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Clinical characteristics of gastrointestinal tract duplications in children

A single-institution series review

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Abstract

Due to the various presentations of gastrointestinal tract duplications (GTD), diagnosing and management for this disease might be varied and difficult. We intend to improve the experiences for these difficult, in terms of the clinical presentations, diagnostic investigations, management.

We reviewed recent literature and retrospectively analyzed 72 pediatric patients with enteric duplication. Diagnosis was confirmed by surgery and pathological examination for imaging characteristics and clinical and pathological features.

The ages of patients ranged from one month to 12.5 years. The clinical presentations of the patients included 57 cases with abdominal pain, followed with nausea or vomiting, abdominal distension, etc. All of the patients were diagnosed by ultrasonography, and most of them presented as intra-abdominal cystic masses. Four cases were diagnosed with the cysts other than GTDs, like, mesenteric cyst, chledochal cyst and abscess, and so on. Computed tomography was performed on 65 patients. X-rays and barium meal showed the outline of the cyst structure, with intestinal displacement due to the pressure from the cyst. Among the 72 cases of enteric duplication, 45 were located with ileocecal area, 41 were ileal and 8 were colonic duplications.

Enteric duplication is very rare in children and is prone to misdiagnosis. The preoperative diagnosis of enteric duplication can be improved through comprehensive analysis of various imaging exams and closely related clinical presentations.

Abbreviations: CT = computed tomography, GT = gastrointestinal tract, GTD = gastrointestinal tract duplications, MR = magnetic resonance, US = ultrasonography.

Keywords: cystic cysts, gastrointestinal tract duplications, ileocecal area, ultrasonography

1. Introduction

Gastrointestinal tract duplications (GTD) are rare congenital abnormalities arising anywhere along the gastrointestinal tract (GT), most encountered in the the small intestine (50%).[1,2,3] The GTD was named according to the structure feature, containing the mucosa of the gastrointestinal tract and sharing the common wall with the GT. Ectopic mucosa (gastric or pancreatic) was reported in 20% to 30% of these cysts structures.[4]

The clinical symptoms vary according to the type and location of the GTD and the different ages of involved patients, like infancy, early childhood or adulthood. It may present with respiratory distress or dysphagia, nausea, vomiting, abdominal distention, pain, mass, Intussusception, bleeding, inflammation, and even perforation per rectum. The preoperative diagnosis is very difficult to make due to lack of specific clinical and imaging manifestations.[5,6] Nowadays, ultrasonography (US) is the most used imaging method of choice to diagnose GTD. Magnetic resonance (MR) and computed tomography (CT) can be required in esophageal or rectal GTDs for planning complicated surgical approach.[7,8]

The purpose of the study was to review the imaging characteristics and the clinical and pathological features in a large cohort of children, with the goal of improving the management experience of pediatric GTD.

2. Methods

Between January 2000 and January 2019, we retrospectively investigated a series of 72 consecutively hospitalized patients who had been surgically and pathologically confirmed for enteric

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duplication at the Department of General Surgery, Chongqing Medical University, China (an urban tertiary care teaching hospital). The study was approved by the ethics committee of children's hospital, Chongqing Medical University. We excluded patients with other gastrointestinal abnormalities (anorectal malformation, intestinal atresia, Meckel diverticulum or Hirschsprung disease).

We retrospectively retrieved the institutional computerized medical records and radiology reports of all included patients, focusing on age, gender, clinical manifestations and the results from ultrasonography and computed tomography. Patient background demographics included age, sex, previous major abdominal surgery, and comorbidities. The clinical history and physical examination included the onset and course of nausea/vomiting, crampy pain, distension, fever, and bowel sounds. The original radiographic studies at admission included ultrasound, plain abdominal films or computed tomography (CT) scans when appropriate. The surgical feature and the pathological characteristic were also noted.

2.1. Diagnosis algorithm

As per our clinical algorithm, ultrasonography was primarily used when the patients presenting with abdominal complaints. Depending on these examinations, computed tomography (CT) was performed for further confirmation. MRI was used only for a limited number of patients. Scintigraphy was seldom used, only for studying the ectopic mucosa. We recorded mass location, size, shape, edge, wall thickness, mobility and the relationship with the intestinal canal and surrounding structures. All of the patients were diagnosed postoperatively by pathological examination.

