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CASE REPORT

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A rare cause of neonatal feeding intolerance: congenital segmental intestinal dilatation—a case report

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Abstract

Background Congenital segmental intestinal dilatation (CSID) is a rare cause of neonatal bowel obstruction. Patients can present in the early newborn period with feeding intolerance and abdominal distension. Diagnosis is challenging due to its rarity and non-specific clinical and radiological findings in a well neonate. Surgical excision remains the mainstay treatment for this disease.

Case presentation We report a case of a newborn male with congenital segmental ileal dilatation, who presented with abdominal distension and feeding intolerance. He was born preterm at 32 weeks and was treated for presumed sepsis due to a premature rupture of the membrane. The patient developed persistent abdominal distension upon initiation of feedings with large amount of gastric aspirates. Otherwise, there was no delay in the passage of meconium. Serial abdominal radiographs showed persistent focal bowel dilatation on the right side. He was initially diagnosed with small bowel atresia. Ultrasound abdomen and lower gastrointestinal contrasted study showed focal ileal dilatation with no evidence of pneumoperitoneum. Intraoperative findings revealed a segmental ileal dilatation with no evidence of mechanical obstruction. The abnormal dilated bowel was excised and sent for histopathological examination. He recovered well after surgery.

Conclusion Diagnosis of CSID can be challenging due to its clinical polymorphism and non-specific radiological findings. CSID should be suspected in neonates with feeding intolerance and focal intestinal dilatation without clinical features of sepsis, peritonitis, or mechanical obstruction. Surgical excision is curative with a good outcome.

Keywords Segmental, Bowel, Dilatation, Obstruction, Pneumoperitoneum, Case report

Background

Congenital segmental intestinal dilatation (CSID) is a very rare entity with unknown pathoetiology [1, 2]. It can occur anywhere from the duodenum to the distal large bowel [1]. Swenson and Rathauser first described a segmental dilatation of the colon in 1959 [3]. Sjölin and Thoren reported a case of segmental ileal dilatation in 1962 [4]. In 1973, Rossi and Giocomoni first described a case of segmental dilatation of jejunum [5].

CSID is characterized by a limited (circumscribed) bowel dilatation with a three to fourfold increase in diameter, an abrupt transition between normal and dilated

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segments, neither intrinsic nor extrinsic perilesional obstruction, as well as the presence of normal ganglionic cells of the gastrointestinal autonomous plexus [6]. Children can have a spectrum of clinical polymorphism, with almost 50% presenting in the neonatal period [2]. Involvement of the ileum is most common [1].

Treatment requires complete surgical excision. Histopathological examination of the involved bowel often demonstrates normal neurological innervation with thickening or thinning of the muscular layer [7]. Prognosis is good after complete surgical excision.

Case presentation

A newborn male presented with feeding intolerance and abdominal distension on day 2 of life. The patient was a first twin, born preterm via spontaneous vaginal delivery at 32 weeks with a birth weight of 2.14 kg. Antenatally, the mother had anemia during pregnancy and was on iron supplements. She was given progesterone for the history of threatened miscarriages in two of her previous pregnancies. The antenatal ultrasound was unremarkable. The patient was treated for presumed sepsis as the mother presented in labor with preterm premature rupture of membranes.

At birth, the patient had mild respiratory distress syndrome and required oxygen therapy. He passed meconium within 24 h and was started on nasogastric tube feedings on day 1 of life. Subsequently, he developed abdominal distension and large amounts of gastric aspirates. Despite the distension, the patient was still passing meconium daily. On examination, the abdomen was distended but soft with no signs of peritonism. Full blood count, renal profile, and C-reactive protein were normal. Blood culture was negative. Serial abdominal X-rays revealed a persistent lucency on the right side of the abdomen (Fig. 1). An initial diagnosis of small bowel atresia was made. Further imaging with an ultrasound abdomen showed a segmental bowel dilatation with no peristalsis. The lower gastrointestinal contrast study showed reflux of contrast into a dilated segment of the small bowel, and the colon was normal in caliber with a normal rectosigmoid ratio (Fig. 2).

