CASE REPORT

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Intrauterine Volvulus

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ABSTRACT Although volvulus is mostly seen in infancy, rare cases of intrauterine volvulus have also been reported. Fetal ultrasonographic (USG) examination of a 32-year-old woman at the 24th gestational week, showed focal dilatation of the intestines with an increased wall echogenity of the dilated segments. At the subsequent visits USG revealed progressive dilatation of the intestines. The patient presented with preterm labor at the 32nd gestational week. Cullen's sign developed on newborn's abdominal skin over time. During the operation, there was a stiff, yellow-colored, fibrinous pouch, jejunal and ileal atretic ends (25 cm of jejunum was remaining). We started total parenteral nutrition (TPN) because of short bowel syndrome. The patient died due to TPN-related sepsis at age 40 days. Timely diagnosis and treatment of intrauterine volvulus cases is very difficult because there are mostly non-specific signs, which have many differential diagnoses. This fact negatively affects mortality and morbidity for intrauterine volvulus cases.

Keywords: Intrauterine; volvulus; atresia; fetal; preterm labor

Intestinal volvulus occurs when the intestines rotate around its mesenteric axis, which includes the superior mesenteric artery. The most important reason that increases mortality and morbidity in intestinal volvulus is the delay in diagnosis and treatment, therefore volvulus is a surgical emergency.[1] Although volvulus is mostly seen in infancy, rare cases of intrauterine volvulus have also been reported. Herein, we report the prenatal, postnatal and operative findings and course of a case with intrauterine volvulus.

CASE REPORT

A 32-year-old woman, (gravida 6, para 2, first trimester abortions 3) with an abnormal sonographic view of the fetal abdomen was referred to the perinatology clinic at the 24th gestational week. She was thyroidectomized because of thyroid cancer and use of levothyroxine. Ultrasonographic (USG) examination showed focal dilatation of the intestines with an increased wall echogenity of the dilated segments (Figure 1). Amniocentesis was offered to exclude karyotype abnormalities, but the couple refused this invasive test. Sonographic reevaluation was performed 3 weeks later; dilatation of the intestines was progressed, and cervical length was measured 27 millimeters (mm). The patient was consulted to a pediatric surgery clinic for differential diagnosis of intestinal atresia, Hirschsprung disease, or volvulus. Because there was no finding of fetal distress and no definitive diagnosis, waiting until the 32nd week of gestation was decided by considering the risk of prematurity complications, especially pulmonary complications. At the subsequent visits at 28 weeks and 5 days and at 31 weeks and 1 day of the gestation, the patient reported decreased fetal movements; again, sonographic examination revealed progressive dilatation of the intestines with a maximum transverse diameter of the dilated segment 30 mm, polyhydramnios with a deepest vertical pocket of 93 mm, and cervical length of 20 mm (Figure 2, Figure 3).

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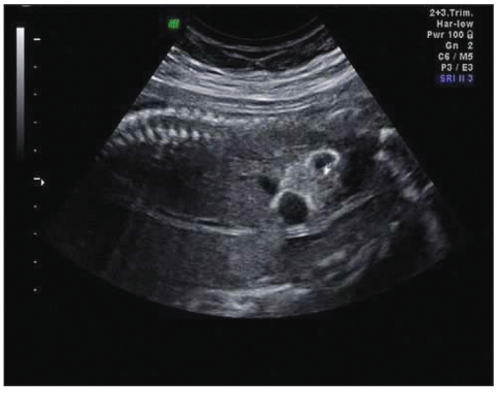

FIGURE 1: Antenatal ultrasonographic: focal dilatation of the intestines with an increased wall echogenity (24th week of gestation).

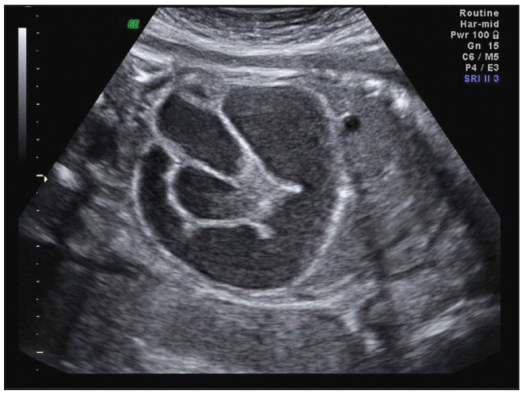

FIGURE 2: Antenatal ultrasonographic: dilated fetal intestines (28 weeks 5 days of gestation).