2.2. Surgical indications

The selection criteria for the surgical approach were the clinical presentations of the patient and the size and location of the cyst (s). In general, emergency surgeries were performed in pediatric patients with enteric duplication when fever, vomiting, abdominal distension, peritonitis or other specific physical signs were present. Resection of the enteric duplication and the primary intestinal anastomosis was usually considered the principle method of treatment for these patients. When patients manifested non-specific symptoms and imaging results indicated cystic lesions, perioperative preparation was considered adequate. Following the exclusion of other neoplastic diseases, exploratory laparotomy was then scheduled to remove the lesions.

Descriptive statistical analysis was performed to summarize the clinical characteristics of the current patients.

3. Results

3.1. Clinical manifestations

A total of seventy two patients with histopathological diagnosis of GTDs were identified to be retrospectively analyzed in this study. The median age of the patients was 4.8 years old (ranged from one month to 12.5 years). Among them, 45 were within 1 year old, 11 were between 1 and 3 years, 10 were between 3 and 7 years and 6 were older than 7 years. The 72 patients consisted of the predominance of 47 males (65.3%) patients.

The symptoms at admission varied based on the cysts location. All the details are summarized in Table 1. Fifty-two patients (72.2%) had at least two symptoms, most commonly abdominal pain, nausea or vomiting and followed with abdominal distension, abdominal mass, and so on. Nineteen patients (26.4%) presented with typical symptoms of acute/chronic intestinal inflammation, manifesting as diarrhea with or without hemafecia. Five of these patients were misdiagnosed with acute/chronic gastritis. Thirteen patients (18.1%) presented with peritonitis accompanied by full abdominal pain and abdominal guarding. Four of these patients were misdiagnosed with acute appendicitis. Ten (13.9%) patients presented with atypical symptoms at admission and incidental diagnosis of GTD was made during abdominal examination or ultrasonography for other unrelated causes. Twenty-three patients presented with signs and symptoms consistent with acute appendicitis, eight with an intussusception, and seven with significant gastrointestinal hemorrhage secondary to the presence of ectopic gastric mucosa. Emergency operative interventions were required for 17 patients.

Table 1

Summary of Patients at admission.

Variables	
Age (yr)(range)	4.8 (range: one month–12.5 yr)
Male: female	47:25
Clinical symptoms, n (%)	
Abdominal pain	57 (79.2)
Nausea or vomiting	38 (52.8)
Abdominal distension	29 (40.3)
Mass	15 (20.8)
Gastrointestinal hemorrhage	13 (18.1)
Diarrhoea or constipation	17 (23.6)
Respiratory distress	6 (8.3)
Acute small bowel obstruction	11 (15.3)
Atypical symptoms	26 (36.1)

3.2. Diagnosis

Ultrasonography (US) is the primary imaging method in the diagnosis of GTDs and were performed in all the current cases. A cyst adjacent to the gut with double-wall was the classical presence of GTDs under US (Fig. 1). Among the 72 patients, 58 cases (80.6%) presented with an intra-abdominal cystic mass, with intestine-like thickened walls wrapped by adjacent peristaltic intestines. These patients were primarily diagnosed with GTD. Six patients presented with an intra-abdominal cystic mass with intestinal disposition due to pressure. These patients were diagnosed with mesenteric cyst. Seven patients had a thick-walled cystic mass with blurred margin. The size of cystic mass was reduced after anti-inflammatory therapy, and the presence of an abscess was considered. In some cases, barium meal radiography might demonstrate a mass marked off by filling barium small intestine in fight inferior belly (Fig. 2).

Computed tomography were conducted in sixty five patients (65/72, 90.3%), whereas diagnoses were incidentally confirmed during surgery in 7 cases. A unilobular cystic mass with certain mobility was found in fifty six of the patients. A contrast-enhanced abdominal CT scan revealed a strengthened signal from the cystic wall and suggesting enteric duplication or the presence of a mesenteric cyst (Fig. 3). Some cases exhibited a cyst structure with blurred margin, which was confirmed during surgery to be caused by inflammation. A small cystic mass was found in nine cases in the right lower abdomen and was accompanied with

