

Antenatal Ultrasound Finding of a Cystic Back Mass in a Fetus with Terminal Myelocystocele

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An ultrasound of a twenty-three weeks fetus showed a cystic mass having a “cyst within a cyst” arising from the distal spine. The posterior fossa was normal with no hydrocephalus. Postnatal MRI confirmed the presence of terminal myelocystocele (TMC).

TMC should be considered in a fetus with a cystic mass over the lower spine especially when a “cyst within a cyst” appearance is present. We present the antenatal ultrasound and postnatal MRI appearance of TMC. As the definitive diagnosis of TMC may be difficult to make on antenatal ultrasound other cystic masses, such as, meningocele, myelomeningocele and cystic sacrococcygeal teratoma should be considered.

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Terminal myelocystocele (TMC) is a rare form of spinal dysraphism. This lesion is characterized by dilation of the distal central canal which extends through a defect in the posterior neural elements and is surrounded by an expanded dural covering or meningocele^{1,2}. TMC should be considered in the differential diagnosis of any fetus who presents with a cystic mass in the distal spine.

The aim of this report is to present a rare case of terminal myelocystocele (TMC) in a fetus discovered during antenatal ultrasound as a cystic back mass.

THE CASE

A twenty-five years old, gravida 2 para 1, female presented for routine antenatal ultrasound at 23 weeks gestation. Figure 1 showed a cystic lesion arising from the tip of the distal sacrum measuring 4.5 x 3.8 cm. The cyst was anechoic with thin wall and contained a second smaller thin walled cyst that measured 1.2 cm within it, giving an appearance of “a cyst within a cyst”, see figure 2. No intrapelvic extension, solid tissues or calcifications were seen. Fetal skull shape was normal, the cerebellum and cisterna magna appearance were normal, no ventriculomegaly. Movement of the lower limbs was observed. Amniotic fluid volume was within normal limits.

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Figure 1: Sagittal Ultrasound Image of the Lumbosacral Spine Showing Cystic Lesion Arising from the Tip of the Distal Sacrum 4.5x3.8 cm. The Cyst is Anechoic with Thin Wall and Contains Second Smaller Thin Walled Cyst 1.2 cm



Figure 2: Transverse Ultrasound Image through the Cystic Lesion Demonstrating “Cyst within a Cyst” Appearance

A live male infant was delivered at 39 weeks gestational age by elective Cesarean section for what was thought at that time to be a sacrococcygeal teratoma. On clinical examination, there was a 9 x 9 cm soft, fluctuant, skin covered mass along the sacral area. There was no evidence of lower extremity neurological deficit or fecal incontinence.

Postnatal MRI of the brain and spine confirmed the presence of 8 x 3 cm elongated, skin covered, cystic mass dorsal to the level of the sacrum, see figure 3. The fluid in the cyst was similar to CSF on T1WI and T2WI's. The lesion was composed of larger outer cyst and smaller inner cyst. The smaller inner cyst was continuous with a dilated central canal of the spinal cord. The outer cyst communicated with the subarachnoid space. The spinal cord was low in position and terminated into the upper part of the cystic lesion. Absence of the overlying posterior elements and partial sacral agenesis were also present. No fatty components were noted. Appearance was consistent with TMC. MRI of the brain was normal. Surgery was performed on the seventh day of life. Two cystic lesions were identified with the inner cyst containing CSF fluid and the outer cyst containing xanthochromic fluid. Both cysts were excised without complications.

Histologically, the lesion consisted of skin and adipose tissues with sheets of meningotheelial cells deep in the dermis and subcutis. Occasional glial cells were seen. Microscopic examination of the xanthochromic fluid from the outer cyst showed meningotheelial cell clusters and inflammatory cells with no evidence of atypia.

After 4 years follow-up, the child had no neurological deficits and had normal verbal communication with the parents.



Figure 3: Postnatal T1 WI Sagittal Image of the Spine Showing 8x3 cm Elongated, Skin Covered, Cystic Mass Dorsal to the Level of the Sacrum (Larger Outer Cyst Indicated by the Arrow and the Inner Cyst by an Asterisk)

DISCUSSION

Neural tube defects are common occurring congenital anomalies in 1:1,000 births³. Spinal dysraphism is classified as either open or closed, depending on whether the back mass is open to the air or covered by skin. TMC is an uncommon form of spinal dysraphism accounting for about 4% to 7% of lumbosacral skin covered back masses and is more commonly found in females^{4,5}. Literature search in MEDLINE revealed only 2 previous reported cases of antenatal ultrasound findings of TMC^{6,7}.

Clinically, the affected infant presents with a skin-covered, midline cystic mass over the lumbosacral area. TMC may present as an isolated finding as in our case but often they are associated with the OEIS complex which consists of omphalocele, exstrophy of the bladder, imperforate anus and sacral agenesis⁸. Isolated TMCs are usually not associated with neurological deficits. There is, however, a higher risk for neurological deficits when ventral wall defects are present⁹.

