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Early intestinal perforation secondary to congenital mesenteric defects

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ABSTRACT

Gastrointestinal perforation (GIP) in preterm neonates may be idiopathic, due to necrotizing enterocolitis (NEC), or mechanical obstruction. The predominant cause of GIP in the neonatal period is NEC. Differential diagnosis with congenital malformations, including mesenteric defects leading to internal hernias, is mandatory if the onset is early. We describe two newborns with trans-mesenteric herniation resulting in GIP, and we discuss the presence of possible additional risk factors such as prematurity and predisposing vascular disruption in connective tissue disorders (Ehlers-Danlos syndrome), twinning, and use of assisted reproductive technologies. These cases prompted us to review our exploratory laparotomies performed for intestinal obstruction, complicated/or not with perforation, to identify the frequency of neonatal trans-mesenteric hernias in a referral hospital. The prevalence of GIP and of internal hernia was 25% and 3.3%, respectively. In conclusion, time-onset and particular conditions associated with GIP should lead to a high index of suspicion for internal hernias in order to achieve appropriate diagnosis and therapy.

Gastrointestinal perforation (GIP) may occur perinatally mainly in term newborns who sustained hypoxic-ischemic events, while after the second week of life it is more common in preterms, mostly caused by necrotizing enterocolitis (NEC) of multifactorial origin. An inverse relationship between gestational age and age at onset of NEC is documented [1], so when GIP occurs early in preterm neonates, other contributing causes for wall's necrosis should be sought. Attention should be focused on congenital structural anomalies causing gastrointestinal occlusion, including unexpected internal hernias through mesenteric defects.

We report on two newborns with internal hernia secondary to congenital mesenteric defects, causing early GIP. We discuss possible risk factors and diagnostic peculiarities of trans-mesenteric hernias in the neonatal period.

1. Case reports

1.1. Patient 1

A 30-weeks-gestation female neonate was born by caesarean section for abnormal fetal heart rate and intrauterine growth restriction. Apgar scores were 7 and 9 at 1 and 5 min, respectively. Primary resuscitation, mechanical ventilation, and total parenteral nutrition were required. Meconium passed normally. The patient's mother, suffering from Ehlers-Danlos syndrome (EDS), had undergone to surgery at birth because of GIP. As well as the mother, the neonate had an EDS evocative phenotype. She was symmetrical small for gestational age (weight 720 g), with partial blepharophimosis, occipital angioma, ligamentous laxity, reduced spontaneous motility and muscular hypotonia. An echocardiogram revealed a wide foramen ovale, mitral insufficiency, and left ventricular hypertrophy.

On the second day of life sudden clinical deterioration with onset of tense abdomen was detected. Sonography showed suspected free air suggestive of perforation, and multiple dilated hyperperistaltic small-bowel loops suggestive of obstruction. Plain

Abbreviations: ART, Assisted reproductive technology; EDS, Ehlers-Danlos syndrome; GIP, Gastrointestinal perforation; NEC, Necrotizing Enterocolitis.

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radiogram (Fig. 1) confirmed GIP, so she underwent urgent surgery. An incomplete ileal volvulus with the cecum in subhepatic position was found. Part of the ileum was herniated through a mesenteric gap between the right colic artery and the ileocolic artery axis. There was an ileal perforation 5 cm proximal to the ileocecal valve. Suture of ileal perforation and mesenteric defect were performed. The tissues appeared extremely friable and easily bleeding. Local hemostasis was obtained, while plasma, platelets, and packed red blood cells were transfused.

Orthostatic plain abdominal radiogram shows pneumoperitoneum and multiple air-fluid levels with loops of distended bowel resembling an inverted "U" that was later revealed to be trans-mesenteric hernia. Gas is absent in the rectum. The mother's family had a history of Ehlers-Danlos syndrome and the baby had a cytogenetic diagnosis of this syndrome.

After surgical intervention a pneumothorax required drainage. Three months later a plastic mesh repair for incisional hernia was performed.

