Cervicothoracic nonterminal myelocystocele with mature teratoma

Case report

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Nonterminal myelocystocele is a rare type of spinal dysraphism characterized by a closed defect with an underlying CSF-filled cyst, either contiguous with the central spinal canal or attached to the spinal cord by a fibrovascular stalk. The authors report the unusual case of a neonate with a prenatal diagnosis of cervicothoracic nonterminal myelocystocele who underwent postnatal surgical untethering of the lesion. Pathological analysis of the excised lesion revealed neuroglial tissue with an ependymal lining associated with a mature teratoma. Three months after surgery, the patient has normal lower-extremity sensorimotor function and no evidence of bowel or bladder dysfunction. To the best of the authors’ knowledge, this is the first report of a patient with a nonterminal myelocystocele found to have an associated mature teratoma.

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Key Words • teratoma • nonterminal myelocystocele • myelomeningocele • meningocele • neural tube defect • Chiari Type 2 malformation • spine • dysraphism

Assman and colleagues8 first introduced the term myelocystocele in 1968 to describe an occult lumbar neural tube defect with extreme hydromyelia leading to herniation of the posterior wall of the spinal cord through defects in the dura and posterior bony elements of the spinal canal. Myelocystoceles may be further characterized as terminal (that is, lumbar or lumbosacral) or nonterminal. Nonterminal myelocystocele is a rare form of occult spinal dysraphism that most commonly occurs in the cervical or cervicothoracic region.18 It is thought to arise from focal nondisjunction during primary neurulation and consists of a posterior midline CSF-filled cystic mass arising from the spinal cord and expanding the dura to form a meningocele, with varying amounts of overlying fat.14–17

Rossi and colleagues17 characterized nonterminal myelocystocele into two distinct types. Type 1 is a forme fruste, characterized by a CSF-filled meningocele connected to the spinal cord by a fibrovascular stalk. Type 2 is a completed nonterminal myelocystocele, in which the cyst is continuous with the central canal of the hydromyelic spinal cord and the posterior wall of the spinal cord is contained in the meningocele.17 Surgical drainage of the meningocele, spinal cord untethering via resection of the stalk and separation from the overlying subcutaneous tissues, and reconstruction of the dura remains the mainstay of surgical treatment.22

Teratomas are the most common perinatal tumors and contain tissue from all 3 embryonic germ layers.7 They occur most frequently in the sacrococcygeal region, followed thereafter intracranially and in the cervical spine. Rarely, teratomas have been reported in association with neural tube defects, including myelomeningocele, diastematomyelia, and lipomyelomeningocele.4,6,10 To our knowledge, this is the first report describing a nonterminal myelocystocele found to contain a mature teratoma.

Case Report

History. This female infant was born via scheduled cesarean section to a 19-year-old G1P0 woman who initially came to neurosurgical attention for prenatal consultation at 25 weeks’ gestation, when her second-trimester ultrasound revealed intrauterine growth restriction and a neural tube defect. Magnetic resonance imaging of the fetus demonstrated a skin-covered neural tube defect ex...
tending from C-6 to T-2, with the splayed spinal cord extending into a skin-covered cyst (Fig. 1). The posterior fossa demonstrated crowding, with inferior displacement of the tonsils consistent with Chiari Type 2 malformation, raising concern that the skin overlying the defect might be thin or perforated. No other congenital anomalies were noted. There were no other gestational or perinatal complications, and the patient’s 1- and 5-minute Apgar scores were 8 and 9, respectively.

**Examination.** The patient was admitted to the neonatal intensive care unit. Physical examination revealed a microcephalic infant with a 4 × 5–cm posterior midline bulge at the cervicothoracic junction covered by dysplastic skin; there was no CSF leakage. The patient’s anterior fontanelle was flat, and she displayed spontaneous vigorous motion of all extremities without evidence of weakness, sensory loss, or deformity. An abnormal red reflex noted in right eye was consistent with Peters’ anomaly, a spectrum of congenital opthalmological abnormalities involving a central corneal opacity. No additional physical abnormalities were identified on examination. Postnatal brain and spine MRI demonstrated a hydromyelic cervical central spinal canal continuous with the cystic neural tube defect, consistent with a Rossi Type 2 nonterminal myelocystocele. The imaging findings confirmed the presence of a Chiari Type 2 malformation and a borderline low-lying conus terminating at L2–3, but no other tethering lesions were identified (Fig. 2).

