Primary segmental volvulus of the ileum in a fetus: Case report

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Abstract

Primary intestinal volvulus in a fetus without an underlying cause has been rarely reported. We report a case of fetal primary segmental volvulus of the ileum. A 27-year-old woman was referred at 32 weeks of gestation for evaluation of decreased fetal movement. A fetal ultrasound detected a single, markedly-dilated bowel loop (coffee bean sign), and a fetal MRI revealed the ‘whirl-pool sign’. An emergency cesarean delivery and laparotomy revealed segmental volvulus of the ileum without any causative factors. Segmental resection resolved the condition without sequelae. Fetal primary segmental volvulus can occur without any underlying pathology, and rapid diagnosis and prompt intervention is mandatory for a good outcome.

Fetal intestinal volvulus is a rare but fatal condition that can cause fetal demise if unrecognized and timely intervention does not occur. Most of the cases of fetal intestinal volvulus develop secondary to various predisposing factors, but primary intestinal volvulus is extremely rare. We report a case of primary segmental volvulus of the ileum in a fetus without an underlying cause.

1. Case report

A 27-year-old multiparous woman was referred for evaluation of decreased fetal movement at 32 weeks of gestation. Her prenatal course had been uneventful with a routine antenatal care. Maternal history was unremarkable. Fetal heart rate monitoring revealed recurrent late decelerations (>50% of uterine contraction in 20-min window). A single, markedly-dilated bowel loop was noted on fetal ultrasound (‘coffee bean sign’), and color Doppler sonography showed no blood flow in the bowel loop (Fig. 1A), suggesting a segmental intestinal volvulus. Subsequent fetal magnetic resonance imaging (MRI) revealed the ‘whirlpool sign’ (Fig. 1B), suggesting a midgut volvulus. An emergency cesarean delivery was performed on the date of referral (32 weeks of gestation) with a presumptive diagnosis of fetal bowel strangulation; after routine neonatal resuscitation, infant underwent exploratory surgery on the first day of life; a segment of ileum was noted to have strangulated due to volvulus (Fig. 2). There were no gross anatomical bowel anomalies, such as malrotation, atresia, or mesenteric defect. A segmental bowel resection and end-to-end anastomosis was performed. Pathological examination revealed a transmural ischemic infarct without any diagnostic abnormalities. The postoperative course was uneventful and the patient was discharged 2 weeks postoperatively.

2. Discussion

Malrotation, intestinal atresia, or mesenteric defects have been known to be the predisposing factors for an intrauterine fetal volvulus [1–3]. However, in extremely rare instances, a primary intestinal volvulus has been reported in which no underlying anomalies could be identified. Chung et al. reported a case similar to ours, in which the gross anatomical abnormalities could not explain the occurrence of the fetal primary small bowel volvulus [4]. However, in frozen sections, they noted immature ganglion cells in the descending and sigmoid colon; thus, they performed an end ileostomy instead of a primary anastomosis. They concluded that the complication of meconium-related ileus due to immature ganglion cells or hypoperistalsis played a role in the etiology of the fetal primary small bowel volvulus. Similarly, Kurashige et al. suggested distension and increased peristalsis could form a primary loop and twist the bowel [5]. Although no macroscopic or microscopic abnormalities were found in our case, such abnormalities are not preventable and can only be elucidated after laparotomy.

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Therefore, we suggest that clinical efforts should be focused on the timely diagnosis and treatment when a primary volvulus is suspected in a fetus.

Due to the non-specific nature as well as the wide spectrum of presenting symptoms and signs of fetal intestinal volvulus, the condition is difficult to diagnosis at an early stage as well as to predict the survival of the affected fetus [6–8]. Polyhydramnios, fetal distress, decreased fetal movement, and abnormal fetal cardiotocographic findings are usually seen in most cases, and the ‘whirlpool sign’ or ‘coffee bean sign’ is the definitive ultrasound finding suggestive of fetal intestinal volvulus [8–10]. In our case, although a fetal MRI suggested a different diagnosis, the deteriorating fetal condition and fetal ultrasound findings were adequate for a diagnosis; moreover, they directly indicated a surgical emergency, and an immediate laparotomy resolved the problem with a good outcome. Therefore, we recommend early intervention in the presence of definitive sonographic findings and fetal distress. In addition, we recommend the omission of time-consuming diagnostic modalities such as a fetal MRI under such conditions. Prompt diagnosis followed by emergent delivery and laparotomy are key elements to treat fetal intestinal volvulus with a favorable outcome. For the treatment of a fetal intestinal volvulus, some surgeons perform a diverting enterostomy after resection [4,11]; however, an end-to-end anastomosis is currently preferred for most cases [12–15]. An enterostomy has been chosen in the presence of immature of ganglion cells in the distal colon [4] and/or a bowel perforation complicated by meconium peritonitis [11]. An enterostomy is an accepted treatment in the presence of diffuse bacterial peritonitis after volvulus perforation [16]; however, it is currently unresolved whether to obtain intraoperative frozen sections of bowel distal to the volvulus to determine the status of the ganglion cells. Therefore, to the best of our knowledge, an end-to-end anastomosis should always be attempted as a primary option in the absence of bowel perforation and diffuse peritonitis.

3. Conclusions

In summary, we successfully treated an extremely rare case of primary segmental volvulus of the ileum in a fetus, in which no macroscopic or microscopic abnormalities were found in the intestine. Fetal intestinal volvulus should always be considered when such prenatal symptoms occur. Early diagnosis, prompt cesarean delivery and laparotomy are of paramount importance for a good outcome.

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References


