japan
[4]Department of Neurosurgery, Graduate School of Medical Sciences, Kyushu University,
Fukuoka, Fukuoka, Japan

Abstract

Subpial lipomas, which are also known as nondysraphic intramedullary spinal cord lipomas, are not associated with spinal dysraphism resulting from the failed primary neurulation. Retained medullary cord (RMC) is a newly defined entity of closed spinal dysraphism that originates from the late arrest of secondary neurulation. We treated a 6-year-old boy presented with myoclonus of the lower limbs, who had subpial lipoma at the lumbar cord, just rostral to the low-lying conus, which was tethered by a cord-like structure (C-LS) continuous from the conus and extending to the dural cul-de-sac. Following cord untethering from C-LS and minimal debulking of the lipoma, the myoclonus was improved. Histological examination of C-LS revealed a large central canal-like structure in the neuroglial core and the diagnosis of RMC was made. Subpial lipomas can be incidentally coexistent with spinal dysraphism resulting from the failed secondary neurulation, such as RMC.

Introduction

Most spinal lipomas are thought to occur in association with spinal dysraphism resulting from the failed primary neurulation. When the cutaneous ectoderm prematurely separates from the neuroectoderm prior to closure of the neural tube, the surrounding mesoderm, such as subcutaneous adipose tissue, gains access to the surface of the developing neural tube, forming spinal lipomas, which communicate with the subcutaneous adipose tissue through the spina bifida.[1,2] In contrast, subpial lipomas, which are also known as nondysraphic intramedullary spinal cord lipomas, are not associated with spinal dysraphism and have intact meninges and posterior vertebral elements.[3,4] These lipomas are rare and the precise embryogenesis is unknown. However, similar mechanisms of mesenchymal migration into the developing neural tube are postulated because these lipomas are almost always located over the dorsal aspect of the cord.[3,4]

It has never been demonstrated that the subpial lipomas developed in association with the spinal dysraphism resulting from the impaired secondary neurulation. The retained medullary cord (RMC) is a newly defined entity of closed spinal dysraphism that is thought to originate from an almost complete arrest of apoptosis during the last phase of secondary neurulation.[5] Pang et al.[6] described seven patients with a redundant nonfunctional "cord-like structure (C-LS)" continuous from the true conus and extending to the dural cul-de-sac, which produced neurological deficits by tethering. We also reported five cases with RMC extending to a sacral subcutaneous meningocele.[7–9]

The present report describes the first documented case with subpial lipoma at the lumbodorsal region associated with RMC. We herein describe the details of this case and discuss the surgical procedures.

Case Report

A 6-year-old boy presented with paroxysmal myoclonus of the lower limbs, predominantly of the left ankle, lasting several minutes, which was first noted at 1 year of age. Recently, the frequency increased to 1–2 times a week. Except for these movements, his neurological findings were normal. He had a dimple in the groove just above the gluteal cleft (Fig. 1A).

Magnetic resonance imaging (MRI) of the brain and electroencephalography failed to reveal abnormalities. However, spinal MRI demonstrated a lipoma at the right dorsal side of the lumbar cord, just rostral to the low-lying conus at the vertebral level of L3-4, which was tethered with the structures continuous from the conus and extending to the dural cul-de-sac (Figs. 1B–1E).

Surgeries for both lesions were performed by skip laminotomy/laminectomy. First, a typical subpial lipoma was exposed following laminoplastic laminotomy of L2 and upper quadrant of L3 laminectomy (Fig. 1F). The subpial lipoma showed exophytic growth from the dorsal cord but did not show the tethering effect. Second, the C-LS, with a 3-mm diameter, was exposed following the lower half of L4 and upper half of L5 laminectomies. Stimulation of the C-LS with an intensity of 1.0 mA did not evoke compound muscle action potentials (CMAPs) of the legs and anus while the stimulation of the lumbar cord evoked the CMAPs. The C-LS was severed at the rostral (Fig. 1G) and caudal side of the operative field and resected as a column. The caudal severed end descended markedly down while the rostral end ascended slightly up. Finally, the lipoma was minimally debulked and the pial surface was reconstructed with sutures (Fig. 1H).

