Case Report

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Thoracic Meningocele, Meningomyelocele or Myelocystocele? Diagnostic Difficulties, Consequent Implications and Treatment

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Key Words
Spina bifida cystica · Meningocele · Meningomyelocele · Myelocystocele · Abortion

Abstract
Spina bifida cystica is a closing disorder of the neural tube which infrequently occurs in the thoracic region. A rare lesion called myelocystocele is a variant of spina bifida cystica and is associated with syringomyelia, Chiari type 2 malformation and hydrocephalus. Usually the patient has no neurological deficit, but future deterioration can occur due to posterior tethering of the spinal cord by adhesions. The prenatal diagnosis by ultrasound study can be misleading and in order to attain the correct diagnosis, especially if abortion is considered, a prenatal MRI scan should be done before the parents are counselled, and should be repeated prior to operative treatment. Surgical correction of myelocystocele is not only for cosmetic reasons, but also to untether the spinal cord prophylactically to prevent future neurological deterioration. In this case report, we present a child born with a thoracic myelocystocele, the diagnostic difficulties, consequent implications and surgical treatment.

Introduction

Spina bifida cystica, also called spina bifida aperta or open spina bifida, is a congenital closing disorder of the neural tube, in which a sac composed of dura and arachnoid filled with cerebrospinal fluid is herniated through a defect in the posterior spine. Basically, spina bifida cystica is classified into meningocele, in which the herniated dural sac is filled with cerebrospinal fluid, and meningomyelocele, in which the sac also contains parts of the spinal cord and nerve roots. Spina bifida cystica most frequently occurs in the lumbosacral region. A thoracic location is rare [1–3].

Myelocystocele is a rare variant of spina bifida cystica which is consistent with a closed neural tube disorder since it is covered with normal skin. The exact incidence and spinal distribution of myelocystocele is not known, but to our knowledge there is no case report published of a thoracic myelocystocele.

In contrast to meningomyelocele, children born with myelocystocele usually have no neurological deficit. The prenatal diagnosis by ultrasound study can be misleading. In order to attain the correct diagnosis, especially if abortion is considered, a prenatal MRI scan should be done before the parents are counselled and should be repeated

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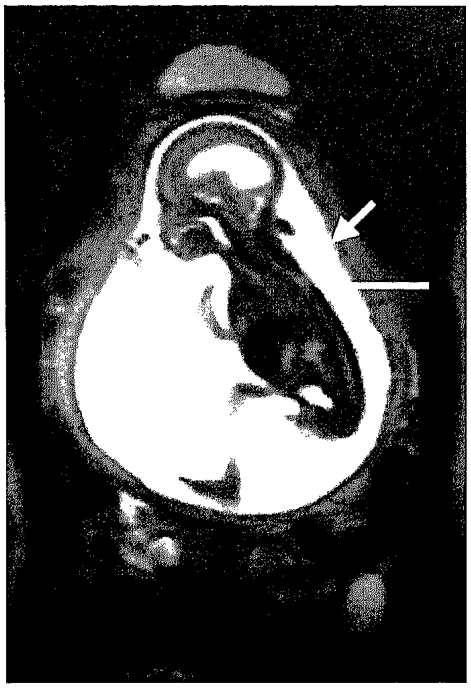

Fig. 1. Prenatal MRI scan at 21 weeks of gestation showing a midthoracic dorsal cele filled with cerebrospinal fluid and containing no neural elements (small arrow). At the level of the cele, the spinal cord is slightly tethered dorsally (large arrow).

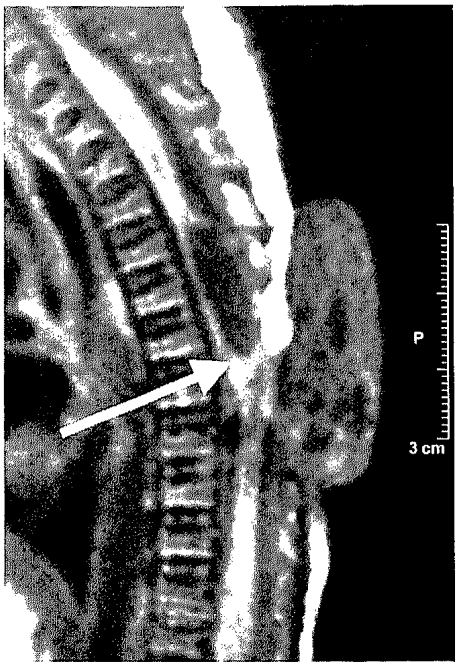

Fig. 3. Postpartum MRI scan of the spine showing a cystic lesion at the level Th5–Th6 where the spinal cord has a normal craniocaudal extension, but at the level of the cele, the cord is tethered posteriorly. Also, at the level of the lesion, a syrinx is visible which is in continuity with the cele and extends in both cranial and caudal directions (arrow).