1.2. Patient 2

An 1870 g male neonate was born at 34 weeks of gestation from a triplet pregnancy achieved through Intra-Cytoplasmic-Sperm-Injection (an assisted reproductive technology-ART). Apgar scores at 1 and 5 min were both 8. Mild respiratory distress required nasal Continuous-Positive-Airway-Pressure for 2 h. Clinical examination was unremarkable except for two small tuberous angiomas.

On the second day of life he had prominent abdomen, painful on palpation, and X-ray revealed a conspicuous pneumoperitoneum. Urgent exploratory laparotomy revealed twisted small intestine not associated to malrotation or atresia. There was meconium peritonitis due to ileal perforation, 8 cm proximal to the ileocecal valve, in an entrapped bowel segment through a congenital mesenteric defect. The patient underwent minimal resection of the perforated tract, suture of the mesenteric gap, and temporary ileostomy.

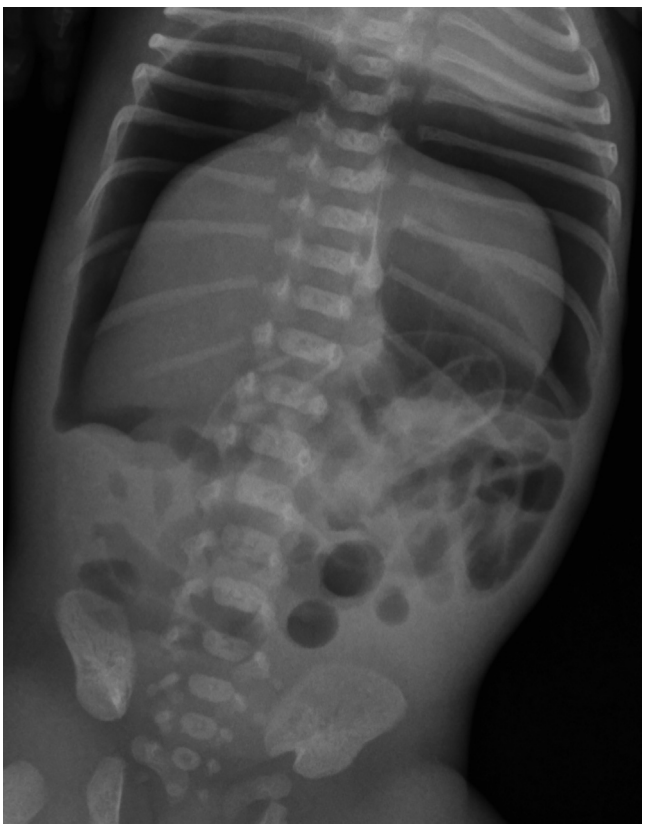

Fig. 1. Abdominal radiogram of patient 1.

2. Discussion

Internal hernias of ileal loops are rare but extremely serious causes of acute or intermittent intestinal obstruction. Published literature estimates an incidence of 0.6–5.8% of all small-bowel obstructions at any age [2,3]. Congenital and acquired forms are reported, the latter resulting from incomplete closure of mesenteric defects due to surgery or inflammation. There are various types: paraduodenal, pericecal, through foramen of Winslow, trans-mesenteric, trans-mesocolic, intersigmoid, and retro-anastomotic [3].

Trans-mesenteric hernias are generally only 5–10% of internal hernias, but they are the most common type in children (35%). They occur when part of the bowel passes through a defect in the dorsal mesentery of small-bowel or colon [4], most frequently localized in the ileocecal region or the sigmoid mesocolon. Treves described a part of mesentery near the terminal ileum, circumscribed by the ileocolic branch of the superior mesenteric artery and its anastomosis with the last ileal artery. This so-called Treves' Field doesn't contain fat, is highly susceptible to injury during development, and is a minor resistance area for hernias [2]. As no hernia sac is present, a considerable length of bowel can protrude through the defect [4]. Recently, Saka et al. [5] analyzed all pediatric cases of congenital Treves' field trans-mesenteric hernias reported in literature: 34 cases were found, 10 of them (29.4%) were in neonatal age.