The postoperative course was uneventful and the myoclonus disappeared. Histopathological examination revealed the lipoma to consist of a mature fibroadipose tissue covered by a fibrocollagenous tissue (Fig. 2A). The filum consisted of fibrocollagenous tissue embedding a large central canal-like structure lined by ependymal cells and surrounded by glial fibrillary acidic protein immunopositive neuroglial tissue (Fig. 2B, C).

Discussion

While the subpial lipoma and RMC were located nearby, the conus existed between these two in the present case, indicating that their coexistence was incidental. The clinical course of the subpial lipoma is generally indolent and surgery including the lipoma debulking and decompressive laminectomy will be needed only when the cord compression is apparent.[3,4] Although the cord compression was minute in the present case, we thought that surgical exploration was needed to confirm the noninvolvement of the tethering. As a result, we performed minimal debulking.

In the diagnosis of the filar lesions resulting from the impaired secondary neurulation, Pang et al.[5] raised the idea that entities such as RMC, filar or terminal lipomas, and thickened filum terminale (TFT) can be considered members of a continuous spectrum of regression failure occurring during late secondary neurulation. They speculated that these pathologies differ from each other in their respective timing and severity of apoptosis failure. The main distinguishing feature of RMC is the dominant presence of a central canal-like ependymal lined lumen with surrounding neuroglial core in the C-LS.[5,6] In filar or terminal lipomas, fibroadipose tissue is the main constituent.[5,10] TFT have dense fibrocollagenous or elastic tissues in the filum.[5,11–13] In the present case, histological examination of C-LS revealed a large central canal-like structure and the diagnosis of RMC was made.

Preoperatively, the low-lying conus was apparently tethered with the C-LS, and untethering surgery was indicated. Surgery for TFT involves a relatively straightforward resection of the filum. In contrast, for RMC, the exact border between the functional true conus and nonfunctional C-LS should be defined by tracing the evoked CMAPs with stimulation starting from the former then on to the latter.[5,6,9,14] However, the extended laminotomy/laminectomy is needed to expose the border.[8] To minimize the extent, the skip technique[15,16] was useful.

Based on the fact that myoclonus in the present case was improved following untethering and minimal debulking of the lipoma, the myoclonus was thought to be of spinal origin. Spinal myoclonus has been linked to a variety of underlying pathologies; however, the exact pathophysiology is poorly understood. For example, there may be enhanced anterior horn neuronal excitability or spinal interneuron dysfunction.[17] Although some cases with spinal dysraphism, such as repaired myelomeningocele[17,18] and large filum terminale,[19] have been reported, the relationship with the tethering has not been documented.