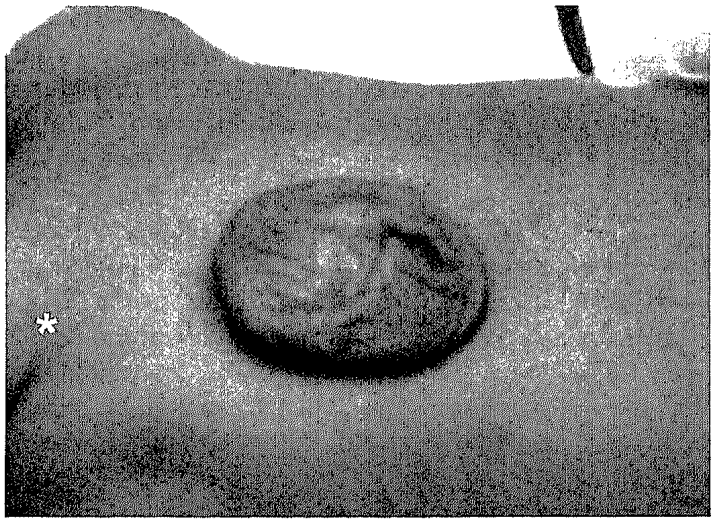

Fig. 2. Physical examination showing a 2 × 3 × 4 cm dorsal midline midthoracic mass covered with normal skin. The asterisks show the site of the head.

prior to operative treatment. Surgical correction of myelocystocele is not only for cosmetic reasons but also to untether the spinal cord prophylactically to prevent future neurological deterioration. In this case report, we present a child born with a thoracic myelocystocele. Discussed are the diagnostic difficulties, consequent implications and surgical treatment.

Case Report

A 22-year-old woman, gravida 3 para 0, underwent a routine ultrasound examination at 16 weeks for the determination of gestation. The ultrasound of the fetus showed ventriculomegaly and a thoracic lesion suspect of meningomyelocele. Because of the expected high paraplegia of the child, the ethical issue of terminating the pregnancy was discussed with the parents. Since a second ultrasound was inconsistent, a prenatal MRI scan was made at 21 weeks of gestation. This showed a midthoracic dorsal cystic lesion filled with cerebrospinal fluid and containing no neural elements. Remarkably, at the level of the lesion, the spinal cord was slightly tethered dorsally (fig. 1). The diagnosis of meningomyelocele was rejected and therefore the

