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Case Report

Refractory CSF leakage following untethering surgery performed 10 months after birth for enlarging terminal myelocystocele associated with OEIS complex

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ABSTRACT

Background: Terminal myelocystocele (TMC) is an occult spinal dysraphism characterized by cystic dilatation of the terminal spinal cord in the shape of a trumpet (myelocystocele) filled with cerebrospinal fluid (CSF), which herniates into the extraspinal subcutaneous region. The extraspinal CSF-filled portion of the TMC, consisting of the myelocystocele and the surrounding subarachnoid space, may progressively enlarge, leading to neurological deterioration, and early untethering surgery is recommended.

Case Description: We report a case of a patient with TMC associated with OEIS complex consisting of omphalocele (O), exstrophy of the cloaca (E), imperforate anus (I), and spinal deformity (S). The untethering surgery for TMC had to be deferred until 10 months after birth because of the delayed healing of the giant omphalocele and the respiration instability due to hypoplastic thorax and increased intra-abdominal pressure. The TMC, predominantly the surrounding subarachnoid space, enlarged during the waiting period, resulting in the expansion of the caudal part of the dural sac. Although untethering surgery for the TMC was uneventfully performed with conventional duraplasty, postoperative CSF leakage occurred, and it took three surgical interventions to repair it. External CSF drainage, reduction of the size of the caudal part of the dural sac and use of gluteus muscle flaps and collagen matrix worked together for the CSF leakage.

Conclusion: Preoperative enlargement of the TMC, together with the surrounding subarachnoid space, can cause the refractory CSF leakage after untethering surgery because the expanded dural sac possibly increases its own tensile strength and impedes healing of the duraplasty. Early untethering surgery is recommended after recovery from the life-threatening conditions associated with OEIS complex.

Keywords: Collagen matrix, Duraplasty, Ependyma, Gluteus muscle flap, Hydrodynamic pressure, Subarachnoid space

INTRODUCTION

Terminal myelocystocele (TMC) is a rare closed spinal dysraphism characterized by cystic dilatation of the terminal part of the central canal, surrounded by a subarachnoid space that then herniates dorsally to an extraspinal region through a posterior spina bifida.[5,6,13,15,20,21] TMC is one of the

common spinal deformities associated with OEIS complex, which is a congenital condition consisting of omphalocele (O), exstrophy of the cloaca (E), imperforate anus (I), and spinal deformity (S) with an estimated incidence of one in 100,000–400,000 live births.[3,7] Because the first three conditions are life threatening and may determine prognosis, they are treated by pediatric surgeons and urologists on a priority basis. Neurosurgical intervention for closed spinal dysraphism, which is covered with normal skin, is deferred until other lifesaving procedures have been performed.[15,19,24] Some types of closed spinal dysraphism, including TMC, accompany the extraspinal portion filled with cerebrospinal fluid (CSF), which may progressively enlarge, leading to neurological deterioration, and early untethering surgery is recommended.[9,11,20]

We present a case of TMC associated with OEIS in which the untethering surgery was deferred until 10 months after birth because of the delayed healing of the giant omphalocele and the respiratory instability due to hypoplastic thorax and increased intra-abdominal pressure. The TMC massively enlarged during the waiting period, and CSF leakage occurred after the untethering surgery, which required three surgical interventions. The optimal timing of the untethering surgery for TMC and the mechanism and treatment of the CSF leakage are discussed.

CASE REPORT

A healthy woman had a natural pregnancy. Prenatal magnetic resonance images (MRIs) examination performed at 34^{+4} weeks of gestation revealed a lumbosacral mass filled with CSF and an abdominal wall defect [Figure 1a]. A boy weighing 2274 g was delivered at 37^{+0} weeks of gestation through an emergency cesarean section due to preterm rupture of the membranes. His Apgar scores were 5–8. He was intubated after birth because of respiratory disorder. Physical examination revealed omphalocele [Figures 1b and c], exstrophy of the cloaca [Figure 1c], and imperforate anus [Figure 1c]. A soft mass, sized 4 × 3 × 1.5 cm and covered with normal skin, was noted at the lumbosacral region [Figure 1d]. The baby had clubfeet [Figure 1b] with severe motor dysfunction below the L2 level on both sides. The omphalocele was so giant (defect >6 cm, with liver herniation), and staged closure was attempted. On day 0 after birth, silo construction, ileostomy, and closure of the urinary bladder were performed. The omphalocele was tentatively closed with prosthetic patches 22 days after birth; however, adequate epithelialization of the omphalocele surface required more time [Figure 1e]. The baby's hypoplastic thorax and excessive intra-abdominal pressure due to the visceroabdominal volume disproportion caused limited expansion of the diaphragm, which required mechanical ventilation and tracheostomy.