**Operation.** The patient underwent surgical repair of the defect on her 5th day of life. She was positioned prone on gel rolls; the dorsal midline was incised superior and inferior to the defect, and an epidural plane was developed. The dysplastic neuroglial wall of the cystic lesion was identified as protruding from incompetent dura. Because the herniated neural tissue appeared grossly dysplastic, it was considered nonfunctional. Further intraoperative characterization with neurophysiological stimulation was not attempted, and the lesion was sharply dissected and cut from the dorsal spinal cord. The open central canal of the spinal cord was visible after resection, confirming the imaging diagnosis of a Type 2 nonterminal myelocystocele. The wound was copiously irrigated and closed in layers.

**Histopathological Examination.** Histopathological analysis of the excised lesion demonstrated neuroglial tissue with ependyma consistent with the posterior wall of the hydromyelic spinal cord. In addition, there was an associated mature teratoma composed of disorganized tissue from all 3 germ layers including cartilage, smooth muscle, adipose, ganglion cells, mucinous glands, squamous epithelium, and ciliated respiratory epithelium (Fig. 3). The teratoma was not evident on preoperative imaging or as a discrete lesion intraoperatively. There was no evidence of immature elements or malignant transformation.

**Postoperative Course.** After surgery, the patient was
neurologically intact and had no new deficit. Follow-up ultrasound of the head did not show progressive ventricular enlargement or hydrocephalus, and the patient did not develop any signs or symptoms of raised intracranial pressure. She was discharged to home at 14 days of age. Three months after discharge, she continues to do well with vigorous activity in bilateral lower extremities and no evidence of hydrocephalus, neurogenic bowel, or neurogenic bladder.

Discussion

Terminal (that is, lumbar or lumbosacral) and nonterminal myelocystoceles are thought to arise from distinct embryological errors. Terminal myelocystocele is postulated to arise from a failure of secondary neurulation. Specifically, obstruction of CSF flow in the primitive neural tube is thought to lead to expansion of the terminal ventricle into a cyst. This subsequently disrupts surrounding neural elements, preventing spinal cord ascent and leading to tethering.9 Patients with terminal myelocystocele commonly have associated hindgut and genitourinary anomalies including imperforate anus, bladder extrophy, sacral dysgenesis, and caudal regression syndromes.11,14

Conversely, nonterminal myelocystocele is postulated to occur as a result of focal myeloschisis due to incomplete fusion of the neural folds during primary neurulation,14 preventing normal separation of ectoderm and neuroectoderm. This results in a persistent connection between the dorsal spinal cord and the overlying skin and subcutaneous tissue via a fibrous stalk extending through an incompetent dural tube and bony spinal canal. Cerebrospinal fluid pulsations from the spinal subarachnoid space are thought to dissect along the stalk and create a cyst, either by herniation of the posterior dura through a defect in the posterior elements of the spinal canal (Type 1) or the expansion of

![Histopathological slides.](image)

Fig. 3. Histopathological slides. A: Myelocystocele (asterisk) composed of mature neuroglial tissue with portions of ependymal lining (inset) associated with a mature teratoma. B and C: The mature teratoma contains disorganized foci of squamoid (sq), ciliated (cil), and foveolar (fov) epithelium, smooth muscle cells (sm), ganglion cells (g), cartilage (c), adipose tissue (a), and peripheral nerves (n). H & E, original magnifications ×20 (A) and ×100 (B, C, and inset).
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a focal hydromyleia with herniation of the posterior aspect of the spinal cord as well as the dura (Type 2).14,17 Non-terminal myelocystoceles may be associated with Chiari Type 2 malformations, as in our patient, as well as with hydrocephalus and diastematomyelia.13,12,14,22 Consistent with previous reports of nonterminal myelocystoceles, the herniated neural tissue was considered dysplastic and was excised.1,11,17,22 While intraoperative neurophysiology may provide further information regarding its functional status, our identification of a teratoma within the dysplastic neural tissue may provide caution to the surgeon considering a strategy other than excision.

Most patients with nonterminal myelocystoceles do not have neurological symptoms at birth, whereas those with terminal myelocystoceles commonly exhibit lower-extremity motor and sensory dysfunction and neurogenic bowel and bladder.11,14 As with other types of occult (that is, skin-covered) neural tube defects, patients with a nonterminal myelocystocele tend to have a better prognosis than those with open defects such as myelomeningoceles. The neural tissue is not exposed to the amniotic fluid and the majority of the neural tissue remains within the spinal canal. Because these lesions are covered with skin and varying amounts of overlying adipose tissue, they typically do not leak CSF and are at low risk for progressive injury due to mechanical trauma.11,14 Surgical indications for repair of nonterminal myelocystoceles include untethering of the spinal cord, cosmetic as well as functional improvement (many infants are unable to lie supine), and resection of a secondary tethering lesion such as a spiculum in the context of split cord malformation.14 Surgical principles involve resecting the myelocystocele at the dura along with any neuroglial or fibrous stalk to avoid retethering and late-onset neurological deterioration.11,15 Superficial, limited excision of the sac is not recommended because of the high risk of persistent tethering with subsequent neurological degeneration.11,15 Pathological examination may reveal a fibrovascular stalk in the case of Type 1 myelocystoceles or neuroglial tissue with ependymal lining in Type 2 myelocystocele, as in the present case.19

