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Case Report

Two cases of retained medullary cord running parallel to a terminal lipoma

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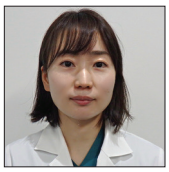

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ABSTRACT

Background: Retained medullary cord (RMC) is a newly defined entity believed to originate from the late arrest of secondary neurulation. Some RMCs contain varying amounts of lipomatous tissues, which need to be differentiated from spinal lipomas, such as filar and caudal lipomas (terminal lipomas).

Case Description: We surgically treated two patients with a nonfunctional cord-like structure (C-LS) that was continuous from the cord and extended to the dural cul-de-sac, and ran parallel to the terminal lipoma. In both cases, untethering surgery was performed by resecting the C-LS with lipoma as a column, under intraoperative neurophysiological monitoring. Histopathological examination confirmed that the central canal-like ependyma-lined lumen with surrounding neuroglial and fibrocollagenous tissues, which is the central histopathological feature of an RMC, was located on the unilateral side of the resected column, while the fibroadipose tissues of the lipoma were located on the contralateral side.

Conclusion: Our findings support the idea proposed by Pang *et al.* that entities such as RMC and terminal lipomas are members of a continuum of regression failure occurring during late secondary neurulation, and the coexistence of RMC and terminal lipoma is not a surprising finding. Therefore, it may be difficult in clinical practice to make a distinct diagnosis between these two entities.

Keywords: Retained medullary cord, Secondary neurulation, Terminal lipoma

INTRODUCTION

Retained medullary cord (RMC) is a newly defined entity believed to originate from an almost complete arrest of apoptosis during the last or degenerative phase of secondary neurulation.[8] Pang *et al.*[9] first described seven RMC patients with a redundant nonfunctional cord-like structure (C-LS), in place of the filum terminale internum, continuous from the true conus and extending to the dural cul-de-sac, resulting in neurological deficits due to spinal cord tethering. As a nonfunctional RMC is indistinguishable from the functional conus on neuroimaging and in the intraoperative view, confirming the presence of a nonfunctional C-LS with intraoperative neurophysiological monitoring (IONM) is essential for the diagnosis of RMC.[1,3,8-10] The central feature in the histopathology of RMC is the predominant presence of a

central canal-like ependyma-lined lumen with a surrounding glioneuronal core.[6,8,9,11]

Although RMCs are not considered very rare,[8,9] the number of RMC patients tends to be underdiagnosed.[1] This is probably because RMCs have been recently defined, and their pathophysiology is not been fully understood.[1] Another reason is that some RMCs contain varying amounts of lipomatous tissues[1,5,8,9,11] and need to be differentiated from spinal lipomas, such as filar and caudal lipomas (terminal lipomas),[1] which are also believed to be caused by secondary neurulation failure.[4] We surgically treated two patients who had a nonfunctional C-LS, with a predominant presence of central canal-like structure, running parallel to the terminal lipoma. We report the detailed clinicopathological findings of the cases and discuss the relationship between RMC and terminal lipoma.

CASE REPORT

Patient 1

Patient 1 was a boy without neurological deficits. At birth, he had a small tail-like appendage overlying a subcutaneous mass on the right side of the gluteal cleft, which was resected by a plastic surgeon a few days after his birth. His parents noticed that the subcutaneous mass gradually increased in size, and he was referred to us at the age of 1 year and 6 months. His gluteal cleft deviated to the left due to the subcutaneous mass [Figure 1a]. Magnetic resonance images (MRI) demonstrated that the caudal spinal cord remained thick with minimal tapering and, with a minimal increase in diameter, extended as a C-LS to the dural cul-de-sac at the S3-4 vertebral level [Figure 1b-d]. Lipomatous tissue was associated with the right half of the C-LS [Figure 1c, e1-3]. The epidural C-LS was located at the center of the epidural fat [Figure 1e-4].

Untethering surgery was performed at the age of 1 year and 8 months. Laminoplastic laminotomy of L4-bifid S2 revealed that the caudal spinal cord and C-LS extended to the dural cul-de-sac without an intervening terminal filum. The lipoma was observed at the right side of the C-LS below the L5-S1 level [Figure 1f-1 and 2, and g-1]. The border between the true cord and C-LS was determined with IONM, by tracing the evoked compound muscle action potentials of the legs and anus with stimulation, beginning from the functional cord [Figure 1h-(1)(2) and (3)] and proceeding to the nonfunctional C-LS [Figure 1h(4)(5) and (6)]. The C-LS with the lipoma was severed at this border and at the caudal most side of the operative field, and it was resected as a column [Figure 1f-3]. The rostral and caudal severed ends ascended and descended, respectively. The remnant lipoma was minimally debulked and the pial surface was reconstructed with sutures [Figure 1f-4, and g-2].

