CASE REPORT

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SURPRISING IDENTIFICATION OF MECKEL'S DIVERTICULUM WITHIN A FETAL OMPHALOCELE DURING PRENATAL CARE

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ABSTRACT

Omphalocele is a rare congenital abdominal wall defect that results in herniation of bowels, stomach, and even liver for inside the umbilical cord. Herein we present a case of a Meckel's diverticulum (MD) which was unexpectedly identified inside an omphalocele of a fetus. The mother was a 25-year-old pregnant woman referred due to a fetal omphalocele, which was first visualized at 16 weeks of gestation. On ultrasonography at 21 weeks and 6 days, there was an apparently abnormal circular-shaped image inside the umbilical cord, suggestive of an omphalocele. Fetal MRI confirmed this diagnosis and revealed that some small intestine segments, and portions of mesentery and transverse colon were in an extra-abdominal situation. Moreover, a hyperintense image in T1 and T2 sequences, measuring about 3.9 x 3.8 x 3.6cm, was observed inside the omphalocele, which was compatible with a MD. The child was born by cesarean section, at 40 weeks gestation, weighing 4,030g. The omphalocele was surgically closed, with resection of the MD followed by a terminoterminal anastomosis of the ileum, on the second day of life. The pathology was also compatible with the diagnosis of MD. This report highlights the importance of the proper determination of the omphalocele content still during the prenatal period, since this may present unexpected abnormalities, such as a MD, and thus modify the gestational management as well as birth planning and postnatal care.

KEYWORDS: MECKEL'S DIVERTICULUM; PRENATAL DIAGNOSIS; OMPHALOCELE; ULTRASOUND; MAGNETIC RESONANCE IMAGING

INTRODUCTION

Omphalocele is a rare congenital defect of the abdominal wall that results in the herniation of intestines, stomach, and sometimes even the liver, into the umbilical cord. It has been reported in 3.38 out of 10,000 pregnancies[1]. Although omphaloceles can occur as isolated anomalies, up to 70% of them are associated with other malformations or syndromes[2]. Their diagnosis can often be made prenatally, typically through ultrasound[1]. The presence of intestinal loops or protrusion of the liver and/or stomach into the umbilical cord after 11 weeks is considered non-physiological. After the diagnostic confirmation, the herniated contents should be evaluated, and magnetic resonance imaging (MRI) can be used for better visualization of anatomical details[2].

Here we present a case of Meckel's diverticulum (MD) unexpectedly identified within a fetal omphalocele.

CASE REPORT

A 25-year-old woman in her first pregnancy was referred for evaluation due to a fetal omphalocele visualized at [21] weeks. She reported smoking (about 5 cigarettes/day) and alcohol consumption in the first month of pregnancy. Additionally, she experienced frequent vaginal bleeding from the 2nd to the 4th month, some of them in large amounts, along with contractions in the third month, requiring bed rest. The husband was a 33-year-old, healthy, non-consanguineous man. There was no family history of congenital defects or genetic diseases.

The image of the omphalocele was first visualized at [16] weeks of gestation. Previous ultrasound examinations at 8 and 11 weeks had not described this finding. At [21] weeks and 6 days of gestation, an apparently abnormal circular-shaped image was observed within the umbilical cord (Figures 1A and 1B). However, at 30 and 34 weeks, only intestinal loops were seen inside the omphalocele. In the last examination at 37 weeks, the omphalocele measured 5.4 x 5.3 x 4.6 cm. Subsequent fetal MRI revealed the defect in the closure of the anterior abdominal wall, located in the umbilical region, apparently covered by a membrane, measuring about 1.7 cm. These findings were consistent with an omphalocele. In addition, there were some segments of the small intestine, along with a small portion of the mesentery and a segment of the transverse colon, in an extrabdominal position. An asymptomatic and hyperintense image was observed on T1 and T2 sequences, measuring about 3.9 x 3.8 x 3.6 cm, indicative of a Meckel's diverticulum (MD) located within the

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omphalocele (Figures 1C and 1D). The fetal karyotype (46, XY) and Doppler echocardiography were normal.

