A Case of a Preterm Infant with Prenatally Diagnosed Intrauterine Midgut Volvulus

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Intrauterine midgut volvulus is a rare and potentially life-threatening congenital disease that can lead to intestinal ischemia, sepsis and peritonitis caused by bowel perforation and meconium obstruction. Early detection and immediate treatment is crucial to improve the outcome. Herein, we report a preterm infant of 30 weeks of gestation with intrauterine midgut volvulus associated with meconium peritonitis who survived after cesarean delivery and immediate postnatal surgical intervention. The outcome of in-utero intestinal volvulus depends on optimal delivery timing and adequate postnatal treatment. Therefore, prompt multidisciplinary consultation and planning with obstetricians, neonatologists and pediatric surgeons is necessary to reduce the morbidity and mortality associated with fetal midgut volvulus, especially in the preterm period.

Key Words: Intrauterine midgut volvulus, Meconium peritonitis, Antenatal diagnosis, Preterm infant

Fetal volvulus is an abnormal twisting of the gastrointestinal tract that predisposes to midgut volvulus and can lead to ischemic bowel infarction, perforation and peritonitis. This condition is extremely rare and has high rates of morbidity and mortality.¹ Early detection and immediate postnatal management is imperative for a better outcome.

Most cases of volvulus are diagnosed in the postnatal period. However, an increasing number of reports of prenatal ultrasound diagnosis have been reported recently with high-resolution ultrasound technology imaging, especially with detection of the "whirlpool sign."² We report a case of fetal bowel dilation and intestinal volvulus presented by the whirlpool sign with meconium peritonitis at 30 weeks of gestation. The patient survived after delivery by cesarean section and appropriate surgical intervention.

Case report

A 29-year-old primigravida in the 29¹⁄₂ week of gestation was admitted to department of obstetrics and gynecology of Seoul National University Hospital because the fetal ultrasound showed a mildly dilated and echogenic bowel with maximal diameter of 32.4 mm without any ascites. Ultrasound showed normal fetal development, and there was no evidence of congenital infection.

Ultrasound at 29⁴⁄₄ weeks revealed a markedly dilated bowel loop with a maximum diameter of 36.3 mm and an amniotic fluid index of 260 mm. In addi-
volvulus with a typical whirlpool sign was observed in twisted bowel loops containing meconium (Fig. 1). Thereafter, induction of labor for delivery was attempted for two days, with failure to deliver. Ultrasound at 30 weeks of gestation showed bowel dilation with echogenic spots and ascites, raising the suspicion of intestinal perforation. This prompted the decision for emergency caesarean delivery.

A male newborn weighing 1,860 g was delivered with Apgar scores of 4, 6 and 7 at 1, 5 and 10 minutes. The baby initially did not cry, and his heart rate was below 100 beats per minute. Positive pressure ventilation (PPV) was applied, but respiratory distress remained with a distended and tense abdomen. Ecchymosis was observed on the abdominal skin with visible peristalsis (Fig. 2). An investigational work-up, including blood culture, chest x-ray, and arterial gas, was performed, and wide-spectrum intravenous antibiotics were started. Immediate ultrasonography of the abdomen confirmed the diagnosis of intestinal obstruction with perforation. An emergent laparotomy was carried out three hours after birth. Intraoperative findings showed a volvulus of the small bowel with meconium-stained turbid fluid occupying the abdomen and the twisted bowel segment 10 cm distal to the ligament of Treitz (Fig. 3). There was no intestinal malrotation, mesenteric defect or atresia. A 6 cm necrosis of the ileum was resected, and an ileostomy was performed. Macroscopic histopathology revealed a gangrenous segment of the small bowel measuring 6 cm in length and

![Fig. 1](image1). Fetal ultrasonography at 29+4 weeks of gestation showed the ‘whirlpool sign’ (arrow).

![Fig. 2](image2). At birth, the abdomen was distended, and the skin was discolored with visible peristalsis.

![Fig. 3](image3). Intra-operative finding showed a midgut volvulus with necrosis and perforation (arrow).
a central segment dilated to a maximum diameter of 3 cm. Microscopic histopathology included a few immature ganglion cells in the submucosa with an absence of mature ganglion cells.

The postoperative course was uneventful. Six weeks later, ileostomy closure was performed. Two weeks later, the patient was discharged with full enteral feeding and adequate weight gain.

