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Midgut volvulus and meconium peritonitis induced non-immune hydrops

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ABSTRACT

Meconium peritonitis (MP) is a rare cause of nonimmune hydrops and only few of those cases have been reported in literature. Here is to report on a case of FH secondary to perforated midgut volvulus and MP with special interest in the discussion of pathophysiology and clinical strategies. We report on a preterm male neonate diagnosed with FH at 29th week of gestational age (WGA) and EXIT at 30 WGA. In addition, he presented with abdominal mass, respiratory distress syndrome and hypoglycemia. Emergent endotracheal intubation and mechanical ventilation rescued the baby. All maternal and infant viral panel, Toxoplasmosis, Sars-Covid 2 and indirect/direct coombs tests were negative. . At laparotomy, we found a midgut volvulus interesting nearly all the jejunum but sparing the ileum which appeared hypoplastic. One of intestinal loop was completely perforated with diffuse MP. Distal to the first jejunal loop, the jejunum appeared necrotic and then resected with bowel exteriorization (terminal jejunostomy and ileostomy). The postoperative course was complicated by worsening of respiratory distress, acute tubular necrosis and renal failure unresponsive to medical therapy and fatal outcome. In conclusion, we believe that hydrops secondary to midgut volvulus and meconium peritonitis has its own pathophysiological and clinical characteristics that make this disease at high risk of unfavourable outcome when compared with prenatal cases of midgut and/or meconium peritonitis without hydrops.

1. Introduction

The diagnosis of fetal hydrops (FH) is uncommon and associated with a poor prognosis. Nowadays, because of prophylactic isoimmunization, non-immune FH has become the most common cause of several maternal and fetal conditions [1]. The incidence of non-immune FH has been estimated as 1.4–3:10,000 live births [2,3]. In these neonates, it has been reported that perinatal mortality ranges from 35 to 43% [3]. Although older studies considered many cases to be idiopathic [4–6], a more recent, larger series and a systematic review report that a cause can be found in nearly 60% of cases prenatally [7] and 85% when postnatal detection is included [8]. Meconium peritonitis (MP) is a rare cause of non-immune hydrops, and only a few of those cases have been reported in the literature [9]. Here we report a case of FH secondary to perforated midgut volvulus and MP and discuss pathophysiology and clinical strategies.

2. Case report

We report on a preterm male neonate diagnosed with FH at the 29th week of gestational age (WGA) and EXIT at 30th WGA. He presented with abdominal mass, respiratory distress syndrome, and hypoglycemia. All previous prenatal ultrasounds were normal until the 25th WGA, and the mother had received isoimmunization because of Rh maternal-fetal incompatibility. Prenatal diagnosis of hydrops was made at the 29th week of gestational age by observing abdominal fluid collec-

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tion and skin oedema (Fig. 1). Then, the mother was immediately transferred to our tertiary centre. At delivery, polyhydramnios was also noted. Histological examination of the placenta was normal. Bodyweight at birth was 2.2 kg. The Apgar score was 3 at 1 min, 5 at 5 min, and 6 at 10 min because of the absence of cardiac and respiratory functions. Emergent endotracheal intubation and mechanical ventilation rescued the baby. There were no cardiac anomalies. All maternal and infant viral panel, toxoplasmosis, SARS-COVID-2, and indirect/direct Coombs tests were negative.

On the following day, physical examination showed diffuse oedema, cardiac and respiratory functions within limits, and abdomen distended because of oedema but treatable and not painful. The infant had a syndromic aspect characterized by abnormal ears implant and transverse palmar fold. Genetic studies did not show chromosomal or gene anomalies. At admission, the main findings at laboratory analyses were albuminemia within normal range, hypoglycemia, hyperleptinemia with polyuric renal failure, and hyperkalemia. Plain abdominal x-ray showed calcification suggestive of MP and limited air only in the first jejunal loop (Fig. 2). Abdominal ultrasound confirmed the presence of diffuse calcification and fluid collection on the left quadrant. Furthermore, at the ultrasound, we observed diffuse hyperechogenic lesions in the renal cortex of both kidneys (suggestive of arterial calcifications).

