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TERMINAL MYELOCYSTOCELE: A RARE VARIANT OF SPINAL DYSRAPHISM. CASE SERIES AND REVIEW OF THE LITERATURE

Terminal Myelocystocele: A Rare Variant of Spinal Dysraphism. Case Series and Review of the Literature

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Abstract

Background Data: A terminal myelocystocele occurs at the distal (terminal) end of the spine from the lumbar to coccygeal regions. It is characterized by a skin-covered mass with spinal dysraphism of the associated spinal cord. Within this herniated sac is the distal spinal cord with a large cystic dilatation of its central canal that occupies the posterior aspect and CSF and arachnoid membrane forming a meningocele lying anterior to the terminal cyst of the central canal.

Purpose: To report our clinical results with repair and untethering of five cases with terminal myelocystoceles.

Study Design: A descriptive cross section retrospective study.

Patients and Methods: This is a retrospective analysis of five cases of terminal myelocystoceles treated between February 2008 and March 2015. All patients underwent neurological examination and magnetic resonance imaging (MRI) of the spine. One patient (older one) had presented with weakness of feet dorsiflexion and plantar flexion bilaterally while 4 (80 %) patients had no deficits at all. Bladder bowel involvement was evident in all cases.

Results: A total of five patients were included. The mean age of intervention was 5 months. Three of five patients were females. All the patients had lower lumbar and upper sacral mass. The mean operative time of the study group was 150 minutes and the mean operative blood loss was 70CC. The mean hospital stay was 8.4 days. The status of all patients with no pre-operative deficits remained unchanged. The patient with motor weakness had improvement during follow up.

Conclusion: Surgical correction of terminal myelocystocele is not only for cosmetic reasons but also to untether the spinal cord to prevent future neurological deterioration. During surgical corrections, it is important to perform a thorough intradural inspection to transect all adhesions. (2015ESJ102)
Keywords: Terminal myelocystocele, Spinal dysraphism, Occult Spinal Dysraphism

Introduction

A terminal myelocystocele (TMC) occurs at the distal (terminal) end of the spine from the lumbar to coccygeal regions.[4,8] It constitutes approximately 5% of skin covered lumbosacral masses.[8] They are defined as spinal dysraphism with a localized, cystic dilatation of the central canal of the spinal cord herniated posteriorly through spina bifida. Terminal myelocystocele can be associated with anomalies of the anorectal system, lower genitourinary system, and vertebrae.[5] A terminal myelocystocele is characterized by a skin-covered mass with spinal dysraphism of the associated spinal cord. The back mass is a sac that contains intraspinal contents herniated through this area of spinal dysraphia. Within this herniated sac is the distal spinal cord with a large cystic dilatation of its central canal that occupies the posterior aspect and cerebrospinal fluid (CSF) and arachnoid membrane forming a meningocele lying anterior to the terminal cyst of the central canal. The sac is covered by dura mater, a large amount of fat that is continuous with the subcutaneous fat of the back and skin.[4,8,10,12]

Because a terminal myelocystocele is such an unusual and complex congenital malformation, it often is confused with other spinal dysraphic conditions, i.e., lipomyelomeningocele and meningocele or a cystic teratoma.[12]

This study analyzes five cases of terminal myelocystoceles and compares them to the literature to better delineate the entity of terminal myelocystocele and to understand the clinical presentations and the surgical management.

Patients and Methods

This study was designed as descriptive cross-sectional retrospective clinical study. Between February 2008 and March 2015, at Neurosurgical department, Suez Canal University Hospital (Ismailia, Egypt) a total of five consecutive patients was recruited for this study. All patients had a swelling in the lumbosacral region with a healthy skin cover. All patients had preoperative MRI of the dorsolumbosacral spine and the brain. The aim of this study is to document our clinical results with repair and untethering of five cases with terminal myelocystoceles.

Preoperative Assessment:

The preoperative assessment included General and neurological assessment. General assessment includes search for associated congenital anomalies in the infant. Neurological assessment by watching for spontaneous movement of the lower limbs (good spontaneous movement correlates with better later functional outcome") and assessed the lowest level of neurologic function by checking response of lower limbs to painful stimulus. One patient (older one) had presented with weakness of feet dorsiflexion and plantar flexion bilaterally, while 4 (80 %) patients had no motor deficits at all. Bladder bowel involvement was evident in all cases as depicted from the preoperative MRI that showed distended bladder and anal exam.

