CASE REPORT

Neonatal Small Colon Syndrome in Infants of Diabetic Mothers: Is It Always a Transient Condition?

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Abstract

Objective: We describe an infant of diabetic mother (IDM) with an unusually severe, extensive, and persistent neonatal small colon syndrome (NSCS).

Case presentation: We report a 36-week-gestation female IDM who developed signs of intestinal obstruction at about 6 hours after birth. A contrast enema showed a small-caliber distal small intestine and colon. There was no clinical improvement over next 2 weeks, and so an exploratory laparotomy was performed; the involved bowel contained viscous meconium with pellets. Histopathological examination showed normal bowel histoarchitecture with an appropriate morphology/number of ganglion cells. A double barrel enterostomy was created, and the distal gastrointestinal tract was regularly flushed. She has since shown good improvement and has been discharged on full, exclusive breastfeeds. Laboratory investigations, including blood counts and chemistries, thyroid function, and screening for cystic fibrosis (CF) were reassuring. Our working diagnosis is an unusually severe/extensive NSCS. We have followed this infant for gastrointestinal symptoms now for 3 months since discharge.

Conclusion: Neonatal small colon syndrome may not always show prompt, spontaneous resolution. It should be included in the differential diagnosis of a newborn infant with unusually prolonged signs of intestinal obstruction. Some infants may require surgical management with ostomy formation.

Keywords: Case report, Contrast enema, Cystic fibrosis, Double barrel stoma, Gestational diabetes, Hirschsprung disease, Infant, Intestinal obstruction, Maternal diabetes, Meconium pellets, Neonate.

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Key Points

1. We describe the clinical course of an infant of diabetic mother (IDM) with an unusually severe and prolonged neonatal small colon syndrome (NSCS).
2. This infant had a prolonged period of feeding intolerance with abdominal distension, tenderness, bilious vomitings, and hypoactive bowel sounds. Radiographs showed dilated bowel loops with no rectal gas. A contrast study showed a small-caliber distal small bowel and colon.
3. An exploratory laparotomy was performed at 2 weeks of postnatal age. The involved bowel contained viscous meconium with pellets. A double barrel stoma was created and distal bowel was serially flushed. The infant improved gradually and has been discharged on full, exclusive breastfeeds.
4. We need to note that NSCS may not always show prompt, spontaneous resolution. It should be included in the differential diagnosis of a newborn infant with unusually prolonged functional intestinal obstruction.

Introduction

We recently treated an IDM with NSCS. She did not respond to conservative management, and an exploratory laparotomy had to be performed after 2 postnatal weeks. This infant had been evaluated for many differential diagnoses, but the clinical course was a reminder that NSCS should be considered as a possibility in infants with prolonged functional intestinal obstruction that extends beyond the left colon even if they are born at near-term/term gestation.[1]

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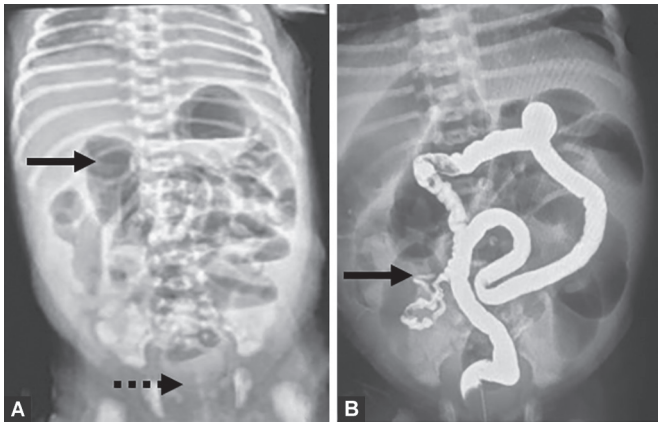
Case Description

We recently treated a 36^{+3} weeks' gestation female neonate born to a 28-year-old G_1P_0 mother by an elective Cesarean section. The pregnancy was nonconsanguineous; antenatal history was



Figs 1A and B: (A) Serial radiographs showing dilated proximal bowel loops with collapsed distal bowel (arrow) and absence of rectal shadow (broken arrow); (B) Contrast enema study showing a small caliber of the colon and distal small intestine (arrow)

