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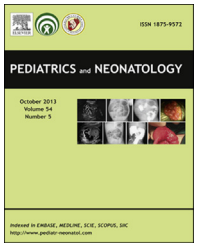

CASE REPORT

Antenatal Ultrasonographic Features Associated with Segmental Small Bowel Dilatation: An Unusual Neonatal Condition Mimicking Congenital Small Bowel Obstruction

Filomena Valentina Paradiso [a], Riccardo Coletta [a], Claudio Olivieri [a], Vito Briganti [a], Lucia Oriolo [a], Romano Fabbri [b], Alessandro Calisti [a,*]

[a] *Pediatric Surgery and Urology Unit, San Camillo Forlanini Hospital, Rome, Italy*
[b] *Radiology Department, San Camillo Forlanini Hospital, Rome, Italy*

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Segmental dilatation of the intestine (SDI) is an unusual condition presenting during the neonatal period, with symptoms of obstruction. Late diagnosed cases are also reported. The clinical polymorphism, and the lack of specificity of radiological investigations, make diagnosis difficult. Prenatal detection of abdominal cystic lesions or bowel dilatation has occasionally been reported to be associated with SDI. We herein report two cases of SDI, with a prenatal ultrasonographic suspicion of intestinal abnormality. In both infants, a dilatation of the ileum was found at surgery, without any evident site of obstruction or abnormal histology. SDI must be taken into consideration when a prenatal alert of possible bowel obstruction is not followed by postnatal clinical signs.

1. Introduction

Segmental dilatation of the intestine (SDI) is an uncommon pathology defined, in accordance with the Swenson and Rathauser criteria,[1] as limited bowel dilatation with 3—4-fold increased size, with an abrupt transition between

* Corresponding author. Pediatric Surgery and Urology Unit, San Camillo Forlanini Hospital, Via Trionfale 7210, 00135 Rome, Italy.
E-mail address: a.calisti@tiscali.it (A. Calisti).

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dilated and normal bowel and without any intrinsic and extrinsic barrier distal to the dilatation or abnormal neurological innervation. A complete recovery is observed after resection of the affected segment. A sharply delimited and markedly dilated ileum is a common intraoperative finding.

The clinical picture in the neonate may be easily confused with other more common mechanical intestinal obstructions, such as atresia and malrotation. Among older children, symptoms may be abdominal pain, growth failure and sometimes chronic gastrointestinal bleeding and anemia. In some cases, association of Meckel diverticulum with SDI has been reported.[2]

SDI associated with prenatal ultrasonographic (US) findings, mimicking prenatal volvulus, cystic mass or bowel stenosis, has rarely been described.[3–8] However, the diagnosis of SDI was confirmed at surgery in all cases.

We report two neonatal cases of SDI where an intestinal obstruction was suspected following US findings of dilated fetal loops at the third trimester. Diagnosis of SDI localized at the ileum was confirmed by postnatal imaging in one case and at surgery in the other. Clinical features and management are discussed.

2. Case Reports

2.1. Case 1

A 31-year-old woman, primigravida, underwent a routine prenatal US examination at 35 weeks' gestation, which showed a male fetus with some slightly distended ileal loops associated with polyhydramnios (Figure 1). There were no signs of ascites or intraperitoneal calcifications and normal intestinal peristalsis was observed. A distal small bowel obstruction was suspected. A 2880 g male baby was delivered by caesarean section, due to maternal reasons, at 35 weeks of gestation. Neither failure to pass meconium, nor biliary emesis, was recorded despite antenatal findings; the abdomen did not distend after meals. The baby was discharged on the 3rd day of life breast feeding regularly but, 2 weeks later, was referred back to our hospital for severe, progressive abdominal distension with conserved canalization and no vomiting. Laboratory results were normal. Physical examination showed a severely distended, soft abdomen. An abdominal plain X-ray revealed some dilated bowel loops without air-fluid levels (Figure 2A). A barium enema and upper gastrointestinal series contributed to evidence of a significant dilatation of the distal ileum, with slow progression of contrast media (Figure 2B and C). A laparotomy was performed due to increasing abdominal distension and failure to thrive, with the suspicion of malrotation and band obstruction. A segment of dilated and aperistaltic ileal segment 30 cm long was found 6 cm above the ileocecal valve. It contrasted with a normally appearing residual small bowel (Figure 3).