In conclusion, subpial lipomas can be incidentally coexistent with the dysraphic lesion resulting from a secondary neurulation failure, such as RMC.
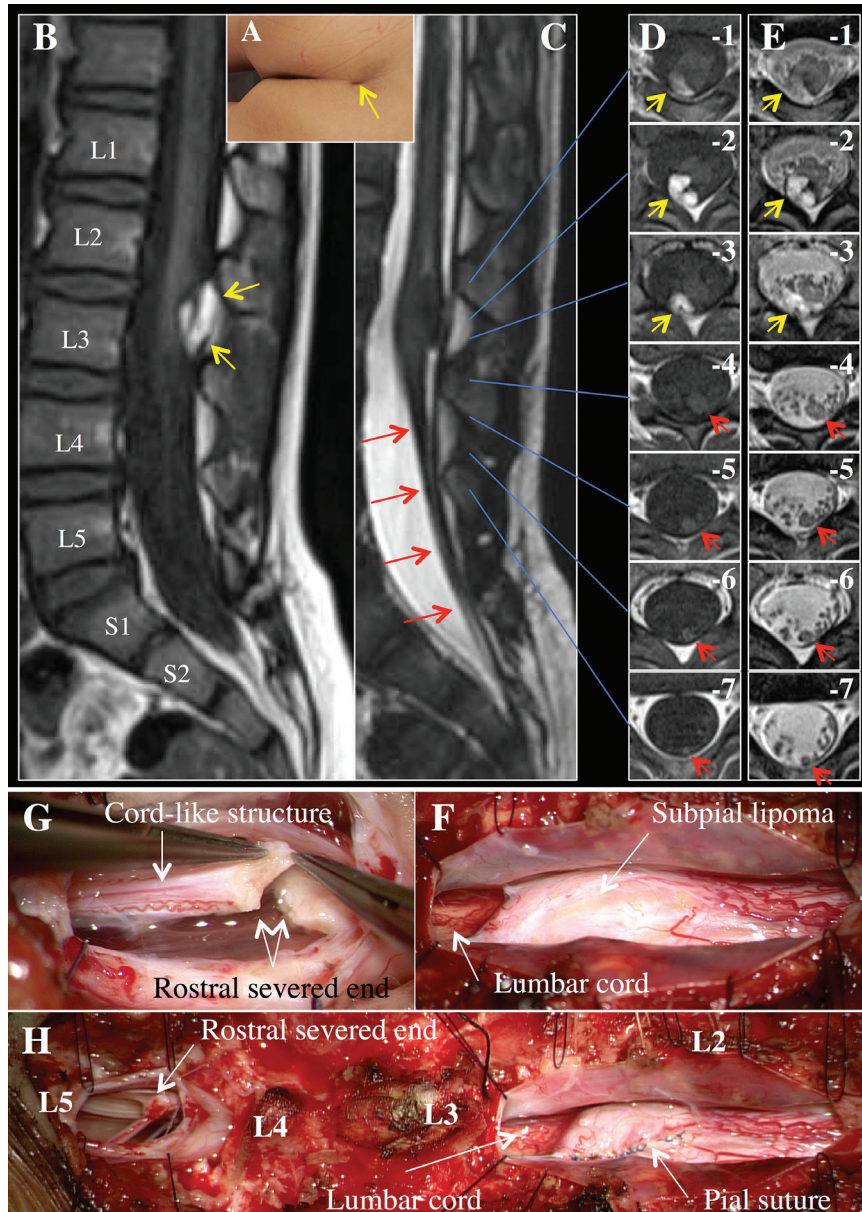
Fig. 1   (A) A dimple (yellow arrow) can be observed in the groove continuous with the gluteal cleft and just to the right of the midline. (B) Sagittal views of a variable flip-angle three-dimensional turbo spin-echo T1-weighted image (2.5 mm lateral to the right from the midline, 1.25 mm in slice thickness) and (C) 3D-hT2WI (midline, 1.25 mm in slice thickness) show a lipoma at the vertebral level of L2-3 (yellow arrows), just rostral to the conus medullaris, and low-lying conus at the L3-4 level, which is tethered with the structures continuous from the conus and extending to the dural cul-de-sac (red arrows). (D) Serial axial views of T1-weighted image (5.25 mm in slice thickness) and (E) T2-weighted image (5.25 mm in slice thickness) demonstrate that the lipoma is located at the right dorsal side of the lumbar cord (yellow arrows in D-1, 2, 3 and E-1, 2, 3). The structure without fat signals starts at the low-lying conus (red arrows in D-4, 5 and E-4, 5) and travels downward within the subarachnoid space (red arrows in D-6, 7 and E-6, 7). Spina bifida is not observed over the lipoma. Each slice level of the axial view is indicated by blue lines on the sagittal 3D-hT2WI of (C). (F–H) Intraoperative microscopic views. (F) Following laminoplastic laminotomy of L2 and upper quadrant laminectomy of L3, the subpial lipoma is exposed. The subpial lipoma shows exophytic growth from the lumbar cord and does not