**Discussion**

Fetal intestinal volvulus is rare, and only a few cases of prenatal diagnosis have been reported. So far, less than thirty cases of fetal midgut volvulus have been reported with only three cases in Korea.\(^3\)\(^-\)\(^11\) In two cases of them, the twisted bowel was observed on sonography, but no diagnostic doppler features were reported. This is the second case of prenatally diagnosed intrauterine volvulus with whirlpool in preterm infant sign in Korea.

Most cases of intestinal volvulus are associated with anomalies of intestinal rotation or congenital developmental defects, such as intestinal atresia, gastroschisis and omphalocele.\(^12\) At approximately 8 weeks of gestation, the midgut rotates an additional 180 degrees counterclockwise, fixing to the posterior wall. Arrest of development at any stage and impaired fixation leaves the bowel at a high risk for volvulus.\(^13\) However, the etiology of intestinal volvulus without malrotation is not well understood; rare predisposing causes have been suggested, such as Meckel’s diverticulum, duplication cyst and meconium plugs.\(^14\)

Sonographic findings of intestinal volvulus were first reported in 1983.\(^15\) The diagnosis of intrauterine midgut volvulus has become easier with high-resolution image technology. Ultrasound findings revealed the hallmark whirlpool sign in which the bowel and accompanying mesentery and vessel wrap around the main superior mesenteric artery.\(^16\) Moreover, polyhydramnios, intestinal dilation and ascites were also helpful in distinguishing bowel obstruction. A recent prospective study of neonates showed that the sensitivity and specificity of the whirlpool sign are as high as 89% and 92%.\(^17\) In our case, the pathognomonic whirlpool sign was detected prenatally, and the diagnosis of midgut volvulus was made before birth.

If the prenatal ultrasound findings show the typical image of the whirlpool sign, delivery should be considered. However, there has been no consensus regarding the timing of delivery in fetuses with a high suspicion of intestinal obstruction, especially in preterm infants. In present case, even though the sonographic abnormalities associated with volvulus are found at 29\(^{+4}\) weeks of gestation, the baby was delivered by caesarean section at 30 weeks of gestation after few days of induction of delivery. For the case was preterm at diagnosis, time for administration of prenatal dexamethasone was required, and there were no abnormal findings on biophysical monitoring and signs of intestinal perforation at the time of prenatal diagnosis. Emergency caesarean section and emergent pediatric surgery are available in our hospital. Careful biophysical monitoring of the fetus and screening of the amount of amniotic fluid and presence of ascites could be helpful.\(^5\) Prenatal emergencies can occur with decreased fetal movement and non-reactive readings on cardiotocography or signs of intestinal perforation. Survival of the affected neonates is influenced by the length of ischemic necrosis bowel, gestational age at birth and the ability to tolerate neonatal surgery.\(^18\)

In summary, the key for successful treatment of the volvulus depends on a high index of suspicion and
clinical skills. A team effort that includes an experienced obstetrician, a pediatric surgeon, a neonatologist and nutritional support is valuable. Herein, we report a case of an infant with intrauterine volvulus with typical findings, who was delivered with prenatal careful monitoring and treated immediately and appropriately after birth.

References

= 국문초록 =

산전 자궁 내 중장 염전 진단 받은 미숙아 1례

서울대학교 의과대학 소아과학전공실, 외과학전공실, 산부인과학전공실
우혜경, 이혜연, 김세연, 정영화, 신승한, 김현영, 김이경, 김한석, 전중환, 최중환

자궁 내 태아의 중장 염전은 매우 드물게 발생하며 장의 허혈, 패혈증, 복막염 등을 유발하여 태아 생존에 위협이 되기도 하므로, 조기에 진단과 즉각적인 치료가 중요한 절차이다. 본 증례에서는 재태주수 30주의 태아에서 태변성 복막염이 동반된 자궁 내 중장 염전이 발생하여 재원질환 분만 후에 즉각적으로 수술을 시행한 후 좋은 예후를 보였던 미숙아를 보고하였다. 자궁 내 장 염전이 발생한 경우 적절한 분만 시점의 결정과 분만 후 치료가 예후를 결정하는 중요한 요소로 알려져 있다. 그러므로 미숙아 분만이 예상될 경우 산과의, 신생아 진료의, 소아외과의의 즉각적인 다학제적인 접근을 통해 태아의 중장 염전으로 인한 이환과 사망률을 줄이려는 노력이 필요하다.

중심 단어: 자궁 내 중장 염전, 태변성 복막염, 산전 진단, 미숙아