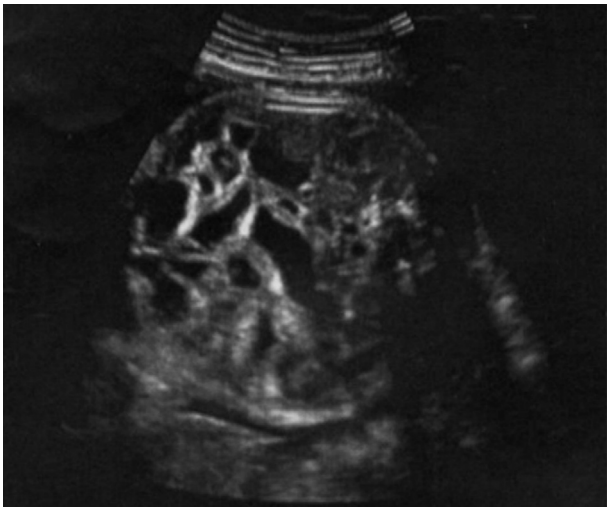

Figure 1 Case 1: antenatal ultrasound scan at 35 weeks' gestation showing distended bowel loops.

No defects of rotation or signs of mechanical obstruction were found. The dilated segment was totally resected and bowel continuity restored by end-to-end anastomosis, followed by routine appendicectomy. Histology findings were apparently normal, despite severe dilatation; ganglion cells were normally represented. Postoperative canalization was delayed. Nasogastric tube aspiration and parenteral nutritional support were required for more than a week. The baby was discharged on day 18. Growth rate is normal at 6 months follow up.

2.2. Case 2

A 2220 g female neonate was delivered vaginally at 37 weeks' gestation. No failure to pass meconium was recorded and oral feeding was regularly started.

Prenatal US findings were normal until 34 weeks' gestation, when a mild fetal growth failure was recorded and an elongated anechoic intra abdominal lesion was recorded, 48 hours before delivery, which was interpreted as small bowel dilated loops. No polyhydramnios was detected (Figure 4).

At 24 hours after birth, bilious vomiting and progressive abdominal distension developed. Laboratory finding showed high levels of C-reactive protein (CRP) (11.4 mg/dL, neonatal range = 0—0.2 mg/dL). An abdominal X-ray revealed some distended bowel loops in the upper abdomen with gasless lower quadrants (Figure 5A). The baby was admitted to our unit for a suspected small bowel obstruction. A barium enema revealed a misplaced, sub-hepatic cecum compatible with malrotation (Figure 5B). Laparotomy was carried out on day 7. Ladd's bands obstructing the third portion of duodenum were resected. In addition, a dilated atonic ileal segment, about 10 cm in length, extending to 5 cm above the ileocecal valve was found, with a threefold increase in diameter in comparison to the remaining small bowel (Figure 6). No evidence of extrinsic compression was found. The dilated segment was resected and an end-to-end anastomosis was performed. On gross pathological examination, the resected segment of the ileum did not reveal intraluminal obstructive lesions. Histology examination of the specimen showed a small heterotopic pancreatic island in the mucosa layer. Normal ganglion cells were found. The postoperative course was uneventful. The baby was discharged after 1 week from

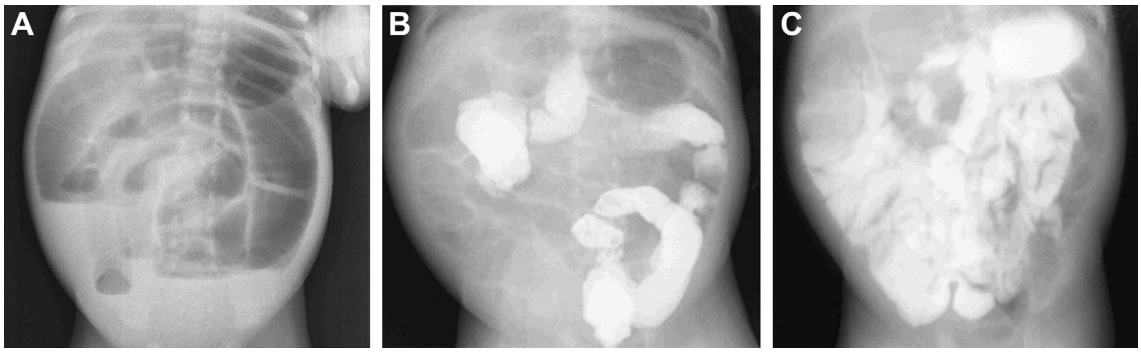

Figure 2 Case 1: (A) plain abdominal X-ray showing massive bowel loops distension; (B) barium enema demonstrating an undistended, normally rotated colon; (C) upper gastrointestinal series shows pooling of the contrast media in distal ileal loops.

surgery and was breast feeding. The baby is well and gaining weight at 6 months' follow up.

