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Case Report

Spinal cord deformity with aggravation of tethering in saccular limited dorsal myeloschisis during the first 2 months of life

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ABSTRACT

Background: Although the optimal timing of prophylactic untethering surgery for limited dorsal myeloschisis (LDM) with intact or subtle neurological findings diagnosed at birth remains undetermined, intentional delayed surgery is commonly used for flat and tail-like LDM. Conversely, for saccular LDM, early surgery is indicated during the postnatal period because it prevents rupture of the sac. We treated a saccular LDM patient, in whom intentional delayed surgery was selected because the sac was thickly covered with normal skin. We describe the clinical course of the case and discuss the optimal timing of the surgery.

Case Description: The patient had a dorsal midline sac in the upper lumbar region. Initial magnetic resonance imaging (MRI) after birth revealed a tethering tract that began at the dome of the sac and joined the lumbar cord. Dorsal bending of the cord at the stalk-cord union and invagination of the cord into the sac were noted. At 2 months, he was neurologically normal; however, the second MRI examination revealed that the cord tethering was aggravated. The cord was markedly displaced dorsally and to the left, with deviation of the cord to the sac out of the spinal canal. Following untethering surgery, the spinal cord deformity markedly improved.

Conclusion: Early surgery may be recommended for saccular LDM when tethering is present, including dorsal bending of the cord at the stalk-cord union and invagination of the cord into the sac observed on detailed MRI examination, even if the sac has no risk of rupture.

Keywords: Limited dorsal myeloschisis, Segmental myelocystocele, Spinal cord deformity, Tethering, Untethering

INTRODUCTION

Limited dorsal myeloschisis (LDM) is thought to originate from a small segmental failure of the dorsal closure of the neural folds during primary neurulation. At the focal limited nonclosure site, the disjunction between the cutaneous and neural ectoderm is impaired. This results in a retained fibroneural stalk linking the skin lesion and the dorsal spinal cord, which results in cord tethering.[15,16,20] The recommended treatment consists of prophylactic untethering of the stalk

from the cord.[15,16,20] Based on the skin manifestation, LDMs were originally classified as either saccular and nonsaccular (flat) LDMs.[15,16] Recently, tail-like LDMs, with a tethering stalk continuing to a human tail-like cutaneous appendage, have been characterized.[1,7,13,18] Between April 2015 and March 2021, 24 Japanese LDM patients underwent initial untethering surgery at our affiliated hospitals. External skin lesions were flat in 13 patients, saccular in six, and tail-like in five.

Although the optimal timing of prophylactic untethering surgery for LDMs with intact or subtle neurological findings diagnosed at birth remains undetermined, intentional delayed surgery is commonly selected for flat and tail-like LDM.[10,11] At our institute, surgery is performed around the age of 3 months and/or attainment of 5 kg in weight, according to the timing of our untethering surgery for lumbosacral lipoma.[6,8] However, early surgery is often indicated during the postnatal period for saccular LDMs because it prevents rupture of the sac or a major hindrance to proper handling of the baby.[2,5,9,11,15-17] Of six saccular LDM cases, five underwent early surgery 1–11 days after birth.[9,11,19] In the remaining case, because the surface of the sac was thickly covered with normal skin at birth and had little to no risk of rupture, intentional delayed surgery was scheduled at 2 months based on the surgery for flat and tail-like LDM. During the first 2 months of life, follow-up magnetic resonance imaging (MRI) demonstrated a spinal cord deformity due to aggravation of the tethering effect with enlargement of the extraspinal sac. Herein, we describe the clinical course of the case and discuss the optimal timing of surgery.

CASE REPORT

The patient was a boy weighing 3055 g delivered through a scheduled repeated cesarean section at 38^{+2} weeks of gestation. The patient had Apgar scores of 8 and 9. Physical examination revealed a dorsal midline sac in the upper lumbar region measuring 30×30×10 mm in length × width × height that was thickly covered with normal skin [Figure 1a]. The patient had no neurological deficits. The initial MRI, including three-dimensional T1-weighted spoiled gradient-recalled echo imaging (3D-T1WI) and three-dimensional heavily T2-weighted imaging (3D-hT2WI),[10,11] was performed 3 days after birth and demonstrated a tethering tract that began at the dome of the meningocele sac, ran caudally in the inner wall of the sac, and joined the lumbar cord at the L3 vertebral level [Figure 1b-e]. The attachment of the stalk to the dome was wide based and contained a small syrinx cavity. With the tethering effect, the cord slightly deviated into the dome and was stuck at the entrance of the dome. The conus medullaris was positioned at the L4 level. The stalk consisting of the anterior wall of the syrinx cavity was almost isointense on T1-weighted imaging (T1WI), while the stalk consisting of the posterior wall was thick lipomatous tissue. No hydrocephalus or Chiari malformations were observed.

