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Technical Notes

Terminal myelocystocele: Surgical management

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ABSTRACT

Background: The authors describe clinical and imaging findings, surgical technique, and outcomes in myelocystocele.

Methods: We describe a surgical procedure performed in six patients, four males and two females, with myelocystocele treated at our hospital. We review the images obtained at the time of diagnosis and after surgery. The patients' age range was 12–56 months and had undergone surgery for terminal myelocystocele between 2015 and 2020. All patients had a large lumbar mass covered with healthy skin and presented spontaneous movements at birth. Two patients presented VACTERL syndrome.

Results: A watertight closure of the soft tissues was performed in all cases. None of the patients presented postsurgical complications, such as cerebrospinal fluid leak or infection. All the patients had undergone excision of the meningocele sacs, the tethering bands were lysed, and the filum was detethered. The mean follow-up period was 34 (12–56) months. A motor deficit was seen in 2 patients (33.3%).

Conclusion: Prenatal diagnosis and early corrective surgical intervention are recommended to prevent deterioration in neurological function. VACTERL association is a common condition and should be investigated.

Keywords: Neural tube defect, Spina bifida, Surgical technique, Terminal myelocystocele

INTRODUCTION

Myelocystocele is a rare form of occult spinal dysraphism, which consists of a cystic dilatation of the central canal of the distal spinal cord, herniated, and reflected through the posterior sac.[12,23] Terminal myelocystocele (TMC) is an anomaly of the caudal cell mass at second neurulation and can be associated with anomalies of the anorectal system, lower genitourinary system, and vertebrae, such as anal atresia, cloacal exstrophy, lordosis, scoliosis, and partial sacral agenesis.[3,8,13,21,23] It should be noted that the TMC is a part of a large group of skin covered malformations such as spinal cord lipomas or meningoceles, which must be clearly differentiated clinically and through imaging methods to establish the appropriate surgical technique. Clinical presentation consists of a large lumbosacral mass with skin cover, most of it lateralized to one side, containing fat, cerebrospinal fluid (CSF), and neural tissue.[12,23]

MATERIALS AND METHODS

We describe the surgical procedure performed by the senior author (S.P.M.) and review the outcomes in six patients with TMC, who were operated on at (hospital's name) between January

2015 and June 2020. Their data, retrospectively collected, include clinical and radiological features, type of surgery performed, clinical and imaging features, and outcomes. All these patients had undergone presurgical magnetic resonance imaging (MRI) [Figures 1 and 2], evaluation of urinary tract functioning by renal scan or ultrasound, and renal function tests. After surgery, patients were regularly followed up, and their clinical status was assessed.

Surgical procedure

The patients were placed in prone position. In our experience, we use an "overlapping flap" technique. After a midline vertical skin incision or a lateral semicircular incision surrounding the lumbar mass, the subcutaneous tissue was separated from the dural plane preserving the sac intact. Once the site through which the sac continued with the normal dural canal was identified, the paravertebral muscle cephalic to the sac entry was incised and dissected, and at least two sets of normal laminae were exposed and removed.

The turgidity of an intact sac had proven helpful during the initial exposure of the myelocystocele. The dura was then opened starting rostrally to the sac in the midline following the normal spinal cord separating the arachnoid adherences, sparing dorsal nerve roots, until reaching the place where distal cord is reflected in the back of the sac. Most often, the content in the cord that corresponds to the central canal was drained. In some cases, the spinal cord was rotated to one side, and when the nerve roots of each side were compared, a difference in development was seen sometimes.