The postoperative course was uneventful. Histopathological examination revealed that the C-LS, which consisted of fibrocollagenous tissue embedding a central canal-like structure lined by ependymal cells and surrounded by glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues, was located on the left side of the resected column [Figure 1i-k]. The lipoma was located on the right side of the column.

Patient 2

Patient 2 was a boy without neurological deficits. At birth, abnormal grooves continuous from the gluteal cleft were noted [Figure 2a]. MRIs, at 26 days [Figure 2b and c] and 3 months [Figure 2 d-g] of age, demonstrated that the caudal spinal cord and continuous C-LS, with a large syringomyelia cyst at the L5-S2 level, extended to the dural cul-de-sac at the S2-3 level. The caudal and right half of the cyst wall comprised lipomatous tissue, which became more evident on the 2nd MRI.

Untethering surgery was performed at 4 months of age. Laminoplastic laminotomy of L5-S3 revealed that the caudal spinal cord and C-LS extended to the dural cul-de-sac [Figure 2h-1, and 2, and i-1]. The cyst wall and lipoma were observed on the right and caudal sides of the C-LS, respectively. IONM failed to determine the border between the true conus and C-LS [Figure 2j(1)-(6)], probably because of the current spread through the small but functional nerve roots, which tightly adhered to the ventral surface of the cord and C-LS [Figure 2i-2]. The C-LS with the lipoma was severed near the dural cul-de-sac and elevated to separate the nerve roots from the C-LS [Figure 2h-3]. A small amount of water-like clear fluid flowed out from the cyst and the cyst collapsed. After confirmation of a nonfunctional C-LS with IONM [Figure 2 j(7) and (8)], the caudal part of the C-LS along with the lipoma was resected as a column [Figure 2 h-4]. The lipoma was minimally debulked to form a decent stoma for cystostomy [Figure 2h-5 and i-4].

The postoperative course was uneventful. Histopathological examination revealed that the C-LS was located on the right side of the resected column [Figure 2k, and l]. The center of the C-LS was a cyst with a large central canal-like structure, lined by ependymal cells, and surrounded by GFAP-immunopositive neuroglial tissue and fibrocollagenous tissue. The lipomatous tissues were located on the left side of the column. The central canal-like structure with surrounding neuroglial tissues and fibrocollagenous tissues was caudally continuous, but its size decreased toward the caudal side [Figure 2 m-q].

DISCUSSION

Although the exact anatomical relationship between RMC and the associated terminal lipoma was not demonstrated,

