



A Rare Case of Antenatal Intestinal Perforation on Top of Primary Volvulus in a Newborn with Congenital CMV

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Abstract

Gastrointestinal perforation during neonatal life is a common condition, carrying a mortality of 17–60%. But antenatal intestinal perforation due to volvulus is a rare condition. Intestinal volvulus is commonly associated with malrotation or atresia. A primary small bowel volvulus is extremely rare. It is a rare but life threatening surgical emergency manifesting after birth and as such rarely diagnosed prenatally. It causes intestinal obstruction and perforation, leading to a sterile chemical peritonitis. The usual site of perforation is the small bowel, the distal ileum being the most frequent site. On the other hand, gastrointestinal involvement in congenital or perinatal CMV infection is infrequent. We present here the case of a preterm baby born with distended, tensed abdomen and bluish discoloration around the umbilicus. CT-scan of abdomen showed presence of large cystic lesion. Exploratory laparotomy showed intestinal obstruction due to volvulus with necrosis of the ileum and perforation causing meconium peritonitis. Also the baby was diagnosed with congenital CMV and received the appropriate treatment. Intestinal perforation rarely occur antenatal, and intestinal volvulus causing perforation usually occurs on top of malrotation. To add to, congenital CMV rarely causes gastrointestinal manifestations.

Keywords: Group B streptococcus (GBS); Cytomegalovirus (CMV); Computed tomography scan (Ct-Scan).

1. Introduction

Intestinal volvulus is a condition in which the small bowel and proximal colon twist around the superior mesenteric artery [1]. It is a rare but life threatening surgical emergency manifesting after birth and as such rarely diagnosed prenatally [1]. Causes of intestinal volvulus includes intestinal malrotation, congenital malformations such as segmental defect of the smooth muscle of the bowel or a primary defect in the mesentery, meconium ileus or bowel atresia [1]. However, in extremely rare instances, a primary intestinal volvulus has been reported in which no

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underlying anomalies could be identified [1]. Intra-uterine bowel perforation leading to meconium peritonitis may be diagnosed prenatally by the presence of generalized fetal ascites, giant pseudo cyst, calcifications, and dilated bowel loops [2]. Gastrointestinal involvement during congenital and post-natal CMV infection is uncommon, in particular in immunocompetent and physiologic newborns. Particularly rare is the onset of these manifestations in the first 24 h of life [3].

2. Case Report

Our case is a 33 weeks gestational age baby boy, born by cesarean section, due to preterm labor to a 27 year old healthy mother (G2P1A1) and a 30 year old healthy father, with no history of consanguinity. Pregnancy was well followed with no complications, no gestational diabetes nor hypertension. TORCH was negative in first trimester, and Group B streptococcus (GBS) test not done. There was no history of any drug intake during pregnancy except iron and folic acid. Baby born APGAR 6 and 7 at 1 and 5 minutes with generalized cyanosis and grunting and started having subcostal retractions when transferred to NICU where he was intubated and ventilated. On the physical exam: the baby was tonic, no dysmorphic features, the abdomen was distended, tense with bluish discoloration around the umbilicus (Fig 1)



Abdominal x-ray showed diffuse opacity and absence of gas in the rectum (Fig 2).



The baby started to have bilious secretions from the nasogastric tube, and failed to pass stool. CT-scan abdomen showed presence of large cystic lesion involving the quasi totality of the right abdomen, measuring 6,5x4,25x6cm of liquid density (28 UH) .The antenatal intestinal perforation should be ruled out (Fig 3).



The laboratory results showed mild increasing in the liver function test with GGT 171 SGPT 34 SGOT 238 and positive CRP 53. Therefore, the surgery team was consulted and exploratory laparotomy showed intestinal obstruction due to volvulus with necrosis of the ileum and perforation causing meconium peritonitis. Enterectomy was done with resection of 10 cm of necrotic ileum and end to end anastomosis. The baby passed through severe

sepsis and DIC. He was treated with broad spectrum antibiotics. Meanwhile direct bilirubin still increasing, being an IUGR baby, TORCH titers ordered and showed positive CMV serology. PCR for CMV also was positive in the urine, blood and cerebrospinal fluid. Note that repeated TORCH titers for the mother after delivery showed also positive CMV serology so the baby was diagnosed having congenital CMV. The baby started on Gancyclovir IV for 14 days, then shifted to Valgancyclovir per os for 6 months, after tolerating per os feeding and passing stool. After treatment with antibiotics and Gancyclovir, Laboratory tests normalize gradually including liver function tests, direct bilirubin, CRP... and baby discharged home on Valgancyclovir per os for total of six months.

3. Discussion

Our case describes a very rare cause of volvulus not associated with malrotation and causing intestinal perforation. Malrotation, intestinal atresia, or mesenteric defects have been known to be the predisposing factors for an intrauterine fetal volvulus [4-6]. However, in extremely rare instances, a primary intestinal volvulus has been reported in which no underlying anomalies could be identified. Chung et al. reported a case similar to ours, in which the gross anatomical abnormalities could not explain the occurrence of the fetal primary small bowel volvulus [7]. Review of literature found 4 similar cases described below. Found two with no definitive cause and two others with hyperperistalsis and hypoperistalsis in the descending and sigmoid colon as a cause. Chung et al, They concluded that the complication of meconium-related ileus due to immature ganglion cells or hypo peristalsis played a role in the etiology of the fetal primary small bowel volvulus [7]. Similarly, Kurashige et al. suggested distension and increased peristalsis could form a primary loop and twist the bowel [8]. A similar article to ours done by Younglim Kima found no etiology for the volvulus and considered it as a primary segmental volvulus occurring at the ileum. We successfully treated an extremely rare case of primary segmental volvulus of the ileum in a fetus, in which no macroscopic or microscopic abnormalities were found in the intestine [9]. In a study of patients without malrotation, Black et al. have shown that segmental mesenteric defects were common in this group and had an etiological role. These occurred either due to excess growth of small segment of bowel or uncontrolled growth of an isolated segment of the mesentery, resulting in a portion of bowel having a small segment of mesentery with a narrow base which predisposes to volvulus [6]. Cytomegalovirus (CMV) is the most common cause of congenital viral infection, affecting 0.2 to 2.3% of all live births in developed countries [10]. Gastrointestinal involvement in congenital or perinatal CMV infection is infrequent [10]. Most of the studies revealed a possible association between CMV infection and enteritis, enterocolitis, and NEC, often with finding of CMV inclusions in histological intestinal samples. It is debatable whether the virus has a role in the pathogenesis of the diseases, either directly or as a secondary super-infection after the acute phase of the enterocolitis [3]. We suggest the hypothesis that an inflammatory process induced by CMV congenital infection may be responsible, in the early gestation, of the intestinal end organ disease, as the intestinal malrotation. [10] CMV infection should always be excluded in full-term infants presenting with colonic stricture or malrotation. [10] What differs from our case is the absence of malrotation, and CMV was not proven positive in histological intestinal samples.

4. Conclusion

Although in-utero intestinal volvulus and perforation is extremely rare, it is a surgical emergency. Even in the absence of malrotation or atresia, a primary volvulus should be suspected to prevent complications. Congenital CMV rarely causes gastrointestinal manifestations and should be confirmed by histology.

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