MRIs at 5 months after birth, including three-dimensional heavily T2-weighted imaging (3D-hT2WI) and 3D-T1-weighted imaging (3D-T1WI),[14,16] revealed cystic dilatation of the terminal spinal cord in the shape of a trumpet (myelocystocele) containing neural placode at its base, which communicated with the rostral hydromyelia [Figures 1f and g]. The myelocystocele was surrounded by a large subarachnoid space that extended posterocaudally into the extraspinal region. Neither Chiari malformation nor hydrocephalus was present. A diagnosis of TMC was made. During this period, the lumbosacral mass increased in size, but no neurological deterioration or CSF leakage was observed.

The surgery for the TMC was delayed until 10 months after birth, when sufficient stabilization of the respiratory status and epithelialization of the omphalocele sac would have been achieved, to ensure that the patient tolerated prone positioning during the operation. The abdomen remained protuberant, and definitive closure of the omphalocele was not performed at this time [Figure 1h]. 3D-hT2WI before surgery revealed further enlargement of the subarachnoid space compared to the myelocystocele, resulting in the expansion of the caudal part of the dural sac [Figures 1i and j]. Brain MRI at the same time revealed no hydrocephalus.

During surgery, the patient was placed prone with special management to protect his abdomen from pressure [Figure 2a]. The pressure on the omphalocele sac was monitored using portable interface pressure sensor (Palm Q, CAPE Co., Ltd., Yokosuka, Japan). The lumbosacral mass was 11 × 11 × 5 cm in size. The myelocystocele sac wall (MCW) was peeled off the overlying subcutaneous fat [Figure 2b], and a neural placode with a dilated central canal was revealed. Stimulation of the neural placode and nerve roots arising from the ventral side showed weak compound muscle action potentials on intraoperative neurophysiological monitoring (IONM) for the leg muscles. The neural placode was untethered by resection of the nonfunctional portion of the MCW and reconstituted with pial suture [Figures 2c and d]. Duraplasty in a water-tight fashion was achieved using fibrous tissue which was in continuity with the dural sac and had composed part of the MCW [Figure 2d]. No graft patches were used. Muscular coverage on the dural sac was achieved only at the rostral portion because there was no sufficient volume of the paraspinous muscles to cover the surgical site [Figure 1j]. Instead, thick subcutaneous fat tissue covered the reconstructed dural sac and reduced the dead space. The surface of the omphalocele sac was intact after 7.5 h of the operation.

Histopathological examination revealed that the inner wall of the myelocystocele sac was lined by an ependymal layer with surrounding glial fibrillary acidic protein-immunopositive neuroglial tissues, and it was histologically diagnosed as TMC [Figure 2e].

Postoperatively, no *de novo* neurological abnormalities were observed. On the 2nd postoperative day (POD), however,

