

Giant chylous cyst and ileal duplication in a young adult

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ABSTRACT Alimentary tract duplications are rare congenital anomalies that usually present in childhood and occasionally in adults. They are most common in the ileum, but can occur anywhere along the alimentary tract from the mouth to the anus. We report a 24-year-old woman who presented with a giant chylous ileum cyst duplication. To our knowledge, there is only one other report of a patient with a giant chylous cyst in the literature.

Keywords: adult, alimentary tract duplications, female, ileum

INTRODUCTION

Alimentary tract duplications (ATDs) are rare congenital anomalies that usually present in childhood and occasionally in adults. ATDs are congenital anomalies that can be encountered anywhere throughout the gastrointestinal tract, from the mouth to the anus. The terminal ileum is the most frequently involved segment.⁽¹⁾ Lined with intestinal mucosa, the duplication shares a common wall and mesenteric blood supply with the adjacent intestine, but usually does not communicate with the gut lumen. More than 80% of patients present before the age of two years with an acute abdomen or bowel obstruction.⁽²⁾ ATDs are frequently associated with other congenital anomalies, particularly in patients with colonic duplication.⁽³⁾ In adults, both cystic and tubular bowel duplications are rare entities.⁽⁴⁾

To our knowledge, only one previous study has reported a patient with a giant chylous cyst in the literature.⁽⁵⁾ Herein, we report a 24-year-old woman who presented with a giant chylous cyst as ileal duplication.

CASE REPORT

This report was approved by the Ethical Board of the Second Military Medical University, Shanghai, China. A 24-year-old woman presented to the general surgical department at Changhai Hospital, Shanghai, China, with a two-week history of abdominal pain. Examination revealed a distended and tender abdomen, with a large, firm, palpable mass extending from above the umbilicus into both the lower quadrants. Laboratory investigations yielded results within normal ranges. However, abdominal ultrasonography revealed a giant cyst in the abdomen. Computed tomography (CT) and magnetic resonance (MR) imaging confirmed the ultrasonographic diagnosis (Fig. 1).

Surgical intervention was decided upon. Although the cysts were initially diagnosed as mesenteric, a peristaltic mass that shared a common wall with the ileum was found during laparoscopic exploration. The exploration revealed a giant cystic

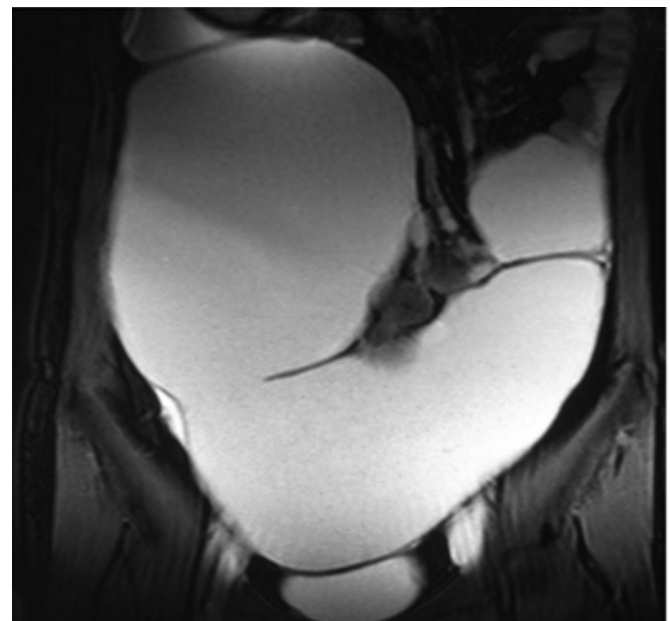


Fig. 1 MR image shows a giant cystic abdominal and pelvic mass.

mass that measured 28 cm × 20 cm in size, occupying almost the entire abdominal cavity. The cyst's stalk was attached to the base of the mesentery of the terminal ileum, approximately 40 cm from the ileocaecal junction (Fig. 2a). The duplication was cystic and shared its muscular layer with the ileum. The cyst contained approximately 4,800 mL of thick, chylous, mucoid material. No connection was identified between the mass and the ileum. The mass and 4 cm of ileum were resected (Fig. 2b), and an end-to-end anastomosis was created. No further masses or anomalies were noted.

Histopathological examination revealed a multilobulated cyst that contained smooth muscle and scattered ganglion cells. The mucosal lining of the cyst was composed of intestinal epithelium and gastric mucosa. The patient's postoperative course was uneventful and she was discharged on postoperative Day 3.