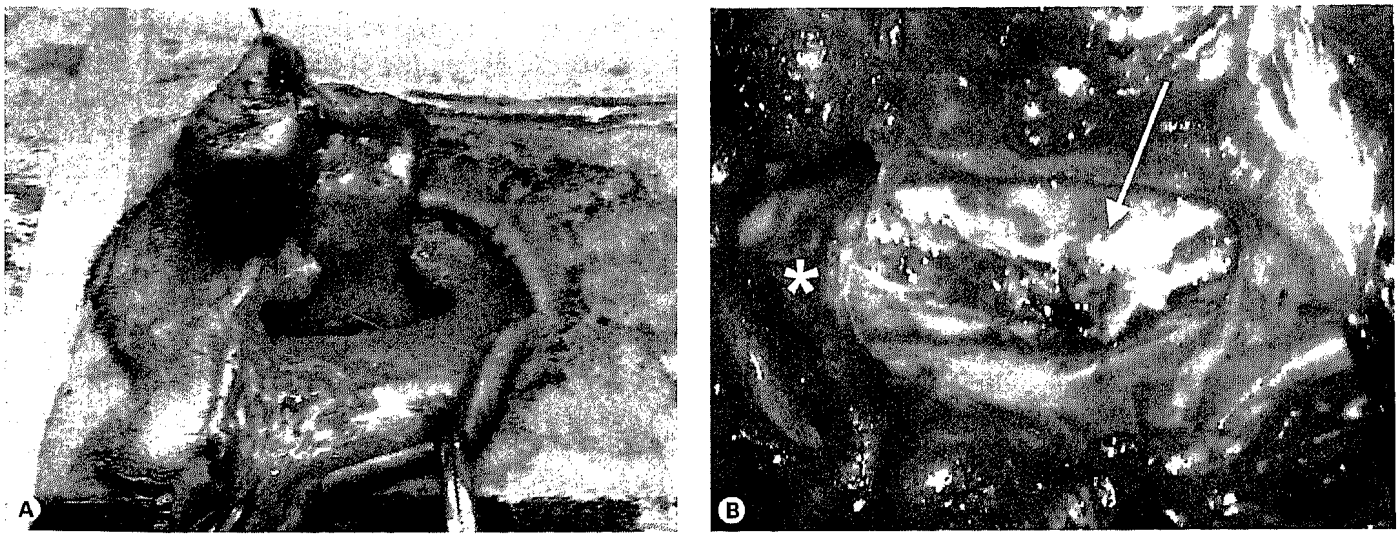

Fig. 4. A Intraoperative image showing the dura of the cele in continuity with the underlying spinal dura. **B** After opening the dura, the spinal cord is untethered due to transecting multiple adhesions extending from the spinal cord to the base of the cele, and transecting the tissue band running from the dorsal aspect of the spinal cord towards the sac (arrow showing coagulated tissue bands). The asterisk indicates the cranial site.

parents decided to continue with the pregnancy. More ultrasounds had been made which displayed no new information. A normal delivery was planned but because of fetal distress, a cesarean section was performed and a boy was born at 37 6/7 weeks of gestation.

On physical examination there was a 2 × 3 × 4 cm dorsal midline midthoracic mass which felt semi-solid, but did not transilluminate. The mass was covered with normal skin (fig. 2). On neurological examination, there were no deficits. A postpartum MRI scan of the spine was performed which showed a cystic lesion at Th5–Th6 where the spinal cord had a normal craniocaudal extension and at the level of the lesion the cord was slightly tethered posteriorly. A syrinx was visible at the level of the lesion which appeared to communicate with the sac and extend in both cranial and caudal directions (fig. 3). Moreover, Chiari type 2 malformation and hydrocephalus were present, which supported the diagnosis of myelocystocele.

At the age of 5 days, the myelocystocele was surgically corrected. After encircling the base of the cele and removing the excess skin, the underlying dura was visible (fig. 4A). The dura was opened and cerebrospinal fluid was discharged. There were no neural elements inside the sac, but the spinal cord appeared to be fixed to the base of the cele due to multiple bands of tissue. After transecting these adhesions, the spinal cord was untethered and some cerebrospinal fluid was discharged from the syrinx. The spinal cord was inspected craniocaudally, and other than some arachnoid adhesions, it showed no abnormality (fig. 4B). The sac was amputated and the wound was closed in layers.

The postoperative neurological examination was normal. A control MRI scan of the spine showed untethering of the spinal cord and reduction of the syrinx (fig. 5). One week later, the head circumference increased due to progressive hydrocephalus, and consequently, a ventriculoperitoneal shunt was placed in the right occipital ventricle. Shortly after, the boy was discharged in good condition with no neurological deficit.

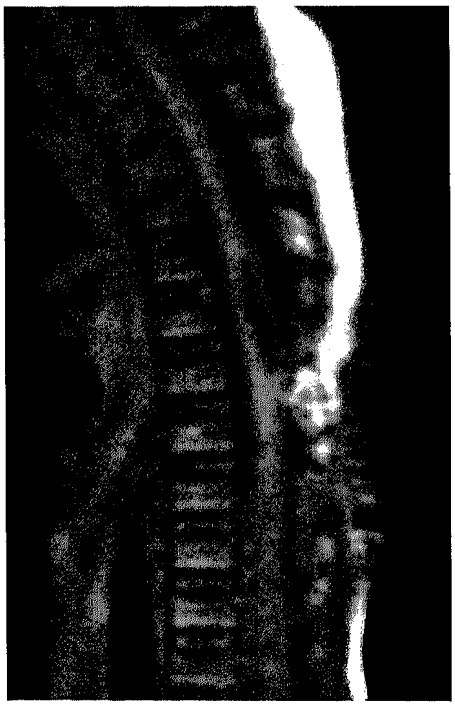

Fig. 5. Postoperative MRI scan of the spine showing the untethered spinal cord and reduction of the syrinx.

Discussion

Spinal dysraphism is a congenital closing disorder of the neural tube reported in 2–4/1,000 live births [4]. Several types of spina bifida, involving protrusion of the spinal cord and/or meninges through a defect in the vertebral arch, are often referred to collectively as spina bifida cystica because of the cyst-like sac that is associated with these malformations. When the sac contains meninges and cerebrospinal fluid, the condition is called meningocele. In case of meningomyelocele, the sac also contains neural elements. The most severe type of spina bifida cystica is called myeloschisis, also known as myelocele. In these cases, the spinal cord is open because the neural folds failed to meet and fuse, and as a result, the spinal cord is flattened [5].

