Terminal myelocystoceles: a series of 17 cases

DEEPAK KUMAR GUPTA, M.CH., AND ASHOK K. MAHAPATRA, M.CH.

Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India

Object. A terminal myelocystocele is a rare form of spinal dysraphism in which the hydromyelic caudal spinal cord and the subarachnoid space are herniated through a posterior spina bifida. In their study of 17 cases, the authors attempt to develop treatment guidelines for patients suffering from this condition.

Methods. Seventeen patients (age range 2 months–5 years) underwent surgery during a 7-year period. Surprisingly, nine of 17 patients had no neurological deficits. Among these nine asymptomatic cases, three patients had a dermal sinus and two had a skin dimple, whereas one had congenital talipes equinovarus deformity (CTEV). Among the eight patients with neurological deficits, seven had lower-limb weakness (two had complete paraplegia), and one patient had urinary incontinence and constipation. Four patients each had a dermal sinus and CTEV; interestingly, one infant had an accessory phallus. Radiologically, eight patients had pure myelocystoceles, and a diagnosis of lipomyelocystocele was made in nine instances on the basis of magnetic resonance images. All patients underwent surgery. The meningocele sacs were excised, and the bands tethering the myelocystocele sac to the meningocele sacs were incised, with as little damage to the roots as possible. Two patients experienced postoperative CSF leakage (one required reexploration), and two others had subcutaneous CSF collection. No patient had deterioration of the neurological condition after surgery.

Conclusions. The authors present the largest study of patients with terminal myelocystocele to date and emphasize that the possibility of this condition must be kept in mind when addressing all congenital lumbosacral masses. The authors assert that myelocystoceles and lipomyelocystoceles should be kept in the same category and recommend early surgery in all cases.

KEY WORDS • spinal dysraphism • lumbosacral mass • terminal myelocystocele • lipomyelocystocele • outcome • pediatric neurosurgery

A MYELOCYSTOCELE is defined as an occult spinal dysraphism (the closed form of a neural tube defect) with a localized, cystic dilation of the central canal of the spinal cord that is herniated through a posterior spina bifida.11,23 Truly an anomaly of the caudal cell mass, a terminal myelocystocele can be associated with anomalies of the anorectal system and lower genitourinary system such as anal atresia, cloacal extrophy, lordosis, scoliosis, and partial sacral agenesis.15,16,29 It can present as a large lumbosacral mass with good skin cover and containing fat, CSF, and neural tissue. The spinal cord terminates at a neural placode, wherein the central canal opens into a CSF-filled cavity. Occasionally the mass attains giant size and is cosmetically disfiguring for the patient, besides causing neurological sequelae from the associated tethered cord. Cervicothoracic myelocystoceles are of unknown cause,27 but clearly they are not the result of disturbances of the caudal cell mass, so these lesions should not be confused with terminal myelocystoceles.27 To the best of our knowledge, only one small study of four cases of terminal myelocystocele has been reported to date.11 Because this report of 17 cases is the largest one so far in the current world literature, we attempt to establish treatment guidelines concerning these patients.

Clinical Material and Methods

Patient Population

Seventeen patients with terminal myelocystoceles who were admitted to our institution from January 1998 to December 2004 are included. During this period, we performed surgery in 540 patients with spinal dysraphism. Table 1 summarizes the clinical details of the 17 patients. All of them underwent MR imaging, evaluation of urinary tract function, and detailed renal function testing. After surgery, patients were regularly followed up, and their neurological status was assessed.

The ages of the 11 girls and six boys ranged from 2 months to 5 years (mean 20.8 months). All patients were the product of full-term normal vaginal deliveries with no significant ante- or postnatal events. An antenatal diagnosis was not made for any of them. All patients presented with a progressively increasing lumbosacral swelling, with over-
Terminal myelocystoceles

