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Multiple-site neural tube defects complicated by multiple-site split cord malformations and thickened filum terminale: experience at a pediatric neurosurgical teaching hospital in Ethiopia. Illustrative case

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BACKGROUND Multiple-site open neural tube defects (MNTDs) and multiple-site split cord malformations (MSCMs) are extremely rare congenital anomalies that are defined by the simultaneous noncontiguous occurrence of more than one neural tube defect (NTD) and split cord malformation (SCM), respectively, in a single case with normal neural tissue in between. This work shows the cooccurrence of MNTDs and MSCMs, which has never been reported in the literature.

OBSERVATIONS A single-stage repair for a 13-day-old female neonate with a preoperative diagnosis of MNTDs (thoracic meningocele and thoracolumbar myelomeningocele) plus an additional intraoperative diagnosis of MSCMs (type 3c) of thoracic and thoracolumbar spine, and thickened filum terminale was done with a favorable smooth postoperative course.

LESSONS The use of intraoperative meticulous surgical technique along with preoperative skin stigmata helped for anticipation, detection, and treatment of associated complex spinal MNTDs, especially in resource-limited settings, where preoperative magnetic resonance imaging is not routinely used. Whether to repair the MNTDs as a single- versus multiple-stage procedure is mainly a function of the patient's tolerance to the duration of anesthesia and the anticipated blood loss for the patient's age. The overall developmental biology and long-term clinical outcome of MNTDs compared to single NTD/SCM is poorly understood and needs further study.

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KEYWORDS meningocele; myelomeningocele; multiple-site neural tube defect; multiple-site (complex) split cord malformation; tethered cord syndrome; thickened filum terminale

Neural tube defects (NTDs) are a common group of central nervous system anomalies that occur from failure of normal neural tube closure due to multifactorial perturbations during the third and fourth weeks of pregnancy.[1,2] The burden of NTDs in developing countries has been reported to be two times higher than in developed countries.[3] Accordingly, NTDs are of major public health importance as estimates show to affect about 300,000 newborns worldwide,[4] resulting in about 88,000 deaths per year.[4]

Multiple-site neural tube defects (MNTDs) are extremely rare congenital anomalies that are defined by the simultaneous occurrence of more than one NTD in a single case with normal neural tissue in between, generally representing only $<$1% of the NTD spectrum.[5,6]

Less than 8% of meningomyeloceles are associated with split cord malformation (SCM) type 1.[7,8] Split cord formations, although a relatively common association with meningomyelocele, rarely occur with MNTDs.[6]

Errors during neurulation may lead to various congenital malformations such as myelomeningocele, meningocele, lipomyelomeningocele, SCMs, the dermal sinus, and intraspinal tumors such as dermoids and epidermoids.[9] Abnormalities that develop during canalization of the tail

ABBREVIATIONS CSF = cerebrospinal fluid; MNTD = multiple-site neural tube defect; MRI = magnetic resonance imaging; MSCM = multiple-site split cord malformation; NTD = neural tube defect; SCM = split cord malformation.

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bud can give rise to the thick terminal filum, terminal myelocystocele, and lipomyelomeningocele.[9] Although the occurrence of multiple split cords itself uncommon, the presence of multiple-site split cord malformation (MSCM) with other open single NTDs like meningocele and myelomeningocele is rare.[9–13] Similarly, to the authors' best knowledge, there is no published evidence on the co-occurrence of MNTDs with MSCM in a single patient. Hence, we present an exceptionally unique case of MNTDs with MSCM and thickened filum terminale. This work shows the challenges encountered in the management of an exceptionally unique case of MNTDs with MSCM and thickened filum terminale. This case also tries to show the uncommon association of primary and secondary neurulation defects.

Illustrative Case

A 13-day-old female neonate born from a para 1 mother by spontaneous vaginal delivery at 38 + 5 weeks, was brought in by her mother with a compliant of thoracic and lumbar area swelling since birth. In addition the baby was unable to move her lower extremities and had poor urinary stream during urination. She did not have discharge from the sites of swelling and no fever. The baby suckled well with no vomiting. The mother had antenatal care follow-up and was reported to have had an uneventful pregnancy.