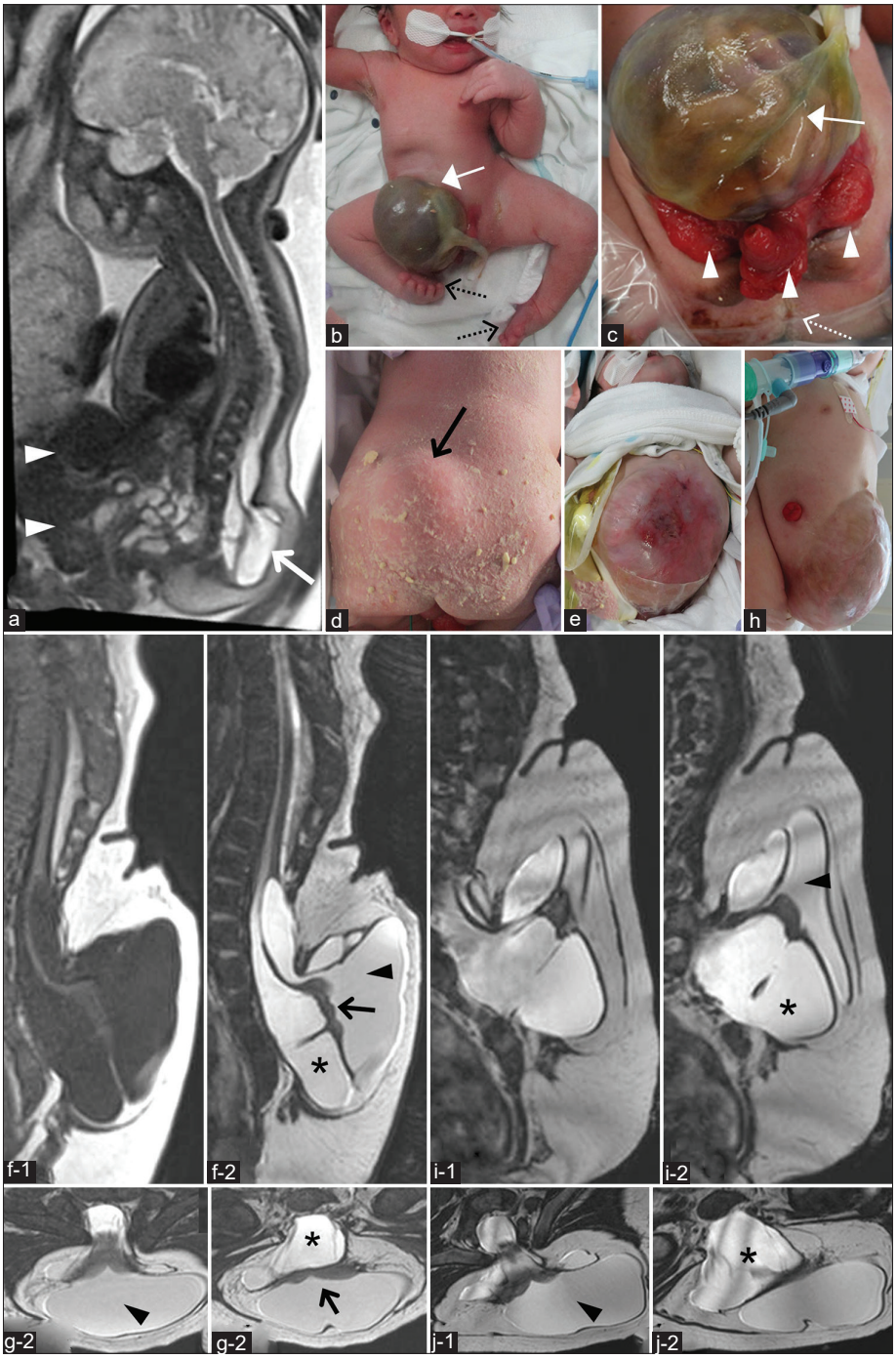

Figure 1: (a) Prenatal half-Fourier acquisition single-shot turbo spin-echo sequence image at 34 weeks of gestation revealing skin-covered CSF-filled mass at the lumbosacral region (arrow) and an abdominal wall defect with omphalocele (arrowheads). (b-d) Photographs of the patient at birth showing omphalocele (white arrow), exstrophy of the cloaca (arrowheads), imperforate anus (dotted white arrow), mass at the lumbosacral region covered with normal skin (black arrow), and bilateral club foot (dotted black arrows). (e) Photograph of the patient at 5 months after birth showing partially inadequate epithelialization of the omphalocele surface. (f and g) MRI at 5 months after birth. 3D-T1-weighted (f-1) and 3D-heavily T2-weighted magnetic resonance imaging (3D-hT2WI) (f-2, g-1 and g-2) revealing a low-lying hydromyelic cord extruding into the extraspinal space with trumpet-shaped cystic cavity (myelocystocele; arrowhead) containing neural placode (arrow) and surrounded by large subarachnoid space (asterisk) extending posterocaudally into the extraspinal region. (h) Photograph of the patient at 9 months after birth showing the epithelialization of the protuberant omphalocele sac. Sagittal views (i) and axial views (j) of 3D-hT2WI at 9 months after birth (just before surgery). Note the more enlarged subarachnoid space (asterisk) compared to the myelocystocele (arrowhead). CSF: Cerebrospinal fluid, 3D-hT2WI: Three-dimensional heavily T2-weighted imaging.

