A neonate with intestinal volvulus without malrotation exhibiting early jaundice with a suspected fetal onset

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Intestinal volvulus without malrotation is a rare disease that causes volvulus of the small intestine despite normal intestinal rotation and fixation. We encountered a neonate with this disease who developed early jaundice and was suspected to have a fetal onset. This patient was characterized by early jaundice complicating intestinal volvulus without malrotation and is considered to have exhibited reduced fetal movement and early jaundice as a result of volvulus, necrosis, and hemorrhage of the small intestine in the fetal period. If abdominal distention accompanied by early jaundice is noted in a neonate, intestinal volvulus without malrotation and associated intraabdominal hemorrhage should be suspected and promptly treated.

Key words: intestinal volvulus, early jaundice, fetal onset.

Intestinal volvulus without malrotation is a rare disease that causes volvulus of the small intestine despite normal intestinal rotation and fixation. While it is observed at all ages from neonates to adults, it occurs most frequently in the neonatal period. The prognosis depends on early diagnosis and treatment, but, in neonates, it lacks characteristic findings and is difficult to diagnose. We encountered a neonate with intestinal volvulus without malrotation showing early jaundice and marked abdominal distention, suspected to have a fetal onset.

Case Report

The patient was a 0-day-old girl who had shown reduced fetal movement since the 35th week of gestation. Since a decrease in variability was noted on cardiotocography at 35 weeks and 6 days of gestation, the infant was born by emergency Caesarean section. The birth weight was 2,136 g, and the Apgar score was 9/10. Since retractive breathing and abdominal distention persisted at 1 hour after birth, the infant was transferred to our hospital.

On arrival, fever, tachypnea, and hypoxemia were noted. The infant was lethargic, the skin of the entire body was pale, and retractive breathing was observed. The abdomen was hard and distended, and no bowel sound was heard. Initial laboratory values revealed a hemoglobin level of 8.9 g/dl, elevated reticulocyte count of 10.7%, increased total serum bilirubin of 8.8 mg/dl, and elevated C-reactive protein of 1.7 mg/dl. Blood gas analysis was pH 7.293, pCO_2 42.8 mmHg, BE -5.3 mEq/L, and HCO_3 18.8 mEq/L. Both the baby and mother's blood type were A and Rh positive. Coomb's test was negative. Plain abdominal radiography 3 hours after birth showed a small volume of intestinal gas and gastric bubble enlargement. Abdominal ultrasonography performed 8 hours after birth showed a dilated bowel loops without bowel movement. No ascites or thinning of the intestinal wall was demonstrated. There was no significant finding in daily radiography and ultrasonography. Barium enema showed neither malrotation nor micro colon. Upper GI contrast could not be performed due to the poor general condition.

After admission, respiratory management was initiated. Gastric intubation was also conducted to reduce the intestinal pressure. While phototherapy was initiated for early jaundice, the total bilirubin level increased to 12.7 mg/dl 10 hours after birth, and exchange transfusion was performed. Abdominal distention and the amounts of bilious drainage from nasogastric tube (NGT) gradually increased. Discharge of a small amount of meconium was observed several times a day. On day 4 after birth, meandering of the intestine became grossly evident, and the abdominal wall had discolored to dark red. Since abdominal radiography also disclosed intestinal distention, and the amounts of bilious drainage from NGT increased, emergency laparotomy was performed. Intraoperative findings included a large amount of bloody ascites and volvulus with extensive necrosis extending from 50 cm distal to the ligaments of Treitz, 45 cm in length (Fig. 1A, B). The perforation was found in part of this area. The necrotic bowel was resected, and ileostomy was carried out. There was no malrotation because the ligament of Treitz and the cecum were in the normal position. The postoperative course was uneventful, enteral nutrition was initiated on day 15 after birth, the stoma was closed on day 42, and the infant was discharged on day 60.