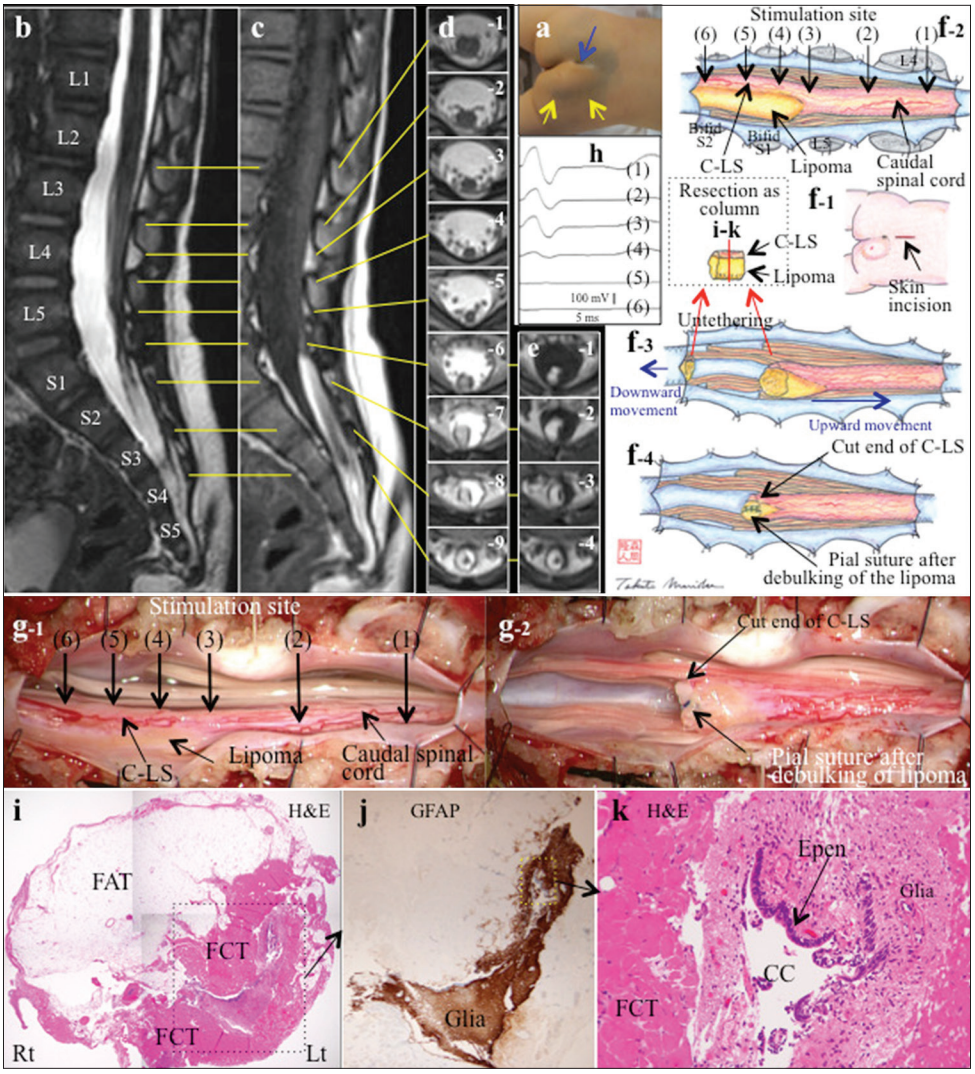

Figure 1: (a) Photograph showing a subcutaneous mass (yellow arrows) on the right side of the gluteal cleft which deviates to the left. Another groove is noted at the rostral side of the gluteal cleft (blue arrow). (b-d) Sagittal views (slice thickness of 1.25 mm) of three-dimensional heavily T2-weighted image (3D-hT2WI) (b) and three-dimensional T1-weighted spoiled gradient-recalled echo image (3D-T1WI) (c) and axial views (slice thickness of 5.2 mm) of T2-weighted image (T2WI) depict that the caudal spinal cord remains thick with minimal tapering (b, c, d-1-5) and, with minimal increase in diameter, extends as a cord-like structure (C-LS) to the dural cul-de-sac at the L3-4 vertebral level (b, c, and d-6-9). (e) Axial views (slice thickness of 5.2 mm) of 3D-T1WI demonstrate that lipoma is associated with the right half of the C-LS (e1-3). The extradural C-LS is located in the center of the epidural fat (e-4). (f) Schematic drawing and (g) microscopic view of the operative findings and (h) intraoperative neurophysiological monitoring (IONM). A lineal skin incision was made on the mid-lumbosacral region (f-1). Laminoplastic laminotomy of L4-bifid S2 revealed that the caudal spinal cord and C-LS extended to the dural cul-de-sac without an intervening terminal filum (f-2, g-1). The lipoma was observed at the right side of the C-LS below L5-S1 level. The border between the true cord and C-LS was determined with IONM, by tracing the evoked compound muscle action potentials (CMAPs) of the external anal sphincter with stimulation of 1 mA (h), beginning from the functional cord (h-(1)(2)(3)) and proceeding to the to the nonfunctional CL-S (h-(4)(5)(6)). Stimulation sites are indicated on (f-2) and (g-1). The C-LS with lipoma was severed at this border and most caudal side of the operative field and resected as a column (f-3). The lipoma was minimally debulked and the pial surface was reconstructed with sutures (f-4, g-2). (i-k) Photomicrograph of cross-sections of the C-LS including lipoma with hematoxylin and eosin staining (HE) (i,k) and immunostaining for glial fibrillary acidic protein (GFAP) (j). The location of section is indicated as red line in (f-3) and the orientation is almost matched with that of (e-2). A higher magnification view of the area is indicated by the dotted square in (i) and (j). The C-LS, which consists of fibrocollagenous tissue (FCT) embedding a central canal (CC)-like structure lined by ependymal cells (Epen) and surrounded by GFAP immunopositive neuroglial tissues, is located on the left side of the resected column. The lipoma, which consists of a mature fibroadipose tissue (FAT), is located on the right side of the C-LS. Original magnification: (i) ×4, (j) ×40, and (k) ×200.

