CASE REPORT

Antimesenteric gastrointestinal tract duplication undergoing non-ulcerative perforation

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Abstract

Duplications of the gastrointestinal tract are rare malformations, most commonly presenting as cystic structures growing within the smooth muscle wall. Very rarely, they are completely detached from the tract. Several theories have been proposed regarding their embryological development, but no single one has been able to account for all of the described variants. The most common type of duplication is related to the small bowel and develops at its mesenteric border, assuming a spherical or tubular shape. Their clinical manifestations vary, depending mainly on their localization and size. Most commonly, they cause subacute abdominal pain and intestinal obstruction in children of less than two years of age. We present a case of an 8.5-year-old girl, investigated for right lower quadrant abdominal pain. On ultrasound scan, a cystic mass indicative of a duplication cyst was discovered and she underwent a laparotomy. A tense cystic spherical mass 2.2 cm in diameter was excised from the terminal ileum, 4 cm from the ileocecal valve. The cyst had the characteristics of a gastrointestinal tract duplication, except from the fact that it was located on the antimesenteric border of the intestine. On the other hand, the lesion did not present the characteristic features of a Meckel's diverticulum. According to our knowledge, this is the first report of an intestinal duplication cyst appearing on the antimesenteric intestinal border.

Keywords: small bowel, malformation, acute abdomen, abdominal cyst, child, intestinal obstruction.

Introduction

Gastrointestinal duplications (GIDs) are rare congenital anomalies, with a reported incidence of 1/4500 [1, 2]. They can occur anywhere from the mouth to the anus. According to data presented in case series by many authors, the most common site is the ileum (17–63%), followed by the esophagus (3–20%), large bowel (5–13%), jejunum (5–10%), stomach (3–7%), duodenum (5–6%) [2–12]. In 7% of cases, multiple GIDs may be present [3]. According to some series, there is a male preponderance from 57% to 70% [4–7].

GIDs that become symptomatic may do so as early as the first day of life [8]. 24–40% are diagnosed during the neonatal period [7, 9], 60% are diagnosed by six months of age [10], 71.4% are symptomatic by the first year of life [5] and 85% are diagnosed before the age of two years [10]. A median of three months of age has been calculated in a large series [7]. With the increasing use of prenatal ultrasound scan, antenatal diagnosis is reached in a significant number of cases [4]. On very rare occasions, they may cause morbidity during adulthood [13].

Symptoms are often related to their location. Oral and esophageal lesions may cause respiratory difficulties, dysphagia, and hematemesis, whereas lower gastrointestinal lesions may cause abdominal pain, nausea, vomiting, and may be the cause of bleeding, perforation, or obstruction, many times mimicking other more common acute or chronic pathologies such as intussusception, volvulus, hypertrophic pyloric stenosis, appendicitis, pelvic abscess, diverticulitis, achalasia, and Hirschsprung's disease [5, 10]. A mobile abdominal mass may be palpated in approximately half of the patients [10].

In this report, we describe the case of an 8.5-year-old girl with an ileal lesion that had the characteristics of a GID. However, the lesion was located on the anti-mesenteric border of the intestine.

Case presentation

An 8.5-year-old, 38 kg girl presented to the Emergency Department with a two-day history of mild colicky lower abdominal pain. Anorexia was reported as an associated symptom, but no fever, changing bowel habits or urinary symptoms were reported. The child had already undertaken blood tests demonstrating normal white blood cell count (6.7×10^9/L, 60% neutrophils, 29% lymphocytes), platelet count (337×10^9/L), erythrocyte sedimentation rate (5 mm/h), C-reactive protein (CRP) level (0.39 mg/dL) and normal serum albumin. Blood biochemistry was normal and so was urinalysis. Abdominal and pelvic ultrasound performed on the day before her admission identified slightly edematous mesenteric lymph nodes.

On examination, body temperature, heart rate and blood pressure were normal. The abdomen was soft without any guarding and demonstrated mild tenderness over the right lower abdominal quadrant and towards the right side of the bladder. Rebound tenderness was not noted. No mass was palpated.