Laparotomy findings revealed an isolated dilated segment of the ileum, 20 cm in length, located 60 cm from the ileocecal valve. The dilated segment was 3×the diameter of the normal bowels. The proximal and distal bowel loops were normal in caliber (Fig. 3). There was no intraluminal mechanical obstruction. Resection of the abnormal segment was done with primary end-to-end anastomosis. Postoperative recovery was uneventful. Feeding was commenced without any problems, and he was discharged home after good weight gain. Histopathological interpretation of the resected specimen showed segmental dilatation of the ganglionic ileum with variable thinning of both layers of the muscularis propria (Fig. 4). He was well and thriving at 6 months of follow-up.

Discussion

Congenital segmental intestinal dilatation (CSID) is a rare entity. It is defined as a focal dilatation of any part of the gastrointestinal tract with no evidence of mechanical obstruction. [8] Swenson and Rothauser established criteria for diagnosis of this condition [3] such as the following: (a) limited bowel dilatation with a 3- to four-fold increase in diameter, (b) abrupt transition between the dilated segment and normal bowel, (c) no intrinsic or extrinsic barrier distal to the dilatation, (d) a clinical picture of intestinal occlusion or subocclusion, (e) normal

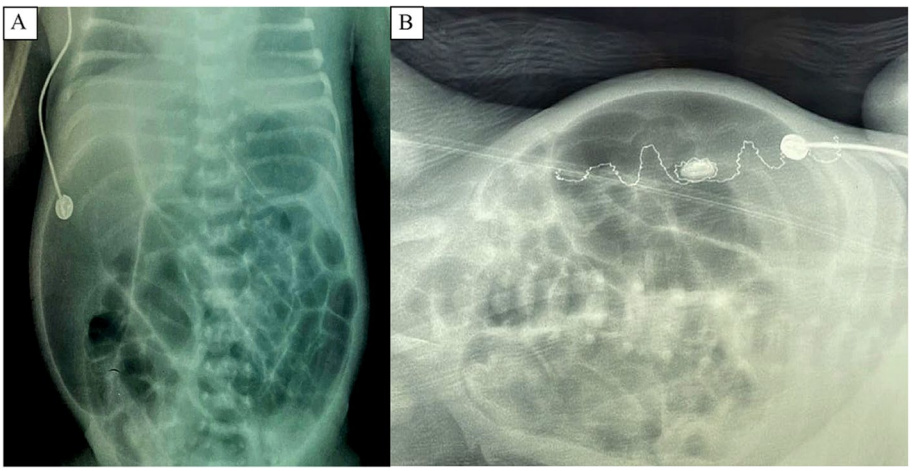

Fig. 1 Abdominal radiograph (**A** in supine position) and (**B** in left lateral decubitus) shows focal bowel dilatation at the right sided abdomen overlapping with other bowel loops mimicking Rigler sign. No intramural air or portal venous gas