Thin fibrotic bands were divided, extra tissue was excised, and conus medullaris reconstruction was done with interrupted 5-0 absorbable pial sutures. If filum terminale was identified, it was cut to detether the cord. The dura is sutured with overlapping flaps so that the terminal cord is not in contiguity with the suture of the dura, avoiding posterior tethering. The same idea is applied to the muscle flaps, with which we use barbed sutures to reduce the tension of the paraspinal muscles, and finally, the excess skin resulting from the sac that no longer exists can be resected. The vascularity and mobility of the flap are, therefore, ideal in providing a soft-tissue interposition between the skin flaps and reconstructed thecal sac or even incorporating this into the dural repair

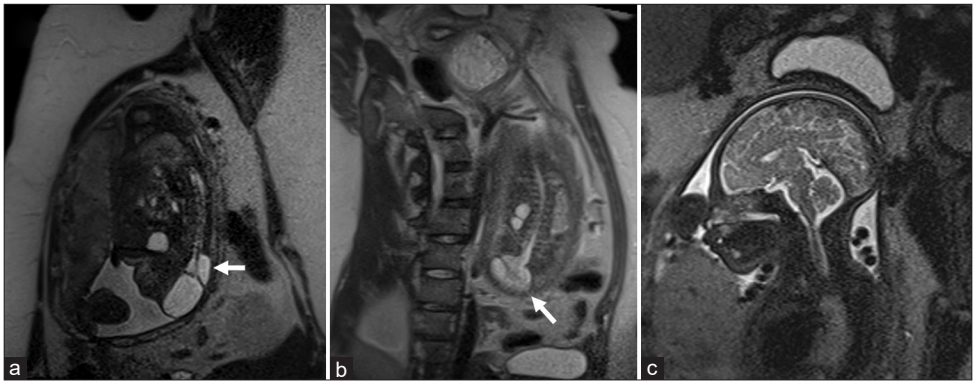

Figure 1: Prenatal MRI. Prenatal MRI of a fetus of 25 week of gestation. (a and b) Sagittal view. An elongated spinal cord can be seen extending dorsally out of the spinal canal, finishing reflected, and tethered in the dorsal part of dural sac, where it expands into a fluid-filled, thin-walled, trumpet-shaped cavity (trumpet-like sign, *white arrows*). (c) Sagittal view of head MRI without hydrocephalus or Chiari malformation.

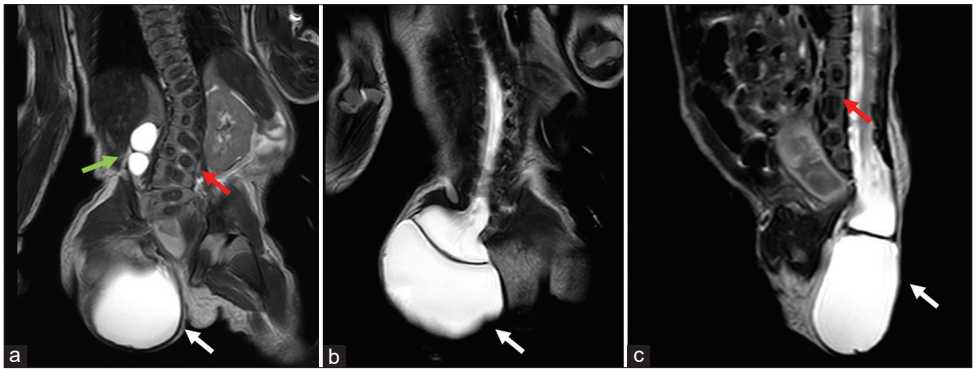

Figure 2: Postnatal MRI. (a-c) Dural sac filled of CSF (*white arrows*), multiple vertebrae (*red arrows*), and renal malformations associated (*green arrows*).

if insufficient dura is available, thus avoiding the need for xenografts or synthetic dural substitutes [Figures 3 and 4].

In the immediate postoperative period, all patients were placed in prone position, and the leg end of the bed was elevated during the first 3–4 days of postoperative period. The patient received intravenous antibiotics for 2 days. The most common complication described is pseudomeningocele formation and CSF leak,[23] which improved with conservative therapy. There are some factors which contribute to the wound-related morbidity, namely, large redundancy of skin, a large defect in the lumbosacral fascia, and an attenuated dural layer limiting the possibility of dural reconstruction and locally altered CSF dynamic.[19] However, if CSF leakage persists, we prefer to place spinal drainage cephalic to the area of surgery and, therefore, the area of leakage, during 5–7 days instead of a second look exploration surgery. In our series, there were no immediate or long-term postoperative complications.

