Sudden Intrauterine Fetal Death Caused by Midgut Volvulus at Term: A Case Report

Dino Pavoković[1], Tvrtko Tupek[2], Anis Cerovac[3,4], Dubravko Habek[5], Mislav Mikuš[6], Vedrana Petrovečki[7]

[1]Department of Gynecology and Obstetrics, General Hospital Virovitica, Croatia

[2]Department of Gynecology and Obstetrics, Sveti Duh, University Hospital Zagreb, Croatia

[3]Department of Gynecology and Obstetrics, General Hospital Tešanj Bosnia and Herzegovina

[4]Department of Anatomy, School of Medicine, University of Tuzla Tuzla, Bosnia and Herzegovina

[5]Department of Gynecology and Obstetrics, Merkur University Hospital

[6]Department of Gynecology and Obstetrics University Hospital Centre Zagreb., Croatia

[7]Department of Forensic Medicine and Criminology, School of Medicine University of Zagreb, Zagreb, Croatia

Dino Pavoković
e-mail: dino.pavokovic@gmail.com
ORCID: 0000-0002-6140-4557

Tvrtko Tupek
e-mail: tvrtko.tupek@gmail.com
ORCID: 0000-0003-4052-4178

Anis Cerovac
e-mail: cerovac.anis@gmail.com
ORCID: 0000-0002-7209-382X

Dubravko Habek
e-mail: dhabek@unicath.hr
ORCID: 0000-0002-7675-7064

Mislav Mikuš
e-mail: m.mikus19@mail.com
ORCID: 0000-0002-1365-8704

Vedrana Petrovečki
e-mail: vedrana.petrovecki@mef.hr
ORCID: 0000-0001-5385-5561

Corresponding author:

Prof. Dubravko Habek, MD, MSc, PhD

Department of Gynecology and Obstetrics, Merkur University Hospital, School of Medicine Catholic University of Croatia
Ilica 244, Zagreb, Croatia

dhabek@unicath.hr

Abstract

Background: Fetal midgut volvulus is a serious finding with a high risk of life-threatening fetal complications.

Aim: To describe a sudden intrauterine fetal death caused by midgut volvulus at term.

Methods: This is a case report of a 28-year-old G2P0 patient with no significant medical history. At 34 weeks of gestation, an ultrasound revealed a cystic formation in the right upper abdominal quadrant of the fetus. In the 38th week, abrupt fetal intestinal dilatation and the absence of bowel peristalsis in what had been a normally developing fetus prompted the decision to induce labor and perform an emergency caesarean section due to terminal bradycardia. Neonatal resuscitation was attempted but unsuccessful (Apgar score 0/0/0 at 1, 5, and 15 minutes).

Conclusion: In this case, the true diagnosis of a fetal midgut volvulus and the cause of fetal death were confirmed by autopsy and a pathologist's finding.

Keywords: midgut volvulus, prenatal diagnosis, stillbirth, autopsy

Introduction

Fetal midgut volvulus (FMV) is a life-threatening condition that arises from intestinal malrotation. The frequency of associated neonatal small bowel complications, such as atresia and obstruction, has been reported to be 1 per 1,500-3,000, with the occurrence of malrotation estimated to be approximately 1 per 6,000 live births (1). This condition refers to a group of intestinal malrotation and fixation abnormalities resulting from intestinal nonrotation, incomplete rotation and impaired intestinal development in the first trimester (2). Midgut volvulus is a disorder in which the small bowel and colon twist around the superior mesenteric artery. In higher grades of bowel obstruction, there can be vascular compromise of the bowel itself, which might lead to infarction and even perforation (3–5). In the literature, two types of volvulus presenting antenatally are well described: the classic and segmental types. The classic type is defined as malrotation of the bowel due to clockwise rotation of the midgut (small bowel and ascending colon) around the superior mesenteric artery without any abnormality predisposing to rotation. Segmental volvulus is twisting of the bowel loops due to an intestinal abnormality, such as mesenteric defects (our case), intestinal atresia, meconium ileus, duplication cysts or congenital diaphragmatic hernia (6).

We present a case report about an autopsy and pathohistologically confirmed ischemic midgut volvulus which led to sudden term intrauterine fetal death.