The baby's general condition was stable, and the vital signs were normal. The head circumference was 33 cm, and the fontanels were flat. On the musculoskeletal system, there were two open neural defects, with a smaller thoracic unruptured meningocele measuring 3 × 4 × 3 cm covered by a dysplastic overlying skin, and a larger unruptured thoracolumbar myelomeningocele measuring 8 × 7 × 6 cm. There was an asymmetrical gluteal cleft and two obvious dimples above the gluteal cleft (Fig. 1). There was no dermal sinus, tuft of hair, or club foot. Neurologically, she was alert but could not move all the key muscle groups of her lower extremities.

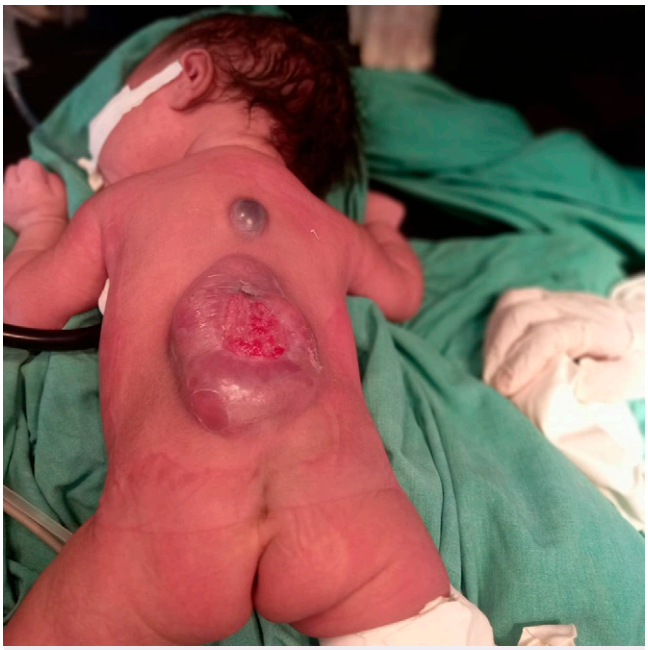

FIG. 1. A clearly visible unruptured thoracic meningocele, thoracolumbar myelomeningocele, and midline supragluteal cleft dimples and asymmetrical gluteal cleft.

With the patient's clinical profile considered, she was preoperatively diagnosed to have unruptured MNTDs of thoracic meningocele and thoraco-lumbar myelomeningocele. Routine basic laboratory investigations and abdominal and transfontanel ultrasound were normal. Imaging of the spine was not performed because of affordability issues, and given the anticipated difficult skin closure, the plastic surgery team was anticipated to be involved in the management of the case. In addition, blood preparation for anticipated blood loss for her small age and weight (4.2 kg) was done. The possibility of the presence of complex spinal defects was also anticipated as the patient had multiple supragluteal dimples.

The patient was taken to the operating room, where she was positioned supine and intubated under general anesthesia. Patient was then repositioned prone and surgical site cleaned with chlorhexidine first and local anesthesia administered. The surgical site again was thoroughly washed with chlorhexidine, alcohol and povidone iodine, and proper prepping and draping done.

A midline skin incision at the junction of dysplastic and normal skin was made first for the thoracolumbar myelomeningocele. Then the dysplastic skin was circumferentially dissected and divided under direct vision. The dysplastic skin was also trimmed sharply from the dysplastic neural tissue and release of the arachnoids was done with sharp dissection (Fig. 2). In doing so, we intraoperatively detected two split cords each separated by a thick bone spur and separate dura (type 1 SCM). The caudal-most part of this split cord merges with the dysplastic neural tissue to become one dysplastic cord while the cranial-most part of it has continuation with a short segment one normal looking spinal cord (Fig. 2). Just caudal to the dysplastic myelomeningocele, there was also thick and short filum that was cut out. The cut-out portion of filum terminale is shown in Fig. 2. Then for this type 1 SCM, the bone spur was sharply