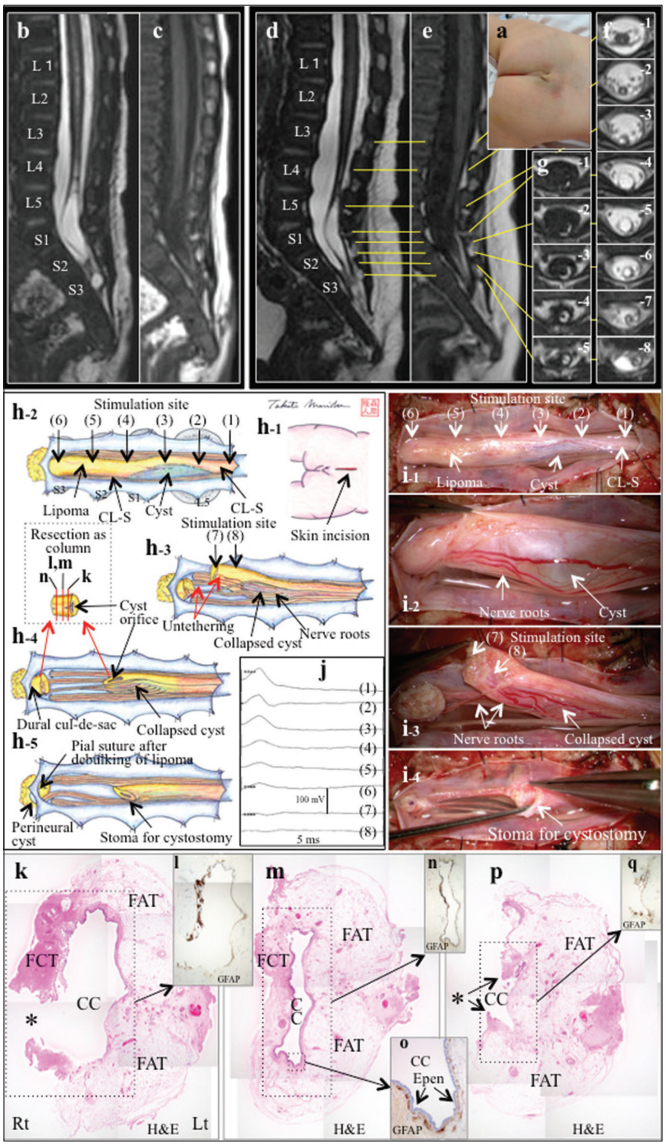

Figure 2: (a) Photograph showing an abnormal Y-shaped groove and another groove, continuous from the gluteal cleft. (b-g) Preoperative images. Sagittal views (slice thickness of 1.25 mm) of 3D-hT2WI (b) and 3DT1WI (c) at 26 days of age, and sagittal views (slice thickness of 1.25 mm) of 3D-hT2WI (d) and 3DT1WI (e) and axial views (slice thickness of 3.9 mm) of T2WI (f) and T1-weighted image (g) at 3 months of age depict that the caudal spinal cord and continuous C-LS, with a large syringomyelia cyst at the L5-S2 level, extends to the dural cul-de-sac at the S2-3 level. The caudal and right half of the cyst wall is lipomatous tissue, which became more evident on the 2nd MRI. A sacral perineural cyst is also noted. (h-j) Schematic drawing (h) and microscopic view of the operative findings (i) and IONM (j). A lineal skin incision was made on the mid-lumbosacral region (h-1). Laminoplastic laminotomy of L5-S3 revealed that the caudal spinal cord and C-LS, including the cyst, extended to the dural cul-de-sac without an intervening terminal filum (h-2, i-1). The cyst wall and the lipomatous tissue were observed at the right and caudal side of the C-LS, respectively. IONM, by tracing the evoked CMAPs of the external anal sphincter with stimulation of 0.5 mA, failed to determine the border between the true cord and C-LS, while the amplitude of CMAPs tended to decrease toward the caudal side (j(1)-(6)), probably due to the current spread through the small nerve roots, which were tightly adhered to the ventral surface of the cord and C-LS (i-2). The lipoma with C-LS was severed at the caudal part and elevated (h-3). A small amount of water-like clear fluid flowed out from the cyst and the cyst was collapsed. After confirming nonfunctional C-LS and lipoma with IONM (j(7)(8)), the caudal part of the C-LS along with lipoma was resected as a column (h-4). The lipoma was minimally debulked to enlarge the cyst orifice and make a decent stoma for cystostomy (h-5, i-4). The caudal remnant lipoma was minimally debulked and the pial surface was reconstructed with sutures. No direct surgical procedure for sacral perineural cyst was performed. (k-q) Photomicrograph of cross-sections of the C-LS including lipoma with HE (k,m,p) and immunostaining for GFAP (l,n,o,q). The location of section is indicated as red line in (h-4) and the orientation is almost matched with that of (g-3,4). A lower or higher magnification view of the area is indicated by the dotted square in (k), (m), and (p). The C-LS, which consists of fibrocollagenous tissue embedding a central canal-like structure lined by ependymal cells and surrounded by GFAP-immunopositive neuroglial tissues, is located on the right side of the resected column. The lipoma, which consists of a mature fibroadipose tissue, is located on the left side of the column. Central canal-like structure decreased in size, moving caudally. *Indicates the destruction of the cyst wall created when preparing the specimen. Original magnification: (k-q) ×4 and (o) ×20.