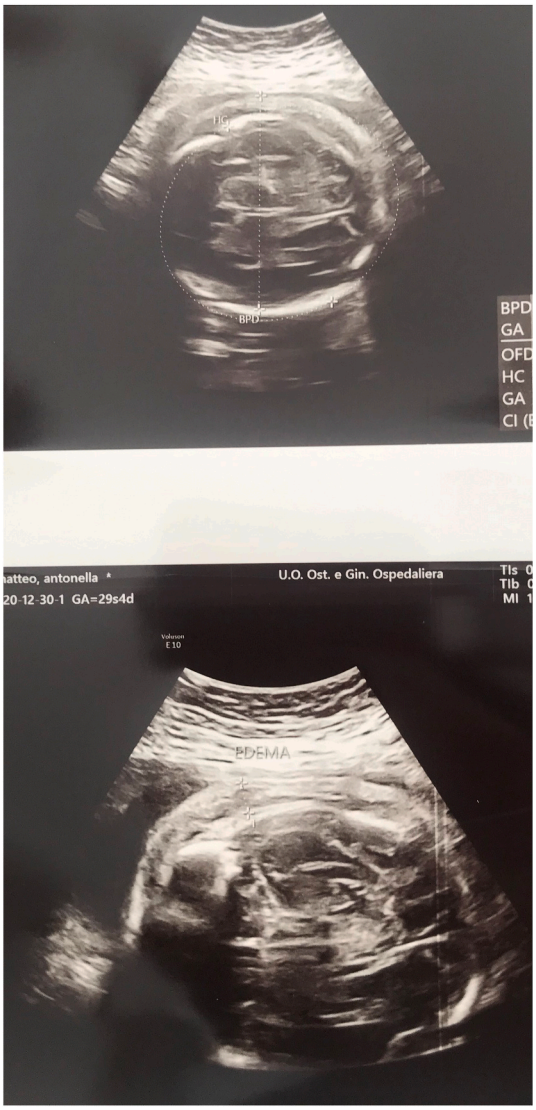

Fig. 1. Prenatal ultrasound at 29 th week of gestational age showing abdominal fluid collection and hydrops.

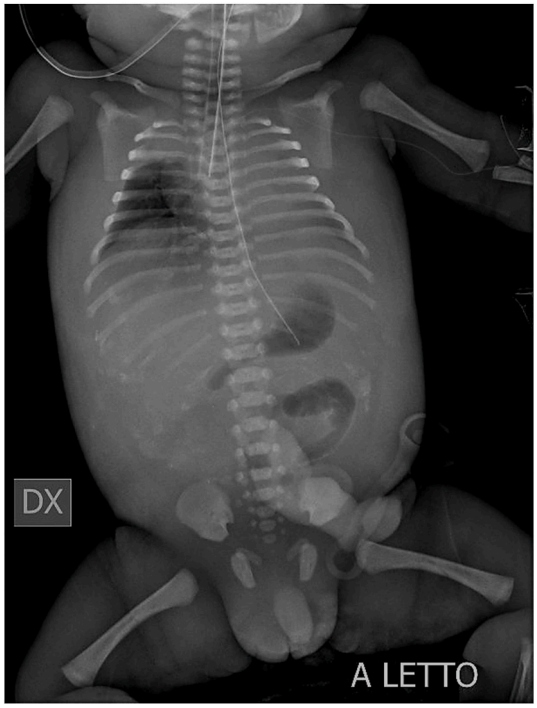

Fig. 2. Plain abdominal x-ray showing jejunal obstruction and calcifications.