**Clinical features of 17 patients with terminal myelocystoceles or lipomyelocystoceles***

<table>
<thead>
<tr>
<th>Age, Sex</th>
<th>Clinical Features</th>
<th>Associated Anomalies</th>
<th>Radiological Findings</th>
<th>Op Details</th>
<th>Follow-Up Status (duration)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6 mos, F</td>
<td>LM (5 × 5 cm), no deficit</td>
<td>none</td>
<td>myelo; conus at S-2</td>
<td>excision of sac, detethering of cord</td>
<td>no deficit (5 yrs)</td>
</tr>
<tr>
<td>5 mos, F</td>
<td>LM (4 × 3 cm), no deficit</td>
<td>none</td>
<td>lipomyelo; conus at S2–3</td>
<td>excision of sac, detethering of cord</td>
<td>no deficit (1 yr)</td>
</tr>
<tr>
<td>1 yr, M</td>
<td>LM (5 × 5 cm), no deficit</td>
<td>skin dimple</td>
<td>myelo; conus at L5–S1</td>
<td>excision of sac, detethering of cord</td>
<td>no deficit (2 yrs)</td>
</tr>
<tr>
<td>3 yrs, F</td>
<td>LM (8 × 7 cm), UI, constipation</td>
<td>none</td>
<td>myelo; conus at S-1</td>
<td>excision of sac, detethering of cord</td>
<td>improved, continent (1.5 yrs)</td>
</tr>
<tr>
<td>2 yrs, M</td>
<td>LM (6 × 4 cm), no deficit</td>
<td>skin dimple</td>
<td>lipomyelo; conus at S2–3</td>
<td>excision of sac, detethering of cord, CSF collection</td>
<td>no deficit (2 yrs)</td>
</tr>
<tr>
<td>3 yrs, M</td>
<td>LM (8 × 6 cm), UI, constipation, lt lower-limb weakness</td>
<td>accessory phallic</td>
<td>lipomyelo; conus at L5–S1</td>
<td>excision of sac, detethering of cord</td>
<td>improved, continent (1 yr)</td>
</tr>
<tr>
<td>2 yrs, M</td>
<td>LM (5 × 5 cm), no deficit</td>
<td>skin dimple</td>
<td>myelo; conus at L-4</td>
<td>excision of sac, detethering of cord</td>
<td>no deficit (3 mos)</td>
</tr>
<tr>
<td>4 yrs, M</td>
<td>LM (4 × 4 cm), UI, constipation, lt lower-limb weakness, sensory deficit</td>
<td>hydromyelencephalos, scoliosis</td>
<td>lipomyelo; conus at S-3, Chiari I, T1–5 syrinx</td>
<td>excision of sac, detethering of cord (CSF leak for which wound reexploration performed)</td>
<td>improved, continent improved (1 yr)</td>
</tr>
<tr>
<td>2 yrs, F</td>
<td>LM (8 × 5 cm), no deficit</td>
<td>DS</td>
<td>myelo; conus at S3–4, SCM II at L3–4</td>
<td>no deficit (18 mos)</td>
<td></td>
</tr>
<tr>
<td>5 yrs, F</td>
<td>LM (10 × 6 cm), UI, rt lower-limb weakness, sensory deficit</td>
<td>none</td>
<td>myelo; conus at L-4</td>
<td>excision of sac, detethering of cord (CSF collection)</td>
<td>improved, continent improved (6 mos)</td>
</tr>
<tr>
<td>1 yr, F</td>
<td>LM (5 × 4 cm), no deficit</td>
<td>blind DS</td>
<td>myelo; conus at S2–4, SCM II</td>
<td>no deficit (6 mos)</td>
<td></td>
</tr>
<tr>
<td>15 mos, F</td>
<td>LM (5 × 4 cm), flt foot &amp; leg weakness, UI</td>
<td>none</td>
<td>lipomyelo; conus at L3–4</td>
<td>excision of sac, detethering of cord</td>
<td>improved, continent improved (1 yr)</td>
</tr>
<tr>
<td>2 mos, F</td>
<td>LM (4 × 4 cm), no deficit</td>
<td></td>
<td>lipomyelo; conus at S3–4, Chiari II, hydrocephalus, dorsal syrinx</td>
<td>no deficit (2 yrs)</td>
<td></td>
</tr>
<tr>
<td>1 yr, F</td>
<td>LM (8 × 5 cm), earlier op, UI, bilateral foot weakness</td>
<td>CTEV</td>
<td>lipomyelo; conus at L-4, Chiari II</td>
<td>excision of sac, detethering of cord w/ VP shunt</td>
<td>worsened initially then improved (6 mos)</td>
</tr>
<tr>
<td>20 mos, F</td>
<td>LM (10 × 6 cm), paraplegia, UI</td>
<td>CTEV</td>
<td>lipomyelo; conus at S2–3</td>
<td>excision of sac, detethering of cord, CSF leakage, meningitis, wound disruption</td>
<td>improved (3 mos)</td>
</tr>
<tr>
<td>3 yrs, M</td>
<td>LM (8 × 4 cm), no deficit</td>
<td>scoliosis, DS</td>
<td>lipomyelo; conus at S2–3</td>
<td>excision of sac, detethering of cord</td>
<td>no deficit (2 mos)</td>
</tr>
<tr>
<td>7 mos, F</td>
<td>LM (8 cm, giant LM (30 × 20 cm), flaccid paraplegia, UI, constipation</td>
<td>CTEV, DS</td>
<td>lipomyelo; conus at S3–4</td>
<td>excision of sac, detethering of cord</td>
<td>unchanged (1 mo)</td>
</tr>
</tbody>
</table>