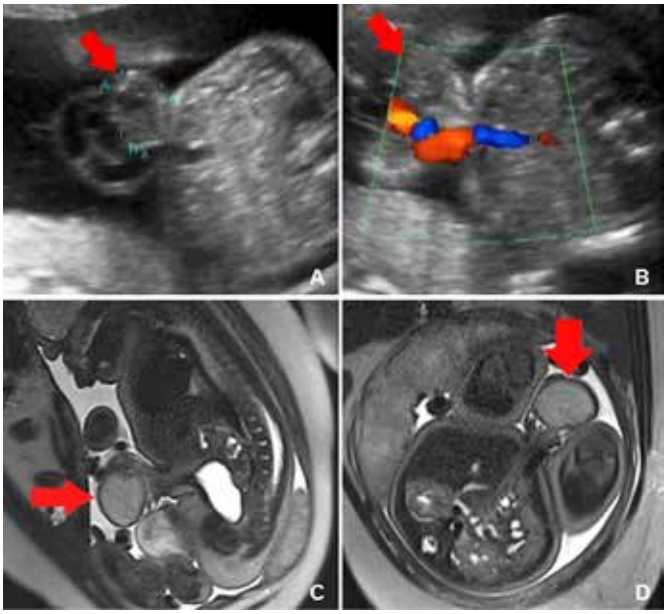

Figure 1. Fetal ultrasound at 21 weeks and 6 days showing the omphalocele with an apparently abnormal circular-shaped image within the umbilical cord (red arrows) (Fig. 1A and 1B). Fetal MRI revealing the omphalocele. There was a hyperintense image inside it, on T1 and T2 sequences, consistent with a Meckel's diverticulum (red arrows) (Fig. 1C and 1D).

The child was born via cesarean section at 40 weeks of gestation, weighing 4,030 g, and had Apgar scores of 10 at the first and fifth minutes. The child underwent omphalocele closure surgery with resection of the Meckel's diverticulum on the second day of life (Figures 2A and 2B). An ileal anastomosis was performed 18 cm from the ileocecal valve (Figure 2C). The pathological examination of the lesion confirmed the diagnosis of Meckel's diverticulum.

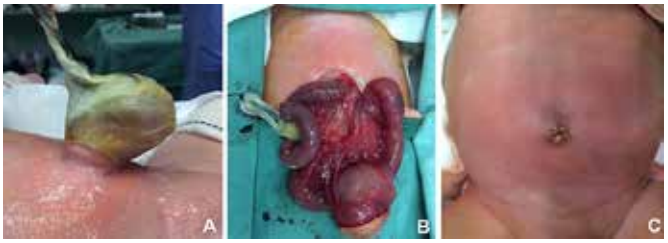

Figure 2. Postnatal appearance of the omphalocele, observed immediately after birth and before surgery (Fig. 2A). In Figure 2B, the contents of the omphalocele, with a focus on the Meckel's diverticulum (MD), can be seen. Figure 2C shows the final appearance of the patient's abdomen after surgery.

DISCUSSION

Meckel's diverticulum (MD) is a congenital anomaly characterized by a true diverticulum that involves all layers of the intestine and is part of the spectrum of anomalies resulting from incomplete closure of the omphalomesenteric duct, which develops during the 6th week of embryogenesis. Its prevalence in the general population varies between 0.3% and 2.9%, with a predominance in male patients in a ratio of approximately 1.5 to 4 times more cases in males than in females.

Meckel's diverticulum (MD) can be located between 7-200 cm proximal to the ileocecal valve (with an average of 52.4 cm) and may have a length ranging from 0.4-11 cm, and a diameter of 0.3-7 cm. Ectopic gastric and pancreatic tissues have been described in patients with MD, and these findings are related to symptomatic cases, primarily due to the occurrence of bleeding[3]. Additionally, MD has been reported in association with omphalocele[4], as observed in our patient.