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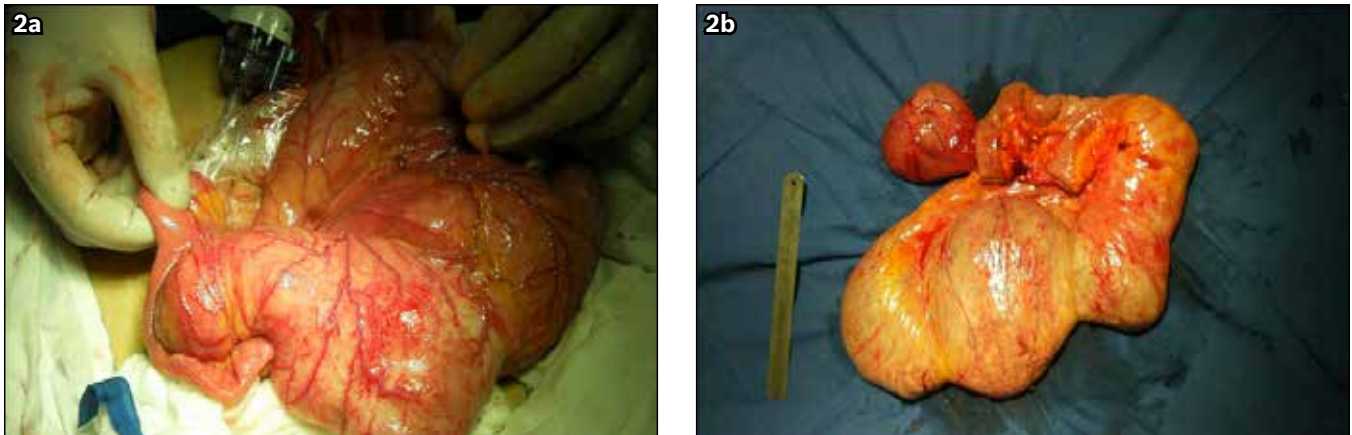


Fig. 2 Operative photographs show (a) part of the ileal duplication cyst before resection and (b) the resected duplication cyst.

DISCUSSION

A total of 28 adult patients (21 men, 7 women) with ileal duplications have been reported in the English literature, including the present study (Appendix I), with the first report published in 1884.⁽⁶⁾ The mean age of patients in the literature is 45.5 (range 20–81) years. The most common symptom reported was abdominal pain (20/28). Other less common symptoms include haematochezia (7/28), vomiting (6/28), nausea (5/28), diarrhoea (2/28), weight loss (3/28), routine control examination (2/28), distension (1/28), and urinary symptoms of frequency and hesitancy (1/28). Of these, 14 patients had undergone segmental ileal resection with end-to-end anastomosis, 4 patients underwent resection of the ileal duplication with a primary anastomosis, while 3 patients required a right hemicolectomy to achieve complete resection of the duplication. A single patient required a transverse colostomy and partial jejunectomy. The ileal duplication was cystic in 17 patients and tubular in 11 patients. Malignancy in the duplication cyst was found at presentation in six patients, (including the patient with mucinous cystadenoma). Our patient, described in the present report, was found to have the largest cyst described among all of the patients published to date.

The cyst in our patient was initially diagnosed as a mesenteric chylous cyst. Mesenteric chylous cysts – also known as retroperitoneal chylomatous cysts, mesenteric lymphangiomas or chylomas of the mesentery – are extremely rare and their nonspecific clinical presentation is known to mimic ATDs.^(7,8) ATDs are defined as cystic or tubular structures found in relation to normal bowel, whose walls resemble intestine and consist of smooth muscle with alimentary tract mucosa. It is hard to differentiate ATDs from mesenteric chylous cysts based on clinical presentation and imaging examinations alone, and a definitive diagnosis is usually only made during surgery.

The clinical presentation of ATDs may vary widely. Symptoms commonly reported by patients with ATDs include vomiting, constipation, abdominal pain, obstruction, abdominal mass and haemorrhage.^(9,10) These symptoms may be either the direct effects of distention of the duplication or

caused by compression of adjacent organs.⁽¹¹⁾ As a result of their rarity and differing modes of presentation, ATDs can be easily misdiagnosed as other disorders, including appendicitis, choledochal cyst, Meckel's diverticulum or Crohn's disease. For ATDs, it is not always possible to obtain a precise preoperative diagnosis.

Plain thoracic and abdominal radiographs, barium meal, enema, ultrasonography, endoscopic exploration, CT, MR imaging or Technetium-99m pertechnetate scintigraphy can all be used for the diagnosis of duplication cysts.^(12,13) If the ATD is a cystic bowel duplication, ultrasonography and MR imaging can be used to identify the true nature, location and extent of the lesion.^(13,14) However, for patients with tubular bowel duplications, endoscopy is the preferred diagnostic tool.⁽⁹⁾ Recently, CT enteroclysis has also been used to diagnose ATDs.⁽¹⁵⁾ While analysing imaging findings, clinicians should remain cognisant of the fact that duplication cyst fluid can produce elevated concentrations of tumour markers, including carcinoembryonic antigen and carbohydrate antigen CA 19.9.⁽¹⁶⁾

Surgical treatment is advocated after the diagnosis of ileal duplication in order to prevent potentially lethal complications such as perforation, volvulus, intussusception, bowel obstruction and enteric bleeding. Associated anomalies, as well as the location and size of the duplication, also influence the surgical management of the patient. Small and medium duplication cysts should be resected with a small segment of the bowel, followed by a primary anastomosis. Our analysis of the literature indicate that this procedure is performed most frequently (Appendix I). Simple excision or enucleation of an enteric cyst is possible in some patients. However, one should bear in mind that local resection of a duplication cyst can compromise the blood flow to the adjacent bowel since both share the same blood supply.⁽¹⁷⁾

