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Urgent surgical management of a prenatally diagnosed midgut volvulus with malrotation

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Abstract. – Midgut volvulus is a life-threatening condition that commonly presents during the first year of life rarely antenatally. Here we report successful urgent surgical management of an unusual case with malrotation exhibiting the sonographic findings of volvulus in utero that leads to premature birth. This 34-wk, 2700 g infant was born via spontaneous vaginal delivery. Prenatal ultrasound showed polyhydramnios and 30 x 40 mm cystic lesion showing whirlpool sign in abdomen that made us think midgut volvulus. The patient had distended abdomen with skin discoloration. An emergency surgical management was performed showing malrotation, volvulus and a 15 cm ischemic necrotic region of terminal ileum.

Key Words:
Midgut volvulus, Whirlpool sign, Intrauterine.

Introduction

Midgut volvulus is a life-threatening condition in which the small bowel or proximal colon twists around the superior mesenteric artery (SMA) and it commonly presents during the first year of life[1,2]. Prenatally diagnosed cases have been reported in the literature, 25% of them may be complicated due to ischemic necrosis[3-6]. The diagnosis of midgut volvulus in infants is facilitated by direct sonographic visualization of the twisted bowel loop but antenatal diagnosis is very difficult[7-9]. Here we report successful surgical management of an unusual case with malrotation exhibiting the sonographic findings of volvulus *in utero* that leads to premature birth.

Case Report

A 25-year-old pregnant woman was on follow-up in our Center during her pregnancy and no problem was reported up to 34th gestational week when she admitted to the Hospital because of preterm delivery. Prenatal ultrasonography performed just before the delivery revealed polyhydramnios and 30 × 40 mm cystic lesion with whirlpool sign in abdomen that made us think midgut volvulus. A 2700 g male infant was born on the same day with spontaneous vaginal route. Apgar scores were 5 and 7 after 1 and 5 minutes, respectively. Positive pressure ventilation was performed at the delivery room and afterwards transported to neonatal intensive care unit (NICU). Oxygen therapy was given with hood. Initial laboratory values included a white blood cell count of 73.000/mm^3, hematocrite 38%, a platelet count of 140.000/mm^3, peripheric blood smear revealed 80% normoblast, C-reactive protein level 39 mg/dl, Interleukin-6 level 220 pg/ml. Blood gas values were normal. A plain supine abdominal graphy did not demonstrate bowel gas, except in stomach. Empirical Penicillin G and netilmycine therapy were begun and oro-gastric decompression was performed. Acute surgical management was planned after the hemodynamic stabilization of the infant. The abdomen was distended markedly and there was a hyperemia on the periumiblical abdominal skin (Figure 1). Immediate ultrasonography and abdominal tomography were performed just before the surgery and revealed a heterogenous 47 × 32 mm cystic lesion lying from the posterior level of the right lob of the liver to the level of umbilicus (Figures 2 and 3). Surgical exploration revealed meconium in abdomen, a volvulus of small bowel with extensive necrosis and perforation at a part of 15 cm of terminal ileum and intestinal malrotation (Figure 4). After detection of midgut volvulus, the involved loop was resected and the proximal and distal ileum edges were juxtaposed and double-lumened stomy was constructed at the right lower quadrant. The patient recovered rapidly and was discharged home on 10th day after surgery. Osteomy was closed on sixth month.

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Figure 1. Hyperemia on the periumbilical abdominal skin.

Discussion

During embryonic development, the gut tube elongates from the stomach to the rectum and it progressively protrudes into the umbilical cord. As the organs develop, the midgut loop rotates

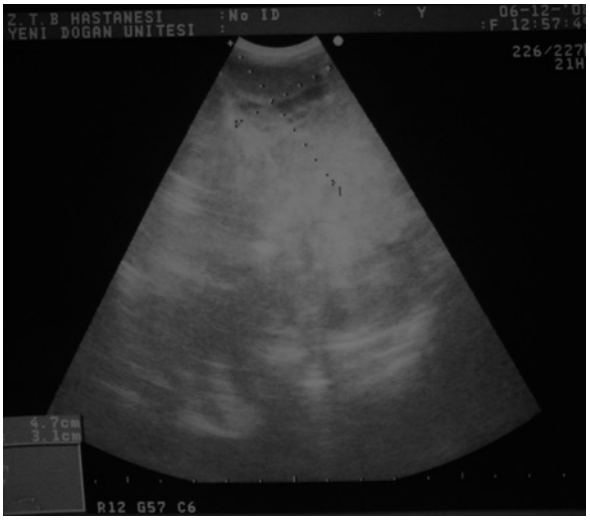

Figure 2. Ultrasonographic appearence of the cystic lesion.

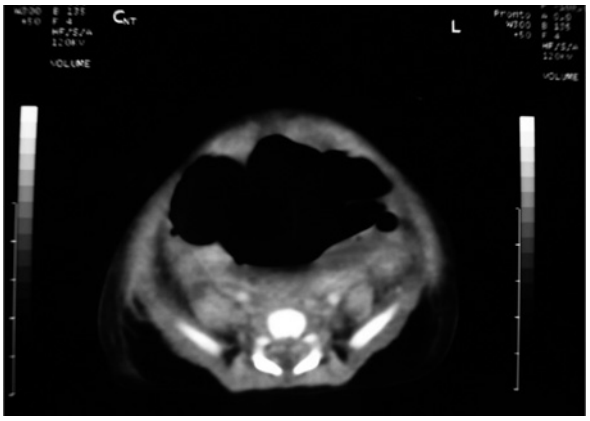

Figure 3. Abdominal tomography showing a heterogenous 47 × 32 mm cystic lesion lying from the posterior level of the right lob of the liver to the level of umbilicus.

90^0 anticlockwise around the axis of SMA which brings the duodeno-jejunal loop to the right and the ceco-colic loop to the left side between the 6th and 10th weeks of fetal development and afterwards intestines return to the intraabdominal position by the 10th week of embryonic life. The proximal and distal loops undergo a further 180° of anticlockwise rotation, a total of 270° rotation and as a result the duodeno-jejunal junction lies in the left upper quadrant and the ileocecal junction in the right lower quadrant of the abdomen[10]. Midgut volvulus occurs when the intestinal loop suspending along the free margin of the mesentery twists around the SMA. Volvulus is most often associated with malrotation that the short basal attachment of the mesentery to the posterior abdominal wall in intestinal malrotation may be the predisposing factor[11].

Figure 4. Surgical exploration of the patient.

Volvulus presents most commonly during the first year of life and fetal intestinal volvulus is extremely rare. Prenatal visualisation of volvulus with ultrasound have been reported in only few cases[5,9,11]. It should be suspected antenatally when polyhydramnios, intestinal dilatation, ascites and/or signs of fetal anemia are present[9]. The definitive diagnostic sign of midgut volvulus is the *whirlpool sign* that is produced by the bowel loop and its accompanying mesentery and mesenteric vessels that wrap around the main SMA[11]. In our case, we demonstrate polyhydramnios and *whirlpool sign* in prenatal USG.

Fetal midgut volvulus is a life threatening condition that may be associated with hydrops fetalis, spontaneous abortion, preterm delivery, fetal growth restriction and intrauterine fetal death[12-14]. The outcome depends on the amount of the compromised part of the intestine and the gestational age at the time of the event. Acute surgical management is necessary for the necrotic part of the intestine. In this report, we present a case with an intrauterine midgut volvulus detected sonographically at the antenatal period. Acute surgical management was performed soon after the birth which lead excellent prognosis.

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