* DS = dermal sinus; lipomyelo = lipomyelocystocele; LM = lumbosacral mass; myelo = myelocystocele; SCM II = SCM Type II; UI = urinary incontinence; VP = ventriculoperitoneal.

Lying intact and healthy skin (Fig. 1). The mean size of the swelling was 7.8 × 5.8 cm (although one patient had a giant terminal lipomyelocystocele measuring 30 × 20 cm). A dermal sinus was noted in four patients, and two patients each had myelocystoceles and lipomyelocystoceles. Three patients had an overlying skin dimple. A CTEV deformity was recorded in four cases, three of which featured lipomyelocystoceles. One boy had an accessory phallus. The anterior abdominal wall and genitalia were normal in all of these cases. None of these children had an associated OEIS complex. Lipomyelocystoceles were seen in nine (53%) of 17 cases. Six patients (35%) had varying degrees of weakness of one or both lower limbs, with associated wasting and sensory disturbances, and two patients (12%) had paraplegia at presentation. Both of the patients with paraplegia had large lipomyelocystoceles. All of the patients who had motor weakness also had associated bladder and bowel involvement. Nine cases (53%) did not involve neurological deficits and featured a lumbosacral mass only. No neurological deficits were observed in patients younger than 2 years of age, who had small myelocystoceles. Of four patients with CTEV, three patients with associated lipomyelocystoceles had motor weakness of the lower limbs and bladder/bowel involvement. Eight patients (47%) had urinary incontinence and seven (42%) had constipation. Scoliotic deformity was recorded in two patients (12%), both of whom had lipomyelocystoceles. Overall, patients with lipomyelocystoceles had both higher incidences and greater degrees of deficits (Table 1).

### Radiological/Neuroimaging Findings

Plain spinal radiographs revealed varying degrees of lumbar and sacral spina bifida (Fig. 2). Two patients (12%) had dorsiolumbar scoliosis. Magnetic resonance imaging of the whole spine and a screening MR imaging study of the brain and craniospinal junction were performed in all instances. The MR imaging study of the lumbosacral spine in all of these cases revealed a cystic lesion in the lumbosacral cord that displayed a trumpetlike flaring of the central canal of the distal spinal cord (Figs. 3–6). A meningocoele or dilated subarachnoid space was located around the dilated central canal, which herniated into the subcutaneous region. In addition, the cord was low lying and tethered posteriorly in all instances (Fig. 7). No cervical thoracic myelocystocele was found in our series of cases. In nine cases (53%), the
overlying subcutaneous tissue contained a lipoma of varying thickness; hence, the lesions were considered to be lipomyelocystoceles. A craniospinal MR imaging study revealed a Chiari malformation in two (22%) of nine patients with lipomyelocystoceles (Fig. 3). Surprisingly, hydrocephalus was seen in only one case in which the patient had associated aqueductal stenosis. Type II SCMs were seen in two (25%) of eight cases involving terminal myelocystoceles. Both of the patients had no preoperative motor weakness and remained neurologically intact postoperatively. No patient with a lipomyelocystocele had an SCM. Dorsal syringomyelia was seen in two patients with lipomyelocystoceles, and both of them had limb weakness and bladder involvement. We also performed renal scanning preoperatively in all cases for evaluation of renal function. Renal tract abnormality was seen only in one patient with a lipomyelocystocele and bilateral hydronephrosis; however, the child’s renal function was normal (Fig. 8 and Table 1).