After fluid resuscitation, albumin replacement, and intense diuretic stimulation to correct hydrops, the baby underwent general anaesthesia and abdominal surgery on the 3rd postnatal day. At laparotomy, we found a midgut volvulus involving nearly all the jejunum but sparing the ileum which appeared hypoplastic (Fig. 3). One of the intestinal loops was completely perforated with diffuse MP. All around, there were dense and organized adhesions with the abdominal wall. Distal to the first jejunal loop, the jejunum appeared necrotic (Fig. 4) and then resected with bowel exteriorization (terminal jejunostomy and ileostomy). There was no ascites or hepatomegaly. Operative time was 160 min without bleeding or intraoperative complications.

The postoperative course was complicated by worsening of respiratory distress, acute tubular necrosis, and renal failure unresponsive to medical therapy. Hemodialysis for preterms is unavailable at our centre, and the baby could not be transferred because of severe respiratory failure, which required high-frequency oscillometric ventilation (low

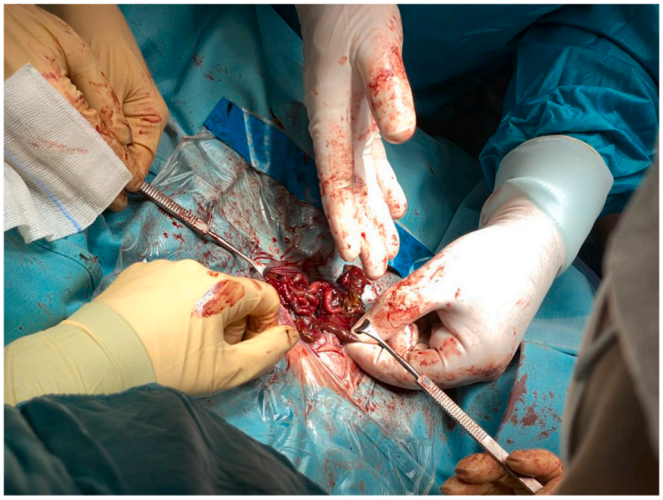

Fig. 3. At laparotomy, we can observe perforated jejunal loop and hypoplastic ileum.

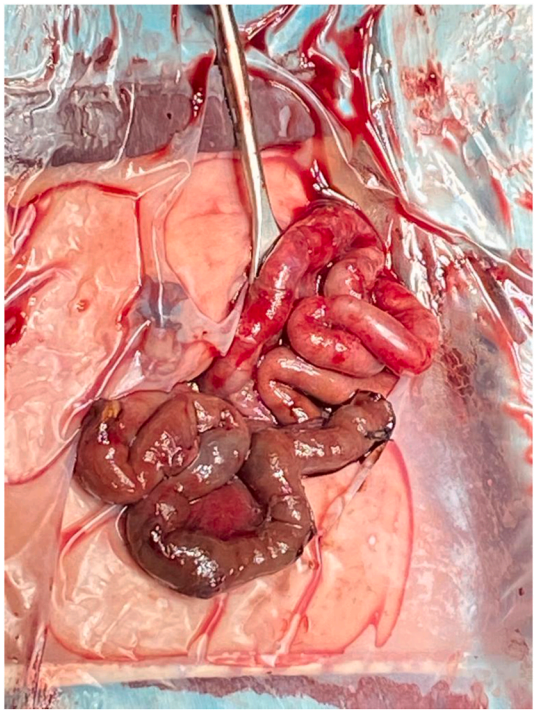

Fig. 4. At laparotomy, extended jejunal segment interested by necrosis and hemorrhage.

volumes-low pressures). We believe that general anaesthesia played a key role in developing acute tubular necrosis and the worsening of renal function. The infant eventually died on the 10th postoperative day. The family did not authorize an autopsy.

3. Discussion

Despite modern advancement in prenatal diagnosis, *in utero* detection of intestinal volvulus remains challenging [9]. Reported prenatal signs of volvulus are dilated intestinal loops and polyhydramnios, while meconium peritonitis is suggested by the presence of cystic abdominal mass, hydrops, and/or polyhydramnios [10]. Timing in diagnosis and surgical intervention in the treatment of midgut intestinal volvulus play a key role in the survival of these unfortunate neonates [11]. In our patient, despite nearly immediate delivery and surgical intervention after hydrops detection and treatment, all efforts we made were unable to reverse clinical conditions because of unresponsive renal failure and severe respiratory distress.

