PHILIPS Ultrasound Clinical case study

Limited dorsal myeloschisis, a challenging differential diagnosis from myelomeningocele

High-frequency transducers

Category

OB spinal assessment

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"Distinguishing limited dorsal myeloschisis from myelomeningocele is important, as the two conditions have very different prognoses."

Pascale Bach-Segura, MD, Radiologist

Overview

Limited dorsal myeloschisis is a distinctive form of spinal dysraphism characterized by a focal midline neural tube defect associated with tethering of the dorsal spinal cord to the overlying skin. Prenatal diagnosis is rare, and yet distinguishing limited dorsal myeloschisis from myelomeningocele is important since the two conditions have very different prognoses. High-resolution scanning available on the EPIQ 7 ultrasound system helps clinicians visualize structures in order to meet this challenge.

Patient history

A 27-year-old primigravida was referred to our center at 23 weeks' gestation to confirm the lumbar myelomeningocele diagnosis suspected on a screening sonogram.

The EPIQ ultrasound system features a range of transducers for high-resolution scanning to meet the challenges of today's most demanding Ob/Gyn practices.

Findings

A lumbosacral meningocele of $15 \times 15 \times 15$ mm was confirmed, but, certain aspects were inconsistent with the diagnosis of myelomeningocele. There was evidence of mild bilateral ventriculomegaly of 12 mm, but, no Arnold-Chiari type II malformation detected

Postnatal examination confirmed a lumbosacral, midline, fluid-filled mass at the level of L5 S1, covered by a thin layer of dysplasic skin, which was not a myelomeningocele but, a limited dorsal myeloshisis.



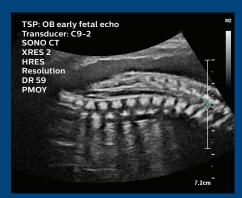


Figure 1 Middle spine sagittal view. Note the cutaneous defect from L5 to the distal sacral area. The spinal cord is low at the L5 level but, is within the spinal canal.

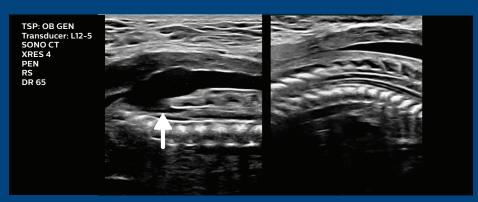


Figure 2 Middle spine sagittal view with high-frequency transducer. The high-resolution Philips L12-5 transducer allows visualization of the spinal cord despite the thick maternal lining. The filum terminale is also seen in the spinal canal (white arrow).

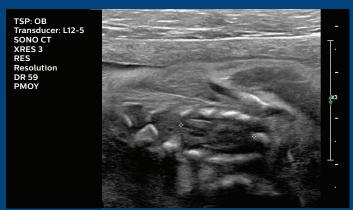


Figure 3 Osseous defect on a lumbosacral coronal view.

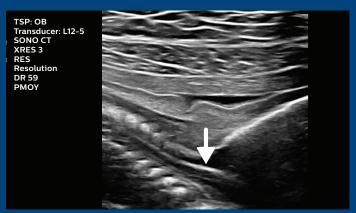


Figure 5 Posterior fossea, sagittal scan. The cerebellum was not displaced inferiorly and there is no evidence of an Arnold-Chiari type II malformation. The cisterna magna is well identified (white arrow).



Figure 4 Osseous defect also well-seen on surface 3D.

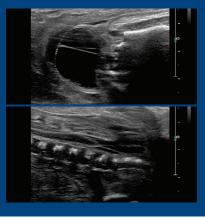


Figure 6 Herniated meninges through posterior vertebral arches defect. Note the linear echogenic stalk within the meningocele.

Conclusion

Despite the markedly abnormal prenatal appearance of limited dorsal myeloschisis with a large meningocele and a tethered fibroneural stalk, it is usually associated with a favorable prognosis. However, it is important to look for associated anomalies, in particular ventriculomegaly and other subtle anomalies of the central nervous system that may negatively affect the prognosis.

Figure 7 Although the meninges herniate through a defect in the posterior vertebral arches, the spinal cord and roots reside within the spinal canal. Two neatly circumscribed lateral thin stalks are quite visible in the meningocele, linking the underlying distal spinal cord to the meningocele dome.



References

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- 2 Pang D, Zovickian J, Wong, ST, Hou YJ, Moes, GS. Limited dorsal myeloschisis: a not-so-rare form of primary neurulation defect. Childs Nerv Syst. 2013;29:1459–1484.

Results from case studies are not predictive of results in other cases. Results in other cases may vary.