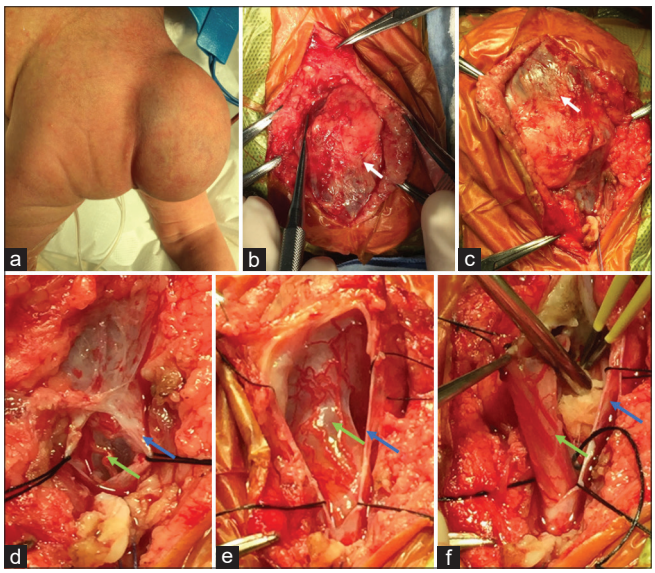

Figure 3: Intraoperative images. (a) Newborn back with a left-sided lumbar mass filled out of fluid. (b and c) Surgical dissection of lumbar mass. Exposition of sac of myelocystocele (*white arrow*). (d-f) Resection of nonfunctional neural tissue, sac closure, and nerve root preservation. After that, watertight closure is performed. *Dural sac: blue arrow, neural tissue: green arrow.*

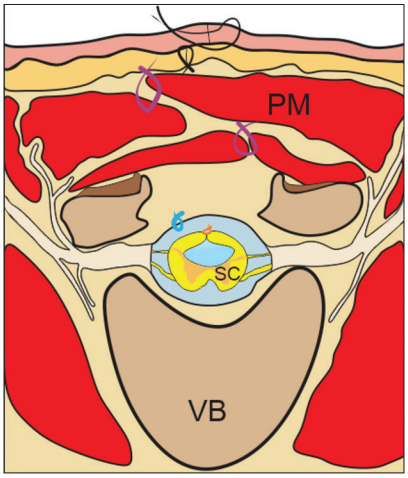

Figure 4: Draw of axial view at lumbar level in an open spinal tube. Axial view at lumbar level with open spinal tube, after performing an overlapped flap closure technique. Vertebral body (VB), paravertebral muscle masses (PM), and closed spinal cord (SC), orange suture: closure of the spinal cord, light blue suture: dural closure, purple suture: muscle closure, and black suture: dermal and epidermal closure.

RESULTS

All the patients were diagnosed prenatally at 12–23 weeks of pregnancy. There were 4 (66.6%) boys and 2 (33.3%) girls. All were born at full term by cesarean section. Patients carachteristics are described in Table 1.

Five patients underwent surgery early after birth, one on the 5th day after birth. We made a complete preoperative multidisciplinary evaluation, due to because it is a skin covered defect, there is no urgency to surgery. In all our cases, the presumptive diagnosis was made during routine obstetric ultrasonography looking for malformations and confirmed with prenatal MRI. All preoperative MRIs show some constant ("obligatory") features[6,18] present in all myelocystoceles, as well as other "optional" findings[18] of variable prevalence. The most important among the obligatory features of TMC is an elongated cord that extended dorsally out of the spinal canal, finishing reflected, and tethered in the dorsal part of the dural sac, where it expanded into a fluid-filled, thin-walled, trumpet-shaped cavity (trumpet-like sign[12,23]) and the "myelocystocele"[6,18] [Figures 1 and 2]. The flared part of the trumpet resembled a three-dimensional cone, with the apex toward the side of the spinal canal and the wide mouth base flatly adherent to the subcutaneous adipose layer of the overlying skin.[18] There was often a thin but visible layer of tissue, continuous with the inner lining of the trumpet, that separated the fluid-filled cavity from the surrounding fat. The most of the flared part of the trumpet is a nonfunctional remnant neural material distal to the conus.[18] Among the optional features of myelocystocele is a continuity of the extraspinal cystic cavity with a variable size of hydromelia within the intraspinal portion of the caudal spinal cord.[6,18] No one showed brain malformations, such as tonsillar herniation or hydrocephalus in MRI prenatal scan. Furthermore, due to characteristics of lumbosacral skin covered malformation, there is no option to consider prenatal correction.

