Intestinal Duplications – Experience in 6 Cases

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Key Words

Intestinal duplications • Gastrointestinal congenital anomalies • Literature review, intestinal duplications

Abstract

Intestinal duplications are rare congenital anomalies, and most of them are detected in the first 2 years of life or antenatally. The clinical presentation depends on location, size, and the presence of ectopic gastric mucosa. Ultrasound scans during pregnancy result in a higher rate of antenatally detected duplications which allows early treatment and avoidance of possible complications. Resection of the duplication without adjacent bowel should be the treatment of choice. En bloc resection or partial excision with mucosectomy is advised in some cases. We present our experience with 6 patients treated in our institution from February 2002 to October 2005.

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Introduction

Intestinal duplications are reported to be present in approximately 1 in 4,500 neonatal autopsies or in 1 in 18,000 livebirths [1]. Regardless of their location, they have three common features: hollow structures, lined

with gastrointestinal tract epithelium, and have a wall of smooth muscle often shared with the contiguous intestinal tract. The duplications are classified according to their location (ileum 30%, ileocecal valve 30%, duodenum 10%, stomach 8%, jejunum 8%, colon 7%, rectum 5%), shape – cystic (>80%) or tubular –, and presence of ectopic tissue within the duplication (gastric mucosa present in up to 50% of the cases) [2]. Additional classification for small-intestine duplications considering vascular supplies to the duplication and the adjacent bowel is also reported [3].

Although considered to be benign, such duplications can have even lethal consequences if not properly treated [4]. The symptoms are unspecific and vary from minor digestive problems to intestinal obstruction, gastrointestinal bleeding, or perforation [5]. The number of antenatally detected duplications is increasing which allows early treatment with fewer complications. If the diagnosis is unclear, some additional imaging procedures can be used: barium swallow, radionuclide scan, or CT scan.

Surgical treatment depends mostly on the location of the duplications in relation to adjacent bowel or other structures like bile ducts and blood vessels. Postoperative complications are rare and unspecific: bleeding, infection, and bowel obstruction. Surgically treated intestinal duplications have a good prognosis.

Patients and Methods

Between 2002 and 2005, at the Department of Pediatric Surgery of the Clinical Hospital Center Rebro, 6 children were surgically treated for intestinal duplications. There were 3 boys and 3 girls with an age range from 27 days to 2 years and 10 months (mean age 13 months; table 1).

Three of our patients had symptoms of intestinal obstruction, 1 child presented with signs of intestinal perforation (pneumo-

peritoneum; fig. 1), 1 had chronic anemia with rectorrhagia and melena, and in 1 case the diagnosis was made prenatally.

We have used the following diagnostic procedures: ultrasound in all cases and barium swallow when needed, CT scan, and radionuclide [^{99m}Tc]pertechnetate macroaggregated albumin scan for detection of gastric mucosa.

The intestinal duplication was highly suspected in 4 cases (in 1 the diagnosis was made prenatally), in 1 case (a 5-month-old girl) the differential diagnosis was an ovarian cyst, and in 1 case Meckel's diverticulum was suspected (chronic anemia, rectal bleeding, positive radionuclide scan). We had two duodenal, one ileal, two cecal, and one colonic duplication.

The surgical procedure was laparotomy, and we have performed complete excision of the duplication without opening the adjacent bowel in 1 case, in 3 cases we underwent en bloc excision with the adjacent bowel, and in 2 cases we performed partial excision with mucosectomy. There were four cystic and two tubular duplications. Communication with the adjacent bowel was confirmed in 2 cases. We had two postoperative complications: one dehiscence of the blind end of the anastomosis and one wound infection.

Mucosa of intestinal type together with gastric mucosa was found in 3 patients, another had pancreatic cells as well as gastric and intestinal mucosae, and the intestine of 1 patient was lined with only intestinal-type mucosa and in 1 patient only with gastric mucosa. During the follow-up period (ranging from 2 months to 4 years) all of our patients did well.

Fig. 1. Cystic duplication resected together with the adjacent part of the ileum. A wooden stick is inserted through the perforated opening of the duplication.

