CASE REPORT

Clinico-Radiological Features of Ileocaecal Duplication Cyst in Children

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ABSTRACT

Enteric duplication cyst (EDC) is a rare malformation of the alimentary tract which can potentially cause intestinal obstruction in children. It can cause obstruction directly either by narrowing the bowel lumen, or indirectly by causing an intussusception. We are reporting three cases of ileocaecal EDC in children who presented with symptoms of distal bowel obstruction; one case of ileal EDC with intussusception and two cases of chronic obstruction by an EDC at caecum and terminal ileum. Ultrasound showed a typical cystic lesion with gut signature sign in two of them. Resection was performed in all cases; primary repair was performed in the intussusception case and diversion stoma with delayed closure was performed in the other two cases with chronic obstruction.

INTRODUCTION

Enteric duplication cyst (EDC) is a rare malformation of the alimentary tract which was described by William Ladd in 1937. It is a well-developed cyst-like structure in close attachment to the alimentary tract and potentially causing an obstruction.[1] EDC can happen anywhere along the gastrointestinal tract and symptoms may vary according to the site. In this series, we are reporting three cases of ileocecal EDC in children less than two years old who presented with symptoms of distal intestinal obstruction. An acute presentation has been contributed by the EDC leading an intussusceptum and another two cases had symptoms of chronic obstruction by the EDC partially obstructing the bowel lumen. All patients underwent resection with different approaches according to the aetiology and histopathological examination of the resected segments of all the cases confirmed the diagnosis.

CASE REPORT

Case 1: AM, a 3-month term baby girl presented with episodes of non-bilious vomiting and passage of fresh blood with mucous per-rectally. A mass was felt at the central abdomen with no features of peritonitis. Ultrasound abdomen revealed a long segment intussusception until left hypochondriac region with presence of a thick-wall cystic lesion (1.5x1.6cm) at the apex suggestive of duplication cyst (Figures 1 a-c). She underwent emergency exploration and confirmed to have a long segment ileo-ileo-colic intussusception with the apex felt at the distal transverse colon. After successful manual reduction, a well-defined cystic lesion was seen at the antimesenteric border of ileum, 30 cm proximal to the ileocaecal valve (Figures 1 e-f). There was no communication of the cyst to the native bowel and the rest of the bowel was healthy after reduction. Wedge resection of the ileum with the cyst was performed and the ileum primarily repaired. She recovered well after the

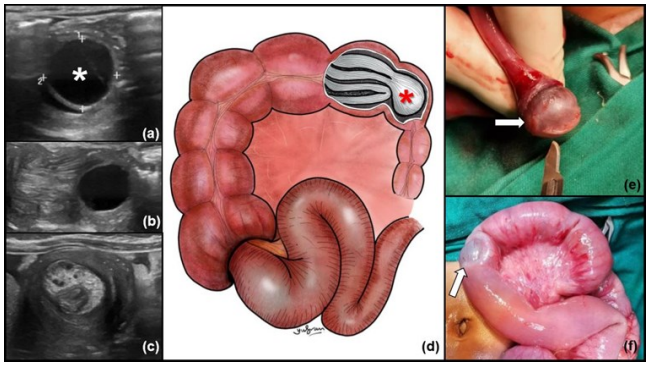

Figure 1: (a) Ultrasound shows cystic lesion with thick-wall in the left hypochondriac region (asterisk), (b) the longitudinal sonographic view of the intussusception with cystic lesion at the apex, and (c) the target sign of intussusception on the axial view. (d) Illustration represents the intussusception before reduction, and operative photographs after reduction (e, f) confirmed an ileal duplication cyst (arrow) found at 30cm from the ileocaecal junction, as a lead point for ileo-ileocolic intussusception.

Case 2: MAF, an 8-month-old boy under paediatric follow up for failure to thrive, presented with bilious vomiting and abdomen distension. At presentation, he was dehydrated and acidotic. His abdomen was distended with visible and palpable bowel loops. There was no peritonism. Plain radiograph showed evidence of distal bowel obstruction. Ultrasound abdomen revealed marked fluid filled small bowel dilatation with the presence of a cystic lesion measuring 2.3 x 3.9 x 4.0cm at the right iliac fossa region showing gut signature sign representing a duplication cyst (Figures 2 a & b). Emergency exploration confirmed a duplication cyst at the caecum, causing obstruction at the ileocaecal junction. The terminal ileum was dilated and thickened (Figure 2 d). Limited right hemicolectomy was performed and the proximal bowel appeared oedematous, not amenable for primary anastomosis. An ileostomy was created and delayed closure was done two weeks later. Postoperative recovery was uneventful.

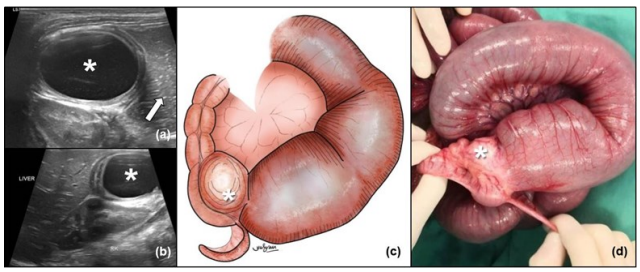

Figure 2: (a) Ultrasound shows a cystic lesion with gut signature sign (asterisk) associated with dilated fluid filled proximal small bowel (arrow) at the right lumbar region (b). (c) Illustration represents cyst at the caecum just distal to the ileo-caecal valve and (d) the operative photograph of the caecal duplication cyst (asterisk) with dilated terminal ileum.

