CASE REPORT

MR Imaging of Fetal Omphalocele: A Case Report

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We report a case of a fetal omphalocele in which the internal structure of the hernia sac was imaged in detail by magnetic resonance (MR) imaging. The hernia sac consisted of amnion, Wharton jelly, and externally, peritoneum. The umbilical vascular channels (2 arteries, one vein) were surrounded by Wharton jelly. Use of MR imaging facilitates a more accurate evaluation of fetal omphalocele with regard to hernial sac, intestinal structure, and differential diagnosis.

Keywords: *MRI, omphalocele, prenatal*

Introduction

An omphalocele is a defect in the fetal abdominal wall¹ that is usually detected on abdominal ultrasound for screening.^{2–5} Although magnetic resonance (MR) imaging clearly delineates the hernia sac of a fetal omphalocele and facilitates differential diagnosis,^{6,7} its use in imaging the hernia's internal structure has not been described. We report a case of an omphalocele and detail the internal structure of the hernia sac.

Case Report

At 22 weeks' gestation, abdominal ultrasound of the fetus of a 22-year-old woman revealed a cyst-shaped echo in the fetal flank. At 23 weeks' gestation, after providing written informed consent, the woman underwent MR imaging for diagnosis of the abdominal tumor and examination of other congenital anomalies. MR study was performed on a 1.5-tesla superconducting system (Signa; GE Medical Systems, Milwaukee, Wisconsin, USA). The parameters of T₂-weighted imaging for single-shot fast spin-echo (SSFSE) sequence were: repetition time (TR), 14000 to 29000 ms; echo time (TE), 183 ms; number of excitations (NEX), 0.5; bandwidth, 31.3 kHz; field of view (FOV), 24 cm; slice thickness/gap, 6.0/1.5 mm; matrix size, 256 × 192. For T₁-weighted imaging for fast multi-planar spoiled GRASS (FMPSPGR) sequence, the parameters were: TR, 150 ms; TE, 4.2 ms; flip angle (FA), 90°;

NEX, 1.0; bandwidth, 31.3 kHz; FOV, 24 cm; slice thickness, 6 mm; and matrix size, 256 × 128. T₂-weighted sagittal images (Fig. 1) showed a cystic configuration (hernia sac) protruding into the amniotic sac from the fetal abdominal midline and the base of the umbilical cord. Bowel with high and low signals in T₁- and T₂-weighted images was present within the cystic structure and contiguous with the abdominal cavity; furthermore, adjacent to the bowel, flow-voids of the 2 umbilical arteries following from the pelvic area were thin and one umbilical vein leading to the liver was thick. The wall of the hernia sac was a dual structure composed of concentric circle-shaped, double, thin walls. The signal of the space between the double walls resembled that of water, low in the T₁-weighted image and high in the T₂-weighted image. The umbilical arteries and vein coursed between the double walls; distal to the hernia sac, they were positioned in the center of the umbilical cord. The omphalocele was vigorously followed throughout the pregnancy, and the patient delivered via cesarean section at 34 weeks' gestation. Abscission of the hernia sac and a choke operation of the hernia orifice were performed just after birth.

Discussion

An omphalocele is a defect in the fetal abdominal wall that occurs in 0.8 to 3.9 cases per 10,000 births.^{1,8} The defect arises in the midline, and the umbilical cord base contains a hernia sac that consists of amnion, Wharton jelly, and externally, peritoneum. The Wharton jelly, which is gelatinous, white, semitransparent connective tissue of

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Fig. 1. Fetal magnetic resonance images of an omphalocele in a 23-week-old fetus (**a-c**: sagittal, **d-f**: axial T₂-weighted images; **g-i**: axial T₁-weighted images). Sagittal and axial T₂-weighted images show a cystic hernial sac protruding into the amniotic fluid from the fetal abdominal midline (white arrowheads). The wall of the hernia sac is a dual structure composed of concentric circle-shaped, double, thin walls (curved black arrows). Flow-voids of the 2 umbilical arteries, which are thin and follow from the pelvic region (black arrows), and one umbilical vein, which is thick and leads to the liver (black arrowheads), traverse the watery space (Wharton jelly) between the double walls. Axial T₁-weighted images show bowel with high and low signals within the hernia sac that are contiguous with the abdominal cavity.

mesodermal origin, contains the 2 arteries and one vein that make up the umbilical vascular channels. Abdominal ultrasonography for screening frequently detects an omphalocele. Gastroschisis, an abdominal wall defect that occurs in the same fetal period, is an important differential diagnosis. Since the 1980s, it has been held that the confirmation of a hernia sac was important for evaluating a structure eviscerated from an abdominal wall defect;²⁻⁴ however, ultrasound has not been able to provide a

detailed image of the configuration of these defects.⁵ In contrast to ultrasound, MR imaging is not suitable for fetal screening but is useful for detailed imaging when an abdominal wall defect is detected by ultrasound, and MR imaging aids differential diagnosis.^{6,7} In addition, chromosomal and congenital anomalies are more frequent for an omphalocele than for a gastroschisis. Compared to abdominal ultrasound, MR imaging is useful for prenatal diagnosis as well as differential diagnosis

in evaluating factors such as size of the abdominal wall defect, quantity of extruded bowel, presence of intestinal atresia, and presence of other congenital anomalies. Umbilical arteries and vein are clearly distinguished and chased continually from a fetus to umbilical cord. In this case, extruded bowel with high and low signals in T₁- and T₂-weighted images in the hernial sac indicates accumulation of meconium.⁹⁻¹¹ The high signal of a T₁-weighted image is useful in visualizing meconium, and at 23 weeks' gestation, extruded bowel is assumed to be small intestine. Liver herniation in the omphalocele is common, but was not recognized in our case. MR imaging also easily depicts liver herniation. The high signal of a T₂-weighted image in a fetal MR image is particularly useful in assessing a fetus suspended in amniotic fluid. Three-direction imaging via SSFSE is used widely; its advantages are short imaging time, high image quality, and good tissue contrast. In this case, it facilitated visualization of internal fetal structures, identification of the umbilical arteries and vein with flow-void, and identification of the intestine in the amniotic fluid. Furthermore, we could confirm the dual structure of the hernia sac and Wharton jelly surrounding the umbilical arteries and vein. In this case, Wharton jelly was surrounded by a thin and uniform amnion and peritoneum; the signal of its content resembled that of water, low in the T₁-weighted image and high in the T₂-weighted image. Wharton jelly consists of gelatinous material, but its specific MR imaging signal was not indicated in this case. The Wharton jelly surrounded the umbilical arteries and vein. Confirmation of a hernia sac via MR imaging has been stressed as an important factor in the differential diagnosis of an abdominal wall defect; however, prior to this report, the detailed structural imaging of the hernia sac and the interior signal was not described. The image quality of current MR imaging equipment permits confirmation of the hernia sac and structural detail of an omphalocele and allows

more accurate diagnosis. These MR imaging studies should be carefully conducted.

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