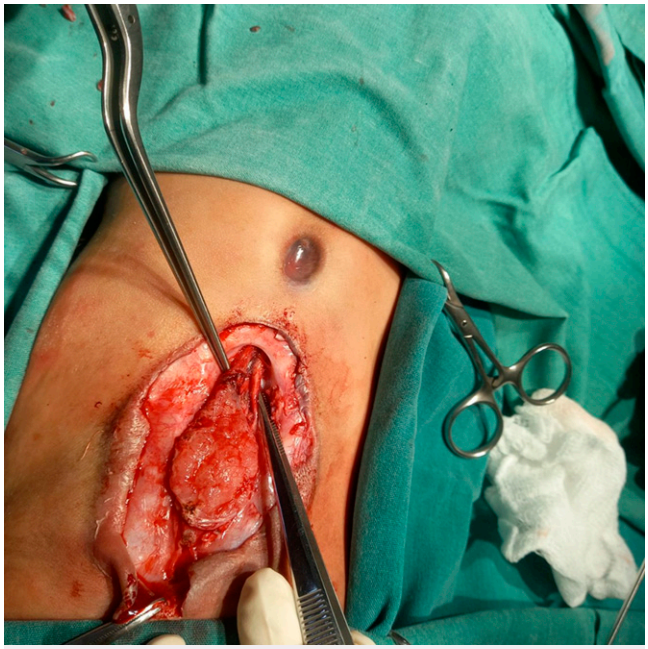

FIG. 2. The placode after the dysplastic skin was circumferentially removed and the arachnoid release was done. Note the first site of the SCM. The cut-out portion of the thickened filum terminale can be seen distally.

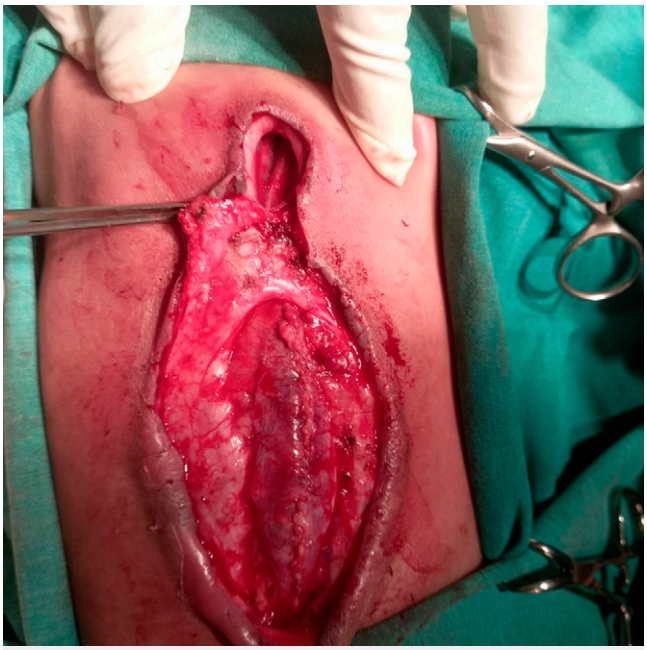

FIG. 3. A communicating skin incision was made between the thoracic meningocele with type 2 SCM and the thoracolumbar myelomeningocele with type 1 SCM to facilitate tension-free primary closure of the skin. Note the water-tight dural closure after the thoracolumbar myelomeningocele placode was tubularized and type 1 SCM bone spur excised.

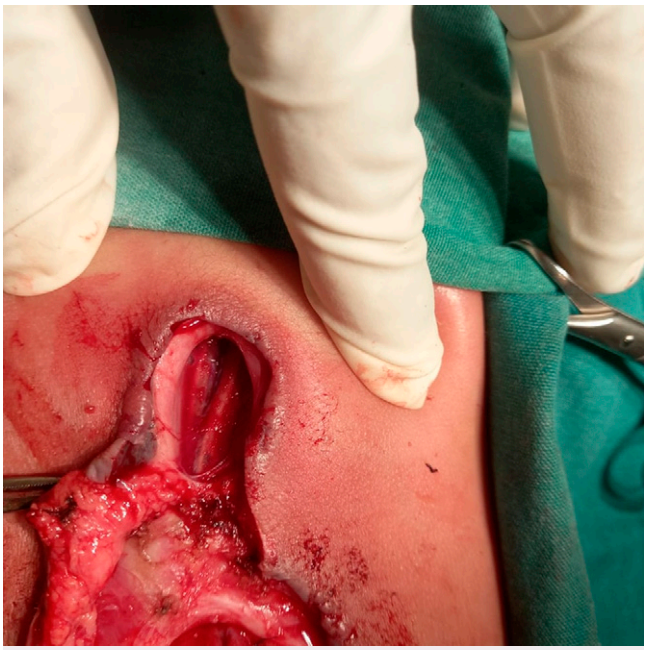

FIG. 4. Type 2 SCM after the dysplastic meningocele sac was circumferentially excised with the caudal part of the split cords merge together to become one normal-looking cord while its rostral split cord extension is uncertain.

dissected from the dura and bone spur was removed. The dysplastic myelomeningocele was tubularized with 5.0 Prolene. Dura was then circumferentially harvested and closed with 5.0 Prolene in a water-tight fashion. Valsalva was checked and no leak identified (Fig. 3).