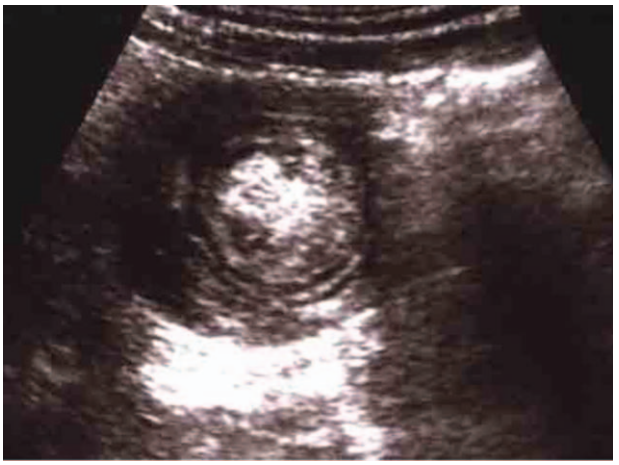

Figure 1. Sonogram of a distal ileal duplication cyst. On tranverse view, an anechoic lesion with typical wall characteristics is seen-inner echogenic mucosal stripe and outer hypoechoic muscle layer (arrow).

abdominal pain. In this case, Mechel diverticulum was considered.

A plain abdominal X-ray was performed in 15 patients, and a gastrointestinal barium exam was performed in 9 patients. In 6 patients, the small intestine or colon was found to be depressed, with a clear cystic mass margin outlined by barium reagent filling the intestinal canals (Fig. 3) (Table 2).

3.3. Classification of GTD

In this group of patients, 45 cases (62.5%) of duplications involved in the ileocecal area, 7 cases of duplication occurred in the jejunum, 41 cases in the ileum, and 9 cases in the colon, with 59 cases located on the mesenteric side and 11 on the contralateral side of mesentery. One duplication was a complete duplication of the colon, including 2 appendixes. The details of the postoperative diagnosis and pathological features of the GTDs were provided in Table 3. In 51 patients, the duplication cysts were connected with the ileum lumen through small pores, with 10 cases of ectopic gastric or pancreatic mucosa. Eight patients had perforation, and 19 patients had complications involving inflammation (including those with ectopic gastric mucosa) and adhesions to the surrounding tissue. Congenital alformations were presented in 6 cases, with cardiac, urinary, spinal defects, etc.

4. Discussion

Gastrointestinal duplication is a rare condition caused by anomalies during embryonic development. We retrospectively reviewed the largest cohort of 72 cases of gastrointestinal duplication managed in our institute to assess the variety features of duplication cysts for further management. The presence of heterotopic tissue, including ectopic gastric, pancreatic tissue, in the current research supported the congenital developmental origin of the alimentary tract. Furthermore, these anomalies occurred anywhere along the length of the alimentary tract, with enteric duplication being the rarest.[9]