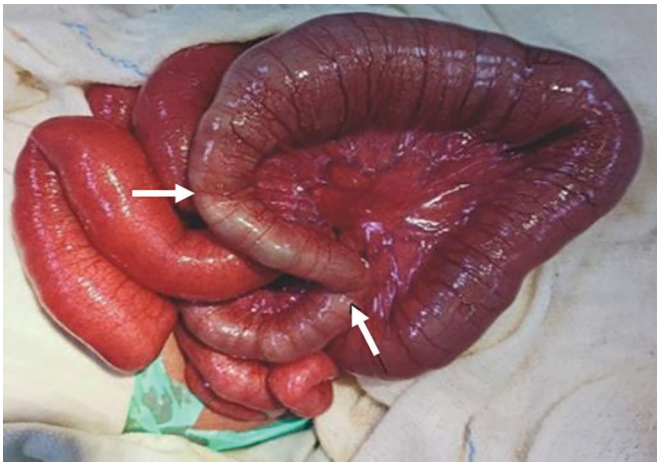


Fig. 2: Intraoperative finding of dilated jejunum and ileum till 40 cm proximal to ileocecal junction, with significant dilatation of the loop of ileum with thickened meconium just proximal to the area of narrowing

marked by gestational diabetes since 31^{+4} weeks (HbA1c = 6.14%, abnormal oral glucose tolerance test). The mother was managed with dietary modifications, but did not require any medications.[2] Her subsequent blood sugar values were within normal ranges. Antenatal sonography showed a normal amniotic fluid index and no fetal abnormalities.[3,4]

At birth, the neonate responded well without the need for active resuscitation; Apgar scores were 9 and 9 at 1 and 5 minutes, respectively. Anthropometric measurements showed that she was large-for-date (weight 3,790 gm; >95th percentile), but the head circumference (34 cm) and body length (50 cm) were appropriate for gestational age.[5] The neonate was roomed-in with the mother and started breastfeeding. However, she developed abdominal distension with bilious vomiting after 6 hours and was transferred to the neonatal intensive care unit. Her vital parameters were all within normal limits. The abdomen was distended, tense, and tender with hypoactive bowel sounds; there was no hepatosplenomegaly, and the rest of the physical examination was unremarkable. The neonate was kept *nil per oral* (NPO) with only intravenous fluids. An orogastric tube was placed, and gastric aspirates were monitored.

She first passed some pellets of meconium at 20 hours after birth but continued to have bilious aspirates. The abdominal radiograph showed dilated bowel loops with no rectal gas shadow (Fig. 1A). Serial radiographs showed minimal change. A contrast enema performed on postnatal day 2 showed a narrow, small-caliber distal small intestine and colon (Fig. 1B). Postenema, the infant intermittently passed a few stool pellets but continued to show abdominal distension, tenderness, and persistent bilious aspirates.

We measured complete blood counts, serum calcium levels, and thyroid function tests, which were all within normal ranges. Considering that we were observing an IDM, NSCS was considered a likely possibility but it was unusually extensive. Meconium plug syndrome was also plausible, but again, considering the extensive radiographic changes, it had to be a diagnosis of exclusion. Cystic fibrosis (CF) with meconium ileus was considered; the estimated incidence of CF in Asia is 1 in 10,000–12,000, but the data from India are limited.[6] In our own experience, it is seen very infrequently. The medical community in India has conflicting views about the incidence of CF in this region, and so we screen all infants with consistent symptoms.[7] This patient tested negative for the immunoreactive trypsinogen assay.[8] Hirschsprung disease was considered, but a clear transitional zone was not seen on the contrast study.[9,10]

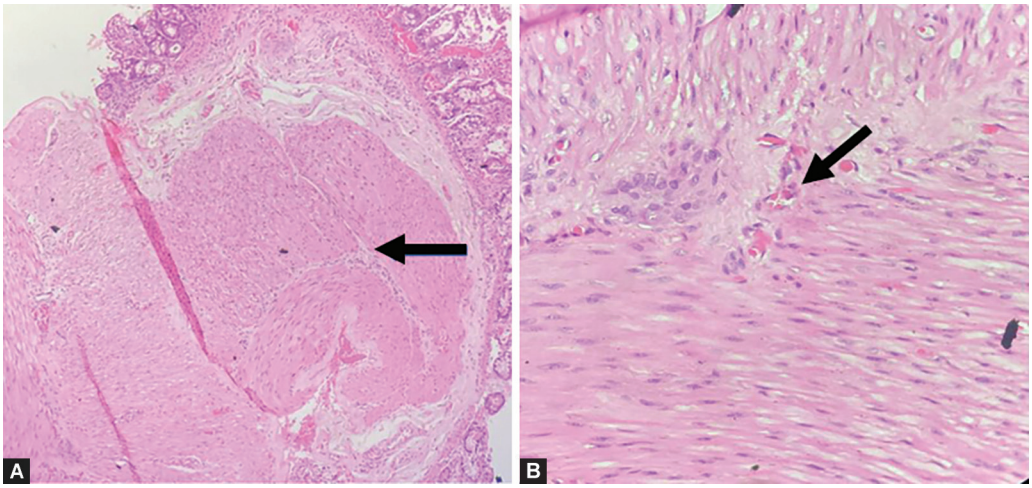
There was no improvement in neonate's condition until 2 weeks. An exploratory laparotomy was performed, which showed a large number of meconium pellets in the colon, ileum, and distal jejunum that had to be gently milked out for extraction. The small intestine showed some dilatation until about 40 cm proximal to the ileocecal junction; the distal bowel segments contained thick meconium and meconium pellets (Fig. 2). A double barrel ileostomy was performed. A punch biopsy drawn from a site distal to the stoma showed normal histoarchitecture (Fig. 3A) with normal-looking ganglion cells in usual numbers (Fig. 3B).