Despite the frequency of Meckel's diverticulum (MD) in the general population and it being the most common congenital anomaly of the gastrointestinal tract (2-3% of the population), its prenatal diagnosis, as reported in the present case, is considered rare[4,5]. This may occur because ultrasound, although widely used for screening during pregnancy, may not be able to adequately distinguish the structures present inside the umbilical cord. Additionally, MD can be associated with other umbilical cord anomalies, including omphalocele4, as observed in our case.

In fetal ultrasound evaluation, MD appears as an ovoid, anechoic structure containing fluid inside, with the appearance of a wall with multiple layers. Doppler imaging does not show flow inside it. The echogenicity of the mass may increase due to debris, becoming hyperechoic in mid-pregnancy and isoechogenic at term due to the presence of meconium inside the intestinal loops. This can make its visualization challenging and, thus, prevent the diagnosis of MD[4-6].

Regarding the differential diagnosis of MD, one should consider cysts and pseudocysts inside the umbilical cord, as well as intestinal duplication, mesenteric cyst, choledochal cyst, and urachal abnormalities, especially in the early second trimester of pregnancy[6,7]. Other relevant differential diagnoses include intestinal obstruction and ovarian cyst, particularly after the twentieth week of gestation[6].

Fetal MRI can be an important complementary exam to ultrasound in cases of omphalocele, as it allows for better visualization of its content[8]. In our literature review, we found a significant shortage of descriptions of MD evaluated through fetal MRI5. The normal fetal gastrointestinal tract typically exhibits the following findings: after 24 weeks of gestation, the colon and rectum appear hyperintense on T1-weighted images and hypointense or with intermediate signal intensity on T2-weighted images (due to the presence of meconium); the jejunum is hyperintense on T2 and hypointense on T1, mainly due to its liquid content, and is usually visualized through T2-weighted images in coronal planes. Normally, duplicated cysts show a hypersignal, similar to fluid, on T2-weighted images and a hyposignal on T1 when visualized through fetal MRI. Meconium cyst, segmental ileal dilatation, and colonic pouch exhibit a hypersignal (meconium-like) on T1-weighted images associated with a

hyposignal on T25. In our case, we observed an unusual cystic image within the omphalocele, which appeared hyperintense on both T1 and T2, indicative of MD.

Despite the description that fetuses with prenatally diagnosed MD are more likely to be born prematurely and have low birth weight[9], our patient did not exhibit such findings. However, it is important to be vigilant for these potential manifestations in order to plan the delivery more effectively.

Most children born with MD are asymptomatic[10], as observed in our patient. However, some newborns may experience lower gastrointestinal bleeding, intestinal obstruction, and local inflammation shortly after birth, which can lead to perforation and, consequently, an increased risk of morbidity and mortality[3].

Hemorrhage in patients with MD can be associated with the presence of ectopic gastric tissue, which is reported in 24.2-71% of symptomatic individuals, who are typically young. The main complication reported, not only in pediatric patients but also in symptomatic adults, is intestinal obstruction (in 35.6-46.7% of cases); hemorrhage and inflammation are also common complications in patients of both age groups[3].

This report highlights the importance of accurately determining the content of the omphalocele, as it may be able to identify anomalies such as MD and modify gestational management. For example, in cases of MD, patients may benefit from birth planning, not only due to the risk of prematurity and low birth weight but also due to the risk of hemorrhage. Additionally, prenatal diagnosis aids in the development of the surgical plan to be carried out after birth. In cases of MD, it is also important that umbilical cord clamping after birth be done away from the base to avoid possible iatrogenic ileal atresia[5]. Supplementary examinations, such as fetal MRI, can also complement the ultrasound assessment and, consequently, help define the correct diagnosis, which implies proper management, follow-up, and treatment[2].

CONCLUSION

In summary, cases of omphalocele diagnosed in the prenatal period should be carefully investigated, as they often do not consist of isolated anomalies. The awareness of additional malformations, such as MD, identified through ultrasound, with or without supplementary examinations like MRI, is of great importance because it directly affects gestational management, birth planning, surgical approach, and postnatal care.

The subsequent steps after diagnosis are crucial to reduce infant morbidity and mortality rates, thereby modifying the prognosis.

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