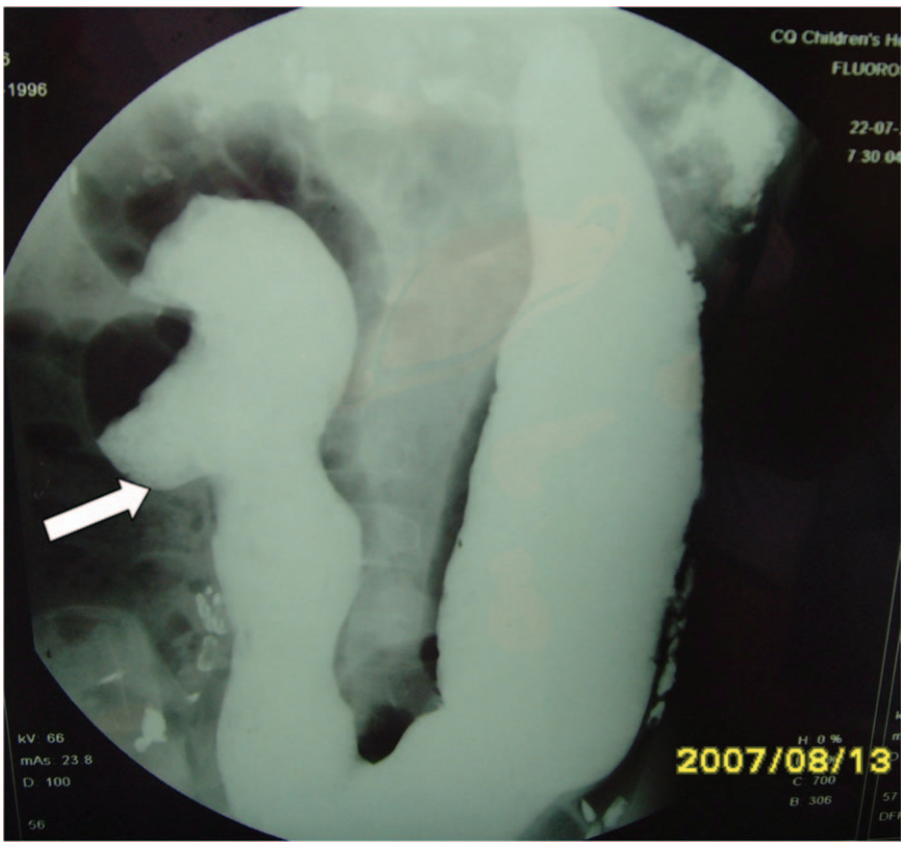

Figure 2. Barium meal radiography of whole digestive tract study demonstrates a mass marked off by filling barium small intestine in fight inferior belly (arrow).

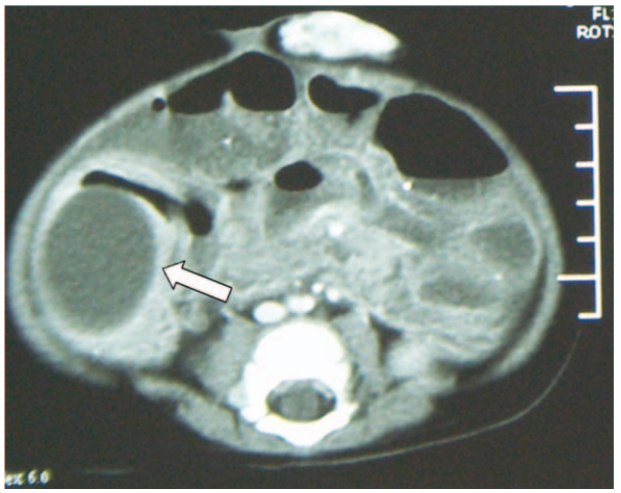

Figure 3. Abdominal CT scan with contrast. It shows a well circumscribed homogeneous cyst (Cy) pushing forward the posterior Ileum wall. The contrast visualized in the Ileum lumen (St) is not evident in the cyst (arrow).

Table 2

Diagnostic investigation and its performances.

Diagnostic investigation	n
Ultrasonography	72
plain abdominal X-ray	15
CT	65
Technetium-99m imaging	23
Gastrointestinal barium exam	9
Pre-operative diagnosis	
GTD	58
Mesenteric cyst	4
Omental cyst	1
Gastric duplication cyst	3
Meckel diverticulum	4

Table 3

Pathophysiological feature of the duplications.

Type of lesion	
Cystic	46
Tubular	21
Complex	13
Site of lesion	
Ileocecal area	45
Oesophageal	2
Gastric	3
Jejunal	7
Ileal	41
Colonic	8
Cecal	1
Ectopic tissue	
Gastric	11
Pancreatic	6
Others	3
Congenital alformations	
Cardiac	12
Urinary	4
Spinal defects	3
Others	5

In our series, the detection age ranged from 1 day to 10 years and nearly two thirds of our patients were infants. The ileocecal area are the most commonly affected sites in our series, accounting for 62.5% of the whole GTDs, followed by the colon, jejunum, stomach and duodenum.[10] Due to the wide spectrum of localization and different signs and symptoms, accurate preoperative diagnosis of enteric duplications was frequently different from the post-operative findings. The major manifestations include vomiting, a characteristic feature of intestinal obstruction, abdominal masses, rectal bleeding, peritonitis and other associated clinical symptoms.[11] The current cases also match the above description, including the age at the onset, site, main clinical presentations and pathological features.

Given its rare incidence and lack of specific imaging indications, the preoperative consideration of enteric duplication is rare, and accurate diagnosis is difficult.[12,13,14] Because of the intra-abdominal nature of these abnormalities, some cases were detected intraoperatively and were not suspected before operation. In the current patients, Intussusception and acute appendicitis in considerable number of cases were later diagnosed with enteric duplications during surgery. Gastrointestinal hemorrhage due to the presence of ectopic mucosa was also presented in 13 cases. Most patients with esophageal duplications were suspected before surgery because the results obtained from contrast radiographic studies uniformly suggested the diagnosis.

