Signature: © Pol J Radiol, 2013; 78(3): 70-73
DOI: 10.12659/PJR.884012

Polish Journal of Radiology
www.PolRadiol.com
CASE REPORT

Received: 2012.12.16
Accepted: 2013.06.31

A huge duplication cyst of the ileum

Mehmet H. Atalar[1], Levent Cankorkmaz[2], Hatice Ozer[3], Gokhan Koyluoglu[2]

[1] Department of Radiology, Cumhuriyet University School of Medicine, Sivas, Turkey
[2] Department of Pediatric Surgery, Cumhuriyet University School of Medicine, Sivas, Turkey
[3] Department of Pathology, Cumhuriyet University School of Medicine, Sivas, Turkey

Author's address: Mehmet H. Atalar, Department of Radiology, Cumhuriyet University School of Medicine, Sivas, Turkey, e-mail: mhatalar@gmail.com

Summary

Background: Massive unilocular intraabdominal cysts in children are rare. Alimentary tract duplications can present diagnostic and therapeutic difficulties. Although they can occur anywhere from the mouth to the anus, they are commonly seen in relation to the ileum. We herein present an unusual case of duplication cyst itself occupying virtually all of the available intraabdominal volume. It appeared to be an enteric duplication cyst of ileal origin.

Case Report: A 3-month-old girl was admitted to our hospital for investigation of progressive abdominal distension and biliary vomiting. Plain radiography of the abdomen showed normal air-fluid level in the stomach and paucity of gases in rest of the abdomen. Magnetic resonance imaging showed a huge, homogenous cyst extending from the xiphisternum down to the pelvis. The cyst was excised completely. Macroscopic examination and histologic findings confirmed the diagnosis of a huge enteric duplication cyst arising from the ileum.

Conclusions: Enteric duplication cyst should be considered in a patient with an abdominal cystic mass. Radiologist must take into account patient age, clinical parameters, and imaging findings to identify the likely etiology of a cystic mass.

Key words: abdominal cystic masses • enteric duplication cyst • magnetic resonance imaging • plain radiography

PDF file: http://www.polradiol.com/download/index/idArt/884012

Background

Gastrointestinal (GI) duplications are rare congenital lesions that can develop anywhere along the alimentary tract. They are characterized by the presence of gastrointestinal mucosal lining. Duplications are most frequently single. They can be cystic (79%) or tubular (21%) and characteristically arise from the mesenteric border of the intestine. Cystic duplications usually present in early childhood as abdominal masses or acute abdomen. Acute onset is due to obstruction or volvulus. However, minority of cases remains unsuspected until adulthood. Huge cystic duplication of the small bowel is a very rare anomaly of the GI system [1–4]. We herein present an unusual case, where the cyst itself occupied virtually all of the available intraabdominal space.

Case Report

A 3-month-old girl was admitted to our hospital for investigation of progressive abdominal distension and biliary vomiting. Patient was born of a nonconsanguineous marriage, at full term, through vaginal delivery. Antenatal history was not significant; she had dilated bowel loops in the antenatal ultrasound. There was no history of recent acute illness or trauma. Her vital signs were normal and she was afebrile. White blood cell count was 11 500/μL (normal range, 4.5–13.5$\times 10^3/\mu$L). Blood tests, including liver function tests, were normal on admission.

Plain radiography of the abdomen showed normal air-fluid level in the stomach and paucity of gases in rest of the abdomen (Figure 1). Ultrasonography (US) showed a huge unilocular cyst extending from the xiphisternum down to the pelvis, with all of the abdominal and pelvic viscera being displaced posteriorly (not shown). There was a large, well-circumscribed, lobulated mass within the abdomen that had no association with other abdominal organs. Abdominal MR studies included axial spin-echo T1-weighted (T1W) (TR/TE=600/15), axial spin-echo T2-weighted (T2W) (TR/TE=2000/80), and axial and

