Giant true dorsal thoracic meningocele in a school-age child

Case report

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✓ A meningocele is a common form of spinal dysraphic lesions, but it is extremely uncommon in children in the upper thoracic region. The authors describe a rare case in which they found a giant true dorsal meningocele in the upper thoracic region in a symptomatic child, which, to their knowledge, is the first such reported case.

A school-age child, who harbored a giant dorsal mass, complained of restriction of function. He underwent successful surgery in which the meningocele sac was totally removed. (DOI: 10.3171/PED/2008/1/5/399)

KEY WORDS • meningocele • spinal dysraphism • thoracic meningocele

Cystic spinal dysraphism is a common term to describe a large group of congenital spinal anomalies, but the lesion is rarely seen in cervical and upper thoracic regions. These locations account for 1–8% of all spinal dysraphic lesions. In general, cystic spinal dysraphism almost always includes myelocystocele and meningocele, which is a well-known anomaly in the cervical region. In particular, true meningoceles are extremely rare in either cervical or upper thoracic regions.

Because meningoceles are repaired when diagnosed in infancy to prevent possible sac-related complications and cervical deformity, they are typically believed to be asymptomatic lesions in clinical practice.

We describe the case of a school-age child who harbored a giant true dorsal meningocele in the upper thoracic region and who underwent surgery for removal of the sac.

Case Report

History. This 7-year-old boy was a villager in Turkey. A cystic posterior dorsal mass had been present since birth and had been growing progressively for 7 years. His family history was unremarkable.

Examination. In August 2005, the patient was admitted to our clinic. He had a giant dorsal sac, which caused mass effect, interfered with dressing, and made lying supine impossible. Physical examination revealed a posterior midline protrusion measuring 10 cm in width, 15 cm in length, and 10 cm in height (Fig. 1 a–c). The sac was oval shaped, hard, not fluctuant, and had a wide base covered with full-thickness skin. The apex of the sac was covered with purplish and poor-quality skin that comprised ~50% of the total surface area of the sac. There was no sign of CSF leakage. Neurological examination demonstrated normal findings.

Plain whole-spine radiography revealed no gross lamina or osseous defects. Thoracic MR imaging showed a fluid-filled, septated cyst communicating with the epidural and subarachnoid spaces at T-1 because of neural arch defect, which was connected with the neck (1 × 1 cm). The cystic lesion was hypointense to neural tissue on T1-weighted images and hyperintense on T2-weighted images (Fig. 2 left), indicating signal characteristics compatible with CSF. The sac contained no nerve tissue and had no connection with the spinal cord (Fig. 2 right). The lesion was thought to be a meningocele. On further MR imaging examination, the spinal cord was observed to be normal, with no Chiari malformation, split cord malformation, low-lying cord, or tight terminal filum. Brain MR imaging revealed no hydrocephalus or other anomalies.

Operation. The thoracic meningocele was excised after induction of general anesthesia. A longitudinal oval skin incision was made around the base of the mass, circumferentially toward the fascia where the mass narrowed into a stalk. After opening the dome, except for some fibrous septa not connected to the spinal cord, no nerve tissue was seen within the cyst, and the sac was amputated at the neck level. The laminar defect at T-1 was enlarged with laminotomy to expose the junction of meningocele neck and the normal spinal dura mater. After extending the dural opening by

Abbreviations used in this paper: CDCTR = cystic dysraphism of the cervical and upper thoracic region; CSF = cerebrospinal fluid; MR = magnetic resonance.
making additional vertical incisions, microscopic magnification was used for intradural exploration. No fibrous bands or dysplastic nerve roots passing from the neck of the sac to the dorsal surface of the spinal cord were seen. The dura mater was closed in a watertight fashion, and the defect was repaired with flaps of paraspinal fascia.

Histopathological examination of a surgical specimen revealed no neural components. The definitive diagnosis was thoracic meningocele.