The basic pathological classifications include intraluminal cysts, extraluminal cysts and tubular and thoracic duplication,[15,16] with the cystic form being the more frequent in our series. Those of tubular configuration tended to be more extensive in our experience. It has a similar structure and shares a common blood supply with the main intestinal canals characteristically located on the mesenteric aspect of the associated native bowel. Intestinal duplication develops in all fully differentiated parietal layers of the alimentary tract, and about 80% of duplication cysts have no connections with the attached gastrointestinal tracts. Approximately 20% to 25% of mucosal layers present with ectopic mucosa.

The imaging presences of uncomplicated GTDs under US included a cyst in relation to the gut with double-wall or muscular rim sign (gut signature sign), which is caused by inner hyperechoic mucosa and outer hypoechoic smooth muscle layer (muscularis propia) and parallel double tubular structures, with cyst-like structures that are surrounded by intestinal canals. Larger duplication always results in intestinal displacement due to pressure.[17] CT scans reveal low-density unilobular cystic masses, mostly spherical in shape and not connected with intestinal canals.[18] In some cases, the duplications are tubular in shape and are connected with the intestinal canals, and the wall of the duplications is close to or thicker than that of the surrounding intestine. Enhanced CT scans reveal a strengthened signal from the cyst's wall. The plain X-ray and barium meal or barium enema examinations might reveal abdominal masses, intestine-filling defects or disposition with pressure, accompanied by possible spinal deformities. If enteric duplications are connected with the main intestinal canal, barium reagent can enter the duplication structure and delay emptying. This presentation is often misdiagnosed as intussusception.

Enteric duplication should be differentiated from other abdominal cystic diseases. Mesenteric and omental cysts can manifest as asymptomatic abdominal masses or present with abdominal pain and intestinal obstruction. CT scans have revealed thin-walled cystic masses with blurred outlines or even

no cystic wall.[19] These cysts are typically unilobular cysts, sometimes containing an internal septum. Omental cysts are located in the anterior abdomen, which posteriorly displaces the intestine. Patients with enteric duplication usually seek treatment due to abdominal masses or other symptoms. The cysts are relatively thick-walled structures that are even thicker than the adjacent normal intestines. Meckel diverticulum typically presents as small cysts in the middle or lower abdomen that are mostly conical or cylindrical in shape and have a diameter of 1 to 2 cm and an average length of 3 to 4 cm. Patients with a Meckel diverticulum are usually admitted for complications. Ectopic gastric mucosa is much less common in enteric duplication. In addition, enteric duplications usually share the blood supply with the adjacent normal intestine, while a separate blood supply has often been observed in Meckel diverticulum cysts. When spiral CT is performed for angiography of the celiac and superior mesenteric arteries, the imaging results can clearly show the superior mesenteric artery and its branches, which can help to differentiate these two diseases.[20] Abdominal abscesses typically present as low-density cystic masses, with walls that have an uneven thickness and blurred edges. Enhanced signal is usually observed from the wall in the scan of the abscess with contrast-enhanced CT. Finally, abdominal cystic teratoma contains mostly cystic components, with soft tissues, adipose tissue or calcification of varying degrees. Pediatric teratoma is most commonly observed in the retroperitoneal region. Enteric duplication can be differentiated from other diseases such as hydronephrosis, megaureter, hydrometrocolpos and ascites based on clinical and imaging characteristics.

Surgical management of enteric duplications depends on the localization and type of the duplication. Those of tubular configuration were more extensive, and at times posed a special challenge to the surgeon. In cases where the cyst has no communication with the native gut, cystectomy is adequate. Others required operative resection with primary end-to-end anastomosis to restore bowel continuity. Selective mucosal excision was reported to be an excellent approach for extensive tubular lesion.[21] We have treated in this manner to avoid extensive resections for three patients with excellent result.

The weakness of the current research is the data collected retrospectively a heterogeneous group of patients for a very long span of time. Outcomes from many patients may not reflect outcomes from current treatment algorithms, there have likely been many practice changes, leading to different care practices between study patients. Another point is that this is an observational research; all the variables were not intervened. We are looking forward to the future prospective clinical trials to provide information about the factors influenced the clinical outcome.

5. Conclusion

In conclusion, enteric duplication lacks specific characteristics in both clinical and imaging presentations. With improved examination methods and comprehensive analysis of various imaging results closely related to the patient's clinical presentation, however, the preoperative diagnosis of enteric duplication can be improved.

Author contributions

Conceptualization: Chunbao Guo.
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Formal analysis: Chunbao Guo.
Methodology: Jiaming Lan, Chunbao Guo.
Software: Bailin Chen.
Writing – original draft: Bailin Chen.

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