Once watertight dural closure was achieved for the thoracolumbar myelomeningocele, a midline skin incision and a circumferential sharp dissection and excision of the dysplastic skin was done for the thoracic meningocele to repair in a single stage since the patient's hemodynamic status was stable. Arachnoid releases were also done. Once we made an arachnoid release, there were two split cords separated by thin fibrous septa (type 2 SCM; Fig. 4). The caudal-most part of it was merged and became one normal looking cord, while its rostral split extension was not possible to ascertain intraoperatively. For this thoracic meningocele, dura was harvested and closed in water-tight fashion.

Finally, an intraoperative diagnosis of MNTDs (thoracic meningocele and thoracolumbar myelomeningocele) plus MSCMs (type 3c, multilevel SCM with mixed fibrous and bony spurs or associated SCM anomalies of thoracic and thoracolumbar spine[14]) and thickened filum terminale was made. Once the dura was closed, skin undermining and communicating the two separate defects were done for tension-free subdermal and skin closure. The skin was approximated tension-free primarily after undermining of the skin and communicating the two defects. Subdermis closed with Vicryl 2.0 in an interrupted fashion. Skin was also sutured with Prolene 3.0 in vertical mattress fashion. Only a short relaxing incision to the left lumbar area was added to achieve tension-free closure. Meticulous hemostasis was achieved with bipolar cautery and total blood loss was about 30 mL. The patient was then transferred to the post-anesthesia unit with stable conditions.

Postoperatively, the patient was followed with neuro-sign chart, serial head circumference, wound status, and presence of cerebrospinal fluid (CSF) leakage. The patient did not require a blood transfusion because her postoperative hemoglobin was in the normal range. The patient also had an uneventful immediate postoperative course and was discharged on the fifth postoperative day (Fig. 5). The patient did not develop postmeningocele and myelomeningocele repair hydrocephalus in the immediate postoperative period and the first 4 months after surgery. However, long-term follow-up is required.

Discussion

Observations

To date, there exist 57 cases of MNTDs reported in the scientific literature, with its occurrence being extremely rare.[10] Similarly, MSCM is very rare and accounts for only 1% of all cases of SCM.[11] rarity of multiple noncontiguous SCM is evidenced by the presence of only 8 cases reported in the scientific community as of 2014. Furthermore, there is only 1 report stating the co-occurrence of MSCM with single open NTDs (e.g., myelomeningocele, meningocele). To the authors' best knowledge, our case is the first reported case that has both multiple noncontagious NTDs and multiple noncontiguous SCMs in a single patient.

Although the presence of MSCMs and thickened filum terminale were ascertained intraoperatively in our case with the use of meticulous surgical technique, it was also anticipated preoperatively for the possible presence of complex associated spinal defects in addition to the already diagnosed MNTDs (in our case meningocele plus myelomeningocele) from the multiple supra-gluteal dimples.