Discussion

In reported neonatal cases of intestinal volvulus without malrotation, primary symptoms were vomiting, bloody diarrhea, and dark discoloration of the abdominal wall.[1] Our patient characteristically showed severe early jaundice, which had not been reported previously. Figure 2 shows the pathological profile of our patient. The small intestine is considered to have been rotated in the fetal period, probably at 35th week of gestation, when a decrease in fetal movement was perceived, leading to a non-reassuring fetal status (NRFS) and abdominal symptoms. Subsequent intestinal necrosis, perforation, and hemorrhage are considered to have caused early jaundice, anemia, peritonitis, and fetal inflammatory response syndrome and resulted in circulatory and respiratory failure. Early jaundice is probably secondary to intraabdominal bleeding in the fetal period.

Patients with intestinal volvulus without malrotation with a fetal onset are classified into mild and severe volvulus groups.[2] In the mild volvulus group, since volvulus persists over a relatively long time, an abdominal mass with polyhydramnios is observed by fetal ultrasonography, but the state of infants after birth is relatively favorable. In the severe volvulus group, volvulus occurs with a sudden onset, causes a decrease in fetal movement and diminished variability on cardiotocography, and requires emergency Caesarean section. Abdominal symptoms often appear from immediately after birth, and the general condition is also poor. Our patient is considered to belong to the severe volvulus group based on the clinical course.

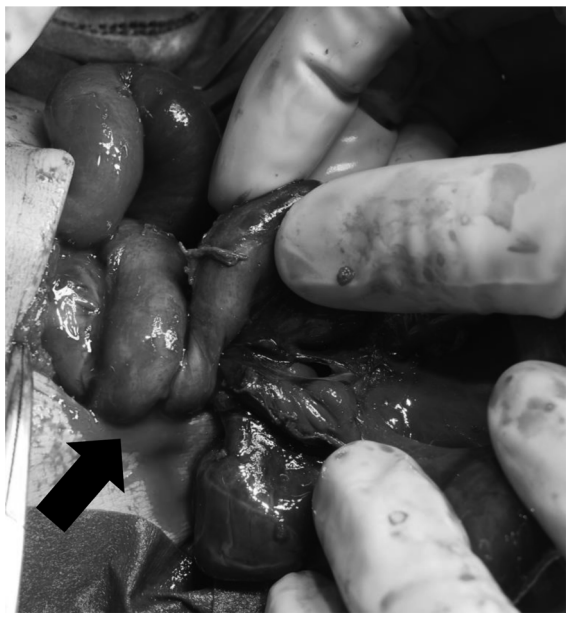

Fig. 1a. Intraoperative photograph showed intestinal volvulus with necrosis. Mesenteric defect could not be identified due to marked damage.

Fig. 1b. Resected necrotic bowel.

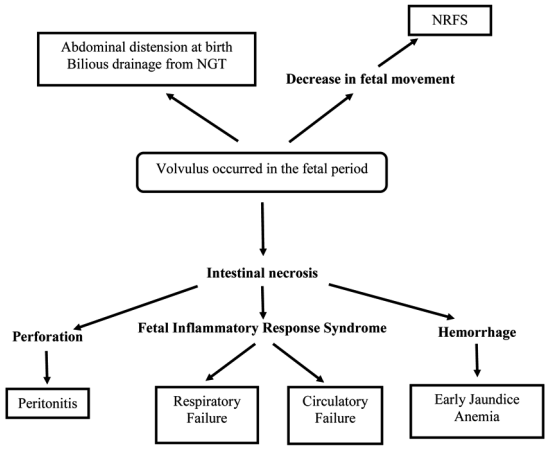

Fig. 2. Pathological profile of the case.

Volvulus of the small intestine has been suggested to be caused by rapid changes in the intraabdominal pressure due to the fetal body position and delivery, and intestinal distention and increased bowel movement associated with the initiation of nutrition after birth. Mesenteric defects, i.e., narrowness of the insertion of the root of the mesentery (basilar defects, in which the entire base of the mesentery is involved) and partial elongation of the long axis of the mesentery (segmental defects, in which only an isolated portion of the mesentery is affected), have also been suggested as anatomical factors (Fig. 3)[3] . Basilar defects have been reported to be difficult to discriminate from intestinal volvulus secondary to malrotation, and segmental defects have been reported to be difficult to discriminate from intestinal atresia. Such anatomical abnormalities are likely to be involved in fetal-onset intestinal volvulus without malrotation. In our patient, the cause of the disease could not be identified due to marked damage caused by necrosis, but the basilar type is considered likely from the clinical course.