Case report

The mother was a 28-year-old G2P0, without any significant medical history. She was referred to our tertiary perinatal center at 34 weeks of gestation for clinical evaluation and management of a small cystic formation in the right upper abdominal quadrant of the fetus that was noted on ultrasound. On our initial ultrasound, we verified a male fetus with normal biometry measurements and normal biophysical profile (BPP) using Doppler sonography. An anechogenic cyst

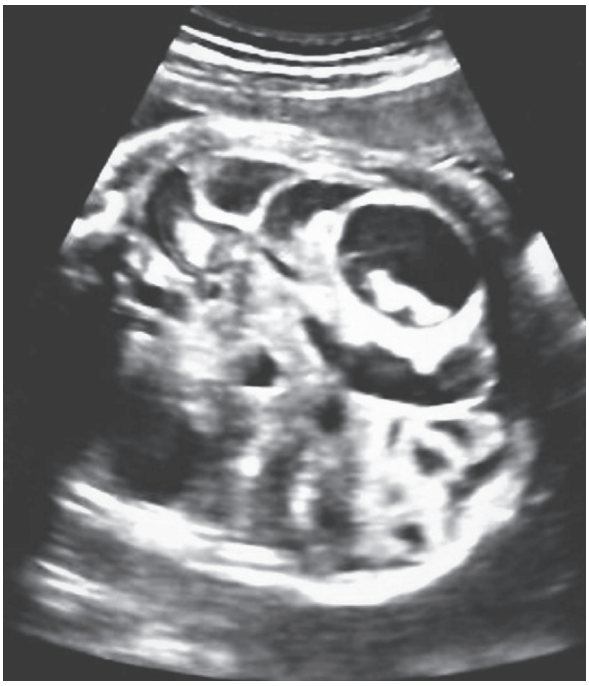

Figure 1. *Ultrasound findings of dilated small bowel*

was positioned under the liver near the right kidney, 31x10 mm in size. Due to the presence of a dilated common bile duct and the absence of hepatomegaly with visible bowel peristalsis, a hydrops of the gallbladder was suspected. The other fetal morphology was normal. Serial ultrasound examinations revealed stable cyst dimensions with normal BPPs. Upon reaching 37 weeks, the mother was admitted to our hospital for fetal surveillance and delivery planning. Despite reassuring daily nonstress cardiotocography (CTG) tests and total BPPs, the cyst continued to enlarge with an associated dilatation of the intestines. On hospital day 7, in the 38th week of pregnancy, the perinatal team decided to proceed with an induction of labor due to ultrasound pictures of increase in bowel dilatation and the absence of bowel peristalsis (Figure 1). Approximately 30 minutes after the decision was made to proceed with labor induction, CTG scan showed fetal terminal bradycardia, which was confirmed with ultrasound. An emergent cesarean section was performed and a eutrophic, hypotonic male neonate weighing 2650 grams was delivered. An unsuccessful neonatal resuscitation was performed (Apgar score 0/0/0 at 1/5/15 minutes).

On fetal autopsy examination, no abnormalities of the liver or gallbladder were noted. In the lower part of the neonatal abdomen, dilated loops of the proximal and middle parts of the small intestine were found with a necrotic, dilated third part of the small intestine. The necrotic loops were dark red and firmly adherent to the adjacent bowel with 210 ml of hemorrhagic fluid. The root of the associated mesentery was partially twisted around the superior mesenteric artery. Segmental overgrowth of the twisted mesentery was noticed and assumed to be the reason for the incident. Upon further inspection of the bowel with dissection in the caudal direction, it was noted that one loop of the distal small bowel was completely twisted around the mesenteric root, leading to complete obstruction of the intestinal lumen (Figure 2).

