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Case report

In utero ileal volvulus and intestinal perforation associated with enteric duplication cyst presenting with preterm labour and acute abdomen in newborn

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SUMMARY

Enteric duplication cysts are a rare cause of intestinal obstruction in the neonatal period. We present the unusual case of an in utero ileal volvulus secondary to an enteric duplication cyst causing an acute abdomen in a 35-week estimated gestational age newborn female delivered to a mother in preterm labour.

BACKGROUND

The incidence of enteric duplication cysts is 1 in every 4500–5000 births, most predominantly affecting males. The most common location of enteric duplication cysts is the ileum (40% of cases).[1–4] First reported by Fitz in 1884[5] and subsequently described by Ladd in 1937 as a phenomenon of 'alimentary tract duplications',[6] 80% of enteric duplication cysts present in the first 2 years of life.[2 7] Duplication cysts are hollow, epithelium-lined, cystic (90%) or tubular (10%) structures surrounded by a smooth muscle layer occurring anywhere along the alimentary tract from the mouth to the anus, often sharing a common blood supply with the adjacent bowel. Many hypotheses exist as to the pathophysiology of duplication cysts including the split notochord theory of traction between gut endoderm and overlying structures resulting in cyst formation, intrauterine ischaemic insult or small diverticula on the anti-mesenteric side of intestine during embryologic development which persist to become a duplication cyst.[8]

CASE PRESENTATION

A 2.2 kg, 35-week newborn female was born via spontaneous vaginal delivery in respiratory distress with a distended and discoloured abdomen. The pregnancy was complicated by preterm labour. However, there were no reported abnormalities during prenatal ultrasounds. Apgar scores were 6 and 7 (1 and 5 min). Meconium was noted in the amniotic fluid at birth. The patient was transported to our neonatal intensive care unit for higher level of care. Resuscitation was initiated en route with a fluid bolus, antibiotics, dextrose infusion (blood glucose was 10–20 mg/dL) and placement of an orogastric tube. Following decompression of 50 mL bilious fluid and 40 mL air from the stomach, the patient's distal perfusion improved. Physical examination revealed a distended, discoloured, tender and tense abdomen (figure 1). Laboratory values revealed haemoglobin 81 g/L, white blood cell 47 000/mm^3 (25% neutrophils, 10% bands, 18% myelocytes, 2% metamyelocytes, 5% blasts), base deficit 12.9, pH 7.15 and bicarbonate 14.4. Supine and lateral radiographs demonstrated a paucity of gas with no air beyond the second portion of the duodenum and no pneumoperitoneum (figure 2).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for a neonate presenting with a bowel obstruction includes malrotation with midgut volvulus, duodenal/intestinal atresia, meconium ileus, alimentary tract duplication, Hirschsprung's disease and anorectal malformation. Physical exam is useful in evaluating degree of abdominal distension and for evidence of imperforate anus and peritonitis. Imaging modalities can also aid with diagnosis. An abdominal radiograph with 'double-bubble' and no distal air is suggestive of duodenal atresia. Pneumoperitoneum can be seen with bowel perforation. An upper gastrointestinal series demonstrating a duodenum failing to cross midline and a 'corkscrew sign' is seen with malrotation with midgut volvulus. A contrast enema demonstrating a microcolon with dilated loops of proximal bowel can be seen with meconium ileus and intestinal atresia. Our case presented with abdominal discoloration/distension, peritonitis, bilious output from the gastric tube and a radiograph with a paucity of gas beyond the proximal bowel. A decision was made to forego imaging and proceed with exploratory laparotomy for definitive diagnosis and treatment given a high suspicion for bowel obstruction secondary to volvulus and ischaemic bowel.