Kim *et al*.[1] showed that, in a caudal lipoma, the tapered low-lying conus is commonly continuous with the distally enlarging lipoma. When the low-lying conus contains a nonfunctional part, it is considered an RMC, as was the case for patient 3 in the first description by Pang *et al*.[9] Therefore, operative exposure with IONM of the critical region is necessary for a definitive diagnosis of RMC.[1,3,8-10]

In the IONM method, stimulation in the rostrocaudal direction is recommended.[3,9,10] This helps identify the exact border when the signal becomes null, and we used this method for patient 1 in our report. To prevent the current spread through the cerebrospinal fluid, a rubber or polyester sheet is commonly placed under the cord/C-LS. In patient 2, however, small but functional nerve roots that tightly adhered to the ventral surface of the cord/C-LS did not permit electrical isolation. Therefore, we followed the method recommended by Kim *et al*.[1] We first cut the safe caudal end and then the CL-S along with the lipoma was gently lifted. After stimulation in the caudo-rostral direction, being careful to avoid the current spread, the nonfunctional C-LS with lipoma was resected as a column; however, we could not define the exact border. Another possible confounding factor contributing to the lack of good IONM findings is patient 2's young age. Although the optimal timing for prophylactic surgery remains undetermined, it is commonly performed within the 1st year of life.[2] At our institute, untethering surgery is performed around the age of 3 months and/or attainment of 5 kg in weight, for a lumbosacral lipoma with intact or subtle neurological findings diagnosed at birth.[2]

In patient 1, neuroimaging and intraoperative findings showed that the nonfunctional C-LS, continuous from the cord, and the terminal lipoma ran on the left and right sides of the column, respectively. Histopathological examination confirmed that the central canal-like ependyma lined lumen with surrounding neuroglial and fibrocollagenous tissues was located on the left side of the column and the fibroadipose tissue on the right. Patient 2 had the same clinicopathological findings as the previously reported cases of cystic RMC with terminal lipoma.[1,5,8] Histopathologically, the C-LS with cystic dilatation of the central canal-like structure, which was continuous from the large syringomyelia cyst caudally, and the terminal lipoma were located on the right and left sides of the column, respectively. Therefore, we concluded that the RMCs ran parallel to the terminal lipoma in the resected column in both patients.

In this study, the RMC was located in the lateral part rather than at the center of the lipoma, as reported in a previous study.[5] This finding was probably caused by the difference between the formation of the primary and secondary neural tubes. Secondary neurulation does not involve folding of the embryonic surface but instead is associated with morphologic changes below the surface ectoderm deep within the body of the embryo.[5,8]

The present findings support the idea raised by Pang *et al*.[8,10] that entities such as RMC and terminal lipomas can be considered members of a continuum of regression failure occurring during late secondary neurulation. They speculate that RMC and terminal lipoma differ from each other in terms of timing and severity of apoptosis failure. Histopathologically, the main distinguishing feature of RMC from other entities is the dominant presence of an ependyma lined canal with a neuroglial core. Fibroadipose tissue is the main constituent in terminal lipomas. Therefore, the coexistence of the RMC and terminal lipoma, as in these cases, is not a surprising finding.[5,7,8] As the terminology for RMC and lipoma was based on different backgrounds, namely, embryological and anatomical backgrounds, respectively, it may be difficult in clinical practice to make a distinct diagnosis between these two entities.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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