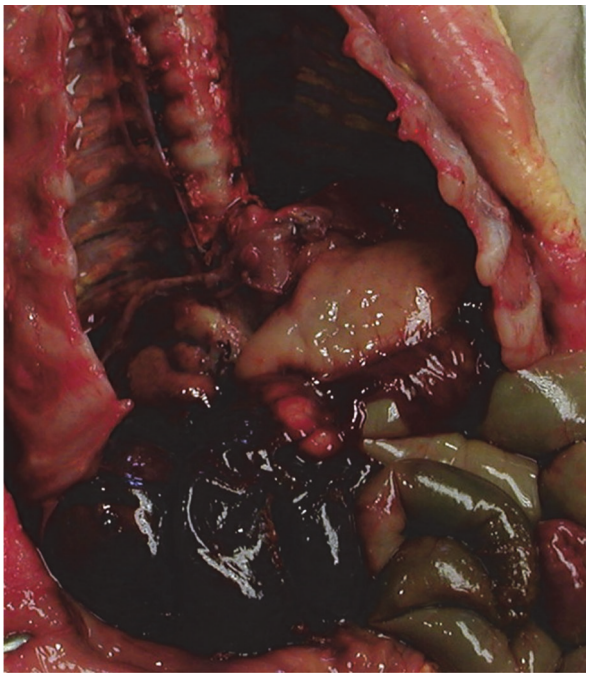

Figure 2. *Pathological examination*

Pathohistological examination of that segment revealed total transmural hemorrhagic necrosis without perforation, with the absence of the mucosal layer and infiltration of neutrophils in the subserous, edematous connective tissue. There was no other focal lesion found that might explain the volvulus. The pathologist gave the diagnosis of FMV with bowel necrosis. The reason for the intrauterine fetal death was assumed to be acute fetal inflammatory biohumoral response with cardiotoxicity and terminal bradycardia to a large segment of bowel necrosis.

Discussion

Fetal intestinal volvulus is a condition in which a delay in diagnosis and surgical intervention leads to high morbidity and mortality. When exhibiting *in utero*, it usually presents with ischemic necrosis of the bowel due to vascular compromise (7, 8).

Volvulus could be suspected during routine ultrasound examination or when non-specific fetal distress symptoms appear. The most frequent symptom is a decrease in fetal movements, which usually accompanies a non-reassuring CTG trace. In the literature, there are many prenatal ultrasound findings that are more or less specific for the diagnosis of volvulus: polyhydramnios, hyperechogenic and dilated loop of the bowel, fetal ascites, peritoneal calcifications as an indirect sign of meconial peritonitis and, finally, as the most specific and pathognomonic, "coffee bean" and "whirlpool" signs (9–11). Doppler studies can demonstrate elevated peak systolic velocity in the middle cerebral artery due to severe fetal anemia, secondary to hemorrhagic ascites (12).

Intestinal atresia and malrotation are almost always the causes of fetal intestinal volvulus (9). In a few case reports, the underlying mechanism of volvulus was a mesenteric developmental abnormality (13). In our autopsy report, we concluded that overgrowth of the mesentery was the reason for the abnormal intestinal movement, which resulted in intestinal volvulus, ischemia and hemorrhagic necrosis. The main cause of volvulus in our case was found to be a mesenteric defect and, thus, the final diagnosis was type 2 volvulus. It is important to emphasize the possibility that bowel perforation and subsequent meconial peritonitis may have led to even more rapid fetal deterioration. In the literature, a few case reports describe intrauterine fetal death due to intestinal volvulus but without intestinal perforation (4,5).

We believe that a combination of pathophysiologic factors contributed to intrauterine fetal demise. Due to arterial obstruction with ischemia and the resulting proinflammatory cytokinemia and hypovolemia, the fetus in our case experienced excessive activity of the parasympathetic nervous system (14, 15), although pathognomonic signs were absent. Prior to the indicated induction of labor, progressive and rapid bowel dilatation suggested the development of intestinal pathology with toxemia. A fetal intestinal volvulus was suspected and definitively diagnosed postnatally at autopsy.

We should always keep in mind that abrupt fetal intestinal dilatation in a previously normally developing fetus with mechanical ileus and decreased fetal movements may indicate intestinal volvulus. With advances in ultrasound technology and its widespread use in the third trimester, the diagnosis of volvulus can be made prenatally.

Conclusion

Without prompt obstetrical intervention, this diagnosis often results in a poor fetal outcome. A multidisciplinary team of obstetricians, neonatologists, anesthesiologists and pediatric surgeons should coordinate a plan that optimizes perinatal outcomes in the setting of fetal intestinal obstruction. However, in this particular case, the true diagnosis and cause of fetal death were confirmed by autopsy and a pathologist's findings.

Declarations

Authors' contributions

DP, TT and DH designed the study, participated in patient treatment and critically reviewed the manuscript, AC and MM designed the study, wrote the main manuscript and critically reviewed the manuscript. VP performed the autopsy and critically reviewed the manuscript.

All authors approved the final version of the manuscript, meet the authorship criteria, and hold rights to the intellectual content.

Ethics consideration

This case report has been approved by the Ethics Committee of the Clinical Hospital Sveti Duh, Decision No. 01-03-2089/4, dated May 12, 2022.

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Competing interests

The authors have nothing to disclose and no conflict of interest to declare.

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