In all patients, we found posterior lumbosacral dimorphism, cord descent at the level of the dysraphism with the presence of myelocystocele, and associate arachnoid sac. One of the

Table 1: The patient characteristics.

Sex	Age at diagnosis	Age at surgery	Clinical features at birth	Associated anomalies	Follow- up	Outcome
M	Prenatal	1 day	Normal Spontaneous movements Skin-covered sac	Ectopic testicle Chiari II	35 months	Neurogenic bladder Walk with assistance
M	Prenatal	1 day	Normal Spontaneous movements Skin-covered sac	Anal Atresia Renal Anomalies Ectopic Testicle (VACTERL Syndrome)	56 months	Neurogenic bladder Walk with assistance
M	Prenatal	1 day	Normal Spontaneous movements Skin-covered sac	None	12 months	Walk without assistance Normal bladder function
F	Prenatal	5 days	Normal Spontaneous movements	Anal Atresia Rectum- vaginal fistula Kidney cyst (VACTERL syndrome)	50 months	Neurogenic bladder Walk without assistance Syringoperitoneal shunt Scoliosis surgery
M	Prenatal	1 day	Normal. Spontaneous movements Skin-covered sac	None	12 months	Walk without assistance Normal bladder function
F	Prenatal	1 day	Normal. Spontaneous movements Skin-covered sac	None	28 months	Neurogenic bladder Walk without assistance

patients presented significant development of subcutaneous fat that adhered to the sac, and all the other patients had prenatal syringomyelia. Four patients had bladder dysfunction. Association with VACTERL syndrome[20] was seen in two patients in our series (33%), both had an antenatal diagnosis. One patient had anal atresia and needed a colostomy and then surgical anatomical reconstruction with anoplasty. Furthermore, he had an ectopic testicle that required surgical treatment and had only one kidney. Another patient also had anal atresia and rectum vaginal fistula and needed a colostomy and surgical reconstruction; renal cyst and lumbar hemivertebrae with congenital scoliosis also needed surgical reparation. A motor deficit was seen in two patients. All the patients had a large lumbar cyst covered by normal skin.

The mean follow-up period was 34 (12–56) months. None of the patients presented postsurgical complications, such as CSF leak or infection.

DISCUSSION

Myelocystocele is a rare congenital malformation,[12] with an incidence of 4–8% of lumbosacral occult spinal dysraphisms.[23] The sacrum and caudal portion of the conus medullaris, filum terminal, lower lumbar, and sacral nerve roots develop from a large aggregate of undifferentiated cells that are remnants of the primitive streak. The embryological processes underlying the development of the caudal cell mass include canalization (secondary neurulation) and retrogressive differentiation.[7,8,12,14,23] In this dysraphism, the ventriculus terminalis becomes dilated and enlarged by the CSF, resulting in rupture of the dorsal mesenchyme.[12] The posterior elements cannot be formed, and as the ventriculus terminalis expands, a meningocele sac is formed.[12] Finally, the reflected distal cord is tethered to the sac preventing its ascent.[12] However, its various denominations in the literature, such as lipomyelocele, lipomyelocystocele, lipomeningocele, lipoma, meningocele, and hydromyelocele, suggest an alternative origin in primary neurulation.[8,18]