**Surgical Procedure**

All of the operations were performed through a vertical midline incision, with the patient prone, and involved laminotomy and excision of the sac, along with cord detethering (Fig. 9). The subcutaneous lipoma was removed piecemeal. The dural sac was identified and opened. Thick intradural arachnoid bands were divided. The terminal cyst, which was actually the dilated terminal central canal, was opened and the contents drained. There were thin, fibrous bands in the myelocystocele, which were divided. The conus was reconstructed with interrupted pial sutures. The filum was also divided and a biopsy taken. The dura mater...
was closed in a watertight fashion, after hydrocortisone was
instilled intrathecally to reduce the chance of future arach- 
noiditis (Figs. 10–13). Postoperatively, all of the patients 
were prone, with the foot of the bed elevated to avoid a CSF 
leak. They received antibiotic agents for 7 days and ace- 
tazolamide for 21 days postoperatively. Ventriculoperitoneal 
shunt surgery was performed in one case for associated hy- 
drocephalus. A CSF leak developed postoperatively in two 
patients (12%). One of them required reexploration of the 
wound and repair of the dural defect, whereas wound resu- 
turing stopped the CSF leak in the second patient. Two pa-
tients (12%) had CSF collection postoperatively, which re-
solved over a 2- to 3-week period with acetazolamide and 
compression dressings. The remaining 13 patients (76%) 
had an uneventful postoperative period.

Follow-Up Status and Long-Term Outcome

All of the patients were followed up in the neurosurgery
outpatient clinic for 1 to 60 months. The mean follow-up 
period was 14.6 months. All nine patients who did not have 
nervological deficits preoperatively remained asymptom-
atic during follow up. Of the eight patients with preopera-
tive motor weakness, five (62.5%) had improvement in 
motor power over a 6- to 18-month postoperative period. 
One patient with preoperative bilateral foot drop worsened 
after surgery; however, he improved subsequently to a pre-
operative motor power level over a 3-month period. Both 
of the patients who were paraplegic preoperatively did 
not improve initially. One of the patients with paraplegia 
who experienced a CSF leak, postoperative meningitis, and 
wound disruption experienced improvement in motor 
power to a Medical Research Council hip and knee grade 
of 3 and 5, respectively, at 1-year follow up. The other pa-
tient with paraplegia also had a giant terminal lipomyelo-
cystocele and recently underwent surgery (1 month previous to this writing) and has not yet shown any improvement in motor power. Of the eight cases involving myelocystoceles, only one patient who had a large (10 × 6–cm) cystocele had preoperative right lower-limb weakness and sensory deficit. This patient also had improvement in motor power and regained continence 6 months after the operation.

Of the nine patients with lipomyelocystoceles, seven had preoperative motor weakness (two of them had paraplegia preoperatively). Four (57%) of these seven patients had improvement in motor power postoperatively, whereas one (with associated conus lipoma) initially worsened but regained preoperative power after 3 months. Of the remaining two patients with lipomyelocystoceles who were paraplegic preoperatively, one had some improvement in motor power at 1-year follow up and the second had no improvement at 1-month follow up. Of the eight patients who had preoperative urinary incontinence, three (37.5%) became continent and three (37.5%) had partial improvement. One patient regained partial continence at 3-month follow up and one patient with a giant terminal lipomyelocystocele remained incontinent (Table 1).

**Discussion**

**Incidence and Epidemiology**

A terminal myelocystocele consists of a skin-covered lumbosacral spina bifida, an arachnoid-lined meningocele directly continuous with the spinal subarachnoid space, and a low-lying hydromyelic spinal cord that traverses the me-

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**Fig. 9.** Preoperative (left; outer sac wall marked with an arrow) and postoperative (right) photographs of a giant terminal lipomyelocystocele.

**Fig. 10.** Schematic drawing of the operative findings. The content of Cyst 1 was clear xanthochromic fluid, and the content of Cyst 2 was clear watery fluid. The pathological findings of a specimen taken from the cyst wall of site 1 (the dilated central canal/cystocele) was as follows: “neuroglial tissue with ependymal lining, meningeal and adipose tissues.” The pathological findings of the specimen taken from the meningocele at site 2 (the extension of spinal subarachnoid space/meningocele) was as follows: “fibroadipose tissue with peripheral nerve twigs.”