Table 1. Intestinal duplications: clinical presentation, location, treatment, and type of mucosa found

Age/sex	Clinical presentation	Working diagnosis	Intraoperative findings and locations	Surgical procedures	Histological findings	Complications	Outcome and follow-up period
8 months/m	abdominal pain, fever	duplication	perforation of ileal duplication	resection en bloc with adjacent bowel, end-to-end anastomosis	mucosa of intestinal type together with gastric mucosa	none	good, 3 years
2 years and 10 months/f	obstruction	duplication	pyloroduodenal duplication	marsupialization	mucosa of intestinal type together with gastric mucosa	none	good, 13 months
15 months/m	gastrointestinal bleeding	Meckel's diverticulum	cecal duplication	excision of the duplication	gastric mucosa	none	good, 17 months
17 months/f	obstruction	duplication or hypertrophic pyloric stenosis	duodenal duplication	marsupialization	mucosa of intestinal type together with gastric mucosa and pancreatic cells	none	good, 9 months
27 days/m	obstruction	duplication	ileocecal duplication	resection en bloc with adjacent bowel, side-to-side anastomosis	mucosa of intestinal type	dehiscence of the blind end of the anastomosis	good, 15 months
5 months/f	none	ovarian cyst	cecal duplication	resection en bloc with adjacent bowel, end-to-side anastomosis	mucosa of intestinal type together with gastric mucosa	wound infection	good, 2 months

Discussion

Gastrointestinal duplications are rare congenital anomalies which are in 85% of the patients detected prenatally or in the first 2 years of life, but can remain undetected until older age [6]. Possible complications like bowel perforation, bleeding, obstruction, and malignant alterations are the reason why all duplications should be surgically treated at the time of diagnosis.

The clinical picture varies according to location, size, and other factors such as the presence of ectopic mucosa within the duplication, communication with adjacent bowel, or inflammation. The most common clinical manifestations are abdominal pain and intestinal obstruction. Rarely, such duplications present with signs of acute abdomen or acute bleeding. Pyloric and duodenal duplications can mimic hypertrophic pyloric stenosis (considered in 2 of our patients with episodes of vomiting and signs of duodenal obstruction) or a choledochal cyst [7, 8]. An ovarian cyst should be considered in female patients, and adolescent patients can sometimes have a presumptive diagnosis of Crohn's disease.

Ultrasound is the imaging modality of choice for the evaluation of an abdominal mass in the neonate. Approximately 30% of the intestinal duplications are diagnosed prenatally [9]. Some ultrasonographic features like hyperechoic mucosa and hypoechoic muscle layer (double-wall sign) and peristalsis when visible are specific for duplications. Additional imaging procedures are sometimes required: barium enema, CT scan, radionuclide scan (HIDA scan for choledochal cyst and pertechnetate scan for Meckel's diverticulum). However, gastric, duodenal, and rectal duplications can lead to a diagnostic dilemma.

Intestinal duplications often require urgent surgical intervention. Secretion of ectopic gastric mucosa causes ulceration of the intestinal mucosa which can lead to massive bleedings or bowel perforation [10]. Most of the authors [11] believe that once the diagnosis is made, an elective surgical procedure should be performed to avoid complications and to perform the procedure in an optimal state of the patient, but few think that only symptomatic duplications should be surgically treated. Our patient who presented with bowel perforation had duplication detected 4 months earlier, but the parents refused the indicated elective surgical procedure.

The recommended surgical procedure is excision of the duplication, and recent studies on vascular supplies for small-intestine duplications show that it is possible to resect them without compromising the adjacent bowel

vascularization. However, it is sometimes not possible (especially in cases with locations like the duodenum) because of shared wall and vascularization with adjacent bowel. Resection 'en bloc' of the duplication with adjacent bowel is then advised. Other procedures like marsupialization with mucosectomy, internal drainage, and Wrenn's operation are also advised when necessary. We had two duodenal duplications (one at the first and the other at the second duodenal segment) without communication with the adjacent bowel. Duodenal duplications rarely communicate with adjacent parts of the intestinal tract, and because of their close contact with bile and pancreatic ducts (can cause icterus or pancreatitis) it is often impossible to perform complete excision [12]. To avoid possible lesions of pancreas or choledochal duct, we have done mucosectomy with marsupialization. Both of these patients were postoperatively well and without complications which makes this procedure recommendable for duodenal duplications to avoid lesions of adjacent structures. In 3 of our patients, we had to perform 'en bloc' resection, and in 1 patient we completely excised the duplication without compromising the adjacent bowel. Laparoscopically assisted resection with or without small laparotomy has been reported and is recommended for uncomplicated cases.

Intestinal-type mucosa is found in most of the cases, and in 16–39% of the patients ectopic gastric or pancreatic cells are found. The mucosa should always be removed because of the malignant potential of ectopic tissue.

Conclusions

Intestinal duplications are rare congenital anomalies which vary in clinical presentation and complication rates such as bowel obstruction, gastrointestinal bleeding, bowel perforation, or malignant alterations. The increasing number of prenatally diagnosed duplications allows early planning and treatment, with the patients being in optimal conditions. Early diagnosis and treatment reduces the risk of dangerous complications like bowel perforation, bleeding, obstruction, and malignant alterations. Resection of a duplication alone is the treatment of choice, and in uncomplicated cases laparoscopic resection is advised. Gastric mucosa is often found in duplications. The prognosis of surgically treated patients is excellent.

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