In intestinal volvulus without malrotation, in which the small intestine alone is partially rotated and ischemic necrosis progresses rapidly, early diagnosis and treatment are more important than in intestinal volvulus with malrotation. However, the diagnosis may be delayed, as in our patient, because the findings on abdominal plain radiography or barium enema are non-specific.[4] While there have been reports that the whirl-pool sign[5] or coffee

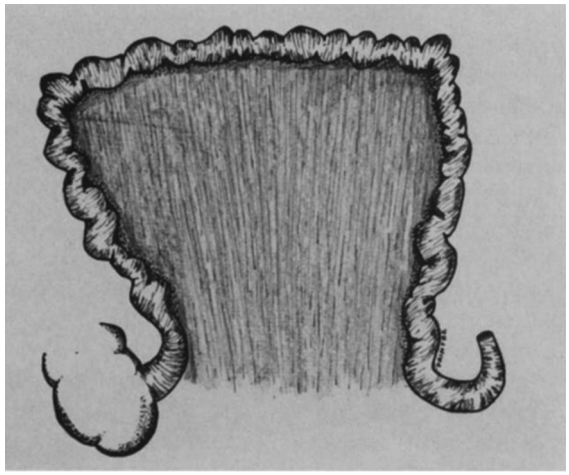

Fig.3. The types of mesenteric defects.
Fig. 3a. Normal, broad-based mesenteric root.

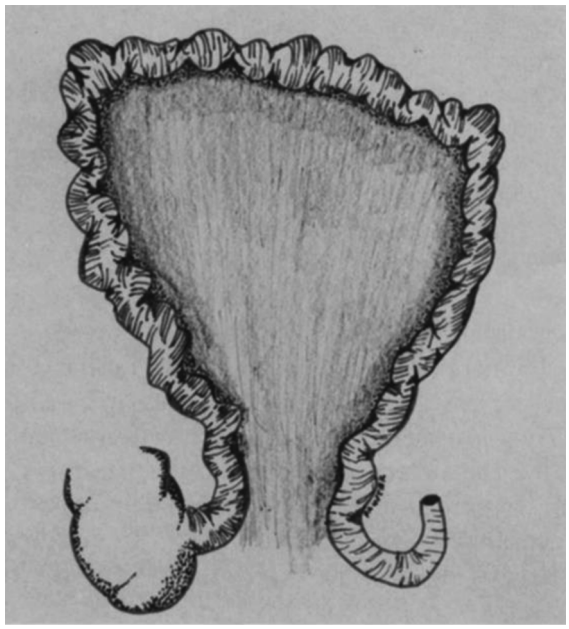

Fig. 3b. Basilar defects. Entire base of the mesentery is abnormally short.

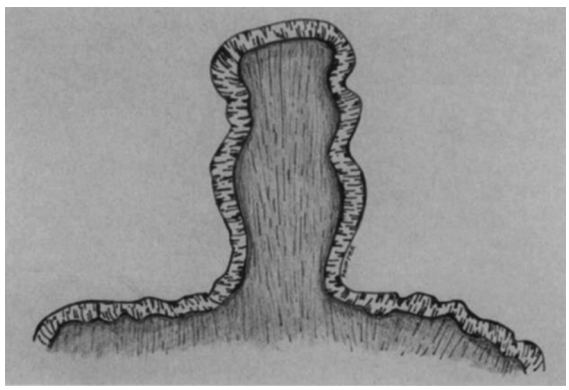

Fig. 3c. Segmental defects. A part of intestine has a small segment of mesentery with a narrow base.

bean sign[6] detected by fetal ultrasonography served as a clue to the diagnosis, they were not clear in our patient. There have also been reports that the whirl-sign detected by CT led to preoperative diagnoses[5], but CT could not be performed in our patient due to the poor general condition. If abdominal distention is noted at birth, intestinal volvulus should be suspected as a differential diagnosis even without malrotation, and early jaundice may be useful as a clue to an early diagnosis.

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