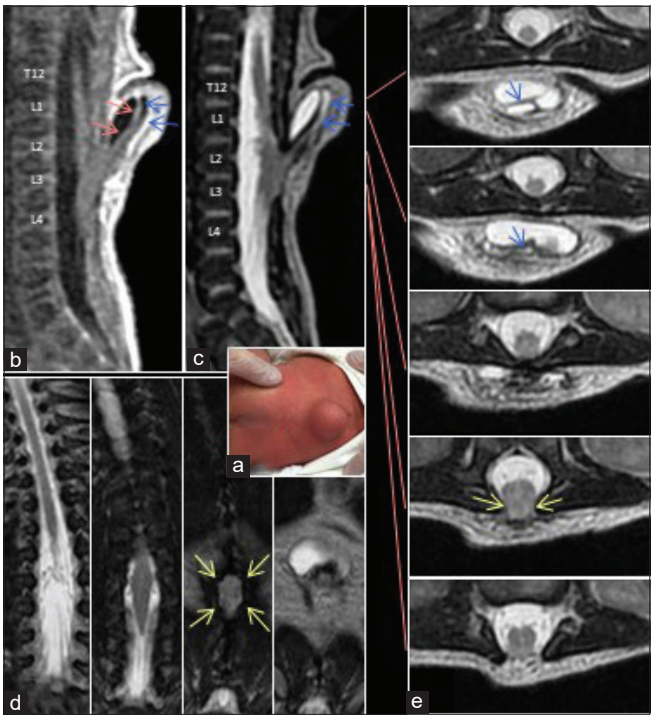

Figure 1: (a) A dorsal midline sac measuring 30×30×10 mm in the upper lumbar region. (b and c) Sagittal views of 3D-T1WI (b) and T2WI (c) demonstrate a tethering tract that began at the dome of the meningocele sac, ran caudally in the inner wall of the sac, and joined the lumbar cord at the L3 vertebral level. The attachment of the stalk to the dome contained a small syrinx cavity (blue arrows). The stalks comprising the anterior wall of the syrinx cavity were almost isointense on 3D-T1WI (red arrows). (d and e) Parallel coronal views of 3D-hT2WI with fat suppression (d) and parallel axial views of T2WI (e) also show that the attachment of the stalk to the dome. The spinal cord slightly deviated into the dome and was stuck at the entrance of the dome, which measured 15× 10 mm (yellow arrows in (d and e)).

At 2 months of age, he weighed 5230 g. The lumbar sac had grown to 50×40×15 mm [Figure 2a]. He was neurologically normal, as before. However, the second MRI examination revealed that cord tethering was aggravated due to enlargement of the sac to the left. The cord was markedly displaced dorsally and to the left, with the deviation of the cord to the sac out of the spinal canal [Figure 2b-d]. In addition to the development of subcutaneous fat, the stalks consisting of the anterior wall of the syrinx cavity became lipomatous tissue.

On the 68th day after birth, untethering and repair surgery of the sac were performed. On opening the sac, the cord emerged from the orifice of the spinal canal, which was 15 mm in diameter, with the tethering of the stalk spread widely on the inner wall of the sac [Figure 2e and f]. The border between the cord and stalk could be distinguished,