have a tethering effect. (G) Following the lower half of L4 laminectomy and upper half of L5 laminectomy, the C-LS is exposed and severed at the rostral side of the operative field. (H) Rostral severed end of the C-LS is observed after untethering. Pia over the lipoma is reconstructed with sutures after the minimal debulking of the lipoma. Note that most of the posterior vertebral elements of L3-5 are preserved. 3D-hT2WI: three-dimensional heavily T2-weighted imaging.
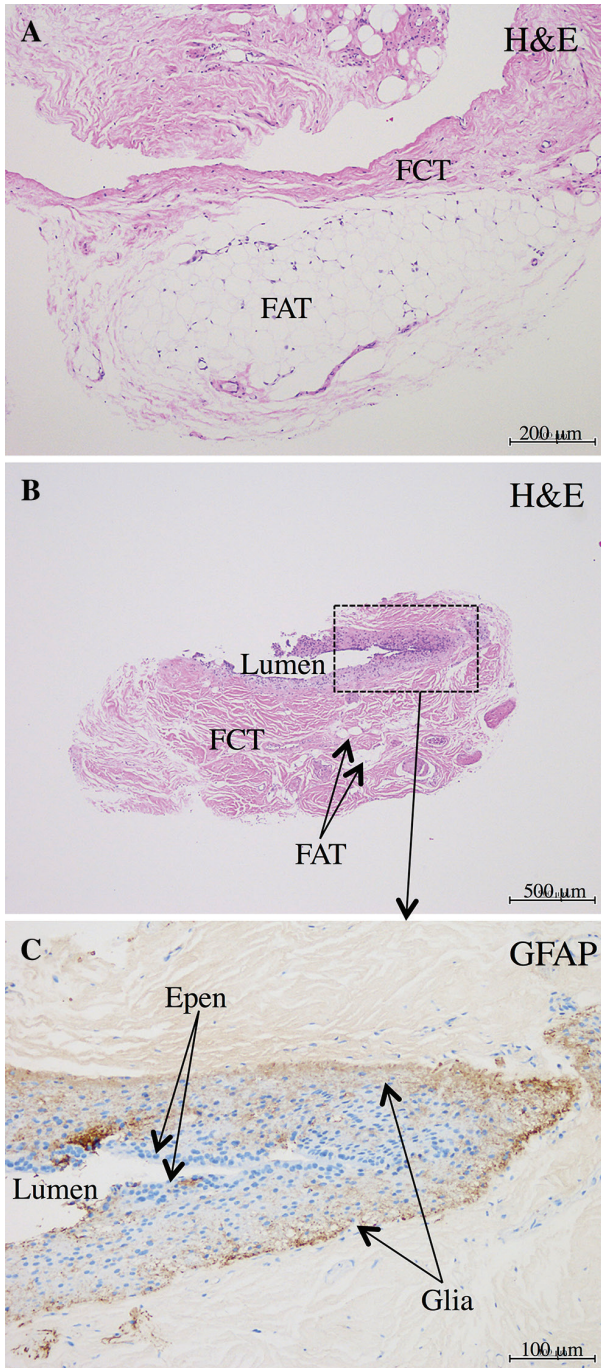
Fig. 2   (A) Photomicrograph of the subpial lipoma stained with H&E shows a mature FAT covered by a FCT. (B and C) Photomicrograph of cross sections of the filum stained with H&E (B) and immunostained for GFAP (C). A higher magnification view of the area indicated by the dotted square in (B) is shown in (C). The C-LS consists of FCT embedding a large central canal-like lumen, lined by ependymal cells (Epen) and surrounded by GFAP immunopositive neuroglial tissue (Glia). A small amount of FAT is seen in the FCT. C-LS: cord-like structure, FAT: fibroadipose tissue, FCT: fibrocollagenous tissue, GFAP: glial fibrillary acidic protein, H&E: hematoxylin and eosin.

Informed Consent

Informed consent was obtained from the family of the infant described in this report.

Conflicts of Interest Disclosure

The authors declare that they have no conflict of interest.

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Corresponding author: Takato Morioka, MD, PhD
Department of Neurosurgery, Harasanshin Hospital, 1-8 Taihakumachi, Hakata-ku, Fukuoka, Fukuoka 812-0033, Japan.
e-mail: takatons1227@gmail.com