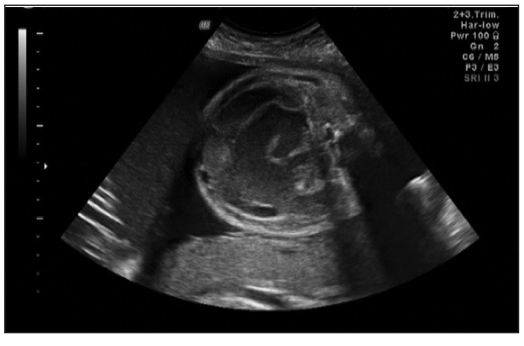

FIGURE 3: Antenatal ultrasonographic: dilated fetal intestines with suspected meconium cyst (31 weeks 1 day of gestation).

Considering the possibility of premature birth, single cure betamethasone treatment was given for fetal lung maturation at the 31st week of gestation. The patient presented with preterm labor at the 32nd gestational week, cesarean section was performed, and a female infant weighing 2,320 g was delivered.

The newborn was intubated because of pulmonary insufficiency. The APGAR scores at the 1st and 5th minutes were 6 and 7, respectively. At the physical examination, there was soft abdominal distention with normal color. A nasogastric tube was placed that drained the bilious content. A plain abdominal graph revealed air-fluid levels. Antibiotic treatment (ampicillin, gentamicin, metronidazole) was started. Postnatal USG was reported as "fluid distention of the stomach and intestines, dilated small intestines without abscess." The colon enema (2nd day) revealed an unused colon (Figure 4). As the intestinal atresia was suspected and abdominal examination was normal, we decided to wait recovery of the baby to a better condition to decrease anesthesia risks for operating for intestinal atresia.

Edema and bruising (Cullen's sign) developed on abdominal skin over time. On the 5th day of life, laparotomy was performed. During the operation, a cystic, stiff, yellow-colored, fibrinous pouch was opened, which was thought to be a meconium cyst (Figure 5). The colon was unused and intact, and the ileum and distal jejunum were not observed. The jejunal and ileal ends were atretic (Figure 6). There was 25 cm of jejunum remaining, and jejunostomy and cecostomy were performed. The pathological examination of the tip of the atretic jejunum was reported as "multifocal calcifications, accumulation of fibrin and necrotic changes on serosal surface." As the upper stoma (jejunostomy) did not permit enteral

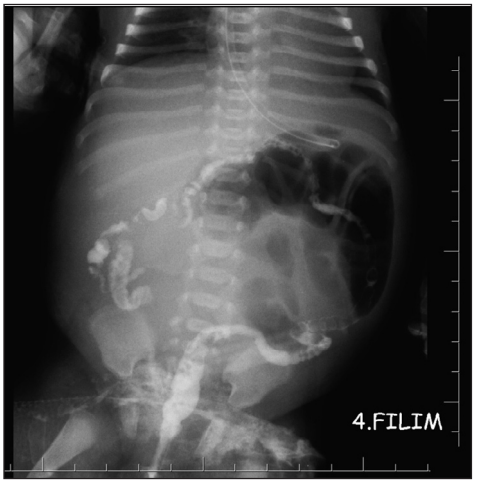

FIGURE 4: Contrast colon graphy: unused colon.

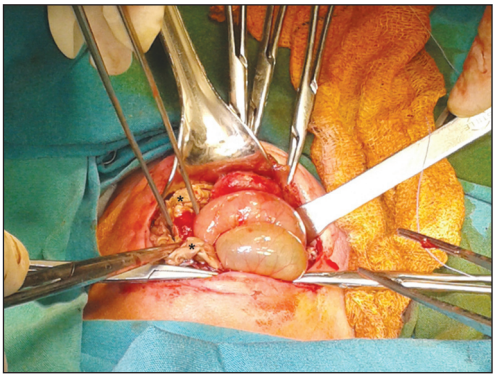

FIGURE 5: Perioperative photography: the wall of meconium cyst that was fibrinous, hard, and yellow colored (asterisks). Dilated proximal intestines were observed.