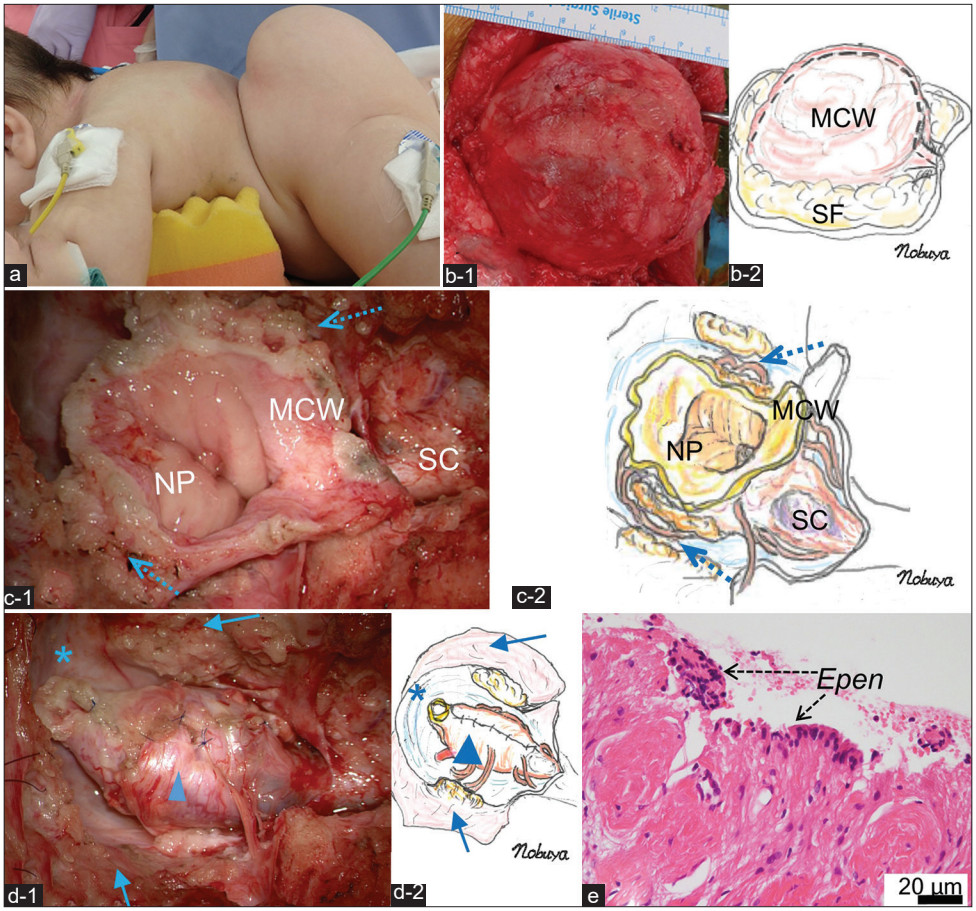

Figure 2: (a) Photograph of the patient in the prone position during the operation. Note the extended lumbosacral mass. (b) Intraoperative photograph (b-1) and schematic drawing (b-2) after stripping the SF from the huge MCW. (c) Intraoperative photograph (c-1) and schematic drawing (c-2) demonstrating the untethered NP including part of the MCW, and the terminal portion of the hydromyelic SC. The untethering site is indicated by dotted arrows. (d) Intraoperative photograph (d-1) and schematic drawing (d-2) after pia-to-pia neurulation of the neural placode (arrowhead) demonstrating lipofibrous layers (arrows) composing a part of MCW and joining to the dural sac used for the subsequent duraplasty. Note the enlarged subarachnoid space in the caudal part of the dural sac (asterisk). (e) Histopathology of the inner wall of the myelocystocele sac demonstrating the ependymal layer (Epen) with the surrounding neuroglial tissues (hematoxylin and eosin staining). MCW: Myelocystocele sac wall, SF: Subcutaneous fat, NP: Neural placode, SC: Spinal cord.

CSF leakage occurred. Surgical intervention revealed a small fistula at the dural sac, and it was sutured. Although ventricular enlargement was not noted, an Ommaya CSF reservoir was placed at the right lateral ventricle to continuously drain the CSF and reduce the pressure in the lumbar subarachnoid space. The intracranial pressure estimated from the ventricular drain was within normal limit.

The patient was placed in a prone or a semi-decubitus position. However, high-pressure CSF leakage continued [Figure 3a]. The second repair surgery for the CSF leakage was performed on POD 10. The terminal portion of the reconstituted placode was severed so that the caudal part of the enlarged dural sac extending posteriorly into the extraspinal region could shortly be reconstructed in the spinal canal [Figures 3a and b]. Although this intervention was initially unsuccessful because the CSF leakage continued, the leakage pressure gradually decreased. MRI at this time revealed that the part of subcutaneous fat had dissolved and disappeared [Figure 3b].