Trans-mesenteric hernias may occur in newborns as early GIP or, in later periods and with larger defects, with painful intermittent obstruction [2]. Jejunoileal atresias are the consequence of intrauterine vascular insults which induce necrosis and then atresia. These atresias are associated with a mesenteric defect. Isolated mesenteric defects are assumed to be incomplete forms of atresia, due to less extensive vascular thrombosis than it occurs with complete ileal atresia [6,7]. The malformation in both of our reported cases can be explained as variant of in utero vascular disruption disorder involving the superior mesenteric artery [7]. Clinical history and imaging are necessary for the diagnosis: ultrasounds show dilated hyperperistaltic loops [8]; X-ray reveals multiple air-fluid levels (differential diagnosis with paralytic ileum) and absent gas in the rectum (differential diagnosis with NEC) [3]. Especially in newborns, the diagnosis is often confirmed only by urgent exploratory laparotomy. Associations are reported with monozygotic twinning, cystic fibrosis, and Hirschsprung disease [2].

In our series, prematurity, structural defects in connective tissue (patient 1), and twinning (patient 2) may have played as aggravating factors and contributed to early-onset herniation and GIP.

In vascular EDS (type IV), because of the bowel wall's structural weakness, a hernia formation tendency and GIP is described [9]. In addition, there are a reduced capacity for wound healing, an increased frequency of incisional hernia, risk for intraoperative bleeding due to vessel's fragility, and spontaneous pneumothorax [10], as observed in patient 1. Vascular disruption disorders are described in vascular EDS.

In patient 2 predisposing factor to internal hernia may have been the twinning itself, since association between monozygotic twins and congenital mesenteric defects has been observed in some case reports [2,11]. Nevertheless, the use of ART increases the incidence of congenital cardiovascular abnormalities [12].

These cases suggested us to review a larger population of newborns with intestinal occlusion with/without perforation. We analyzed retrospectively the medical records of sixty newborns (range of gestational age 24–41 weeks, weight 600–4040 g) admitted for mechanical ileus to this Neonatal Intensive Care Unit

between 2009 and 2013. GIP was present in 25% of cases, mainly in preterm newborns (80%), and was classified in late-onset, occurring all in low-birth-weight preterms (8 NEC, 1 ileal atresia), and early-onset (3 term newborns with small-bowel atresia, 1 prenatal onset cystic fibrosis, and 2 internal hernias). There were no significant pathognomonic laboratory differences. Our prevalence of neonatal GIP due to internal hernias was 3.3% of all intestinal occlusions during the study period. Late-onset GIPs in preterms were mainly due to NEC, whereas early-onset GIPs were all caused by congenital malformations: intestinal atresia (in term infants) or internal hernia through mesenteric defect (in preterms).

Generally, ileal atresia is difficult to diagnose with routine prenatal ultrasound that may only be suggestive that there is an abnormality. A strict postnatal evaluation of the intestinal atresia can help to determine the best course of treatment, reducing the risk of GIP. The prenatal detection of a mesenteric gap is almost impossible, representing a real challenge for the future.

Trans-mesenteric hernias should be considered as differential diagnosis of bowel obstructions above all in early GIP in preterms. Of fundamental importance is to distinguish them from advanced NEC, in which sometimes conservative therapy is chosen, whereas in GIP secondary to internal hernia immediate surgical treatment is mandatory. Since the symptoms are similar and aspecific, the presence of particular predisposing factors (prematurity, collagenopathies, twinning, ART), some radiological features, and the precocity of GIP should further support the neonatologist to suspect them and make the appropriate diagnostic and therapeutic choices.

Given the broad spectrum of pathology, congenital mesenteric defects may often remain underdiagnosed. The formation of internal hernias can thus occur in later periods, until adulthood, when engaging other conditions (pregnancy, trauma, inflammatory processes, rapid weight loss with intraperitoneal fat reduction) cause an increased intra-abdominal pressure or an enlargement of the mesenteric defect.

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Conflict of interest

None.

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