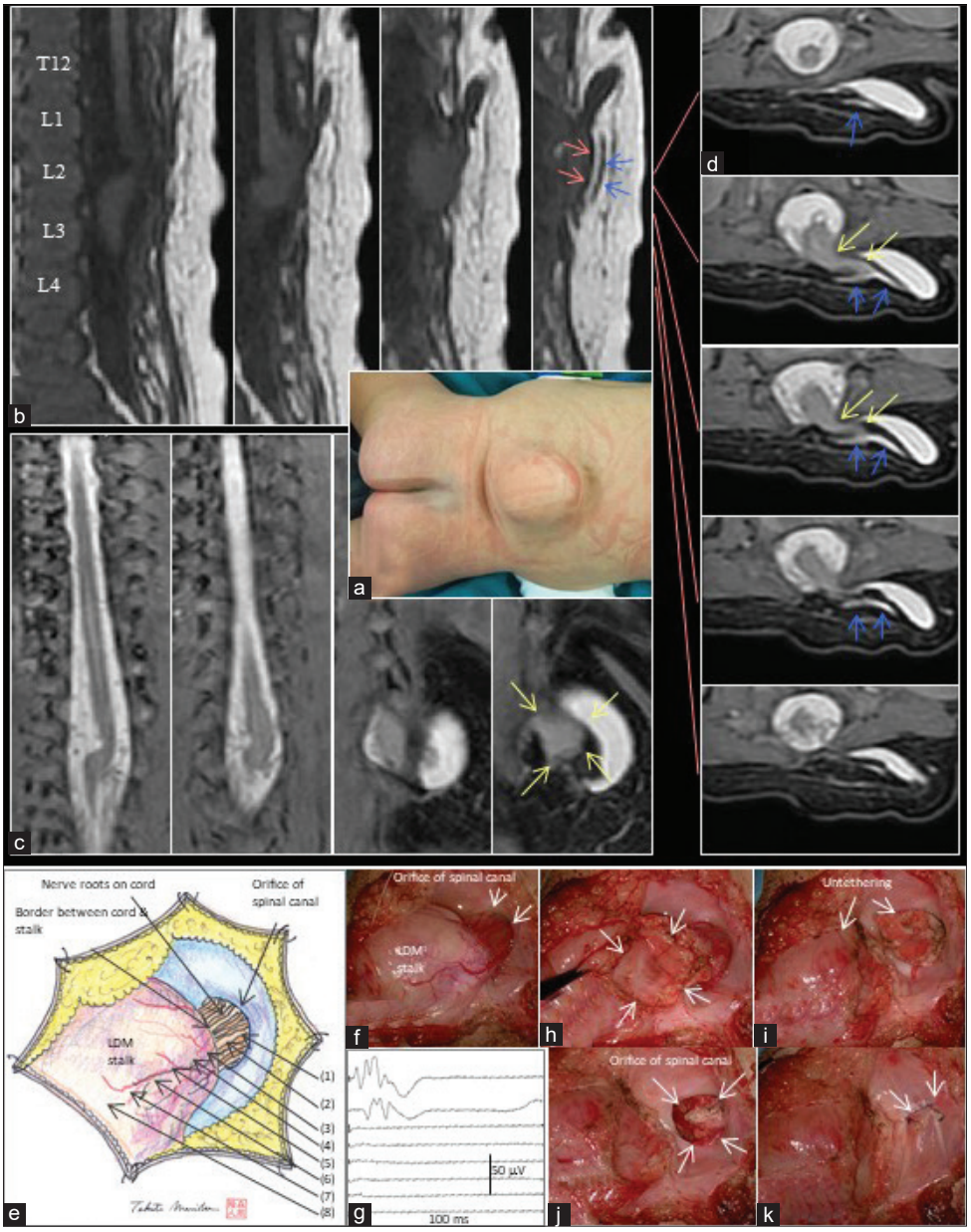

Figure 2: (a) The dome grew to 50×40×15 mm by 2 months of age. (b and c) Parallel sagittal views of T1WI (b), parallel coronal views of 3D-hT2WI with fat suppression (c), and parallel axial views of 3D-hT2WI with fat suppression (d) revealed that cord tethering was aggravated. With the deviation of the cord to the sac out of the spinal canal (yellow arrows in (c and d)), the cord was displaced dorsally and to the left. The stalk comprising the anterior wall (red arrows) of the syrinx cavity (blue arrows) became lipomatous tissue. (e-k) Schematic drawing (e) and microscopic view of the intraoperative findings (f and h-k), and intraoperative neurophysiological monitoring (g). (e and f) The cord emerged from the orifice of the spinal canal. The border between the spinal cord and stalk could be distinguished and nerve roots were found on the cord. (g) This border was neurophysiologically confirmed by tracing the evoked compound muscle action potentials (CMAPs) of the hamstring with direct stimulation with 3 mA intensity starting from the functional cord and continuing to the nonfunctional stalk. The CMAPs were evoked following stimulation at the cord (1-2); no CMAPs were evoked following stimulation at the stalk (3-8). (h) The stalk was severed just distal to the border; the syrinx cavity was opened. (i) The cord was untethered from the stalk. (j) The severed edge was approximated with a pial suture and returned to the spinal canal. (k) The orifice of the cord was tightly closed.

and nerve roots found on the cord. This border was neurophysiologically confirmed by tracing the evoked compound muscle action potentials of the external anal sphincter, hamstring, and gastrocnemius muscles with direct stimulation starting from the functional cord and continuing to the nonfunctional stalk [Figure 2g]. The stalk was severed

immediately distal to the border, the syrinx cavity was opened [Figure 2h], and the cord was untethered from the stalk [Figure 2i]. The severed edge was approximated with a pial suture and returned to the spinal canal [Figure 2j]. The orifice of the cord was tightly closed [Figure 2k] and covered with a flap of the paravertebral muscle.