The patient was admitted for observation. Her condition remained stable during the next day. There were three episodes of colicky pain and the findings on palpation were similar to the day before. A new blood test was performed yielding similar results except that CRP level was 0.6 mg/dL. She had normal bowel movements and no blood was present on stool. An abdominal ultrasound

was performed which demonstrated the presence of a cystic lesion in the right lower quadrant situated between the intestinal loops, showing the typical double layer wall, identical to the intestinal wall. The cyst was sharing a common wall with the distal ileum (Figure 1). The diagnosis of GID cyst was reached.
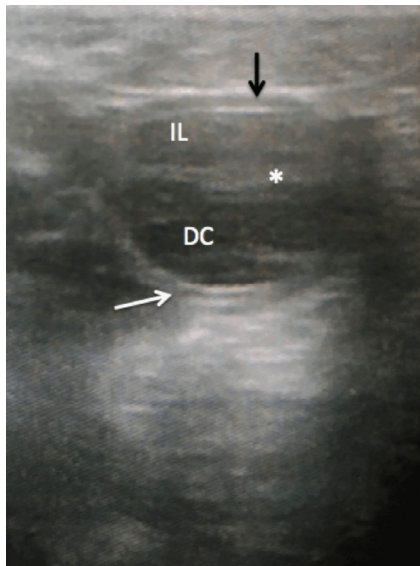
Figure 1 – *Abdominal ultrasound scan. Cystic lesion in the right lower quadrant. DC: Duplication cyst; IL: Ileum; Black arrow: Ileal wall; White arrow: Cyst wall; Asterisk: Common wall.*

On laparotomy, through a transverse incision crossing the McBurney's point, a small quantity of clear peritoneal fluid was found in the peritoneal cavity, which was aspirated. The cecum and distal ileum were exteriorized. The appendix was normal, without any signs of inflammation. A tense spherical cystic mass was observed on the terminal ileum, 4 cm from the ileocecal valve, arising from the intestinal wall, on its antimesenteric side. Part of the mass was protruding outwards and part towards the intestinal lumen (Figure 2). It was of 2.3 cm in diameter. A small perforation of the cyst wall was observed at the extra-luminal part and a drop of mucus appeared at the perforation point, but not pouring out. Apparently, a protrusion of the cyst's mucosa was sealing the perforation. The mass was tense as if almost no mucus was emptied. Its intraperitoneal surface was covered smoothly by serosa and in continuity with the seromuscular wall of the intestine. No mesenteric fold with vessels was reaching towards it. No abnormality was present by the mesenteric border of the native intestine. No Meckel's diverticulum or any other anomaly was present on the small intestine up to 170 cm proximally to the ileocecal valve. The right ovary and adnexa were normal.

The mass was excised with a margin of 1–2 mm of intestinal wall, through a wedge excision, leaving intact the mesenteric aspect of the intestine. No communication of the cyst with the intestinal lumen was present (Figure 3). The ileum was sutured transversally in two layers. Recovery was uneventful.

Histological examination revealed the presence of a cyst filled by mucous and serous fluid, containing some cellular debris, and lined by flattened intestinal epithelium. The part of its wall shared with the native intestine

consisted of a muscular layer lined by the flattened intestinal epithelium on the cyst's side and by normal intestinal epithelium on the intestinal luminal side (Figure 4).

