Case Reports

Ileal duplication in adults

Sulhi A. Al-Sarem, MBBS, *Jehad S. Al-Shawi,* FRCSed, FRCSI.

ABSTRACT

Alimentary tract duplications are rare anomalies usually present in childhood. They are most common in the ileum, but can occur anywhere along the alimentary tract from the mouth to the anus. We report a case of a 23-year-old female presented with severe epigastric and lower abdominal pain of one day duration, and was found to have perforated tubular ileal duplication communicating with the ileum on surgery.

Saudi Med J 2007; Vol. 28 (11): 1734-1736

From the Department of Surgery, King Abdul-Aziz Medical City, King Khalid National Guard Hospital, Jeddah, Kingdom of Saudi Arabia.

Received 15th November 2006. Accepted 25th February 2007.

Address correspondence and reprint request to: Dr. Jehad S. Al-Shawi, PO Box 9515, Jeddah 21423, Kingdom of Saudi Arabia. Tel. +966 050799973. Fax. +966 (2) 624 0000 Ext. 1985. E-mail: alshawi_jehad@yahoo.com

The clinical presentation of intestinal duplication in adults ranges from chronic nonspecific gastrointestinal discomfort to one of acute abdominal pain.[1] Although 60-85% is diagnosed by age 2 years,[2] there have been few reports in adults. Although the exact incidence is unknown, in 1961 there were 2 cases reported in more than 9000 fetal and neonatal autopsies.[1,3] The duplication occurs along the mesenteric aspect of the alimentary tract and is most commonly discovered in the ileum.[1,2] These lesions share patterns of presentation common to many nonspecific intra-abdominal diseases, yet are readily managed by a surgical approach.[1] We report this case to provide an opportunity to review the spectrum of presentations associated with enteric duplication in adults, and the current literature on this topic.

Case Report. A 23-year-old Saudi female presented to the emergency department with sudden onset of severe epigastric and lower abdominal pain of one-day duration. This pain is associated with fever, but no history of nausea or vomiting. No diarrhea or change of bowel habits. She gave a history of recurrent, central abdominal pain, which was not investigated. Past surgical history was negative. Physical examination revealed temperature of 38°C, pulse 110/min, blood pressure 103/45, and she was mildly dehydrated. Abdominal examination revealed generalized tenderness with guarding and positive rebound tenderness. Plain chest and abdominal radiography showed air under diaphragm. Her laboratory investigations were normal. She was rehydrated and resuscitated. An exploratory laparotomy was carried out. There was 300 mls of pus in the peritoneal cavity and perforated tubular ileal duplication communicating with the ileum (**Figure 1**). Resection of the duplicated segment and appendectomy were carried out using linear stapler gastrointestinal anastomosis (**Figure 2**). Histopathology did not reveal any gastric mucosa or malignant tissues. She had an unremarkable postoperative course and was doing well at follow up.

Discussion. This case provide an opportunity to review the current literature regarding the incidence, clinical presentation, and management of alimentary tract duplications in adults. Numerous terms have been used to describe the condition, including enterocystomas, enterogenous cysts, supernumerary accessory organs, ileum duplex, giant diverticula, and unusual Meckel diverticulum.[3] The term intestinal duplication was first used in 1844,[1] but it was not widely used until popularized in the year 1930,[1] with further classifications in 1950s,[1] as either spherical or tubular.[3] Although intestinal duplication are considered as benign lesions, they may result in significant morbidity and mortality if left untreated.[3]

Approximately 75% of duplications have been reported to be located within the abdominal cavity, while the remaining are pure intrathoracic (20%) or thoracoabdominal (5%). Jejunal and ileal lesions are the most commonly encountered (53%), followed by mediastinal (18%), colonic (13%), gastric (7%), duodenal (6%), rectal (4%), thoracoabdominal (2%) and cervical (1%).[3] Enteric duplication by definition compromise a group of lesions that contain a