Meningoceles can occur anywhere on the neural axis, but are located more often in the lumbar and lumbosacral region than in the cervical and thoracic region. In the study of Ersahin et al. [2], 2 out of 22 cases of spinal meningoceles were located in the upper and midthoracic region and Doran and Guthkelch [1] reported 65 children with a spinal meningocele of which 7 were thoracic.

In the cervical region, Steinbok and Cochrane [6, 7] and Steinbok [8] described two types of spina bifida cystica lesions based on the gross anatomic findings, namely myelocystocele and meningocele. The myelocystocele, also called syringocele or syringomyelocele, is characterized by a cyst within a cyst; the outer cyst is in continuity with the subarachnoid space and the inner cyst is in continuity with a distended central canal or syrinx. In other words, at the level of the spinal defect, there is a band of tissue running from the syrinx posteriorly into the inner cyst, containing a lumen filled with cerebrospinal fluid. Around this band of tissue, there is the outer cyst containing cerebrospinal fluid in continuity with the subarachnoid space. Moreover, the lesion is associated with Chiari type 2 malformation and hydrocephalus.

The meningocele, on the other hand, also contains a band of tissue extending from the dorsal position of the spinal cord towards the sac itself. The tissue band does not contain a lumen and the lesion is not associated with a syrinx.

In both types of dysraphic lesions, the spinal cord can be tethered posteriorly due to the tissue band running into the sac, or due to adhesions at the base of the sac. Pang and Dias [9] reported 9 cases of cervical myelomeningoceles and also reported the presence of a band of tissue tethering the spinal cord to the adjacent dural sac.

From an embryological point of view, Steinbok and Cochrane [6, 7] and Steinbok [8] postulated that both meningocele and myelocystocele lesions are part of a spectrum of the same underlying abnormality, namely limited dorsal myeloschisis. Limited dorsal myeloschisis is 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, in the midline, the myofascial tissues do not develop normally and 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 hydromyelia, the central canal in the stalk either stays open (myelocystocele), or regresses (meningocele).

Since the sac of the meningocele and myelocystocele does not contain neural tissue, children born with such a lesion usually have no neurological deficit. However, neurological deterioration can occur due to associated anomalies like tethered cord, thickened fillum terminale, Chiari malformation, hydrocephalus and hydromyelia [2, 10]. As mentioned before, the spinal cord can be tethered posteriorly due to the tissue band running into the sac, or due to adhesions at the base of the sac. Therefore, during surgical correction, it is important to perform a thorough intradural inspection to transect all the adhesions, in order to prevent future deterioration due to spinal cord tethering. Also in the patient we present, the sac contained no neural elements, but the spinal cord appeared to be fixed to the base of the cele due to multiple bands of tissue and was consequently tethered posteriorly. After transecting these adhesions, the spinal cord was relieved and untethered. Moreover, some cerebrospinal fluid was discharged from the band of tissue, which contained a central canal in continuity with the syrinx. These operative findings are consistent with the description of the myelocystocele of Steinbok and Cochrane [6, 7] and Steinbok [8].

The prenatal diagnosis of spina bifida by ultrasound study is well established and can be done in the second trimester of pregnancy [11]. However, the detection of all anomalies by ultrasound is limited and distinguishing the meningocele or myelocystocele from the meningomyelocele can be difficult [12, 13]. Since the former two types of lesions usually have no neurological deficit, it is very important to establish the correct diagnosis when a possible abortion is being discussed with the parents. In this particular case, the parents were being counselled since the diagnosis was thought to be a thoracic meningomyelocele and high paraplegia was expected. Because of the diagnostic dilemma, an additional MRI scan of the fetus

was performed, which rejected the diagnosis of meningomyelocele, and consequently, the pregnancy was continued and an unwarranted abortion was prevented. Therefore, we propose to perform a prenatal MRI scan in any case of diagnostic doubt. Moreover, we postulate that an MRI scan must be made whenever an abortion is being discussed, before counselling the parents in order to obtain solid arguments for the termination of the pregnancy.

Conclusion

We presented a child born with a thoracic myelocystocele, which is a rare lesion. Usually, there is no neurological deficit; however, neurological deterioration can occur due to posterior tethering of the spinal cord by intradural adhesions, or as a result of a tissue band extending from the dorsal spinal cord towards the sac. Surgical treatment consists of resection of the sac, dissecting the tissue band running inside the sac, and transecting the adhesions at the base of the cele in order to untether the spinal cord to prevent future deterioration. The prenatal diagnosis by ultrasound study can be misleading and therefore a prenatal MRI scan should be made and repeated prior to surgical correction. Moreover, we propose to perform a prenatal MRI scan whenever an abortion is being considered, before counselling the parents in order to obtain solid arguments for abortion.

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