As noted above, the last two morphological prenatal ultrasound were obtained at 25th and 29th WGA; interestingly, the first was normal while the second showed clear signs of hydrops and abdominal cystic mass. For this reason, we believe that the timing of development intestinal volvulus occurred in this specific interval period. The findings we observed at surgery support this hypothesis.

There are only a few reported cases of non-immune hydrops secondary to midgut volvulus in the literature, and these are often associated with poor prognosis. Seward et al. reported a full-term infant weighing 2.7 kg born very oedematous, lethargic, and cyanotic. After initial resuscitation, emergent laparotomy, and extended bowel resection, this infant eventually died 34 h after birth [12]. Imai reported a preterm infant diagnosed with complicated polyhydramnios and non-immune hydrops who eventually died on the 2nd postnatal day for respiratory distress [13]. Hasegawa et al. reported a child with very low birth weight and at 19th gestational weeks was diagnosed with polyhydramnios and multiple cystic spaces in the abdomen, which was markedly distended and was compressing the chest. Following intestinal resection and jejunostomy, he died on the 7th postoperative day because of heart failure secondary to severe volume, electrolytes, and acid-base imbalance [14]. Nogami et al. reported a near-term neonate born with generalized oedema and severe abdominal distension by ascites. Despite intensive resuscitation, the neonate died 2 h after delivery. Postmortem examination showed severe midgut volvulus with infarction from the duodenum to distal ileum [15]. Survival in preterm born neonates with fetal hydrops secondary to midgut volvulus is rare [9].

On the contrary, there are several reports of survival in patients with a prenatal diagnosis of meconium peritonitis or midgut volvulus but not associated with hydrops [16–18]. In those children, prenatal diagnosis of midgut volvulus is often possible, and appropriate management with serial ultrasounds and early delivery in a tertiary centre with pediatric surgeons available may significantly increase survival [11]. In our case, the preoperative findings of renal injury demonstrated by abnormal serum creatinine and polyuria together with prolonged operative time played a key role in the development of acute kidney injury following general anaesthesia for major abdominal surgery. In fact, preterm infants with pre-existing kidney injury are significantly more predisposed to develop acute renal failure following general anaesthesia under these circumstances [19].

Recently, neuroaxial anaesthesia has been proposed in preterm infants to limit these adverse reactions following general anaesthesia [20]. Payne and Moore reported the use of continuous spinal plus general anaesthesia for major abdominal surgery in ten patients in the age range of 2–59 months [21]. Somri et al. reported the use of combined spinal and epidural anaesthesia in major abdominal surgery in 28 high-risk neonates and infants [20]. However, although spinal anaesthesia allows the prevention and the reduction of perioperative complications, its duration is an important limiting factor [22]. Because of this limitation, short surgery is the most indicated under spinal anaesthesia. We did not proceed with spinal anaesthesia in our setting because our anaesthetist was inexperienced with spinal anaesthesia in low-weight preterm infants, and surgery was expected to be prolonged.

Finally, the pathogenesis of hydrops in prenatal midgut volvulus remains uncertain. Various factors have been suggested to explain prenatal hydrops, including low oncotic pressure, severe anaemia, and congestive heart failure [15]. However, in their review, Barnes et al. show that none of these factors has been present in all cases [23]. Mayock et al. suggested that giant cysts that develop due to meconium peritonitis compromise the venous return by compressing the inferior vena cava, thereby increasing venous pressure [24]. This pathophysiology could have been possible in our patient.

In conclusion, we believe that hydrops secondary to midgut volvulus and meconium peritonitis has unique pathophysiological and clinical characteristics that increase the risk of an unfavourable outcome compared with prenatal cases of midgut and/or meconium peritonitis without hydrops. In our opinion, to improve the prognosis of these neonates, efforts should be made to detect this disease early, before hydrops develops.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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