As the diagnosis of TMC is difficult on prenatal ultrasound, other cystic masses occurring in the lumbosacral area must be considered. The differential diagnosis of a cystic mass in the fetal lumbosacral area includes myelomeningocele, meningocele and cystic sacrococcygeal teratoma (SCT)^{6,7}. It is important to try and distinguish these lesions from each other as it may affect perinatal management and parental counseling. For example, misinterpretation of a terminal myelocystocele as a neurologically compromising myelomeningocele could lead to elective termination of the pregnancy¹⁰. In this case, the patient had Cesarean section because it was thought to be a sacrococcygeal teratoma (SCT).

If a cystic mass is found along the lumbosacral area on antenatal ultrasound, careful evaluation of the cyst walls, intracystic contents, fetal head and the presence or absence of spinal dysraphism may help to distinguish myelomeningocele, meningocele, cystic sacrococcygeal teratoma and TMC from each other^{6,10}. The cysts were thin, there were no solid components and the fetal brain was normal in the case we presented.

Myelomeningocele accounts for 98.8% of all open spinal defects. It consists of a neural placode elevated beyond the level of the skin by an enlarged CSF filled subarachnoid space¹. Antenatal ultrasound shows that the cystic lesion has a thick wall with open or absent skin and herniation of the spinal cord through the spinal bifida⁶. Because 99% of myelomeningoceles are associated with Chiari II malformations, evaluation of the fetal head might confirm the diagnosis¹¹. Chiari II malformation is a hindbrain defect in which the posterior fossa is small resulting in crowding of the normal structures. The cerebellum appears wrapped around the brainstem with the shape of a banana, hence the “banana sign”. In turn, the CSF filled cisterna magna is small (less than or equal to 2 mm) or obliterated¹². Hydrocephalus is also typically present. In our case, there was no Chiari II malformation and the cyst was covered by skin.

Meningocele is the least common spinal dysraphism. It is a closed form of dysraphism characterized by herniation of the meninges through the normal spaces between the vertebral bodies. As the neural tissues are not involved, neurological function is preserved, although there have been a few cases with tethered cord¹³. Antenatal ultrasound reveals thin walled cystic lesion corresponding to the herniated meninges with intact skin and no spinal cord in the herniated sac⁶. No spinal dysraphism is present which might be challenging to verify on prenatal ultrasound. Chiari II malformation is not present as this is only seen with open spinal defects. There was “cyst within a cyst” in our patient.

Recently, the “lasso sign” has been described as useful in the diagnosis of posterior meningoceles. The “lasso sign” consists of a small cystic lesion floating in the amniotic fluid attached to the lower spine by a very tiny stalk and no vascularity on Doppler examination¹⁴. In our case, the cyst was not floating in the amniotic fluid.

Sacrococcygeal teratomas (SCT) are usually solid or solid-cystic; however, 15% of SCTs are purely cystic with no teratomatous structures and therefore, must be considered in the differential diagnosis^{10,15}. The cyst wall may be thin with no internal echoes or multilocular¹⁵. SCTs are not neural tube defects and therefore, there is no associated spinal dysraphism¹⁰. Our case was thought to be a sacrococcygeal teratoma (SCT).

TMC typically consists of a thin walled cyst covered by skin. The spinal cord remains within the spinal canal with herniation of dilated central canal and the meningeal sac through a boney spinal defect⁶. The cyst may have a funnel shape or a “cyst within a cyst” appearance^{6,16}. In our case, a thin walled lumbosacral cyst with a “cyst within a cyst” appearance was observed. The inner cyst corresponds to the cystic dilation of the caudal end of the spinal cord and is lined by ependyma and the outer cyst corresponds to the skin covered arachnoid-lined meningocele¹⁶. This sign was first described as an antenatal ultrasound finding for a non-terminal myelocystocele¹⁶. This is the first case that we know of in which this sign has been described for a terminal myelocystocele. Non-terminal myelocystoceles occur in the cervical and thoracic areas. While non-terminal myelocystoceles have different embryological origin and prognosis; they are similar to TMCs in consisting of focal hydromyelia of the cord protruding into a meningocele sac^{16,17}.

Ultrafast MRI might be helpful for the work up of congenital anomalies; unfortunately, Ultrafast MRI was not available in SMC. Ultrafast MRI allows for better delineation of

the fetal anatomy and has been shown to be useful in the prenatal work up of cystic back masses¹⁰.

CONCLUSION

We presented a case with terminal myelocystocele diagnosed on antenatal ultrasound which showed a “cyst within a cyst”.

TMC is an uncommon form of closed spinal dysraphism which presents as a cystic lesion along the distal spine. Careful evaluation of the cystic lesion for wall thickness, intracystic contents, presence or absence of spinal dysraphism and evaluation of the fetal head is useful to distinguish TMC from other cystic lesions occurring in the lumbosacral area. When a “cyst within a cyst” sign is found on antenatal ultrasound, TMC should be strongly considered.

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