TREATMENT

Given clinical signs of an acute abdomen with peritonitis, the decision was made to proceed with emergency exploratory laparotomy with a transverse abdominal incision. On entering the peritoneal cavity, a large volume of haemorrhagic succus was noted. A necrotic distal ileal segment with mesenteric volvulus was visualised with dilated proximal small bowel. There was normal intestinal rotation. The distal end of proximal viable small bowel was brought up as a jejunostomy, 35 cm distal to the ligament of Treitz. The remaining 3 cm of distal ileum was oversewn with a prolene suture for later identification and reanastomosis; a mucous

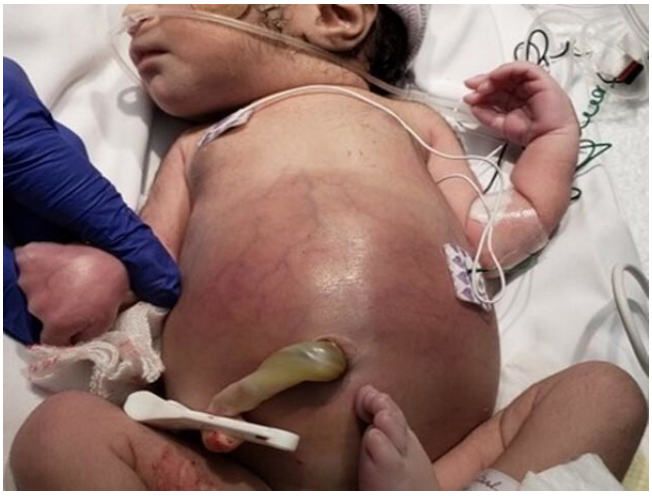

Figure 1 The patient was born with a distended, discoloured, tender and tense abdomen.

fistula was avoided given the short length of this segment and the concern that takedown of the fistula could further shorten the ileum as well as impede our ability to preserve the ileocecal valve. A tunnelled central venous catheter was placed for resuscitation and the anticipated need for parenteral nutrition in the setting of short gut. Histopathological examination of the specimen showed chronic inflammation and necrosis of the small intestine with haemorrhagic congestion. Further evaluation of the necrotic ileal segment revealed two adjacent intestinal lumens. Trichrome staining demonstrated a shared common wall consistent with an enteric duplication cyst (figure 3).

OUTCOME AND FOLLOW-UP

Postoperatively, enteral feeds were initiated, advanced and optimised for her short gut syndrome. Initially, she continued to dump from her stoma and could not be weaned off parenteral nutrition. At 3 months of age, she underwent jejunostomy closure with an end-to-Cheatle jejunoileal anastomosis. Total estimated small bowel length was 38 cm—including 35 cm of jejunum distal to the ligament of Treitz, 3 cm of distal ileum and a preserved ileocecal valve. She was able to achieve goal oral feeds with appropriate weight gain, no longer requiring parenteral nutrition or gastrostomy tube feeds by 4 months of age.

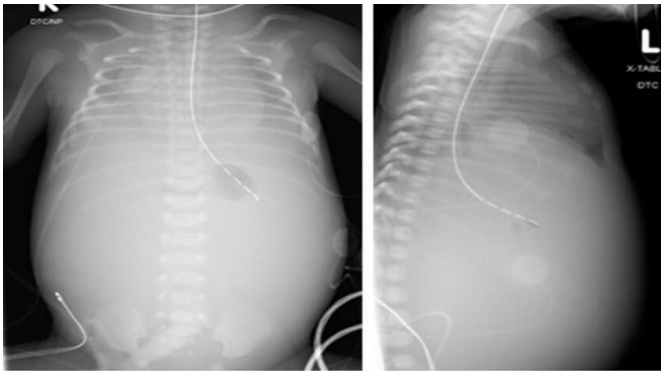

Figure 2 Supine and lateral abdominal radiographs demonstrating paucity of gas and significant abdominal distension with no evidence of pneumoperitoneum.