Postoperatively, the patient did not develop neurological deficits. MRI performed on the 14th day postoperatively demonstrated that the spinal cord deformity markedly improved due to the successful untethering of the cord, although minor hemorrhage was noted at the severed edge of the stalk [Figure 3a and b].

Histopathologically, the resected stalk included a central canal-like lumen lined by ependymal cells. Surrounding glial fibrillary acidic protein (GFAP)-immunopositive neuroglial tissues and fibrocollagenous tissues embedded

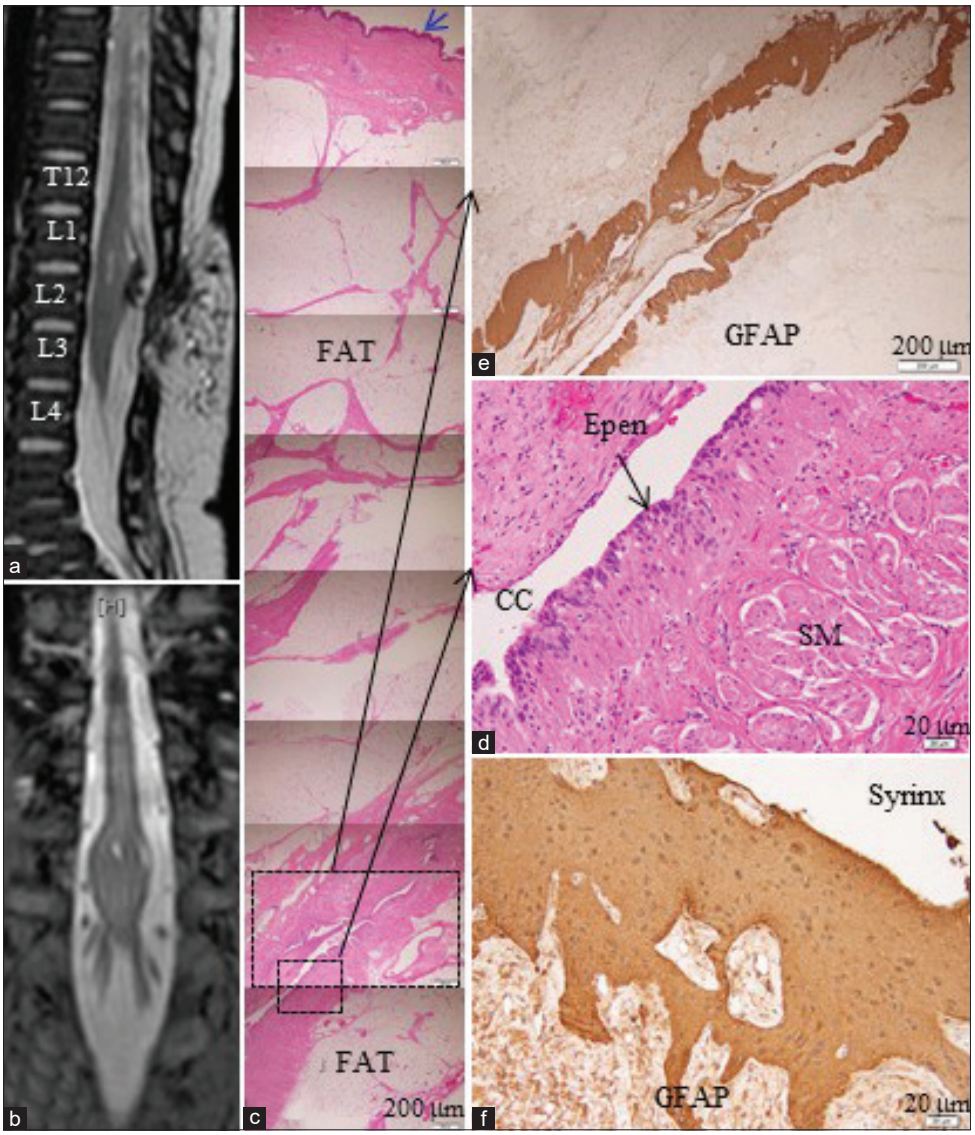

Figure 3: (a and b) Sagittal view of T1WI (a) and coronal image of 3D-hT2WI with fat suppression (b) performed on the postoperative 14th day demonstrated that the spinal cord deformity markedly improved, although a minor hemorrhage was noted at the severed edge of the stalk. (c-f) Histopathological examination of the resected stalk stained with hematoxylin and eosin (c, d) and immunostained for glial fibrillary acidic protein (GFAP) (e and f). Higher magnification view of the area indicated by the dotted square in (c) is shown in (d and e). A central canal (CC)-like lumen lined by ependymal cells (Epen) and surrounding GFAP-immunopositive neuroglial tissues and fibrocollagenous tissue embedded with smooth muscle (SM) fibers was noted in the fibroadipose tissue (FAT). The sac was covered with finely jagged squamous epithelium (blue arrow in (c)). (f) The wall facing the syrinx cavity was composed of GFAP-immunopositive neuroglial tissue including neuronal cells and had no ependymal lining.

with smooth muscle fibers in the fibroadipose tissue were also present [Figure 3c-e]. The wall facing the syrinx cavity was also composed of GFAP-immunopositive neuroglial tissue, including neuronal cells, with no ependymal lining [Figure 3f].

DISCUSSION

Pang *et al*.[15,16] classified saccular LDMs into three types: segmental (nonterminal) myelocystocele, basal neural nodule, and stalk to dome. This case was classified as a segmental myelocystocele because MRI demonstrated a syrinx cavity in the stalk along the wall of the sac, and histopathological examination revealed a central canal-like lumen lined by ependymal cells and surrounding GFAP-immunopositive neuroglial tissues present in the stalk. An ependymal lining could not be identified where the wall faced the syrinx cavity. Despite the expectation of an ependymal lining, as implied by their name, myelocystocele sacs are often devoid of ependymal lining because of their vulnerability.[14] Thus, the LDM stalk in the present case was a segmental myelocystocele attached to the extraspinal sac wall that was caught in the thick subcutaneous lipomatous tissue of the sac wall.

