Case Study: Diagnosis of Diastematomyelia

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These images from a routine ultrasound examination of a 20-week fetus reveal a rare abnormality of the fetal spine.

History and symptoms: A 27-year-old woman underwent obstetric ultrasound imaging at 20 weeks' gestation. She had a history of normal menstrual cycles prior to the pregnancy and has reported no vaginal bleeding since becoming pregnant. She had no history of clinically significant illnesses during the current pregnancy.

Clinical examination: The patient’s blood pressure was within normal limits (122/80 mm Hg), as were her other vital signs. Chest examination did not reveal any abnormalities. The abdominal examination findings revealed a fundal height that corresponded to a gestational age of 20 weeks.

Imaging studies: This routine transabdominal obstetric sonography was performed to exclude any congenital fetal anomalies. The patient had not undergone any sonographic study prior to this scan. The images seen in Figure 1, Figure 2, and Figure 3 show the coronal section of the fetal spine and were taken during this routine sonographic examination.

Sonographic Findings

In Figures 1, 2, and 3, the volume of amniotic fluid appears normal. The fetal abdomen also shows no significant abnormality. The placenta is fundic and posterior. No retroplacental hematoma is seen.

The most prominent anatomy in these images is the fetal spine. The fetal head is to the left of the images, and the fetal lower limbs are to the right of the images. The upper cervical and thoracic spines appear normal. However, there is an abnormality within the lower thoracic vertebrae that appears as an echogenic lesion within the spinal canal. It looks linear and extends within the mid-portion of the spinal canal. We cannot further distinguish the abnormality in this region. What are the diagnostic possibilities based on these images, at this stage?

Shall we call this some form of spinal dysraphism or neural closure defect? The most common anomalies that arise are tethered spinal cord, thickened filum terminale, neurenteric cyst, meningocele, and dermal sinus tract. Also, there are the remote possibilities of diplomyelia and diastematomyelia. Let us examine each of these possibilities.

Meningocele is characterized by spina bifida and herniation of the meninges through a spinal defect, but that does not seem to be the case here. To be sure, let's look at Figure 4, Figure 5, and Figure 6, which are axial images of the fetal spine.

After studying the aforementioned images, there is no obvious herniation of the meninges through a dorsal or ventral defect in the spine. However, there is an obvious anomaly visible in the images of the axial section of the thoracic vertebra, which was also visible in the images of the coronal section of the thoracic vertebrae. The anomaly is some form of division of the spinal canal.

Could this be hydromyelia, which is characterized by dilation of the central canal of the spinal cord? Or is it diplomyelia, which is division of the cord into two complete halves? Diplomyelia is possible, since there is a division of the cord in the region of the affected thoracic spine.

True diplomyelia is characterized by near-total division of the spinal cord, which is not the case here. The division is localized to the affected lower thoracic spines. In addition, there is a vertical echogenic, possibly bony, septum within the affected thoracic vertebrae. This finding suggests the possibility of diastematomyelia. This condition is seen as a partial or complete bony septum or fibrous septum that divides the spinal canal (localized to affected spine), with partial division of the spinal cord in the affected parts only.

To review, the findings in this fetus include the following:
- Midline bony spur or septum in the lower thoracic spine.
- Normal central canal of the spinal cord.
- No significant dilatation of cerebral ventricles.
- Division of the spinal cord localized to the affected thoracic spines.
- Normal spinal cord above and below the affected vertebrae.

The most common anomaly associated with these findings becomes our final diagnosis, which is
Diastematomyelia is a very rare anomaly characterized by clefting of the spinal cord due to a partial or complete bony or fibrous septum within the spinal canal. This results in localized division of the spinal cord, which roots into two parts on either side of the septum. This division may affect one or more vertebrae and usually involves the lower thoracic or upper lumbar spines. However, any part of the spine may be involved. It is said to be caused by separate closure of two hemicords during early gestational stage. Among the associated anomalies are club feet, kyphosis, and scoliosis.

**Prognosis**

The prognosis in this fetus is quite good given the absence of any associated anomalies and the affected region being localized to just two thoracic vertebrae. Surgical removal of the spinal bony septum will usually result in excellent recovery, although some cases may show limited neurological deficits.

References:
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