Case 3: MH, a 15-month-old boy presented with non-bilious vomiting and abdominal distension. He was dehydrated on admission with evidence of acute kidney injury. Abdomen was distended but soft, no mass was felt. He had history of occasional abdominal distension and vomiting since after weaning and care-giver had difficulty to establish solid food intake. Ultrasound abdomen revealed dilated fluid filled small bowel with presence of cystic lesion at the right lumbar region measuring 2.3 x 3.6 x 2.5cm showing gut signature sign representing duplication cyst (Figures 3 a & b). He was brought to the operating theatre for exploratory laparotomy and found to have a dilated small bowel until terminal ileum and the cystic lesion was seen at the anti-mesenteric border of the terminal ileum without a definitive communication with the native bowel (Figures 3 d & e). Limited right hemicolectomy was performed and ileostomy creation was done. He was sent home well and had reversal of the stoma a month later.

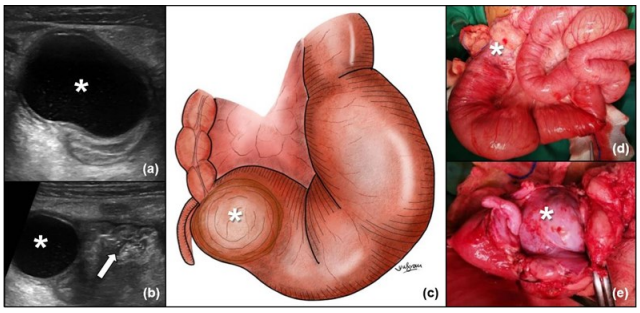

Figure 3: (a) Presence of a duplication cyst (asterisk) at the terminal ileum with a gut signature sign, due to its double wall or muscular ring sign seen on ultrasound and (b) collapsed distal bowel was seen (arrow). (c) Illustration represents the lesion at the terminal ileum just proximal to the ileocaecal valve at its anti-mesenteric border and (d, e) the operative photographs of the terminal ileal duplication cyst (asterisk) causing small bowel obstruction.

DISCUSSION

Alimentary tract duplication is a rare condition with reported cases of one in 4500 autopsy series. The exact cause is not certain; however several major theories were brought forward. Among the concepts proposed are related to partial or abortive twinning, split notochord theory, diverticula and canalization effect and a consequence of environmental insult such as trauma and hypoxia.[2] Enteric duplication cyst (EDC) can present in

either cystic or tubular form and may share a common wall or be entirely separate from the native bowel.[1]

Presentation of EDC varies according to the size, location, local effect, sequelae of having ectopic gastric mucosa or through antenatal detection.[2,3] A report by Sujka et al revealed that prenatal diagnosis was made in 37% of patients with alimentary tract duplication cyst.4 A systematic review by Fahy and Pierro reported 39.6% of patients who had prenatal diagnosis of EDC had ileal and ileocaecal cyst.[5] None of our patients reported in this series had prenatal diagnosis and in fact they presented later beyond neonatal period with symptoms of distal bowel obstruction. Obstructing EDC can be attributed either directly through its mass effect of obliterating the bowel lumen or indirectly by acting as a lead point for an intussusception or volvulus.[1] Intussusception may cause an acute obstruction as in our first case, as compared to the other two older patients who had chronic obstruction due to partially obstructing EDC which caused failure to thrive in one of them.

In general, no specific signs of EDC can be detected clinically, biochemically and on plain radiograph.[6] Ultrasound assessment has been reported to be a useful tool to diagnose EDC. It is radiation free and can be done without sedation. Presence of a cyst in relation to the bowel with double wall or muscular rim sign, or known as gut signature sign is pathognomonic.[7] Performing ultrasound in children with air-filled dilated bowels may be challenging, however in presence of intussusception, or thickened dilated fluid-filled bowel in cases of chronic obstruction, the cyst can be easily appreciated.[7] Ultrasound abdomen was done in all patients in this series to establish the cause of intestinal obstruction and all found to have cystic lesions with two of them showing the typical gut signature sign. None of our patients required more invasive imaging such as computed tomography (CT) or contrast study. CT is not typically performed and reserved for complicated cases or when diagnosis is not clear, for example in cases of tubular type of EDC with communication with native bowel segment.[6]

The mainstay of treatment of EDC, as demonstrated in our series is excision of the cyst with its adjacent bowel.[1] An acutely obstructed bowel as in intussusception, the excision may follow with primary repair without diverting stoma.3 However, in case where proximal bowel is chronically dilated and thickened, staged repair is our preferred approach. Following a period of diversion with a stoma, the proximal bowel is expected to become less oedematous and less thickened to allow a reasonable discrepancy for an end-to-end anastomosis, while giving us some time to improve the nutrition in these relatively malnourished patients. Otherwise more generous resection of the dilated bowel including right hemicolectomy for an EDC around the ileocecal region need to be done if to consider a single stage approach. Catalano et al. suggested preservation of ileocaecal valve for cases of ileocecal duplication cyst through limited excision and enterorrhaphy as reported in their series.[8] They described the technique as performing a total excision of the lesion together with the common wall with ileal lumen, and repairing the ileum transversely to prevent stenosis. In cases of asymptomatic prenatally detected EDC, almost half will become symptomatic within the first year of life necessitating surgery. They may be closely monitored for complications, however option for surgery as a prophylactic resection may be considered before they started to have symptoms which may complicate the surgery later.[5]

CONCLUSION

Enteric duplication cyst is one of the rare causes of intestinal obstruction in children, either directly or indirectly. Ultrasound has been a useful tool to diagnose EDC, especially with the demonstration of a gut signature sign in a cystic lesion. More invasive imaging like CT can be avoided and reserved for more complicated cases. Resection of the cyst is the preferred option to avoid recurrent symptom or subsequent complication.

CONFLICT OF INTEREST

The authors declared that they have no conflicts of interest with respect to either the authorship or publication of this article.

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