The most characteristic morphological finding in this case was aggravation of tethering and deformity of the cord during the first 2 months of life. Kim *et al*.[4] stated that enlargement of extraspinal cysts associated with closed spinal dysraphism, including saccular LDMs, could aggravate tethering by pulling the tip of the cord. In this case, the sac was enlarged to the left, and the cord entered the sac with the aggravation of tethering. According to the original description by Pang *et al*.,[15,16] tension in the stalk is illustrated by a dorsal bending of the cord at the stalk-cord union, which was demonstrated on the initial MRI in our case. Furthermore, the cord deviated slightly from the spinal canal toward the sac. The spreading of the stout LDM stalk on the inner surface of the sac might have contributed to the aggravation of tethering. These findings became clear by imaging with sagittal, coronal, and horizontal sections of 3DT1 and 3DhT2WI, in addition to the conventional T1WI and T2WI, because these images can be visualized with a thin slice.[3,6,8,10-12] In our case, development of neurological exacerbations was prevented by the untethering surgery on the 68th day. However, early surgery could be recommended when there are any stout tethering findings, including dorsal bending of the cord at the stalk-cord union and invagination of the cord into the sac as visualized on detailed MRI examination at birth.

Another notable finding was that the LDM stalk consisting of the anterior wall of the syrinx cavity became lipomatous tissue, and histologically, most of the resected stalk consisted of fibroadipose tissue. LDMs are thought to arise from focal incomplete disjunction between the cutaneous and neuroectoderm during primary neurulation, while spinal lipomas of the dorsal type (dorsal lipomas) arise from premature disjunction. Thus, the simultaneous occurrence of an LDM and a dorsal lipoma has been well documented.[13,15,16] Spinal lipomas are known to have the potential for growth during the 1st months of life.[6] Thus, it is quite possible that the lipomatous component in the LDM stalk also increased, as demonstrated in a previous report.[13]

CONCLUSION

Spinal deformities could occur due to aggravation of the tethering effect with the enlargement of the extraspinal sac. As such, early surgery may be recommended for saccular LDMs when there are any stout tethering findings, including dorsal bending of the cord at the stalk-cord union and invagination of the cord into the sac at birth, even if the surface of the sac is covered with normal skin.

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Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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