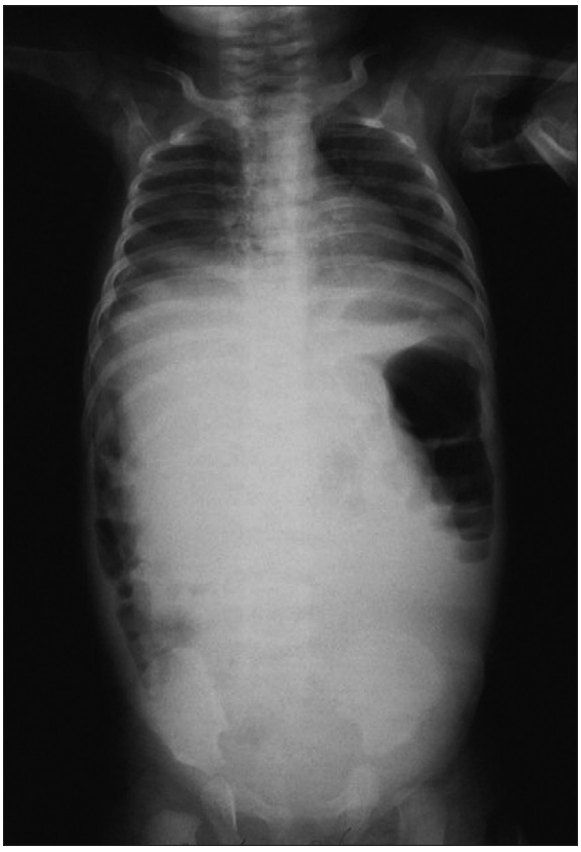

Figure 1. Plain radiograph of the abdomen shows a normal air-fluid level in the stomach and paucity of gases in rest of the abdomen.

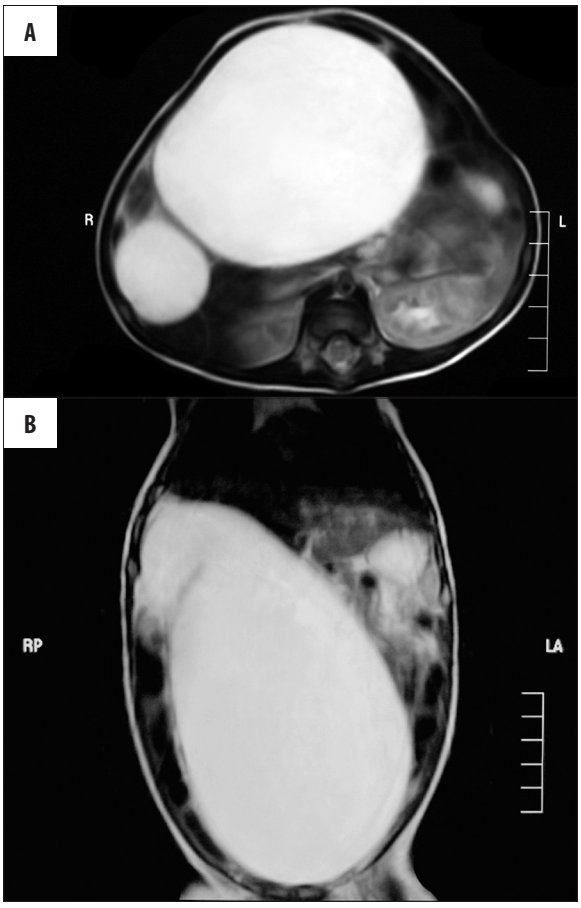

Figure 2. (**A**) Axial and (**B**) coronal T2-weighted MR images show a huge, homogenous cyst extending from the xiphisternum down to the pelvis.

sagittal spin-echo fat-suppressed T1W (TR/TE=600/15) imaging. Magnetic resonance imaging (MRI) with intravenous Gd-DTPA confirmed US findings and showed that the cyst occupies virtually the entire abdominal cavity (Figure 2A, 2B). On post-contrast T1W images, the mass demonstrated peripheral enhancement and uniform central low signal intensity. There were no vertebral or other bony abnormalities on either plain radiographs or MRI. All these findings together strongly suggested the diagnosis of an intestinal duplication cyst.