**Fig. 11.** Intraoperative photograph of a terminal lipomyelocystocele. The probe inside a dilated central canal is marked with an arrow. Forceps pointing at the nerve roots coursing across the outer sac are shown with a dotted arrow.
ningocele and forms a distal sac that does not communicate with the subarachnoid space (Fig. 14). The terminal cyst is lined by ependyma and dysplastic glia, and is directly continuous with the dilated central canal of the cord, probably representing a ballooned terminal ventricle. 15,16 Terminal myelocystoceles constitute 4 to 8% of lumbosacral occult spinal dysraphisms11,12,16,19,23 and can occur, although rarely, in the lower thoracic region.3,18 Epidemiologically, myelocystoceles are reported sporadically, and there is no known familial incidence.2,16 No true preponderance by sex has been described.3,16 Female preponderance (65%) was seen in the present series. Most of the previous literature on this entity is in the form of case reports.3,11,12,16,19,23,27 There have been no large published studies, and, to the best of our knowledge, this is the only series reported in the English literature. Isolated case reports on terminal myelocystoceles are available in the literature.3,9,12,16,17,20,21 McLone and Naidich16 described two of them in a study of 48 cases of skin-covered lumbosacral masses. In 1973, Hayden, et al.,9 reported two cases of terminal myelocystocele associated with exstrophy of the cloaca. Russell22 and Schmidt and Kawakami23 reported on similar cases. Kumar and Chandrak11 described four cases of terminal myelocystoceles, but none of them was associated with OEIS complex. Lemire, et al.,12 reported on two cases of terminal myelocystocele in a review of 31 cases of lumbosacral mass. In the present study we describe 17 cases of terminal myelocystocele but, to our surprise, none of them involved associated OEIS complex.

Embryology of Myelocystoceles

The normal spinal cord has two stages of development. The segment from the medulla to the midlumbar regions develops by neurulation, and the more distal cord, the conus medullaris, and the terminal filament develop by canalization and retrogressive differentiation. Neurulation and tube closure begins at numerous sites and proceeds in both directions. The anterior neuropore closes on approximately the 24th day and the posterior neuropore on the 27th day of intrauterine life. Canalization is the elongation of the neural tube caudal to the posterior neuropore. A caudal mass forms from the fusion of the notochord and neuroepithelium. Microcysts form within this mass at approximately 30 days and eventually combine to form an ependyma-lined tube that becomes united with the neural tube. On the 38th day, the caudal neural tube decreases in size because of cell necrosis. This retrogressive differentiation forms the distal conus medullaris, terminal filament, and ventriculus terminalis.1 Steinbok and Cochrane24–26 postulated that both the meningocele and the myelocystocele lesions are part of a spectrum of the same underlying abnormality (namely limited dorsal myeloschisis, defined as an incomplete fusion of the posterior part of the neural tube) and at this level, the cutaneous ectoderm fails to separate from the neuroectoderm. Consequently, the myofascial tissues do not develop normally in the midline. As a result, a band of tissue (or stalk) containing a central canal extends from the dorsal spinal cord to the skin. Depending on the presence of hydromelia, the central canal in the stalk either stays open (myelocystocele) or regresses (meningocele). Terminal myelocystoceles are thought to arise during the period of secondary neurulation from the caudal mass because of spontaneous closure of the caudal end of the neural tube.15,16,19 McLone and Naidich16 postulated that a terminal myelocystocele arises because CSF is unable to exit from the central canal. This blockage dilates the terminal ventricle, disrupting the dorsal mesenchyma but not the surface ectoderm. Thus, a spina bifida develops beneath the intact skin. Continued growth of the terminal cyst by accumulation of CSF distends the surrounding arachnoid lining of the distal thecal sac, causing formation of a meningocele. Progressive distension of the distal cord causes it to bulge caudally below the end of the meningocele into the extraarachnoid space, where it is covered by fat. The bulk of the cyst also bulges cephalad to expand the distal cord, producing trumpetlike flaring, and prevents ascent of the cord, producing a teth-
eder cord. The cystic dilation remains tethered to the cord, preventing the ascent of the cord.3,4,7,16,28 A few reports are also available in which the authors suggest a possible relationship to teratogens such as hydantoin, loperamide, and retinoic acid, although the exact origin of meningoceles is not yet known.2,21,22 In experimental studies, retinoic acid has been used to produce myelocystoceles in golden hamsters.13,22