Surgical Procedure:

We tend to perform the repair of TMC within the first 6 months to minimize neurological losses from spinal cord tethering. The patient is positioned prone. Through a midline incision, the most caudal intact lamina is identified and removed to localize the normal thecal sac (Figure 1). The myelocystocele sac and dura are then circumferentially dissected away from the surrounding soft tissue. This usually requires release of the dorsal fibrous band that connects the most cranial bifid lamina across the midline.

The normal dura can then be visualized as it continues caudally over the meningocele. The dura and arachnoid are opened at the level of the laminectomy, and the incision proceeds caudally over the myelocystocele sac to allow visualization of the dilated spinal cord, terminal lipoma, and associated nerve roots that traverse the meningocele and reenter the spinal canal. Careful dissection through the fat caudal to the meningocele will reveal the terminal cyst, which can then be opened in the midline, caudal to the emerging nerve roots. At this point, the terminal spinal cord is inspected caudally and dorsally for tethering elements, which are released if encountered. The wall of the cyst and associated lipoma can then be resected distal to the caudal arachnoid reflection. The free edges of the remaining sac may be approximated and the residual dura closed primarily to reestablish the thecal sac. The soft tissues are then closed in layers.

Perioperative Data:

In all patients, duration of surgery, blood loss, and the duration of inpatient treatment were recorded. Intraoperative and perioperative major and minor complications were assessed.

Clinical Follow up:

Patients were followed 1, 3, 6, 9 and 12 months after surgery and then every 6 months. During follow up visit the following data were collected: back wound; neurological deficits, orthotic deformity, sphincters control and motor developmental milestones.

Results

Preoperative Data:

A total of five patients with TMC were included in this study. The demographic data are presented in (Table 1). The mean age of intervention was 5 months (1-9 months). Three of the 5 patients were females. All the patients had lower lumbar and upper sacral mass.

Operative Data:

The mean operative time of the study group was 150 minutes and the mean operative blood loss was 70CC. The mean hospital stay was 8.4 days.

Post-operative Clinical Data:

All patients had undergone excision of the meningocele sacs, the tethering bands were resected, and filum was detethered. The mean follow-up period was 36.2 (14-60) months. The status of all patients with no preoperative deficits remained unchanged. The patient with motor weakness had improvement. The patients remained stable throughout the follow up period and no one had got any symptoms or signs of retethering of the spinal cord.

Complications:

Complications like pseudomeningocele were seen in 4 (80%) patients, cerebrospinal fluid leak in 3 (60%) patients, and surgical site dehiscence in 1 (20%) patient.

Table 1. Patient Characteristics and Clinical Findings in the Study Series

Age (mos) /Sex	Neurological Status	Operative Time/min	Hospital Stay/days	Complications	Follow Up /mos	Outcome
1/F	Distended bladder	160	7	Pseudomeningocele	14	Intact
6/M	Distended bladder	120	4	-	60	Intact
9/M	Distended bladder & weak feet dorsiflexion and plantar flexion bilaterally	180	14	Pseudomeningocele CSF leak wound dehiscence	48	Intact
1/F	Distended bladder	160	8	Pseudomeningocele CSF leak	35	Intact
3/F	Distended bladder	130	9	Pseudomeningocele CSF leak	24	Intact

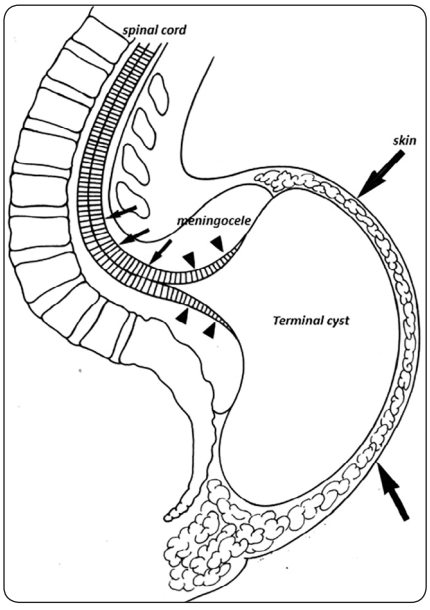

Figure 1. Artistic illustration of the components of a terminal myelocystocele.