Shortly after undergoing these studies, patient was taken to the operating room and underwent a laparotomy. Laparotomy showed a huge (35×10 cm), duplication cyst originating from the mesenteric side of the bowel, sharing a common muscularis layer with the adjacent bowel, without communication to the ileal lumen (Figure 3). She had intestinal malrotation. The cyst was completely excised, the patient made an uneventful recovery and remains well at three years postoperatively. Macroscopic and histologic findings confirmed the diagnosis of a huge enteric duplication cyst arising from the ileum (Figure 4).

Discussion

Duplications of the GI tract are rare congenital anomalies and most reports in the literature are either case reports or small series. GI tract duplications may be found anywhere from the mouth to the anus. In a large collective review of the literature of 495 duplications, 50% involved midgut, 36% foregut, and 12% the hindgut. Esophageal duplications constituted 19%, while of the midgut duplications, ileal duplications were the most common, amounting to 35%. Localized duplications are common in the ileum and jejunum. Some theories explain duplication as a defect in recanalization of the intestinal lumen after the solid stage of embryological development [1–6].

Duplications can present with a variety of signs and symptoms and, depending on their site, can be life threatening, sometimes necessitating urgent surgical intervention. Most duplications present early and, although some can be asymptomatic, the majority will lead to complications such as obstruction, inflammation, or hemorrhage. Hemorrhage is seen in those with heterotopic gastric mucosa, which is present in 29–35% of duplications and may be identified preoperatively by technetium scintigraphy [2–5].

Enteric duplication cysts are hollow, epithelium-lined, cystic or tubular structures that are intimately attached to a portion of the GI tract. GI duplication cyst is a spherical or tubular mass adherent to the GI tract that sometimes communicates with it. It is lined with intestinal epithelium and contains smooth muscle within its wall. The most common location is the terminal ileum, but it can occasionally be seen at the distal esophagus, stomach and duodenum, or

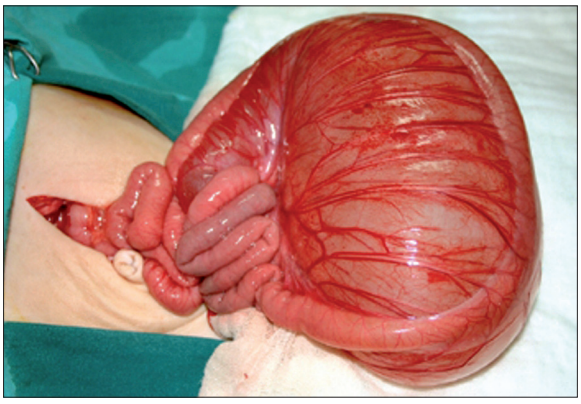

Figure 3. Laparotomy revealed a huge duplication cyst connecting with the ileum.

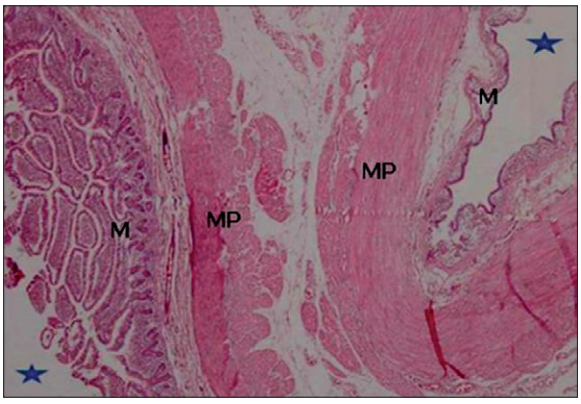

Figure 4. The duplicated segments showing two complete intestinal walls lying side by side. The lumen of each segment is designated by the (asterisk) and the intestinal epithelium is attenuated. M – mucosa, MP – *Muscularis propria*.

elsewhere. Most patients present within the first year of life, with symptoms that include GI obstruction, palpable mass and abdominal distention. To our knowledge, a huge ileal duplication cyst of such dimensions has not yet been reported in the literature [2,4–6].