Postoperative Course. The patient remained neurologically intact (Fig. 1d). Brain computed tomography scanning demonstrated no hydrocephalus. The patient was discharged on the 5th postoperative day. He had no signs of neurological disturbance, and his growth and development appeared normal throughout 12 months of follow-up.

Discussion

Although a great deal has been published about spinal dysraphism, CDCTR is a rare clinical entity. These locations account for only 1–8% of all cases of spinal dysraphism.5–7,9,11,12 Recently, CDCTR has been classified into 3 types according to the structures found inside the cyst: a Type I lesion has a stalk, either neuroglial or fibrovascular, connecting the meningocele sac and spinal cord; Type II is a myelocystocele; and Type III is a true meningocele.9

A meningocele is an unusual herniation of meninges through a defect of the posterior spinal arch. The general term cystic spinal dysraphism is used to describe the lesions that are found in the common locations (thoracolumbar and lumbosacral spine). Upper-level (cervical and upper thoracic) spinal meningoceles, however, differ from those in the common locations in many ways. First, the more common lesions are usually fragile and covered by a thin arachnoid layer, whereas higher spinal lesions are always covered with full-thickness skin. Because of these main structural differences, CSF leakage is unusual in high spinal lesions. Second, the neural compartment of the common lesion is usually a flattened terminal neural placode, whereas in high spinal lesions there is a nearly closed neural tube. Third, patients harboring the common lesions often exhibit serious neurological dysfunction at or below the level of the lesion, whereas patients with high spinal lesion experience mild or no deficits.6,7

Based on our experience with CDCTR, we would now refine the description of meningoceles to include the fact that they are much more circumscribed by full-thickness skin, except for the sac dome. Also, the spinal cord typically remains entirely confined to the vertebral canal; there is no arachnoid exposed and, consequently, no CSF leakage.1,7,9,11 According to the new definition, few pediatric cases of thoracic meningoceles have been reported in the literature.1,4,9 These lesions (especially true meningoceles) may be differentiated from dorsal myelocystoceles either on imaging studies or intraoperatively. In myelocystoceles, the spinal cord may be tented posteriorly, angling toward the posterior wall of the spinal canal and enlarging the ventral subarachnoid space with fibrous bands,8 whereas in true meningoceles the spinal cord may be in a natural position and may not exhibit the MR imaging feature of being pulled to the posterior wall of the spinal canal. Additionally, myelocystoceles always include herniated neural tissue or a stalk,8 whereas true meningoceles have the spinal cord in normal relation to the spinal canal at surgery.

In general, a meningocele is initially not associated with paralysis because it contains only membranes in the sac wall. Because of this structural feature, except for its ex-
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ternal manifestation, a meningocele has been viewed as an asymptomatic lesion. A newborn in whom CDCTR is noted on initial neurological examination, for instance, is usually considered “normal” because an infant has no ability to articulate the presence of symptoms. On the other hand, some meningoceles can be associated with paralysis that occurs later in life as a complication of abnormal tissue growth associated with occult spinal lesions (lipomatous, teratomatous, or dermoid tissue and scarring at the site of repair) causing pressure on a nerve or the spinal cord. In the present case, we described a true dorsal thoracic meningocele in a school-age child. In our opinion, our patient represents the only case of a symptomatic meningocele in childhood, with complaints of restricted functional capacity later due to the progressive growth of the giant dorsal sac. Aside from the true meningocele in the upper thoracic region, the child had no additional anomalies.

To the best of our knowledge, the natural history of CDCTR has not been well known because most of these lesions in children were corrected surgically at the time of diagnosis to protect the child from a tethered cord or serious sac complications. Because of surgical morbidity and the obvious appearance of a meningocele, families sometimes prefer a follow-up course. Thus, our present case underscores the point that the natural course of high spinal meningoceles is a tendency to enlarge over time without treatment.

In conclusion, we have described a school-age child who harbored a giant true dorsal thoracic meningocele, which, to the best of our knowledge, is the first symptomatic case in childhood. These lesions can be repaired successfully with surgical therapy.

References


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