Malignant transformation has been reported in the intestines of some patients diagnosed with ATDs as adults.^(18,19) The incidence of malignancy in the literature among adult patients diagnosed with ileal duplications was 6/28, with one of these patients having a mucinous cystadenoma. Given the risk of malignancy, we advocate the resection of all

duplications. The treatment outcome for surgical resection is good, with low morbidity and mortality.⁽¹⁾

It is not always possible to obtain a precise preoperative diagnosis of ileal duplication cyst. Rather, a definitive diagnosis is often only reached intraoperatively. Surgical treatment should be performed to prevent potentially lethal complications in patients with ileal duplications. This report adds to the literature on the clinical variability of the presentation of ATDs and presents the second patient to be diagnosed with giant chylous cyst.

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APPENDIX

APPENDIX I. Summary of adult patients with ileal duplications in the English literature.

Study (year)	Age (yrs)/ gender	Presenting features	Surgical treatment*	Form	Size (cm)	Malignancy
Fitz (1884) ⁽¹⁾	65/male	Abdominal pain, weight loss, diarrhoea	NA	Cystic	NA	Yes
Nolan (1953) ⁽²⁾	67/male	Abdominal pain, nausea, vomiting	NA	Cystic	NA	No
Thomson et al (1954) ⁽³⁾	26/male	Abdominal pain	NA	Tubular	NA	No
Micolonghi et al (1958) ⁽⁴⁾	31/male	Haematochezia	NA	Cystic	NA	No
Anderson et al (1962) ⁽⁵⁾	31/male	Abdominal pain	NA	Cystic	NA	No
Curt et al (1966) ⁽⁶⁾	69/male	Abdominal pain, nausea, vomiting	3	Tubular	6 × 4	No
Orr et al (1975) ⁽⁷⁾	54/male	Abdominal pain, nausea, vomiting	1	Tubular	NA	Yes
Mair et al (1976) ⁽⁸⁾	81/male	Abdominal pain, nausea, diarrhoea	1	Tubular	6.5 × 1.5	No
Adair et al (1981) ⁽⁹⁾	41/female	Abdominal pain, nausea, vomiting, weight loss	NA	Cystic	NA	Yes
Salameh et al (1984) ⁽¹⁰⁾	30/male	Haematochezia	2	Tubular	11 × 2	No
Marks (1984) ⁽¹¹⁾	62/male	Routine control examination	1	Tubular	7	No
Cecil et al (1990) ⁽¹²⁾	20/male	Abdominal pain	1	Cystic	10 × 20	No
Staunton et al (1990) ⁽¹³⁾	31/male	Haematochezia, abdominal pain	1	Cystic	1.8 × 1.8	No
John et al (1994) ⁽¹⁴⁾	36/female	Abdominal pain	3	Tubular	NA	No
Tanabe et al (1995) ⁽¹⁵⁾	35/male	Haematochezia	1	Tubular	23 × 13	No
Otter et al (1996) ⁽¹⁶⁾	52/male	Urination frequency and hesitancy	1	Cystic	18 × 12	No
Tew et al (2000) ⁽¹⁷⁾	57/female	Abdominal pain, vomiting	1	Cystic	15 × 5	No
Nomura et al (2002) ⁽¹⁸⁾	61/male	Abdominal pain	2	Cystic	5 × 5	No
Kusunoki et al (2003) ⁽¹⁹⁾	72/male	Abdominal pain, distension	4	Tubular	10	Yes
Qasim et al (2004) ⁽²⁰⁾	43/female	Abdominal pain, vomiting	3	Cystic	4 × 3	No
Simsek et al (2005) ⁽²¹⁾	20/male	Abdominal pain, weight loss	1	Cystic	7 × 7	No
Toth et al (2006) ⁽²²⁾	32/male	Haematochezia	1	Cystic	5.5 × 2.5	No
Hosseini et al (2007) ⁽²³⁾	54/male	Haematochezia	1	Cystic	NA	No
Al-Sarem et al (2007) ⁽²⁴⁾	23/female	Abdominal pain	1	Tubular	NA	No
Tomas et al (2007) ⁽²⁵⁾	64/female	Routine control examination	2	Cystic	7.5 × 5 × 3	Yes†
Babu et al (2008) ⁽²⁶⁾	59/male	Abdominal pain	1	Tubular	16 × 2	Yes
Ogino et al (2008) ⁽²⁷⁾	35/male	Abdominal pain, haematochezia	2	Cystic	2.5 × 2	No
Present study	24/female	Abdominal pain	1	Cystic	28 × 20	No

*Surgical treatment was broadly divisible into four procedures. †Mucinous cystadenoma. 1 = segmental ileal resection with end-to-end anastomosis; 2 = ileal duplication resection with primary anastomosis; 3 = right hemicolectomy with primary anastomosis; 4 = transverse colostomy and partial jejunectomy. NA: not available

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