The role of radiological techniques is to demonstrate an abdominal mass and identify the organ, from which the mass originates. The second step in radiological diagnosis is determination of the nature, relationship to surrounding organs and size of the cyst. On imaging work-up, US plays a decisive role in the diagnosis. US differentiates the cystic nature of duplications from solid tumors and also demonstrates an intimate association between the duplication and the bowel wall. Reliable indicators of a duplication cyst include an inner echogenic rim of intestinal mucosa surrounded by a characteristic hypoechogenic rim of muscle in the wall, indicating various layers of the intestine, as well as inner debris or hemorrhage. Inner echogenic contents may develop due to mucous and proteinaceous secretions or to hemorrhage or infection within the cyst. In such cases, establishing ultrasound diagnosis may be difficult because heterogeneous contents or obliteration of the cystic wall mimic other abdominal conditions such as abscesses or tumors. Computed tomography (CT) and MRI can define its precise anatomic location. They can be helpful in difficult cases that require multiplanar approach. MRI can be useful for demonstration of the cystic nature of these masses, as these lesions continue to have characteristically high signal intensity when imaged with T2W sequences regardless of the nature of cystic contents [2–4,6–9].

Because of their common location at the mesenteric border, they may be easily mistaken for mesenteric or omental cysts, only to be identified by mucosal rather than endothelial lining [3–6].

Table 1. Differential diagnosis of a huge abdominal cystic mass.

Duplication cyst
Omental cyst
Mesenteric cyst
Huge diverticulum
Cystic tumor of the pancreas
Hemorrhagic pseudocyst secondary to pancreatitis
Choledochal cyst
Abscess
Cystic/necrotic tumors

When facing such a huge abdominal cystic mass, differential diagnoses including duplication cyst, omental cyst, mesenteric cyst, huge diverticulum, cystic tumor of the pancreas, hemorrhagic pseudocyst secondary to pancreatitis, choledochal cyst, postinflammatory abscess formation and cystic or necrotic tumors secondary to degeneration, should be considered (Table 1) [1,3,5–7,9,10].

The treatment of choice for enteric duplication cysts is surgical excision [2]. In the case reported herein, the patient had an uneventful course of recovery following resection of the duplication cyst.

Conclusions

In conclusion, enteric duplication cyst should be taken into account in a patient with an abdominal cystic mass. Abdominal US can be highly suggestive in the diagnosis of duplication cyst. MR imaging may be useful as a noninvasive technique for the diagnosis and accurate preoperative assessment of the cystic mass. Radiologist must consider patient age, clinical parameters and imaging findings when stating the likely etiology of the cystic mass.

References:

1. Kangarloo H, Sample WF, Hansen G et al: Ultrasonic evaluation of abdominal gastrointestinal tract duplication in children. Radiology, 1979; 131: 191–94
2. Pokorny WJ, Goldstein IR: Enteric thoracoabdominal duplications in children. J Thorac Cardiovasc Surg, 1984; 87: 821–24
3. Robert W: Intestinal duplications, Meckel diverticulum, and other remnants of the omphalomesenteric duct. In: Behrman RE, Kliegman RM, Jenson HB (eds.), Nelson Textbook of Pediatrics. 16th ed. Philadelphia: WB Saunders, 2000; 1137

4. Segal SR, Sherman NH, Rosenberg HK et al: Ultrasonographic features of gastrointestinal duplications. J Ultrasound Med, 1994; 13: 863–70
5. Steiner Z, Mogilner J: A rare case of completely isolated duplication cyst of the alimentary tract. J Pediatr Surg, 1999; 34: 1284–86
6. Mihmanli I, Erdogan N, Kurugoglu S et al: Radiological workup in mesenteric cysts: Insight of a case report. J Clinical Imaging, 2001; 25: 47–49
7. Sakamoto K, Hasegawa S, Yamazaki Y et al: Ileal duplication presenting as perforation: report of a case. Surg Today, 2000; 30: 445–47
8. Karnak I, Ocal T, Senocak ME et al: Alimentary tract duplications in children: report of 26 years' experience. Turk J Pediatr, 2000; 42: 118–25
9. Siegel MJ: Gastrointestinal tract. In: Siegel MJ (ed.), Pediatric body CT. Lippincott Williams and Wilkins, Philadelphia, 1999; 201–25
10. Tong SC, Pitman M, Anupindi SA: Best cases from the AFIP. Ileocecal enteric duplication cyst: radiologic-pathologic correlation. Radiographics, 2002; 22: 1217–22