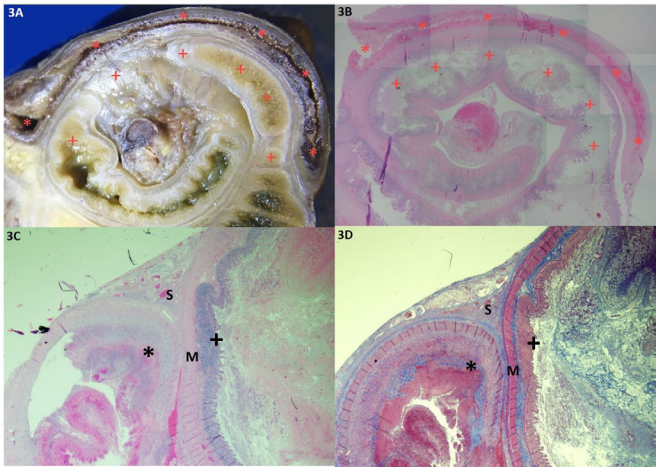

Figure 3 (A) Photograph of cross-section of necrotic ileal mass with lumen of intestinal duplication cyst (*) and adjacent ileum (+) highlighted. (B) Composite micrograph (H&E stain, 5×) of cross-section of necrotic ileal mass with lumen of intestinal duplication cyst (*) and adjacent ileum (+) highlighted. (C) Micrograph (H&E stain, 40×) of shared common wall highlighting serosal (S) and muscular layers (M) (lumen of intestinal duplication cyst (*): lumen of ileum (+)). (D) Micrograph (trichrome stain, 40×) of shared common wall highlighting serosal (S) and muscular layers (M) (lumen of intestinal duplication cyst (*): lumen of ileum (+)). The muscularis mucosae of the duplication cyst possesses only circumferential smooth muscle and lacks longitudinal smooth muscle.

DISCUSSION

Our case study presents a unique case of an enteric duplication cyst causing in utero volvulus leading to intestinal obstruction and perforation in a newborn. The quality of the haemorrhagic succus and the moderately dense intra-abdominal adhesions suggested that the process occurred prior to delivery. The presence of meconium in the amniotic fluid suggested that the obstruction was relatively recent. We speculate that preterm labour was likely precipitated by the intestinal perforation. Despite the short bowel length following intestinal resection, the patient was successfully advanced to goal feeds and weaned off parenteral nutrition.

While enteric duplication cysts can be found on prenatal ultrasound, they are usually asymptomatic and can be followed during the postnatal period with serial ultrasounds and elective resection. Radiographic studies with plain films are often non-specific. Prenatal diagnosis by ultrasound only recognises 20%–30% of enteric duplication cysts, which can often be mistaken for other cystic structures such as ovarian, renal, mesenteric or omental cysts.[9] Postnatal sonographic findings of a cyst sharing a common muscular wall with an adjacent segment of bowel are highly suggestive of an enteric duplication cyst.

There are only a few case series and reports of neonatal bowel obstruction caused by enteric duplication cysts—none of which describe an in utero process of bowel ischaemia and perforation precipitating preterm delivery.[4 5 7 9–11] The majority of neonatal bowel obstructions manifest after birth and are often attributable to intestinal atresia, malrotation or meconium disease. While enteric duplication cysts are an uncommon aetiology of neonatal bowel obstruction, the majority are located in the ileum with a reported mechanism of volvulus or mass-effect causing extrinsic compression of adjacent bowel.[7] The rarity of this in

utero disease process calls for timely recognition and surgical intervention to optimise neonatal outcomes.

Learning points

- Enteric duplication cysts can manifest at any time of life (including the in utero and neonatal period) with a myriad of different complications including bowel obstruction (volvulus, intussusception and extrinsic compression of adjacent bowel), ulceration/bleeding secondary to heterotopic gastric mucosa and infection.
- In non-emergent cases, a diagnostic ultrasound can help support the diagnosis of an enteric duplication cyst if a shared common wall with an adjacent segment of bowel is visualised.
- Management requires complete surgical resection of the cyst and adjacent bowel with primary anastomosis if feasible. In cases where the adjacent bowel cannot be safely resected, the cyst and its mucosal lining should be excised.

Contributors SGK wrote the manuscript and participated in surgery and patient follow-up. MC obtained and formatted gross specimens and histopathology images including captions; read and approved final version of manuscript. DAD operated on the patient, supervised the work, revised and reviewed the manuscript. SF supervised the overall histopathology work; read and approved final version of manuscript.

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