3. Discussion

SDI is a rare event, characterized by a sharply demarcated segmental dilatation of the gastrointestinal tract, without any evidence of mechanical obstruction or abnormal neurologic innervation. More frequently, it involves the ileum, but involvement of the duodenum or jejunum is rare.[9] Its etiology is unclear, although most authors believe that it is caused by an extrinsic prolonged intrauterine compression of a loop of bowel at both ends.[10] Since the first description of SDI by Swenson and Rathauser in 1959,[1] around 150 reports have been published. Approximately 50% of the SDIs are seen in the first days of life. Both sexes are affected with a slight male predominance. SDI in the neonatal period usually presents signs of intestinal obstruction; however, in older children it presents abdominal pain, failure to thrive and anemia, due to chronic bleeding because of the presence of heterotopic gastric mucosa of the dilated loop.[11,12] Associated malformations, such as omphalomesenteric remnants or malrotation, have been reported.[10,11] In our second case, SDI was associated with malrotation.

To the best of our knowledge, only six cases of SDI associated with prenatal intra abdominal lesions have been reported to date.[3–8] In all, antenatal findings were aspecific and could only alert to a possible gastrointestinal tract anomaly.

Clinical onset of SDI is often delayed. Most patients with this anomaly can be discharged after birth on regular oral feeding, without any symptoms demanding deeper investigation, as our first case.[4] However, in our second case, despite prenatal features of dilated bowel, SDI was an occasional finding at surgical exploration, due to duodenal obstruction secondary to Ladd's bands.

On abdominal plain X-ray, SDI may be associated with dilated bowel loops, with or without air-fluid levels, as in other more common forms of congenital intestinal

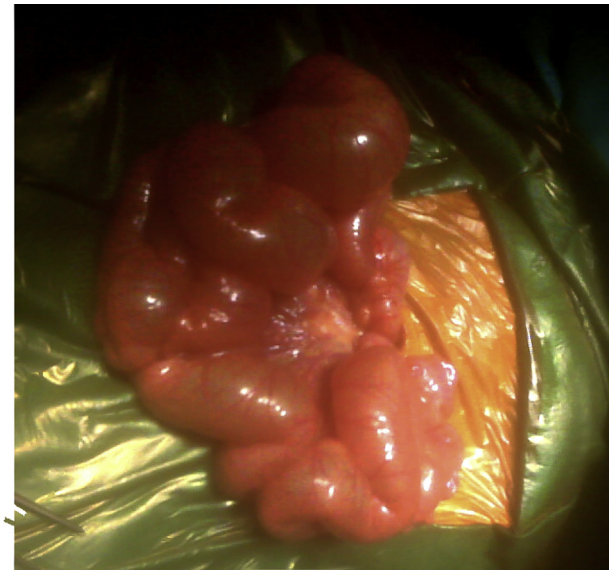

Figure 3 Case 1: intraoperative picture of a 30 cm long segmental dilatation of the distal ileum. Transition of normal bowel on both ends did not show any sign of mechanical obstruction.

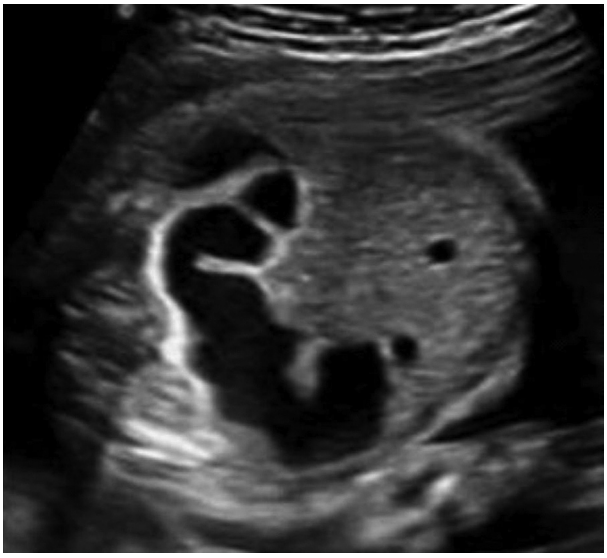

Figure 4 Case 2: antenatal ultrasound scan at 38 weeks' gestation showing an elongated anechoic intra-abdominal lesion, compatible with a distended bowel loop.