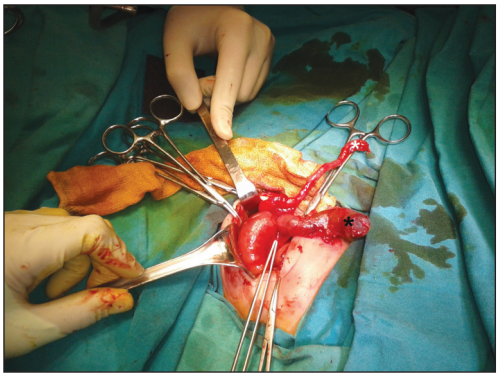

FIGURE 6: Atretic segments; both proximal atretic segment (black asterisk) and distal atretic segment (white asterisk) which were entering the meconium cyst.

feeding, we started total parenteral nutrition (TPN) because of short bowel syndrome. On the 35th day of the postnatal period, the patient's general condition deteriorated, and the patient was intubated. Thrombocytopenia and leucopenia developed. Antibiotics were changed to vancomycin and meropenem. The patient died due to TPN-related sepsis at age 40 days.

Informed consent was obtained from the family of the case.

DISCUSSION

Prenatal USG findings of intrauterine volvulus are dilated bowel loops, polyhydramnios, static abdominal mass, peritoneal calcifications, ascites, and increased abdominal circumference.[2-4] These findings point to a wide range of diagnoses, including intestinal atresia, segmental small bowel dilatations, cystic meconium peritonitis, Meckel's diverticulum, duplication cysts, cystic mass lesions (teratoma, lymphangioma, mesenteric cysts) and omphalomesenteric cysts.[5]

Definitive diagnosis of intrauterine intestinal volvulus can be made by observing "whirlpool or snail configuration" on prenatal ultrasonography.[1,2] The rare "coffee bean sign" also indicates volvulus. These 2 findings are very difficult to detect on prenatal ultrasound and are not always associated with volvulus.[4,6,7] The sensitivity and specificity of the whirlpool sign have been reported in 89% and 92% of intrauterine volvulus cases, respectively.[8] The higher incidence of non-specific findings, including many differential diagnoses, in intrauterine volvulus makes timely diagnosis and treatment difficult.[2,4,6,9-11]

When intrauterine volvulus occurs in the early weeks of pregnancy, it results in meconium peritonitis and/or intestinal atresia as a result of impaired intestinal blood flow.[9,12,13] In our case, there was atresia at the proximal and distal ends, absence of intestine between these sections and meconium cyst formation in between. Similar cases with meconium cysts have been reported by other authors.[2,3,10,14] In one case, the volvulated intestinal loop became a necrotic mass, and in 2 cases a cystic abdominal mass, in one of which the presence of necrotic intestinal remnants within the cyst have been reported.[2,10,14] The differential diagnosis of prenatal abdominal cystic masses includes antenatal volvulus with pseudocyst formation, cystic meconium peritonitis, and segmental intestinal dilatation of the small intestine.[11] In our case, we assumed that necrosis caused by volvulus that started at the 24th week of pregnancy resulted in atresia at the proximal and distal ends of the intestinal segments and a meconium cyst formed between them at 32nd week of pregnancy. Of course as we could not show the volvulated segment which occurred at 24th week of pregnancy directly, we assumed there was intrauterine volvulus according to the prenatal USG and operative findings.

In our patient, we had findings indicating that volvulus worsened at 32 weeks of gestation. We lost most of the small intestine (mid intestine) at that time, a meconium cyst formed and the remaining viable intestinal segments were very short. In our case, though that the mortality and morbidity risks of prematurity

were higher than the risks related to volvulus and the lack of a definite diagnosis prevented us from making an early intervention decision. Sciarrone et al., in one of the cases they reported, they decided to wait till normal-term delivery because they thought that delivery as soon as diagnosis was made (at 27 weeks of gestation) would result in a very premature baby without any improvement in prognosis.[5]

In conclusion timely diagnosis and treatment of intrauterine volvulus cases is very difficult because there are mostly non-specific signs, which have many differential diagnoses and specific symptoms of intrauterine volvulus is seen less. This fact negatively affects mortality and morbidity for intrauterine volvulus.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Nihal Şahin Uysal, Tuğba Acer Demir;* ***Design:*** *Nihal Şahin Uysal, Tuğba Acer Demir;* ***Control/Supervision:*** *Tuğba Acer Demir, Lütfi Hakan Güney;* ***Data Collection and/or Processing:*** *Nihal Şahin Uysal, Müge Sağnak Akıllı;* ***Analysis and/or Interpretation:*** *Tuğba Acer Demir, Müge Sağnak Akıllı;* ***Literature Review:*** *Tuğba Acer Demir;* ***Writing the Article:*** *Nihal Şahin Uysal, Tuğba Acer Demir, Müge Sağnak Akıllı;* ***Critical Review:*** *Nihal Şahin Uysal, Lütfi Hakan Güney.*

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