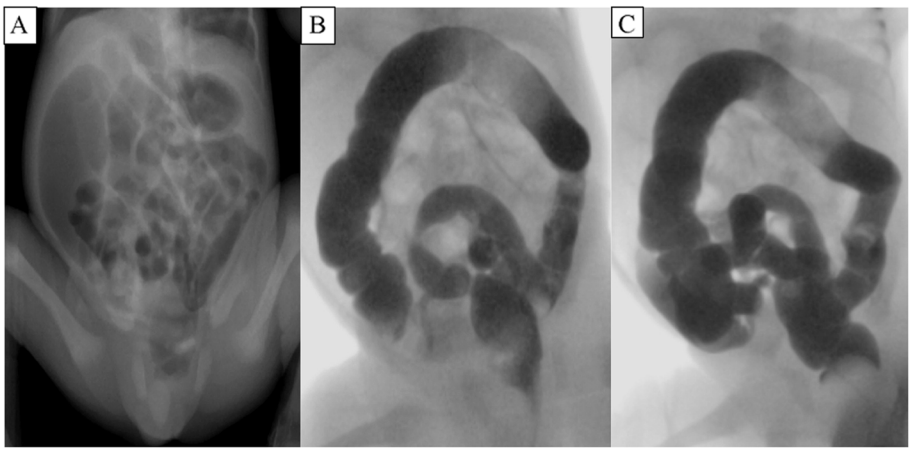

Fig. 2 **A** A preliminary film of lower gastrointestinal contrast study shows focal bowel dilatation of the ileum. The rest of bowel loops were not dilated. No pneumoperitoneum. **B**, **C** Lower gastrointestinal contrast study in supine and right lateral decubitus position showed normal colon with no evidence of microcolon or filling defect seen within

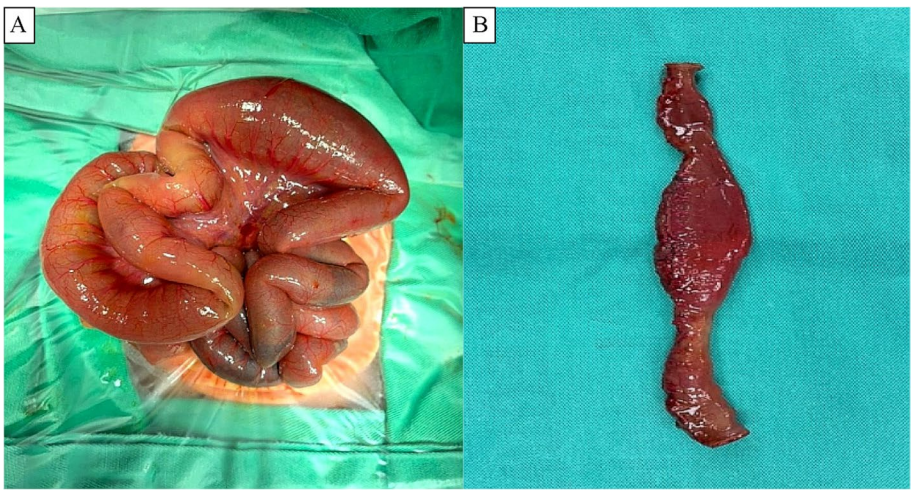

Fig. 3 **A**, **B** Operative findings confirmed a focal segmental ileal dilatation with no extrinsic nor intrinsic barrier. The rest of the bowel loops were normal

neuronal plexus, and (f) complete recovery after resection of the affected segment. Their true pathoetiology remains unknown. Transient embryonic structures such as vitelline vessels and omphalomesenteric bands resulting in external compression to both ends of the bowel have been proposed [9, 10]. The role of the enteric nervous system was investigated, and no neurogenic etiology could be attributed to congenital segmental bowel dilatation [11].

The most frequent site involved is the ileum, followed by the colon, jejunum, and duodenum [6, 12]. This entity occurs in both sexes with a slight male gender preference [1, 12]. Almost 50% of congenital segmental bowel dilatation is present in the newborn period [13]. However, some patients may only become symptomatic during infancy or adolescence [1]. The clinical presentation of segmental bowel dilatation depends on its size, location, and presence or absence of complications. Bowel obstruction is the commonest clinical presentation during the neonatal period. Older children may present with abdominal pain, anemia, and failure to thrive [8]. Heterotopic gastric mucosa could be the cause of anemia in patients with congenital segmental bowel dilatation [1, 13]. Omphalomesenteric remnants and malrotation can be associated with congenital segmental bowel dilatation [1]. Mathur reported a case of segmental dilatation of the cecum and ascending colon, with concomitant high anorectal malformation and colovesical fistula [14]. The