**Clinical Presentation**

A cystic, skin-covered, lumbosacral mass is nearly always visible at birth, varying in size from small to large.8,16 The mass typically occupies and obliterates the intergluteal cleft and extends upward from the perineum for a variable distance. The skin overlying the mass can appear normal or can exhibit hemangioma, nevus, or hypertrichosis.16,21 The intergluteal fold is commonly obscured and distorted by a myelocystocele, but it is preserved with a lipomyelomeningocele.16 The mass can be low lying and can present low in the perineum. Midline abdominal and pelvic anomalies often accompany terminal myelocystoceles.11,12,15,23,29 This constellation of abnormalities is best represented by the acronym “OEIS,” a complex described in the literature as including an omphalocele defect, exstrophy of the bladder, imperforate anus, and spinal abnormalities, all occurring together.2,19 All of our patients presented with progressively increasing lumbosacral cystic masses. One patient had a giant terminal lipomyelocystocele. No patient in our study had OEIS complex. Three patients (17.6%) had an overlying skin dimple and four (23.5%) had dermal sinus. One boy had an accessory phallus. Four patients (23.5%) had foot deformities. The anterior abdominal wall and genitalia were normal in all of these cases. Scoliotic spinal deformity was seen in two cases (12%). In contrast to the previously reported cases, most of our patients presented later and had larger areas of swelling, accounting for the high incidence of neurological deficits seen in our study (eight of 17 patients had preoperative motor deficits). Of the four patients with terminal myelocystoceles reported on by Kumar and Chandra,11 all were younger than 1 month of age and all had lesions less than 2 cm, except for one patient with a 12 × 15–cm swelling. The mean size of the lumbosacral mass seen in our study was 7.8 × 5.8 cm, and the mean age was 20.8 months.

In contrast to children born with meningomyeloceles, children born with myelocystoceles usually have no neurological deficits because the sac does not contain neural tissue in these cases; however, neurological deterioration can occur because of associated anomalies like tethered cord, thickened terminal filum, Chiari malformation, hydrocephalus, and hydromyelia.6,10 In contrast to the reported incidence of minimal neurological deficit in the isolated case reports of terminal myelocystoceles, we noticed a very high incidence of preoperative motor weakness in eight (46%) of 17 of our cases of terminal myelocystoceles (two of these patients had paraplegia at presentation). Of the nine patients with lipomyelocystoceles, seven had preoperative motor weakness (two of them had paraplegia preoperatively). Of the eight patients with myelocystoceles, only one with a large (10 × 6–cm) cystocele had preoperative weakness and sensory deficit of the right lower limb. We thus

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**Fig. 14.** Anatomical drawing depicting a terminal lipomyelocystocele. The trumpetlike flared central canal is surrounded by a meningocele with associated lipomatous tissue (double arrow). The lipoma and subcutaneous fat are shown by a dotted arrow. subcut = subcutaneous.
observed preoperative motor deficits in the majority of patients with lipomyelocystoceles. Six patients experienced varying degrees of weakness in one or both lower limbs associated with wasting and sensory disturbances, and two patients had paraplegia at presentation. Both of the patients who had paraplegia had large lipomyelocystoceles. All of the patients who had motor weakness also had associated bladder and bowel involvement. A few patients have deficits at birth and others are born with normal lower extremities but lose function over a period of time, probably as a result of the tethered spinal cord. Limb asymmetry can be detected clinically and is associated with spinal malformations, such as sacral dysgenesis. Paralysis of the bowel and bladder is complete when motor or sensory deficits are detected, as was seen in the present study.

Radiological/Neuroimaging Assessment

Magnetic resonance imaging is the radiographic modality of choice for patients with myelocystoceles, and it also delineates a number of associated abnormalities. Peacock and Murovic described the MR imaging findings in two cases involving terminal myelocystoceles. Imaging studies demonstrate direct continuity of the meningocele with the subarachnoid space and the presence of a cyst that is continuous with the central canal of the spinal cord. The lesion’s distinctive appearance on MR images is characterized by trumpetlike flaring of the distal cord central canal into an ependyma-lined terminal cyst. Possible abnormalities of the vertebral column include lordosis, scoliosis, and agenesis of the sacral parts. Compression of the spinal cord and the meningocele by a fibrous band can also occur. Because the cord does not communicate with the meningocele, myelography, if performed, will demonstrate the meningocele sac only. Imaging studies demonstrate that the clinically apparent mass is a second, thin-walled cyst with no internal structure. The ependyma-lined cyst is frequently the larger of the two cysts; it is typically situated posteriorly and inferiorly to the meningocele and occasionally extends rostrally outside the meningocele. Hence, it appears that the tethered cord and disruption of the caudal motor segments produce symptoms that may be present at birth or that may appear later and progress. To make a correct diagnosis, especially if abortion is being considered, a prenatal MR imaging procedure as well as a routine ultrasonography evaluation should be performed before the parents are counseled and should be repeated prior to operative treatment. We routinely perform MR imaging of the entire spine and a screening MR imaging study of the brain and the craniospinal junction in all patients with spinal dysraphism. The cord was low lying and tethered posteriorly in all of the patients in our study. No case featured a cervical thoracic myelocystocele in the present study. In nine cases, the overlying subcutaneous tissue contained a lipoma of varying thickness; hence, the lesion was considered a lipomyelocystocele. Craniospinal MR imaging revealed a Chiari malformation in 22% of cases with lipomyelocystoceles. Hydrocephalus was seen in only one case (this patient had associated aqueductal stenosis). Type II SCMs were seen in 25% of cases with terminal myelocystoceles.