Teratomas are traditionally thought to arise from errant primordial germ cells from the yolk sac. They are subclassified into the following: the mature or adult type, which is characterized by well-differentiated tissues and low or absent mitotic activity; the immature or fetal type, which has less well-differentiated tissues; or the malignant type, which involves frank neoplastic tissue.5,16 Koen and colleagues4 reported on a series of 4 patients with teratomas that were associated with a variety of dysraphic lesions, and they proposed a dysembryonic mechanism for the development of teratomas. Specifically, they suggested that pluripotent cells of the caudal cell mass may give rise to these tumors through various types of cell-signaling dysfunction.5,6 The patient described in our case had a cervical nonterminal myelocystocele containing a teratoma located above the caudal cell mass. It has been postulated that these midline tumors may originate from rests of pluripotent cells at sites of early neural tube closure.10 Habibi and colleagues4 reported the largest series of patients with myelomeningocele associated with teratomas. In their series of 13 patients with teratomas and 2 with teratoid tumors, most lesions occurred in the lumbosacral spine. Three cervical and one cervicothoracic lesions were included in the series. All of the lesions were treated with standard excision of the myelomeningocele sac, including the teratoma, and closure of the neural tube defect. All teratomas were of the mature type, and none of the patients experienced tumor recurrence during the reported follow-up period. The authors reported common features—including a “finger-like appendage” protruding from a cutaneous lesion and 2 lesional cysts that did not communicate with the spinal canal in 4 of the cases with cervical involvement—and they concluded that these stigmata may be indicative of an associated teratoma.4 None of these findings were present in our case.

Great variation exists in the nomenclature used to describe cervicothoracic spinal dysraphic lesions. Some authors have suggested that previously described cervicothoracic meningoceles actually represent nonterminal myelocystoceles.1,21 The series reported by Pang and Dias4 originally described the pathological condition as “cervical myelomeningocele,” but numerous authors later deemed the lesion consistent with a myelocystocele.11,14,17 While teratomas have been reported in association with myelomeningocele, diastematomyelia, and lipomyelomeningocele (Table 1), to our knowledge this is the first re-

**TABLE 1: Representative studies illustrating the spectrum of dysraphic lesions associated with mature teratoma**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Dysraphism (no. of lesions)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reid &amp; Mickle, 1985</td>
<td>1</td>
<td>MMC</td>
<td>lumbosacral level</td>
</tr>
<tr>
<td>Koen et al., 1998</td>
<td>3</td>
<td>MMC (1), diastematomyelia (2), lipomyelomeningocele (1)</td>
<td>1 patient w/ both diastematomyelia &amp; MMC</td>
</tr>
<tr>
<td>Ozer &amp; Yüceer, 1999</td>
<td>1</td>
<td>MMC, dermal sinus tract, diastematomyelia</td>
<td>thoracolumbar level</td>
</tr>
<tr>
<td>Semerci et al., 2001</td>
<td>1</td>
<td>MMC, absence of thoracic, lumbar, &amp; sacral vertebrae</td>
<td>30-wk-old fetus, also w/ imperforate anus, Potter facies, bilateral renal agenesis, ven-tricular septal defect</td>
</tr>
<tr>
<td>Muthukumar, 2003</td>
<td>1</td>
<td>Type 2 split cord malformation w/ meningocele</td>
<td>initially thought to be lipomyelomeningocele</td>
</tr>
<tr>
<td>Habibi et al., 2007</td>
<td>13</td>
<td>MMC</td>
<td>2 additional patients w/ teratoid tumor</td>
</tr>
<tr>
<td>Parelkar et al., 2012</td>
<td>1</td>
<td>ruptured MMC</td>
<td>L5–S1 level</td>
</tr>
</tbody>
</table>

* MMC = myelomeningocele.
ported case of a nonterminal myelocystocele associated with a teratoma. We cannot confirm whether earlier reports of other dysraphic lesions associated with teratomas may actually represent myelocystoceles.

Conclusions
We report what, to our knowledge, is the first case of an infant with a nonterminal myelocystocele containing a mature teratoma. We did not observe the cutaneous stigmata that have been postulated to be associated with the presence of a teratoma. We recommend excision of the myelocystocele with the tumor.

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Disclosure
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 to the study and manuscript preparation include the following. Conception and design: Bollo. Acquisition of data: Gressot, Mohila. Analysis and interpretation of data: Gressot, Mohila. Drafting the article: Gressot. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Bollo. Study supervision: Bollo.

References

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