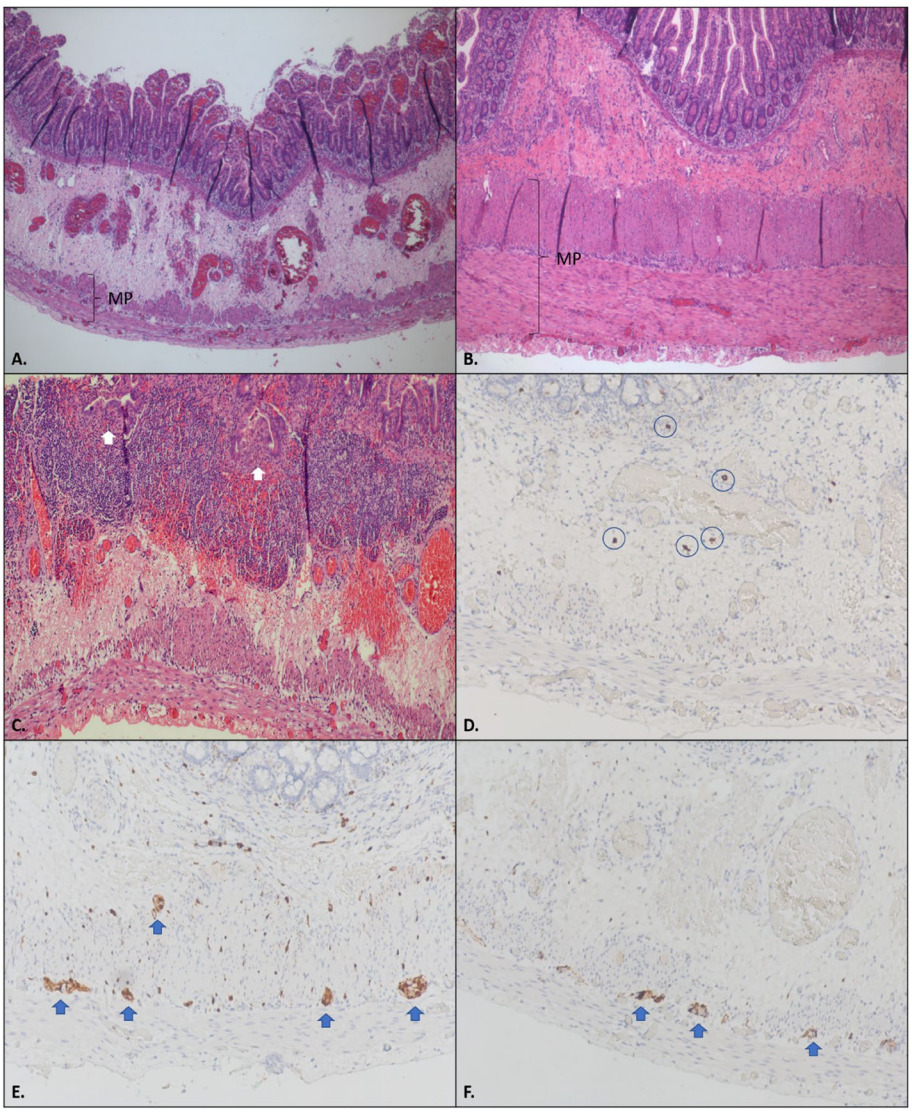

Fig. 4 Dilated small bowel segment with intact but variably thinned layers of the muscularis propria (MP) (**A**) as compared to non-dilated areas with normal thickness muscularis propria (MP) (**B**). **C** Prominent submucosal lymphoid follicles observed in the dilated segment with presence of cryptitis/crypt abscess (white arrow). Paucity of spindled interstitial cells of Cajal within the muscularis propria as demonstrated by lack of c-kit (CD117) immunostain. **D** Mast cells in the submucosa and lamina propria are highlighted (circles). Submucosal and intermyenteric ganglion cells (blue arrow) are present and normal in distribution highlighted by antibodies for S-100 (**E**) and Calretinin (**F**)

large bowels distal to the dilatation were normal, differentiating it from a congenital pouch colon. Sergi reported a case series of 4 patients diagnosed with congenital segmental bowel dilatation, associated with congenital heart disease [15].