The outer layer of the intraperitoneal part was covered by a seromuscular layer. Evidence of inflammation was present around the region of the perforation (Figure 5).
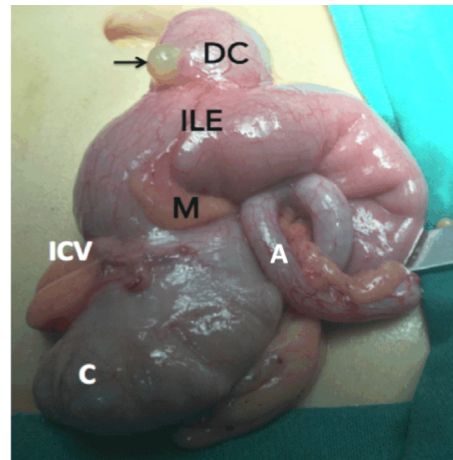
Figure 2 – *Cecum (C) and terminal ileum (ILE) are exteriorized. A duplication cyst (DC) arising from the terminal ILE on its antimesenteric side. ICV: Ileocecal valve; M: Mesentery; A: Appendix; Arrow: Site of perforation.*
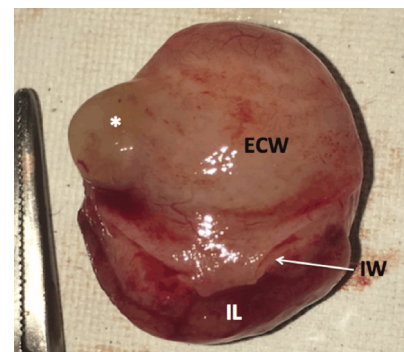

Figure 3 – *Duplication cyst resected. IL: Ileal side of the common wall; IW: Intestinal wall; ECW: External cyst wall; Asterisk: Perforation site.*
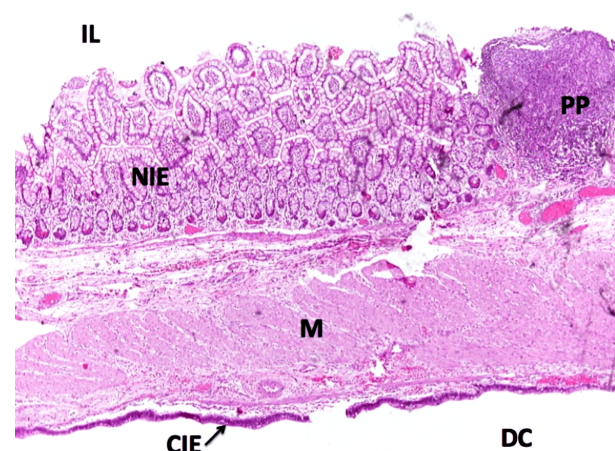
Figure 4 – *Common wall, between native ileum and cyst. IL: Lumen of the ileum; NIE: Normal intestinal epithelium; PP: Peyer's patch; M: Muscular layer; CIE: Cystic intestinal epithelium (flattened); DC: Interior of the duplication cyst. Hematoxylin–Eosin (HE) staining, ×40.*

A **B**
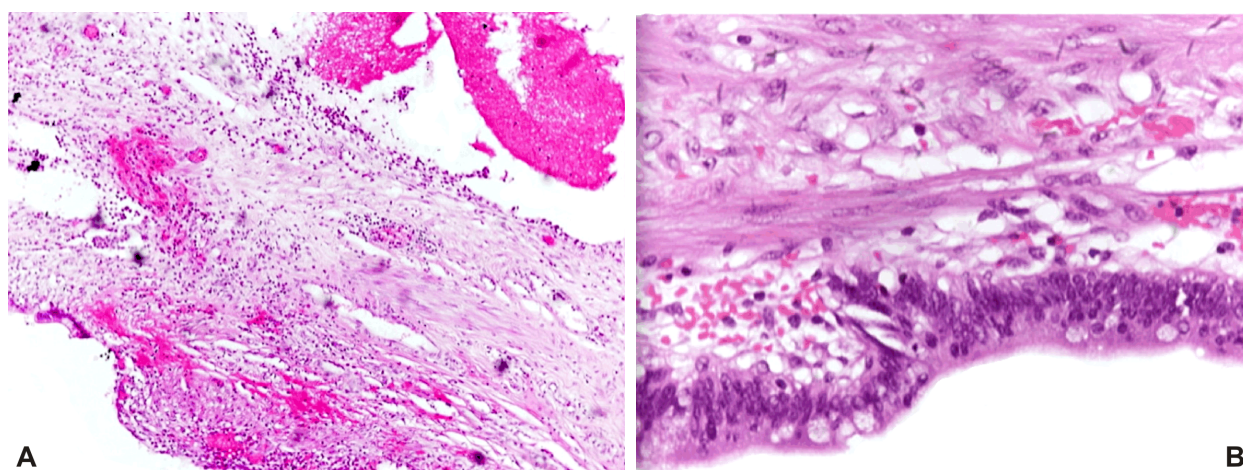
Figure 5 – *(A and B) Cyst wall, near the perforation site. Polymorphonuclear infiltration. HE staining: (A) ×100; (B) ×400.*