All our patients had a skin-covered cystic mass, which is typically lumbosacral, obliterates the intergluteal cleft, and extends upward from the perineum for a variable distance. In the patients with neurological deficit at birth, the mass was slightly lateralized to the side of the leg with the worse motor deficit. It is important to know that the intergluteal fold is obliterated and distorted in myelocystocele but preserved in lipomyelomeningocele.[12,23]

MRI is the imaging study of choice for patients with neural tube malformations. In this case, it can demonstrate direct continuity of the meningocele with the subarachnoid space.[6,12,20,23] It is imperative to perform an MRI of the whole spine, including the craniovertebral junction, and to examine the brain to rule out other congenital anomalies.[6,19,23] Furthermore, we consider that a postnatal MRI is necessary to assess more accurately the anatomy of the defect. Differential diagnosis includes sacrococcygeal teratomas and overgrowing fatty tissue in caudal regression, whose location is more caudal, that is, at or below the intergluteal crease.[24]

Caudal dysgenesis is the term applied to the partial or complete absence of the sacrum, usually including the spinal cord, anorectal, and urinary malformations. The spectrum of syndromes and associations with caudal dysgenesis includes Currarino syndrome;[15] caudal regression syndrome;[1] sirenomelia;[22] the OEIS complex;[11] and VACTERL[21] associations.[7]

The incidence of VACTERL syndrome is estimated at approximately one in 10,000–40,000 live-born infants,[21] which is typically defined by the presence of at least three of the following congenital malformations: vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities.[3,13,21] Spinal dysraphism is reported in 10–40% of VACTERL cases, but they are not a part of the definition.[3] Antenatal diagnosis can be challenging, as some of the component features can be difficult to ascertain before birth.[21] The management of patients with VACTERL association typically focus on the surgical correction of the specific congenital anomalies in the immediate postnatal period. If optimal surgical correction is achievable, the prognosis can be relatively positive[21] and (in our cases) determined by neural tube defect, because they may be responsible for motor weakness and/or sphincter disorders.

The main goal of the surgery is the release of the tethered spinal cord. The watertight closure of the soft tissues above the defect has always been a surgical challenge. Several surgical techniques have been proposed for soft-tissue closure of various spinal defects, including primary closure, split-thickness skin grafts, local skin flaps, uni- or bilateral muscle flaps, and composite flaps such as fasciocutaneous or myocutaneous flaps.[4,5,10,16,17,19,25] All techniques aim at providing durable, tension-free, and watertight closure with well-vascularized tissue to reduce the complications rate.

The surgical procedure can be done using intraoperative electrophysiological monitoring,[2,9] which is aimed at distinguishing between functioning nerve roots, spinal cord tissue, or nonfunctional tethering structures that are fixing the cord. Therefore, the lower limb evoked potentials, electromyograms, and bipolar nerve root stimulation has been used. However, the neurological deficit that the patients present at birth and the preoperative electrophysiological studies used to decide the timing and characteristics of the surgical procedure may not correlate with what happens during surgery. Because of this, it is very important to have a proper knowledge of the embryology and anatomy of these congenital spinal defects.

CONCLUSION

TMC is a congenital defect mostly dependent on secondary neurulation. The diagnosis can be made in the prenatal stage, and it is necessary to confirm it after birth. Therefore, defects such as anal atresia, bladder exstrophy, sacral agenesis, or defects in the perineal floor may also be present like VACTERL. These malformations may require surgery before the myelocystocele. The characteristics of this malformation at the level of the distal spinal cord are complex, for which the surgical treatment requires an appropriate embryological knowledge that can differentiate it from other malformations such as spinal cord lipomas or simple sacral meningoceles. CSF leakage, the most frequent complication in these surgeries, can be avoided with an adequate closure of the soft tissues. It is also important to keep in mind that these are skin-covered lesions; therefore, there is no urgency to repair them when they are in association with other major anomalies such as cloacal exstrophy, omphalocele, and imperforate anus. Instead, initial care needs to be directed toward these anomalies rather than the myelocystocele.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

The authors declare that they have no conflicts of interest. Artwork was created by Dr. Montivero using CorelDraw®.

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