Giant ileal duplication cyst presenting as malrotation

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Abstract

Introduction: Alimentary Tract Duplications (ATDs) are rare anomalies. The ileum is the most common site, while rectal, duodenal, gastric and cecal duplications are extremely rare [1,2].

Ileal duplication cysts (IDCs) are enteric cysts that are hollow, epithelium-lined, cystic or tubular structures. IDCs may or may not communicate with a portion of the gastrointestinal tract. If there is no communication with the gastrointestinal tract, these cysts are considered sequestered or isolated. Ileal duplication cysts most frequently present with clinical features of a bowel obstruction but can also present with bleeding or perforation.

We discuss a rare case of an IDC presenting with radiographic and clinical features of malrotation in a six-day-old female infant. This is the first case of IDC presenting as clinical and radiographic malrotation described in the literature.

1. Presentation of a case

A six-day-old term female infant born at 40 weeks and 2 days was presented at our institution with 48 h of abdominal distention followed by three episodes of bilious vomiting. She was exclusively breast fed and had passed meconium within 2 h of birth. Prenatal history and imaging at 20 weeks gestation were completely normal. Her parents were both healthy, and her 2-year old sister was born with no congenital abnormalities.

At presentation she weighed 3810 g. Her vitals were as follows; rectal temperature 37.7, pulse rate 132, blood pressure 109/70 mm Hg, respiratory rate 42 and oxygen saturation 96% on room air. She appeared well. There were no signs of dehydration. She was not in obvious pain. Her cardiorespiratory exam was unremarkable. Her abdominal exam revealed a soft, non-tender, distended abdomen without a palpable dominant mass. There were no hernias. We inserted a nasogastric tube and it drained minimal amounts of non-bilious gastric contents.

Hematologic and biochemical parameters were normal except for a mixed picture of acute respiratory alkalosis and metabolic acidosis presumed secondary to her abdominal pathology.

Abdominal X-rays (AXR) (Fig. 1) demonstrated a paucity of bowel gas in the right lower quadrant with mildly dilated loops of bowel. The abdominal films were abnormal but nonspecific. AXR was followed by upper gastrointestinal (UGI) series (Fig. 2), which demonstrated many of the radiographic features of malrotation without obvious volvulus. These features included duodenal redundancy, duodenojejunal junction malposition, a nonascending
duodenojejunal junction, stomach and duodenal distention, and gastroesophageal reflux. Based on the contrast study, a diagnosis of malrotation without volvulus was made.

The significance of the UGI could not be overlooked and thus we decided to explore the abdomen immediately. However, given the patient’s stable condition and a benign abdomen, we felt comfortable with the laparoscopic approach as an initial procedure.

At laparoscopy we found excessive amounts of serous fluid in the pelvis and chyle in the mesentery of the small bowel. On running the bowel we discovered two loops of small bowel physically attached to each other: one loop appeared pink, healthy and decompressed while the other had a distended appearance. The rotation of the bowel could not be confidently assessed at laparoscopy. A decision to convert to laparotomy was made due to the limited abdominal domain and inability to identify the rotation of the bowel.

Once the bowel was eviscerated we determined that the rotation was normal but immediately discovered an isolated ileal duplication cyst (Fig. 3). A 30 cm long cyst starting 45 cm from the ileocecal valve was observed (Fig. 4). The cyst was intimately attached to the normal ileum. With careful dissection we were able to separate the isolated duplication cyst.

The postoperative period was uneventful. The patient was discharged home on postoperative day six, and the child is well at three months of age.

The histopathological examination showed histologically normal intestinal wall. It showed congested serosa, areas of flattened and dilated mucosa. There were no tumors present. Focal areas of mucosal squamous metaplasia were noted (Fig. 5).
Fig. 5. Histopathological section demonstrating histologically normal intestinal wall, congested serosa, areas of flattened and dilated mucosa, and absence of tumors.

2. Discussion

Alimentary duplications are rare cystic lesions that occur at any point in the alimentary tract. Roughly half are small bowel duplications with most of these being ileal duplication cysts [4,5]. Although ileal cysts of over 60 cm in length have been reported [6], most are less than 10 cm [6].

Patients with ileal duplication cysts can present with vomiting, constipation, or abdominal distension and are often diagnosed following development of complications including gastrointestinal hemorrhage, intussusception, volvulus, or bowel perforation [7].

Our patient presented with bowel obstruction and subsequent radiologic investigations suggested this was secondary to malrotation and intermittent volvulus. We suspect the weight and volume of the isolated duplication cyst changed the position of the normal bowel as well as the duodenojejunal junction, which led to a misdiagnosis of malrotation.

Although histologically benign, the epithelial lining of an IDC contains ectopic acid-secreting mucosa in more than 50% of cases and this can be a cause of ulceration, bleeding, or transmural erosion [8]. Other abnormalities reported in association with small bowel duplication cysts include omphalocoele, intestinal atresia, intestinal stenosis, and malrotation of the midgut [9,10].

The diagnosis of IDC can be made by ultrasound, which is reliable in differentiating them from mesenteric cysts, which have a thinner wall [11]. Prenatal US diagnosis has also been reported [6]. Plain abdominal X-rays, upper GI series, and barium enema can differentiate this condition from a volvulus, intestinal atresia, stenosis, or other congenital bowel obstruction [12,13].

Treatment consists of cyst resection with preservation of the normal bowel whenever possible, although this can be extremely difficult. Normal bowel is often resected en bloc with the cyst, followed by Anastomosis of the normal bowel. Different surgical techniques have emerged depending on the size of the cysts and whether or not gastric mucosa is present within the cyst [4,14,15].

3. Conclusion

We identify a case of an ileal duplication cyst in a six-day-old female infant who presented with both clinical and radiographic features of small bowel malrotation and intermittent volvulus. Small bowel duplication cysts can be misdiagnosed as malrotation because the weight and volume of the duplication cyst can change the position of the duodenojejunal junction and the remaining small bowel. Until now, this has been an undescribed presentation of an isolated ileal duplication cyst.

Consent statement

Written informed consent was obtained from the parent for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interests statement

The authors declare that there are no conflicts of interests.

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