After surgery, the neonate was continued on parenteral nutrition. Feedings were started cautiously on postoperative day (POD) 10. The stool output initially remained limited to thick pellets, and so we irrigated the stoma several times every day with *N*-acetylcysteine.[11] She gradually began to tolerate feedings to reach full volumes by POD 14. She was discharged at 1 month after birth on full, exclusive breastfeeds. She has now been followed up for 3 months and has shown normal growth and neurodevelopment. An end-to-end anastomosis with stoma closure is being planned.

Discussion

Neonatal small colon syndrome is a condition characterized by features of large bowel obstruction in an otherwise healthy neonate; 40–50% of these patients are IDMs.[12] The pathophysiology is believed to be related to the smooth muscle constricting effects of glucagon and increased sympathetic activity resulting from hypoglycemia at birth. Additionally, functional immaturity of the ganglion cells and abnormalities in the autonomic nervous system may contribute to this condition.[13]

Early descriptions of meconium-related functional intestinal obstruction differentiated between two different presentations. Symptoms related to a meconium plug located in the colon were usually relieved after the meconium was excreted following conventional treatment. The presence of more viscous, sticky meconium in the small intestines with poor response to conventional treatment, consequently requiring surgical intervention, has also been described.[14,15]



Figs 3A and B: Normal histopathology of tissue biopsy samples: (A) Full-thickness biopsy showing an intact myenteric plexus between the muscularis externa layers (40×, hematoxylin and eosin); (B) Higher-magnification photomicrographs (400×, hematoxylin and eosin) showing normal morphology and number of ganglion cells

Our index case was a near-term infant. Her clinical features of prolonged nonobstructive intestinal dysfunction due to viscous meconium have previously been seen more often in very premature infants with a maternal history of hypertension, magnesium sulfate therapy, Cesarean delivery, and diabetes mellitus.[15,16] Yamoto et al. noted an association with intrauterine growth retardation.[17] Okuyama et al. found twin pregnancies, prolonged rupture of membranes, and the use of maternal steroids as risk factors.[18] In their cohort, low birth weight, fetal distress, maternal DM, and the maternal use of steroids independently increased the risk of these clinical features.

Neonatal small colon syndrome often shows clinical features suggestive of lower intestinal obstruction, similar to those seen in meconium plug syndrome, meconium ileus, CF, and Hirschsprung disease.[1] Except for those with Hirschsprung disease or CF, these infants begin to show spontaneous clinical improvement in 1–2 weeks following a contrast enema. Some infants may find additional benefit with a trans-anal catheter for drainage. In a case series of 105 IDMs, five had NSCS, and all showed resolution of clinical symptoms with contrast enema.[12]

The absence of a transition zone on imaging and during intraoperative examination, presence of ganglion cells that showed normal histomorphology and number, and the reassuring postoperative course have largely excluded the possibility of Hirschsprung disease.[19] Cystic fibrosis was less likely because of ethnic origin and normal screening tests; we will continue to follow and request for genetic tests if needed.[6,7] This case report brings up the dilemma of a clinician who is treating an infant with bowel dysfunction related to a well-documented, long narrow-caliber segment of the intestine, and is not showing clear evidence of recovery; it is difficult to decide between conservative management vs early surgical exploration in these cases.

To summarize, we report an IDM with an unusually severe and extensive NSCS, where signs of nonobstructive bowel dysfunction persisted for 2 weeks. An exploratory laparotomy was then performed, and viscous stool and pellets were seen in the distal small and the entire large intestine. An enterostomy with regular irrigations helped, and the infant began to feed normally after about 10 days. She has now been discharged home and has shown normal tolerance to oral feeds, growth, and development. We plan to close the stoma soon and will follow this infant closely for 3 years.[12,20] If problems recur or if she does not develop normal bladder control, we will request for genetic testing.[21]

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