Journal of Pediatric Surgery 55 (2020) e1–e3

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Small left colon syndrome in 3 sisters

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ARTICLE INFO

Article history:
Received 17 February 2012
Received in revised form 3 July 2012
Accepted 8 July 2012

Key words:
Small left colon
Neonatal bowel obstruction
Meconium plug syndrome
Siblings
Diabetes
Genetic disease

ABSTRACT

Neonatal small left colon syndrome is a rare cause of bowel obstruction. Its etiology remains unknown, but a significant association has been noted between maternal diabetes and small left colon. No reported cases within the same family could be found in the literature, excepting 2 sets of twins. We report 3 cases of small left colon syndrome in 3 consecutive sisters born of a nondiabetic mother. This raises the question of a genetic factor in its etiology.

Meconium plug syndrome, small left colon syndrome (SLCS), functional neonatal bowel obstruction, and Hirschsprung disease are all causes of distal bowel obstruction in the neonate. The clinical features are similar: abdominal distention, bilious vomiting, and failure to pass meconium. Neonatal SLCS is considered a rare functional lower intestinal obstruction, characterized by a narrowed left colon with an abrupt transition zone at the splenic flexure on contrast enema. The etiology is unknown, but a significant association has been noted between maternal diabetes and SLCS [1]. The clinical course of SLCS varies in severity from mild symptoms, relieved by enema, to severe obstruction associated with colonic perforation. Several contrast enemas are sometimes necessary to avoid operative intervention. If bowel obstruction persists after the passage of meconium, a rectal biopsy should be performed. We report 3 cases of SLCS in 3 consecutive sisters born of a nondiabetic mother.

1. Case reports

1.1. Mother

The mother was treated with levothyroxine 125 mg/d for hypothyroidism and smoked 5 cigarettes a day during pregnancy. Thyroid function was normal during the pregnancies. Glycemia at the beginning of the pregnancies was normal, and she had an O'Sullivan test around 28 weeks of gestation for the last 2 pregnancies, which showed a glycemic level lower than 1.35 g/L. The parents were not consanguineous, and the mother had an abortion between her second and third daughters.

1.2. Case 1

The first sister was born in 2006 at 38 weeks' gestation by cesarean delivery because of abnormal fetal cardiac rhythm. Her birth weight was 2500 g. At 48 hours, she developed abdominal distention with bile-stained vomiting, and no meconium had been passed. Plain abdominal radiograph showed distended bowel loops. A rectal examination was done, and a small mucus plug and meconium were passed. An undiluted gastrografin enema at 7 days showed a narrowed left colon up to the splenic flexure. A rectal suction biopsy showed normal ganglion cells. Cystic fibrosis was ruled out. Thyroid function was normal (free thyroxine [FT4], 25.9 pmol/L; thyroid-stimulating hormone [TSH], 1 mUI/L). She was discharged at age 19 days after having recovered normal bowel function. She is now 4 years old and has had no bowel dysfunction since she was discharged. On clinical examination, she is an 18-kg healthy girl with no abdominal distension.

1.3. Case 2

The second sister was born in 2007 at 34 weeks of gestation by cesarean delivery because of abnormal fetal cardiac rhythm. Her birth weight was 2130 g. She needed supplemental nasal oxygen for a few days and had transient unexplained axial hypotony. At 72 hours, she developed abdominal distention with bile-stained vomiting and had not passed

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https://doi.org/10.1016/j.jpedsurg.2012.07.038
0022-3468/

meconium. She had dilated bowel loops on plain abdominal radiographs. An undiluted gastrografin enema at 4 days showed the characteristic patterns of a neonatal SLCS. The enema led to the evacuation of several plugs of sticky meconium. After the enema, bowel dysfunction persisted with vomiting and abdominal distention. A rectal suction biopsy excluded a Hirschsprung disease. Results from immunoreactive trypsinogen and thyroid function studies were normal (FT4, 14 pmol/L; TSH, 2.5 mUI/L). The enema procedure was repeated twice. The abdominal distention resolved, and she progressed to a regular formula diet at the term of 38 weeks. She was discharged at 42 days of age. She is now 3 years old, weighs 16 kg, and has normal bowel function with mild abdominal distention.

1.4. Case 3

The third sister was born in 2010 at 34 weeks of gestation by cesarean delivery because of abnormal fetal cardiac rhythm. Her birth weight was 2460 g. At 48 hours of age, she developed abdominal distention and vomiting. She had not yet passed meconium. Plain abdominal radiograph showed distended bowel loops. A rectal examination was performed initially, and she passed a small meconium plug. Undiluted gastrografin enema was done at age 3 days. Neonatal SLCS was confirmed (Fig. 1). After colon filling, symptoms resolved for 2 days, but abdominal distention and vomiting then recurred. Despite the 3 repeat gastrografin enemas, intermittent partial intestinal obstruction persisted for several weeks. A rectal suction biopsy was obtained to exclude aganglionosis and showed normal ganglion cells. Cystic fibrosis and neonatal hypothyroidism were also ruled out (FT4, 21 pmol/l; TSH, 2 mUI/L). The addition of *N*-acetylcysteine per orogastric tube at a dose of 5 mL of a 10% solution every 6 hours was not efficient. Erythromycin was given orally at 10 mg kg^{-1} d^{-1} for 3 weeks and then at a dose of 40 mg kg^{-1} d^{-1} for 10 days. Bowel movements subsequently improved. A colostomy had been considered but not performed based on the previous favorable experience without surgery in both of her older sisters. The abdomen remained distended, but the neonate eventually progressed to a regular formula diet with normal bowel motility at age 52 days. She was discharged at age 63 days. The erythromycin had been stopped a few days before. At 4 months of age, she weighs 6 kg and has a normal bowel function with no abdominal distention.