smooth-muscle wall and enteric mucosa and are found only on the mesenteric border of the intestine.[1] These features differentiate intestinal duplications from the more common Meckel's diverticulum, which is found on the antimesenteric border. Intestinal duplication may be divided into non-communicating and communicating types. The noncommunicating type is more common and cystic, usually attaching to the mesenteric aspect of the intestine by a common seromuscular coat. The communicating type is rare and tends to be more tubular. A communication is found between the duplication and the intestinal lumen in only 19% of cases. The communication may be proximal, distal, or both.[4] The following criteria for the diagnosis of duplication were suggested in previous study:[2] 1. duplications of the alimentary tract are hollow structures, which posses a muscular coat, usually of 2 layers, and are lined with epithelium similar to that found in some portion of the gastrointestinal tract, 2. these lesions are always contiguous to some portions of the alimentary tube, and 3. the type of epithelium lining the duplication does not necessarily correspond to that part of the alimentary tract to which the structure is attached.[5] An understanding of the embryologic development of enteric duplication may provide further insight into its clinical presentation. The "enteric bud" theory of Lewis and Thyng[2] states that buds of the intestinal epithelium protrude into lamina propria and enlarge to establish a connection with the bowel lumen. The "solid lumen theory" of Bremer[2] postulate that the rapidly enlarging embryonic gastrointestinal tract outgrows the celomic cavity and causes accumulation of intestinal epithelial cells that then vacuolize, coalesce and form the lumen of the intestinal duplication. Previous study[2] theorized that hindgut duplication represents partial twinning, whereas others have postulated that an adhesion in the presomite embryo between the ectodermal and endodermal germ layers results in splitting or deviation of the notochord, which leads to alimentary tract duplications in association with vertebral anomalies. Each of these theories falls short in explaining both the presence heterotropic mucosa and the consistent mesenteric location.[1] Foregut duplications are most commonly associated with epigastric pain and dysphagia, the latter due to impingement of the cystic mass in the esophageal lumen.[2] Duplication of the midgut is generally associated with abdominal pain; obstruction and intussusceptions have been reported.[1] The presence of heterotropic gastric mucosa predispose to bleeding and perforation. Duplications of the hindgut may be seen in association with intestinal obstruction and tenesmus and may be confused with a colorectal neoplasm.[1]

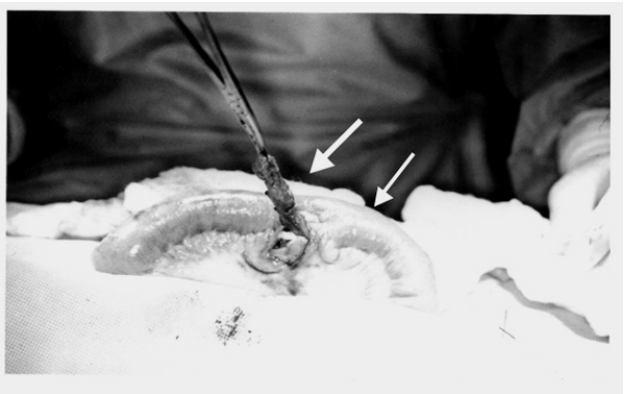

Figure 1 - Perforated ileal duplication (thick arrow) and bowel (thin arrow).

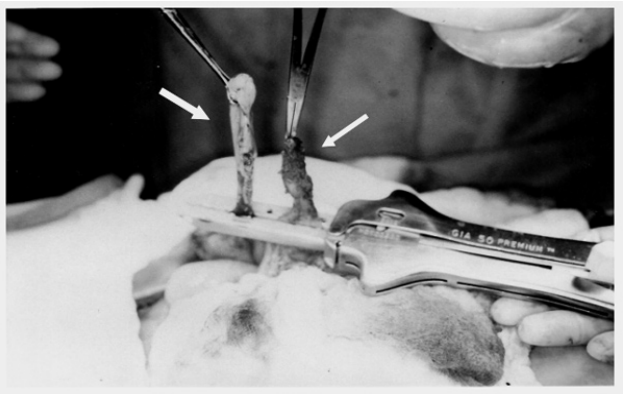

Figure 2 - Resection of the ileal duplication (thin arrow), and the appendix (thick arrow) using gastrointestinal anastomosis.

The most common preoperative symptoms were abdominal pain, nausea and vomiting; less common symptoms included weight loss, hematochezia, melena, and diarrhea.[2] Tender abdominal mass in the right lower quadrant may be noted on physical examination. The clinical presentation of enteric duplication in adults is variable and has undergone a transition with time. In the past, patients were operated on with complications of these lesions, such as bleeding, perforation, intussusception, bowel obstruction from adjacent pressure or mass effect, volvulus and associated malignancy.[6] Today, the diagnosis is made on the basis of ultrasound (US) and computed tomography (CT) for persistent, mild gastrointestinal pain.[1] This symptom complex is dictated chiefly by the location and size of the lesion.[1,7] Radiological diagnosis with US can be made with visualization of a 2-layered wall; the characteristic location adjacent to the bowel, and separation from other organs where cystic masses commonly occurred.[7] On US, the inner echogenic layer represents mucosa and the outer hypoechoic layer represents muscle.[7]

The diagnosis can be made with technetium scanning if associated with heterotropic gastric mucosa.[3,7] The CT scan can also play a role in localization of the duplications.