⊟ Discussions

GIDs appear to result either from a localized aberration in the development of the intestinal wall of the or as part of a more generalized malformation. Several theories have been postulated to explain their formation, but no single one has been able to account for all of the known variants, their various characteristics and the accompanying malformations [3, 10, 11]. Partial or abortive twinning aims to explain the association of GID with doubling of other body parts. The "split notochord" theory is the prevailing explanation for the enteric-lined cysts located in the posterior mediastinum, abdomen, or spinal canal, which are called neurenteric cysts, and are associated with adjacent vertebral, skin, central nervous system and gastrointestinal tract anomalies. Among GIDs, particularly those originating from the esophagus have a high prevalence of associated vertebral anomalies [3, 11]. Other congenital malformations associated with GIDs are intestinal atresias, malrotation and genitourinary malformations. Coexisting malformations may be present in one third of the cases [8].

A localized aberration in development of the intestinal wall is most widely accepted as a cause for the malformation when the problem remains local, especially for midgud duplications, such as the one described in this report. According to "aberrant luminal recanalization theory", the "solid stage" of intestinal development is followed by a defective luminal cell apoptosis, which can result in divisions and developing of enclosed spaces within the future muscular intestinal wall, which develop in duplications [3]. At this stage, a possible alternative mechanism could be the sequestration of islands of endodermal cells at the time of formation of the circular muscle coat. However, these theories fail to explain the presence of heterotopic gastric mucosa, which occurs in as many as one third of these lesions, or the frequency of mesenteric positioning [10]. Their provenance from congenital diverticula has also been postulated, as well as environmental stresses exerted on the embryo causing ischemic–hypoxic trauma [10].

The prototype GID case occurs as a single thin-walled cystic structure intimately related to the bowel wall. It appears as if the muscular layer of the intestine is split and the space created in between is lined by intestinal epithelium, usually arranged as a single flattened layer. The two halves of the split muscular layer form the wall of the cyst, one part protruding within the lumen of the bowel and another extraluminally, at its mesenteric side, extending between the layers of the mesentery [11]. The intraluminally protruding cyst wall is shared with the intestine itself, constituting its wall as well, and is covered, at its intestinal luminal side, with normal intestinal epithelium corresponding to the region.

Morphologically, most GIDs (71–90%) are spherical in shape, and the rest are tubular [3, 5, 9–11, 13]. Spherical cysts have a diameter of less than 10 cm, usually varying in size from 0.5 cm to 2.5 cm in diameter [13]. Tubular duplications are rarely longer than 20 cm, but lesions even longer than 100 cm have been encountered [9]. Tubular cysts usually communicate with the lumen of the native intestine, while spherical lesions do not [3–8]. In only one series, such communications have been reported, in 10 out of 50 spherical GIDs, but without reporting their site. In the same series, 17 out of 25 tubular lesions communicated with the lumen of the alimentary tract [7]. In all other series reviewed, which included 182 jejunoileal GIDs, no such mention was made. Another exception we encountered in literature was a case of a communicating spherical duplication of the duodenum [14].

All 200 jejunoileal GIDs in the series which we reviewed, originated from the mesenteric border of the intestine.

A structure protruding from the antimesenteric aspect of the intestine, especially in the range from 15 cm to 167 cm proximally to the ileocecal valve would most likely be a Meckel's diverticulum. However, this is a true diverticulum, with its lumen communicating with the intestinal lumen and which is fed by the remnant of the omphalomesenteric artery carried by a dedicated mesenteric fold [15, 16].

Symptoms caused by intestinal duplications can be elicited through various mechanisms. Distension of the lesion due to accumulation of secretions can cause intense pain and obstruction by compression of the adjacent bowel lumen. Intussusception can also be elicited. Heterotopic gastric mucosa may cause peptic ulceration, resulting in occult blood loss, hematochezia and perforation [7, 10].