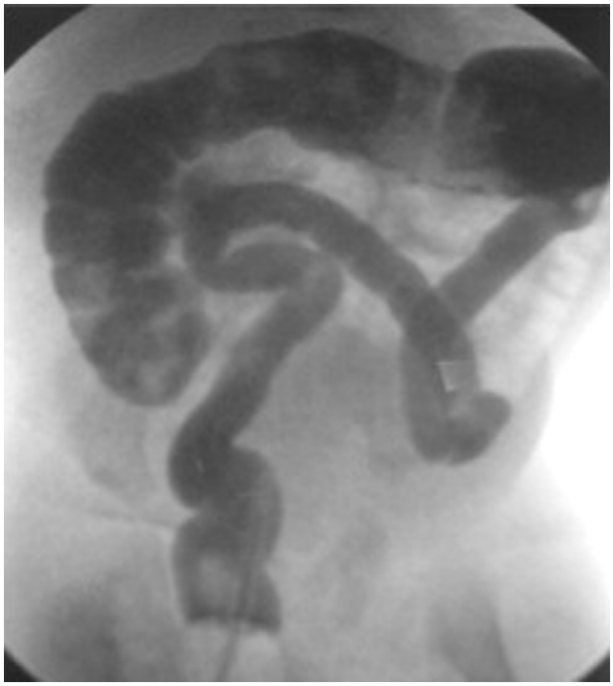

Fig. 1. Contrast enema of the third sister.

2. Discussion

Small left colon syndrome is a rare cause of neonatal bowel obstruction. Its etiology remains unknown. In our literature search, we found only 2 cases of SLCS within the same family: a set of twins born of a diabetic mother [1] and another set of twins born of a nondiabetic mother [2]. To our knowledge, no specific genetic studies have been performed. Cohen et al [2] suggested an intrauterine etiology for SLCS. Although the etiology is unknown, a significant association has been noted between maternal diabetes and SLCS, with an incidence of 40% to 50% of reported cases being progeny of diabetic mothers [1,3]. Philippart et al [4] suggested that neonatal hypoglycemia causes glucagon release and sympathoadrenal and vagal stimulation causing a decreased motility of the jejunum and left colon.

Although neonatal hypothyroidism is known to be responsible for a transient intestinal dysmotility, our research in the literature did not find any studies that have shown any association between maternal hypothyroidism and bowel dysfunction in neonates. In our series, maternal thyroid function remained stable, less than 15 mUI/L (TSH), during the 3 consecutive pregnancies.

Transient intestinal dysmotility is often described in premature newborns, particularly in severe premature neonates. Colonic biopsies were taken in premature newborns with low colonic obstruction and revealed immaturity of the colonic enteric nerve plexuses [5,6]. In our series, 2 of the 3 sisters were born prematurely at 34 weeks of gestation; however, their symptoms seemed to be too significant to be explained only by prematurity and gastrografin enema demonstrated typical features of SLCS.

The persistence of symptoms of partial intestinal obstruction for days after the evacuation of all meconium strengthens the hypothesis that this syndrome results from dysmotility rather than intraluminal obstruction [4]. Our 2 younger sister's clinical features support this assumption.

Gastrografin enema is frequently curative in SLCS. However, caution should be taken using a full-strength enema, given the risk of local tissue injury. The addition of *N*-acetylcysteine has also been used to resolve obstruction by meconium plug [7]. In our series, this treatment was inefficient, in accordance with an obstruction suspected to be secondary to an intestinal dysmotility rather than a sticky meconium. Oral erythromycin was more successful. It is widely used in low or high dose for preterm infants, considering its prokinetic effect to promote gastrointestinal motility. However, the evidence of its efficacy has not yet been proven [8].

Observations of cecal perforation or persistent obstruction after gastrografin enema, which required colostomy, were reported in the literature [4]. All authors agree that if bowel function after the passage of meconium is not entirely normal, a rectal biopsy should be performed to rule out Hirschsprung disease. In case of severe persistent intestinal obstruction, intestinal diversion should be considered. In our series, the favorable evolution without surgery in both the previous sisters helped us maintain a conservative attitude with close follow-up for the third sister. Despite the persistent intermittent obstruction, no colostomy was performed and the symptoms decreased progressively.

This is the first report of an SLCS in 3 consecutive sisters born of a nondiabetic mother. It adds to 2 previous reports of SLCS in the literature of affected twins. In addition to environmental factors (ie, maternal diabetes) described as a possible etiology of SLCS, these case reports raise the question of a genetic factor. To date, to our knowledge, no gene mutation has been described in this syndrome.

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