The third repair surgery on POD 20 revealed CSF oozing from the dural sac without any apparent fistula. A collagen matrix (DuraGen) was placed on the dural sac, and muscle turnover flaps employing the superficial layer of the gluteus maximus were used to cover the dural sac and reduce the dead space. Postoperatively, the CSF leakage decreased despite discontinuation of the external CSF drainage on POD 32 and eventually resolved. MRI 4 months after the untethering surgery revealed neither CSF leakage [Figure 3c] nor hydrocephalus. For the 1 postoperative year up to the

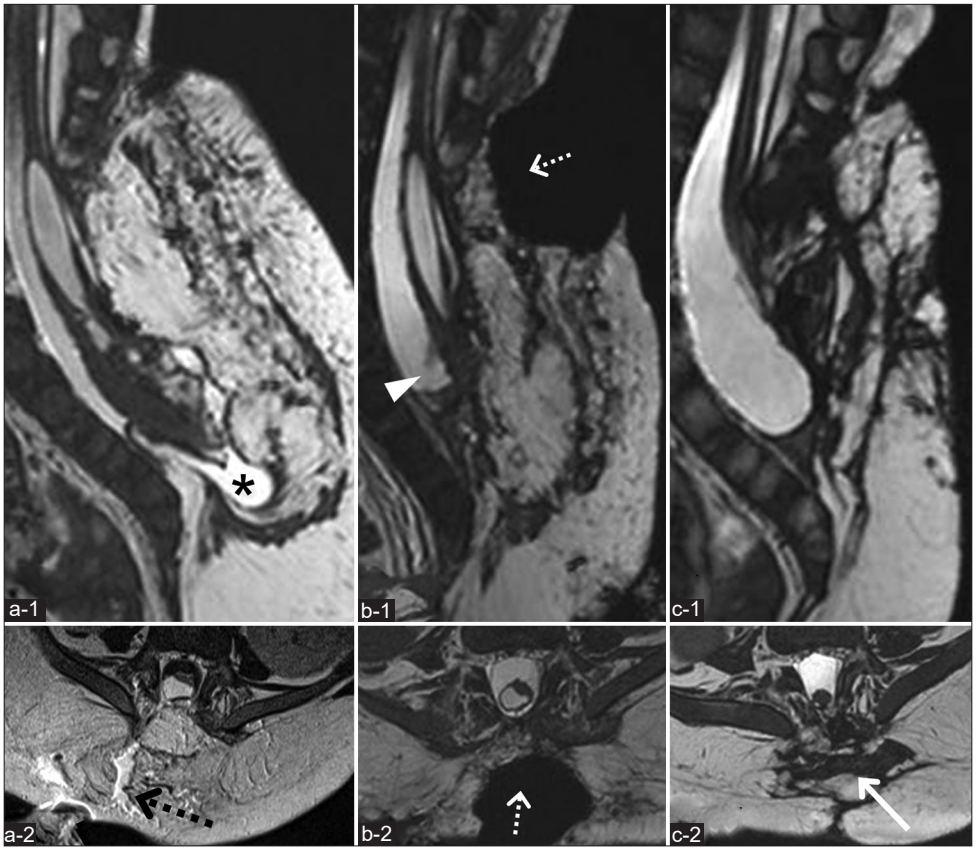

Figure 3: (a) MRI after the first repair surgery for the CSF leakage showing the caudal subarachnoid space in the dural sac (asterisk) (a-1; 3D-hT2WI) and CSF leakage through the gaps between the subcutaneous tissue (black dotted arrow) (a-2; T2WI). Sagittal (b-1) and axial (b-2) views of 3D-hT2WI after the second repair surgery for CSF leakage revealing the deficit of the subcutaneous tissues and the dorsal aspect of the dural sac, which was covered with only thin tissue without muscle layers (white dotted arrow). Note the shortened terminal part of the reconstituted spinal cord and the dural sac (arrowhead). Sagittal (c-1) and axial (c-2) views of 3D-hT2WI 3 months after the third repair surgery (4 months after the untethering surgery) demonstrating the dorsal aspect of the dural sac covered with flaps of the gluteus muscles (arrow) without CSF leakage. CSF: Cerebrospinal fluid, 3D-hT2WI: Three-dimensional heavily T2-weighted imaging.

time of writing, neither recurrence of CSF leakage nor hydrocephalus was noted. The motor function of the lower extremities did not worsen, and the level of cognitive development was considered appropriate for his age.