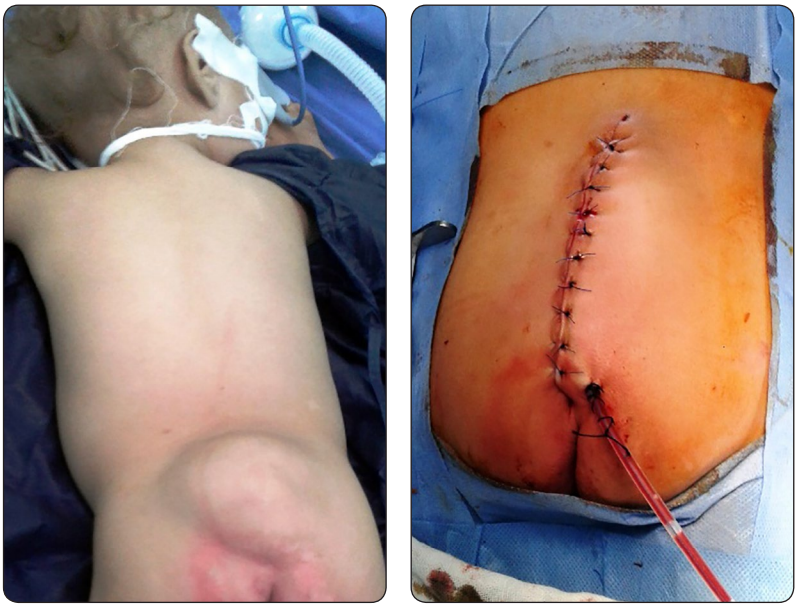

Figure 2. Case N0 1. Lumbosacral mass above the inter-gluteal cleft in the pre and post-operative views

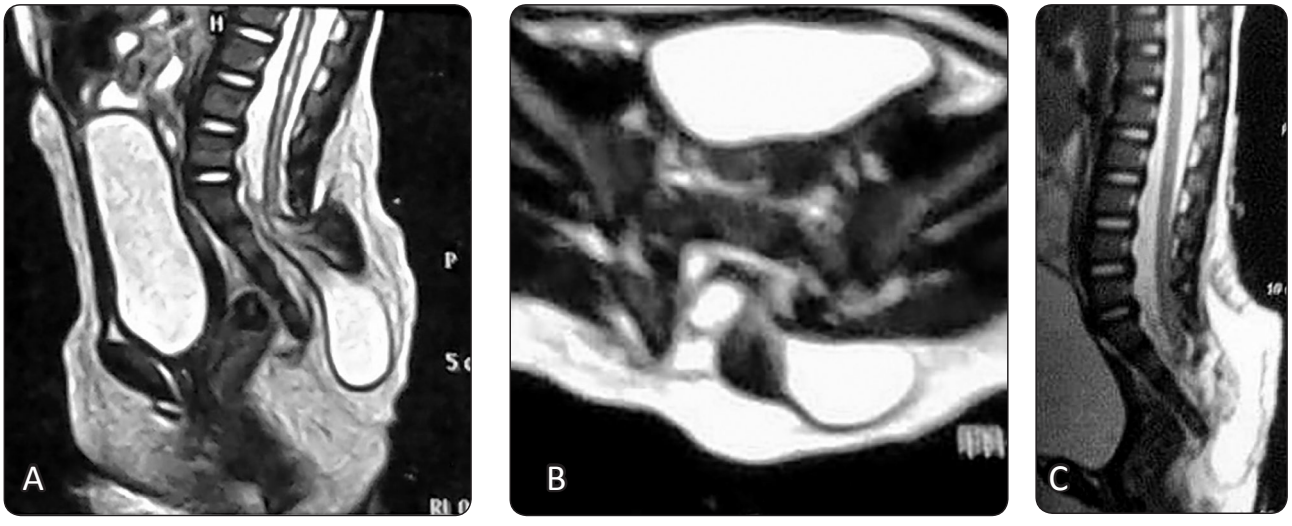

Figure 3. Case No 4. MRI lumbar spine (A) sagittal T2 showing tethered cord, spina bifida, lipoma at the spine defect and meningeal sac consisting with Terminal lipomyelocystocele. (b) axial T2 of the same patient (c) postoperative MRI lumbar spine showing untethering of the spinal cord and pseudomeningocele at the operative bed.