The pathological diagnosis is based on the presence of an organized muscular coat with myenteric plexus.[7] Once the diagnosis of an enteric duplication is made, surgical correction is warranted for 3 reasons. First, the majority of the patients require surgical intervention for the relief of symptoms. Second, surgery removes the otherwise persistent risk of perforation and bleeding caused by heterotropic mucosa in the unresected cyst. While our patient did not have gastric mucosa, however, still she had perforated ileal duplication may be due to inflammation. Third, there have been reports of the development of carcinoma in the lining of cyst, and the most common cell type was adenocarcinoma.[1,5] Surgical treatment of alimentary tract duplications is largely dictated by the anatomic location of the lesion and its relation to normal anatomic structure.[3] As duplication shares a portion of its wall with the adjacent small intestine and a common blood supply, complete resection of the duplication is the treatment of choice.[2] Internal drainage via fenestration or Roux-en-Y anastomosis may be necessary if the duplication is extensive in length or if contiguous vital structure are involved.[2,3] Complications related to surgical intervention are typically nonspecific and include post-operative bleeding, infection, and bowel obstruction. However, in patients with large tubular duplications, injury to the normal intestine with resultant short bowel syndrome must be considered.[8] Of the small number of ileal duplications presenting in adults, malignancy occurred in 23% of patients.[2] The evidence of epithelial instability might suggest a tendency toward malignant transformation in long standing duplications.[7] Hence, complete resection of duplication is the most appropriate method of treatment.[2,3]

In conclusion, although ileal duplications are extremely rare it should be seriously considered in the differential diagnosis of acute surgical abdomen. Although CT and US can show the duplication segment, the diagnosis is difficult preoperatively. We also believe that junior surgical staff must be aware of these conditions in the management of the acute surgical abdomen.

References

1. Hackam DJ, Zalev A, Burnstein M, Rotstein OD, Koo J. Enteric duplication in the adult, derived from the foregut, midgut and hindgut: presentation, patterns and literature review. ***Can J Surg*** 1997; 40: 129-133.
2. Johnson JA 3rd, Poole GV. Ileal duplications in adults, presentation and treatment. ***Arch Surg*** 1994; 129: 659-661.
3. Michalsky M, Besner G, Alimentary tract duplications. eMedicine Journal, June 18, 2002, 3: 6. Available from: URL: http://author.emedicine.com/PED/topic2922.htm
4. Ohba S, Fukuda A, Kohno S, Takeuchi S. Ileal duplication and multiple intraluminal diverticula: Scintigraphy and barium meal. ***AJR Am J Roentgenol*** 1980; 136: 992-994.
5. Kusunoki N, Shimada Y, Fukumoto S, Iwatani Y, Ohshima T, Arahi E, et al. Adenocarcinoma arising in a tubular duplication of the jejunum. ***J Gastroenterol*** 2003; 38: 781-785.
6. Otter MI, Marks CG, Cook MG. An unusual presentation of intestinal duplication with a literature review. ***Dig Dis Sci*** 1996; 41: 627-629.
7. Tew K, Soans BK, Millar EA. Adenocarcinoma in an ileal duplication cyst: ultrasound and computed tomography findings. ***Australas Radiol*** 2000; 44: 228-231.
8. Holcomb GW 3rd, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. ***Ann Sur*** 1989; 209: 167-174.

Related topics

Mohammed AA, Al-Gadi MA. Neonatal Staphylococcal scalded skin syndrome complicating ileal atresia. ***Saudi Med J*** 2003; 24: 538-541.

Abduljabbar AS, Abdul Rahman MM. Restorative proctocolectomy with ileal reservoir. ***Saudi Med J*** 2003; 24: 234; author reply 234.

Rahman MM, Shihed MH. Restorative proctocolectomy with ileal reservoir. ***Saudi Med J*** 2002; 23: 667-671.