Early detection of CSID in this patient was a challenge due to the non-specific nature of symptoms. Abdominal distension was present with large amounts of gastric aspirates after feeding with no abdominal tenderness or guarding. The abdominal radiograph showed dilated bowel loops which can also occur in patients with necrotizing enterocolitis, Hirschsprung's disease, ileal atresia, or meconium ileus [8]. Congenital segmental ileal dilatation can also mimic total colonic aganglionosis. This diagnosis is unlikely as our patient has passed normal meconium regularly since birth.

The abdominal radiograph in this patient showed focal bowel dilatation with overlapping gas-filled bowels making the wall contours more striking, mimicking Rigler's sign. Pneumoperitoneum has been ruled out from clinical examination and ultrasound abdomen which showed a segmental small bowel dilatation with no free air or free

fluid. The lower gastrointestinal contrast study revealed the flow of contrast into a segment of dilated small bowel dilatation with no microcolon. These findings exclude a small bowel atresia. Computed tomography should be avoided in neonates to reduce radiation exposure. The diagnosis of CSID is challenging and not easy during the intrauterine or postpartum period. Antenatal ultrasound may show minor cystic changes in the fetal abdomen. Differential diagnosis includes duplication cyst, meconium pseudocyst, mesenteric cyst, meconium ileus, or duodenal atresia [15]. In most instances, congenital segmental bowel dilatation is identified incidentally during a surgical operation.

Complete resection of the dilated segment with end-to-end anastomosis remains the main treatment. In the majority of patients, the prognosis is good after surgical resection with full recovery. Usually, histopathological evaluation of the resected segments revealed no significant abnormalities [8]. In this patient, there was segmental dilatation of the ganglionic ileum with variable mural thinning, partial loss of ICC, focal acute ileitis, and prominent Peyer's patches. This is consistent with reports of other investigators which showed hypertrophied or very thin muscle layer in the affected segment [8]. Focal acute ileitis with the presence of cryptitis/crypt abscess in the involved segment of CSID may represent non-specific inflammatory changes. In this patient, there was no local vacuolization in the smooth muscle layer, mucosal abnormality, or supernumerary intestinal muscle coats [16]. Our case report suggests the role of myopathy as the etiology of CSID. Other causes may include ectopic gastric mucosa which causes an interruption in the neural and muscular network, presence of heterotopic esophageal mucosa or cartilaginous foci, and partial loss or complete loss of ICC in the dilated bowel [8, 17, 18].

Conclusion

Congenital segmental ileal dilatation is a rare entity in the pediatric age group. Diagnosis can be challenging in view of its clinical polymorphism and non-specific radiological findings. Radiological features of a segmental ileal dilatation with no microcolon may differentiate it from other more common causes of neonatal bowel obstruction. The knowledge of CSID may be helpful in early detection and intervention. Surgical resection with histopathological examination confirms this diagnosis. The outcome is favorable after local resection of the involved segment with end-to-end anastomosis.

Abbreviations

CSID Congenital segmental intestinal dilatation
DCDA Dichorionic diamniotic twin pregnancy
PPROM Preterm premature rupture of the membranes
CPAP Continuous positive airway pressure
ICC Interstitial cells of Cajal

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Nil

Authors' contributions

THH: conception, design of the work, acquisition, analysis of the data, and preparation of the manuscript. TSK: acquisition and analysis of the data, drafting, and critical review. RAH: analysis of the data and critical review. MYMY: analysis of data and critical review. PP: analysis of data and critical review. TPI: analysis of data and critical review. The authors have read and approved the final manuscript.

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written consent had been obtained from the parent of the study participant.

Competing interests

The authors declare that they have no competing interests.

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