The nature of underlying CSF disturbance is uncertain. Because hydrocephalus is extremely uncommon in patients with terminal myelocystoceles, we suggest that the disturbance is local rather than global and that the patients have normal intellectual potential. In our study of 17 cases of terminal myelocystoceles, only one patient had hydrocephalus, and that was associated with aqueductal stenosis.

Surgery and Outcome

Early corrective surgery in terminal myelo- and lipomyelocystoceles is the recommended treatment. Surgery was performed in all patients in our series. All of the patients who were neurologically intact preoperatively remained healthy in that regard postoperatively. Most of the patients who had preoperative deficits also had neurological improvement following surgery. Surgical correction of myelocystoceles is not done only for cosmetic reasons but also to detether the spinal cord prophylactically to prevent future neurological deterioration. The spinal cord can be tethered posteriorly because the tissue band runs into the sac or because of adhesions at the base of the sac. It is therefore important to perform a thorough intradural inspection to transect all adhesions during surgical corrections so that future deterioration due to spinal cord tethering can be prevented. Surgery per se is uneventful in the majority of these cases. No neurological deterioration has been reported in the isolated case reports of terminal myelocystoceles available so far. In our experience, CSF leakage is a frequent complication after surgery, but it often subsides with conservative treatment. Only one patient who had CSF leakage suffered from meningitis postoperatively. Neither Kumar and Chandra in their study of four patients nor Sim, et al., reported any CSF leakage or meningitis during 2 to 8 months of follow up. There were no operative deaths. All nine patients who did not have preoperative neurological deficits remained asymptomatic during 2 to 60 months of follow up. Of the eight patients with preoperative motor weakness, five had improvement in motor power over the 6- to 18-month postoperative period. One patient with preoperative bilateral foot drop worsened after surgery initially but improved subsequently to preoperative motor power at 3 months. Of the two patients with paraplegia, one had partial improvement 1 year later and the one who had flaccid paraplegia remained unchanged at the 1-month follow up. Overall, most patients in this study had a good outcome. Terminal myelocystoceles have been overlooked in the past because of inadequate diagnostic techniques. This entity should be considered when pediatric lumbosacral masses are evaluated. Undoubtedly, additional cases in the literature are incorrectly described as other disease entities.

Conclusions

A terminal myelocystocele is a rare and distinct form of spinal dysraphism. All patients should undergo an MR imaging study of the entire spine with a screening MR imaging study of the brain and the craniospinal junction. The outcome is better in myelocystoceles and lipomyelocystoceles (because they do not contain any neural elements) than it is with myelomeningoceles. Lipomyelocystoceles are as common as myelocystoceles. Associated with a poorer outcome, they usually present late and with preoperative motor and sensory deficits. Foot deformities are more commonly seen with lipomyelocystoceles. Early surgery
improves the outcome in the majority of patients, including those with preoperative deficits. Most of the patients present with associated congenital anomalies and neurological deficits. Surgical treatment consists of resecting the sac and transecting the adhesions at the base of the cystocele to detether the spinal cord to prevent future deterioration. Although uncommon, myelocystoceles should be included in the differential diagnosis of congenital lesions presenting as a lumbosacral mass. Terminal myelocystoceles should be surgically repaired as early as possible to prevent progressive neurological deficit. We strongly suggest surgery even in asymptomatic patients, not only for cosmetic reasons but also to prevent future neurological deterioration from the tethered cord.

References


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D. K. Gupta and A. K. Mahapatra

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Address reprint requests to: Ashok K. Mahapatra, M.Ch., Department Of Neurosurgery, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India. email: akmahapatra_22000@yahoo.com.