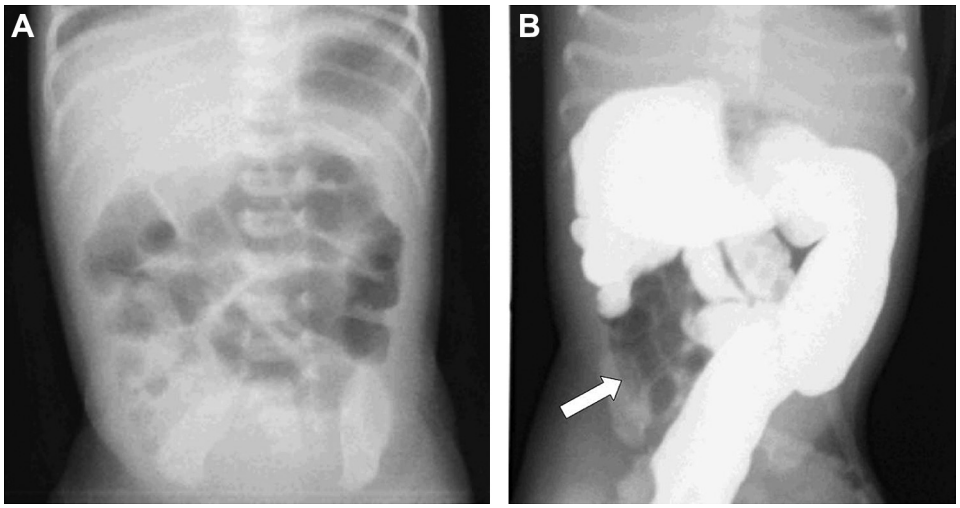

Figure 5 Case 2: (A) upper abdominal distended bowel loops and no gas in the lower abdomen; (B) barium enema study showing incompletely rotated colon with a sub-hepatic cecum. No reflux into the distal ileal loops, which appear distended by gas (arrow).

obstruction. Preoperatively, barium enema studies can identify the dilated segment, as in our first case. Radiological investigation was omitted at birth, in the absence of symptoms, but was performed 2 weeks after, following the onset of clinical signs of suspected occlusion. Radiological findings included marked segmental dilatation of the small bowel.

SDI is occasionally found at surgical exploration in patients with an unexplained obstructive intestinal pattern, as in our second case. Segmental dilatation of the small bowel is retrospectively correlated with nonspecific prenatal findings and equivocal postnatal symptoms.

Histology of the resected specimen is usually reported to be normal; some anomalies are referred, such as hypertrophied muscular layer[3] or heterotopic esophageal or gastric mucosa or cartilaginous foci.[10] An ectopic island of pancreatic tissue was found in the dilated intestinal segment of our second case, without any resemblance to Meckel diverticulum.

Treatment of SDI is simple, consisting of the resection of the dilated segment and end-to-end anastomosis, followed by an uneventful postoperative course.

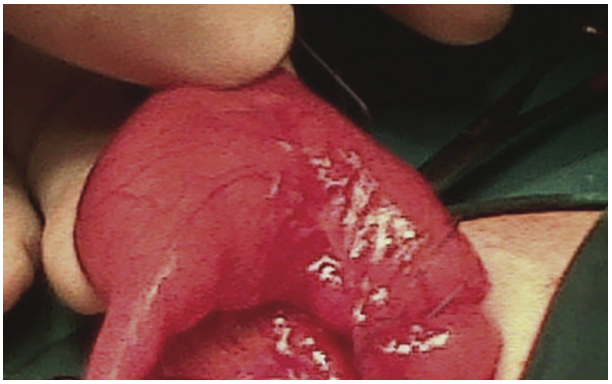

Figure 6 Case 2: intraoperative finding of sharply ending segmental intestinal dilatation.

In conclusion, SDI is often represented by an increasing, intermittent, intestinal dysfunction that requires surgical exploration to be clearly differentiated by other most common mechanical obstructions. Limited forms of SDI may remain undiagnosed for a long time, since the symptoms are scarce and discontinuous. Our two cases are added to the small reported series of SDI associated to aspecific prenatal US findings of small bowel dilatation. These signs are not helpful in discriminating SDI from other more common forms of mechanical obstruction preoperatively. Clinicians may be misled by the delayed clinical onset of SDI and can omit a thorough diagnostic work up, despite prenatal alert. This approach may be justified in asymptomatic cases; however, parents must be advised to refer to the hospital if any symptoms appear after discharge.

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