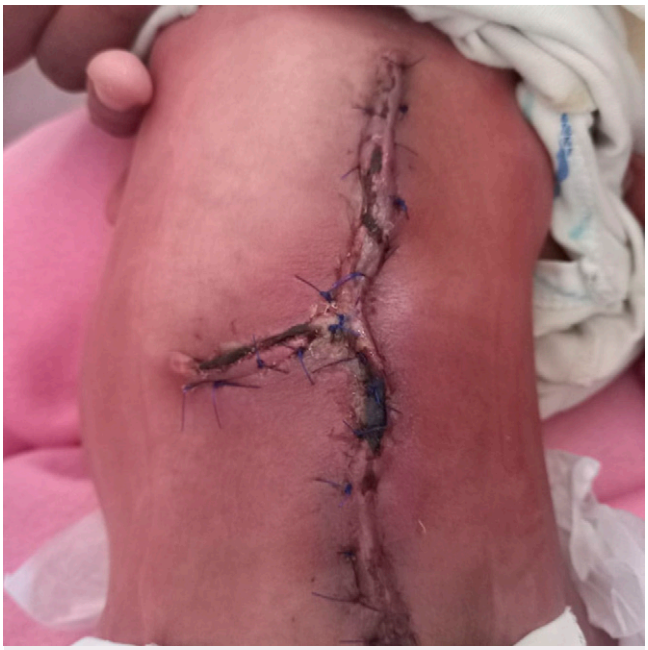

FIG. 5. A well-healing incision at the 5th postoperative day.

Hence, some skin stigmata like dimples can be used for possible presence of associated NTDs of any type, whether single or multiple NTD, in addition to what is evidenced. This is especially helpful for resource-limited settings where routine magnetic resonance imaging (MRI) is not obtained and in the case of repair of ruptured NTDs on an emergency basis where addressing the infection concern takes priority over getting an MRI to screen for associated neuraxis spine dysraphism.

Although the origin of single-site SCM malformation is partly explained by the widely accepted theory that proposes that the origin of all SCMs originate from one basic ontogenic error occurring around the time of closure of the primitive neurenteric canal that leads to formation of an accessory neurenteric canal, which is considered as an abnormal fistula that causes regional splitting of the notochord and the overlying neural plate,[12–14] there is paucity of evidence regarding how and why MSCMs exist. Similarly, the issue of zipper-like closure versus multiple-site closure model for closure of be single- or multiple-site NTDs is still controversial, and a third theory (the rezippering closure model) that makes use of the strength of the two prior theories and address their limitation has been proposed.[5,15,16] The co-occurrence of the noncontiguous open MNTDs and MSCMs in a single patient adds complexity to the already intriguing embryogenesis of each; hence this paves the way for future further study.

A patient who has multiple-site defects can be repaired as a single one-time or multistaged procedure with an uneventful postoperative course. Whether to repair the MNTDs as a single- versus multiple-staged procedure is mainly a function of the patient's tolerance of the duration of anesthesia and the anticipated blood loss for the patient's age. Accordingly, single-stage repair of defects is safe and not associated with increased postoperative complications. The repair for our case was done as a single-staged procedure with an uneventful postoperative course, which is consistent with other case reports.[10,17] However, the long-term outcome and associated complications (the risk of postdefect-repair hydrocephalus and the need of CSF diversion, the risk of spinal cord tethering, etc.) for patients with MNTD and MSCM compared to single-site NTD and SCM is not known well due to paucity of evidence and requires further study. According to a comparative study done among 46 patients with SCM who were grouped into those with SCM alone versus SCM with myelomeningocele to assess the operative outcome, progressive neurological deficit was higher in those with SCM with myelomeningocele in comparison to the group harboring SCM without myelomeningocele. Accordingly, this study underscored the thorough screening of neuraxis (by MRI) to treat all treatable anomalies simultaneously for desired outcome.[18,19]

Lessons

The co-occurrence of MNTDs and MSCMs is an extremely rare event. The use of meticulous intraoperative surgical technique along with preoperative skin stigmata for prediction of associated complex spinal defects is of paramount importance in the anticipation, detection, and treatment of complex MNTDs in particular and NTDs in general, especially in resource-limited settings, where preoperative MRI imaging is not routine due to affordability issues. Whether to repair the MNTDs as a single- versus multiple-staged procedure is mainly a function of the patient's tolerance of the duration of anesthesia and the anticipated blood loss for patient's age. The overall developmental biology and long-term clinical outcome of MNTDs compared to the ordinary single NTD/SCM is poorly understood, which is the main limitation of this study. Hence, further research needs to be conducted in this area to understand and treat it better.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Shiferaw, AT Aklilu. Acquisition of data: Shiferaw, Worku. Drafting the article: Shiferaw, AT Aklilu. Critically revising the article: Shiferaw, YB Akililu, T/Mariam, AT Aklilu. Reviewed submitted version of manuscript: Shiferaw, YB Akililu, T/Mariam, AT Aklilu. Approved the final version of the manuscript on behalf of all authors: Shiferaw. Administrative/technical/material support: Shiferaw. Study supervision: Shiferaw, Worku, AT Aklilu.

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