All cases of GID perforation that we were able to trace in literature were due to peptic ulceration. However, in our case, no ectopic gastric mucosa was present and no gross or histological signs of peptic ulceration were observed.

Conclusions

The lesion described in our case exhibited all the typical characteristics of the spherical GID, *i.e.*, usual localization on the ileum, absence of communication with the lumen of the native intestine and appropriate configuration of its wall, both macroscopically and histologically, albeit located on the antimesenteric border of the intestine. Its clinical manifestation can also be considered as typical with the exception of the age of the patient. The pathology by which the symptoms were elicited, *i.e.*, the non-ulcerative perforation, may also be considered very rare.

Conflict of interests

The authors declare that they have no conflict of interests.

References

[1] Jeziorczak PM, Warner BW. Enteric duplication. Clin Colon Rectal Surg, 2018, 31(2):127–131.
[2] Schalamon J, Schleef J, Höllwarth ME. Experience with gastro-intestinal duplications in childhood. Langenbecks Arch Surg, 2000, 385(6):402–405.
[3] Macpherson RI. Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. Radiographics, 1993, 13(5):1063–1080.
[4] Rattan KN, Bansal S, Dhamija A. Gastrointestinal duplication presenting as neonatal intestinal obstruction: an experience of 15 years at tertiary care centre. J Neonatal Surg, 2017, 6(1):5.
[5] Olajide AR, Yisau AA, Abdulraseed NA, Kashim IO, Olaniyi AJ, Morohunfade AO. Gastrointestinal duplications: experience in seven children and a review of the literature. Saudi J Gastroenterol, 2010, 16(2):105–109.
[6] Sharma S, Yadav AK, Mandal AK, Zaheer S, Yadav DK, Samie A. Enteric duplication cysts in children: a clinico-pathological dilemma. J Clin Diagn Res, 2015, 9(8):EC08–EC11.
[7] Stringer MD, Spitz L, Abel R, Kiely E, Drake DP, Agrawal M, Stark Y, Brereton RJ. Management of alimentary tract duplication in children. Br J Surg, 1995, 82(1):74–78.
[8] Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowan MA, Martin LW. Duplications of the alimentary tract. Clinical characteristics, preferred treatment, and associated malformations. Ann Surg, 1988, 208(2):184–189.
[9] Karnak I, Ocal T, Senocak ME, Tanyel FC, Büyükpamukçu N. Alimentary tract duplications in children: report of 26 years' experience. Turk J Pediatr, 2000, 42(2):118–125.
[10] Stern LE, Warner BW. Gastrointestinal duplications. Semin Pediatr Surg, 2000, 9(3):135–140.
[11] Forshall I. Duplication of the intestinal tract. Postgrad Med J, 1961, 37(432):570–589.
[12] Kuo HC, Lee HC, Shin CH, Sheu JC, Chang PY, Wang NL. Clinical spectrum of alimentary tract duplication in children. Acta Paediatr Taiwan, 2004, 45(2):85–88.
[13] Huang ZH, Wan ZH, Vikash V, Vikash S, Jiang CQ. Report of a rare case and review of adult intestinal duplication at the opposite side of mesenteric margin. Sao Paulo Med J, 2018, 136(1):89–93.
[14] Uzun MA, Koksal N, Kayahan M, Celik A, Kilicoglu G, Ozkara S. A rare case of duodenal duplication treated surgically. World J Gastroenterol, 2009, 15(7):882–884.
[15] Androulakis JA, Skandalakis LJ, Kingsnorth AN, Colborn GL, Weidman TA, Richardson DD. Small intestine. In: Skandalakis JE (ed). Skandalakis' surgical anatomy: the embryology and anatomic basis of modern surgery. Paschalidis Medical Publications, Athens, 2004, 791–839.
[16] Sagar J, Kumar V, Shah DK. Meckel's diverticulum: a systematic review. J R Soc Med, 2006, 99(10):501–505.

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