DISCUSSION

The pathoembryogenesis of TMC is considered to be a retained "terminal balloon" attached to the skin, which normally regresses, but can remain because of late arrest of the secondary neurulation before the degenerative phase.[12,20,21,26,27] Some TMC cases may show early rapid enlargement, causing the development of neurological symptoms during the 2–6 weeks of waiting for surgery.[9,11,20] Kim *et al*. explained the deterioration by aggravated traction of the spinal cord tethered to the extraspinal cyst wall as the cyst enlarges, based on IONM observation of a case with lipomyelomeningocele, and recommended urgent surgery when the extraspinal cyst enlarges.[8] At our institutes, untethering surgeries for TMC cases with OEIS have been performed immediately after recuperation from abdominogenital repair, and the postoperative course has been uneventful.[15] In the present case, we performed surgery for untethering and duraplasty in the same manner as before, except that the time of neurosurgery was delayed to 10 months after birth. Therefore, the deferment of neurosurgery may be the principal factor for postoperative complication. There might have been a chance to perform neurological surgery for TMC at 7 months after birth when primary epithelialization of the omphalocele sac was achieved, but the benefit had to be weighed against the risk of omphalocele rupture and respiratory compromise, which can cause death in patients with omphalocele.[18] Another reason for the delay of the neurological surgery was that the patient already had severe neurological impairment after birth in the lower extremities, urination, and defecation and did not clinically worsen during the 10 months.

The extraspinal CSF-filled portion of TMC consists of the terminal cystic cavity of the spinal cord (myelocystocele) and the surrounding subarachnoid space. Hydrodynamic pressure and pulsation effect are assumed to cause enlargement of the extraspinal CSF-filled portion of closed spinal dysraphism.[9,10,23] In the present case, excessive intra-abdominal pressure due to the visceroabdominal volume disproportion could increase the hydrodynamic pressure, resulting in further enlargement of the extraspinal subarachnoid space and the caudal part of the dural sac than that of the myelocystocele during the waiting period. A larger inner surface area of the dural sac is likely to be influenced more by the hydrodynamic pressure and pulsation effect. According to the Laplace's law employed by Early and Fink to explain ventricular dilatation,[2] an increasing radius of the dural sac can raise the tensile strength of the sac itself. Therefore, the expanded dural sac itself might have impeded the healing of the duraplasty, even after the untethering surgery. Based on this hypothesis, we reduced the size of the caudal part of the dural sac at the second repair surgery by severing the electrophysiologically functional placode with nerve roots. This was justified by pre-existing severe neurological state of the patient. This procedure had no immediate effects, but it might have contributed to the healing of the duraplasty because no apparent fistula in the dural sac was observed at the third repair surgery. A few TMC cases with or without OEIS complex require a VP shunt for progressive hydrocephalus after untethering surgery,[1,4,12,20] which might indicate that TMC itself or its repair surgery is relevant to the dynamics CSF. External CSF drainage in the present case also had no immediate effect, but it might have helped in the last stage of the healing of the duraplasty by reducing the CSF pressure.

Corrective approaches for CSF leakage after untethering surgery include wound repair, fistula closure, and cystoperitoneal shunt.[25,28] A fascial patch graft is used to fill the gap in case of a dural or fascial defect. The role of the overlying muscle fascia in compressing the extraspinal sac of lipomyelomeningocele has been reported.[8] In the present case, the paraspinous muscles were primarily located away from the midline due to the spina bifida at the surgical site, which was further displaced laterally by the enlargement of the TMC extruding into the extraspinal region during the waiting period. Therefore, the lack of paraspinous muscles at the surgical site to compress the extraspinal dural sac was another factor that impeded the solution. Although gluteal muscles can be a strong vascularized cover over the dural repair,[22] we hesitated to use them because it could have worsened the motor deficit of the patient's lower extremities. Collagen matrix (DuraGen) has been reported to be an effective tool for dural repair;[17] however, we were concerned about infections. Eventually, both worked effectively without complications. In hindsight, reduction of the size of the caudal dural sac, and use of the gluteus muscle flaps and the collagen matrix might have been performed in combination in the early phase of the CSF leakage in the present case.

CONCLUSION

The present case demonstrates that early untethering surgery for TMC is recommended after recovery from the life-threatening conditions of OEIS complex, because the enlargement of the extraspinal subarachnoid space during the waiting period may cause postoperative CSF leakage as well as neurological deterioration.

Ethics statement

The authors confirm that written informed consent was obtained from the family of the infant described in this report.

The authors declare that this work complies with the guidelines for human studies, and the research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

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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