Discussion

Terminal Myelocystoceles are defined as localized cystic dilatation of the central canal of the spinal cord. Terminal Myelocystocele (TMC) constitutes 4-8% of lumbosacral occult spinal dysraphism. Epidemiologically, myelocystoceles arise sporadically; there is no known familial incidence. No true sex preponderance has been described.[7, 9, 10]

The TMC is composed of a low-lying conus medullaris with cystic dilatation of caudal central canal, a surrounding meningocele, and a lipoma that extends from the conus to a

subcutaneous fat collection. The terminal cyst is lined with ependymal and dysplastic glia and does not communicate with the subarachnoid space.[13]

TMC can be deconstructed into essential and nonessential features. Essential features are present in all TMCs and constitute the core malformation, comprising an elongated spinal cord extending extra spinally into a csf-filled cyst that is broadly adherent to the subcutaneous fat. The functional conus resides in the proximal cyst or within the intraspinal cord and the caudal myelocystocele wall is nonfunctional fibro neural tissue. Non-essential features include variable measures of hydromyelia, caudal meningocele, and fat, present in only some patients.[11]

The myelocystocele is due to a defect in secondary neurulation. The caudal neural tube forms via canalization and regression, beginning at day 48. If an insult disrupts retrogressive differentiation, regression fails to occur and an abnormally low conus results.[13] According to Pang et al., the core structure of TMC strikingly resembles a transitory stage of late secondary neurulation in chicks in which the csf-filled bleb-like distal neural tube bulges dorsally to fuse with the surface ectoderm before focal apoptosis detaches it from the surface and undertakes its final dissolution. Author theorizes that TMC results from a time-specific paralysis of apoptosis just before the dehiscence of the cystic distal cord from the future skin, thereby preserving the embryonic state.[11]

The majority of children present with a skin covered mass at the lower back, in the intergluteal cleft and may be of variable size. Midline abdominal and pelvic anomalies often accompany the TMCs. This constellation of abnormalities is best represented by the acronym "OEIS," a complex which is described in the literature as including an omphalocele defect, exstrophy of the bladder, imperforate anus and spinal abnormalities, occurring together in the same patient.[6]

Many newborns with TMC have no neurological deficits but develop them with age. As the spinal cord is tethered in TMC, a slow neurological deterioration is expected with age, but some infants have precipitous loss of leg and bladder functions in early life coinciding with rapid enlargement of the myelocystocele.[11]

A myelocystocele can be diagnosed prenatally with high resolution fetal ultrasonography. MRI is the best noninvasive modality to diagnose all of the components of a TMC and the associated central nervous system findings.[1, 2] MRI appearance is distinctive and is characterized by a "trumpet-like" flaring of the distal cord central canal into an ependyma-lined terminal cyst. Abnormalities of the vertebral column may occur and include lordosis, scoliosis and agenesis of the sacral parts.[8]

TMC should be repaired within the first few months of life or once the diagnosis is made regardless of whether there are preexisting deficits. The aim of the TMC repair is to create an internal milieu least conducive to resticking of the remaining neural tissue, with maximal elimination of any nonfunctional wall through the use of reliable intraoperative electrophysiology.[3, 11]

Conclusion

Surgical correction of myelocystocele is not only for cosmetic reasons but also to untether the spinal cord prophylactically to prevent future neurological deterioration. During surgical corrections, it is important to perform a thorough intradural inspection to transect all adhesions, in order to prevent future deterioration due to the spinal cord re-tethering.

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الملخص العربي

القيلة المائية النخاعية أسفل الظهر: شكل نادر من خلل رفاء العمود الفقري

البيانات الخلفية: تقع القيلة المائية النخاعية أسفل الظهر في نهاية الفقرات القطنية. وتتميز بوجود كتلة مغطاة بالجلد الطبيعي مصحوبة بخلل في رفاء الفقرات مع شد على الحبل الشوكي.

الغرض: توضيح النتائج الجراحية لجراحة إصلاح القيلة المائية النخاعية أسفل الظهر مع تحرير الحبل الشوكي.

تصميم الدراسة: دراسة لحالات أكلينيكية على ٥ مرضى يعانون القيلة المائية النخاعية أسفل الظهر.

الطرق و المرضي: تم إجراء الجراحات من ٢٠٠٨ إلى ٢٠١٥ . تم عمل أشعة رنين مغناطيسي لجميع المرضى عانى مريض واحد فقط من ضعف في الأطراف قبل الجراحة.

النتائج: عانى ٥ مرضى من القيلة المائية النخاعية متوسط العمر ٥ شهور. متوسط زمن الجراحة ١٥٠ دقيقة متوسط فقد الدم ٧٠ سم متوسط فترة النقاهة في المستشفى ٨ أيام و بعد الجراحة أظهر جميع المرضى تحسن ملحوظ واستمر هذا التحسن طوال فترة المتابعة.

الاستنتاج: جراحة إصلاح القيلة المائية النخاعية أسفل الظهر مع تحرير الحبل الشوكي يحتفظ بنسبة نجاح عالية فى الشفاء.