Duplications of the Gastrointestinal Tract

Hagith Nagar, MD

Department of Pediatric Surgery, Tel Aviv Sourasky Medical Center, Tel Aviv, Israel

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Abstract

Background: Gastrointestinal duplications are rare, benign congenital lesions that may occur at any location along the alimentary tract and generally require surgical intervention. Presenting symptoms may be quite varied even among patients with the same anomaly.

Objective: To review the clinical presentation of gastrointestinal duplications and present our experience with such lesions over the past decade.

Methods: The records of all patients treated for gastrointestinal duplications at a tertiary hospital during 1987 through 1996 were collected, and relevant published literature reviewed.

Results: In the nine patients with gastrointestinal duplications, six were in the small bowel and one each in the cecum, colon and esophagus. Presenting clinical features were varied and often subtle. Perinatal ultrasonography, radioscintography and computerized tomography were useful in some cases, while in others the correct diagnosis was established only at surgery.

Conclusions: Alimentary tract duplications are uncommon, and may present as solid or cystic tumors, intussusception, perforation or gastrointestinal bleeding. A high index of suspicion is required when dealing with such cases. Appropriate investigations, including imaging techniques, should be directed toward adequate and planned surgery.

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Gastrointestinal duplications are rare benign congenital lesions that may occur at any location along the alimentary tract and generally require surgical intervention. The normal and duplicated segments usually share a common muscular wall and blood supply, with the normal segment in proximity to the antimesenteric aspect of the duplication. Most such anomalies are cystic in nature, and tubular duplications account for only a small minority. Presenting symptoms may be quite varied, even among patients with the same anomaly, and a high index of suspicion is required when dealing with such cases. Clinical features may mimic tumor, obstruction, hemorrhage or intussusception. Initial signs may also include respiratory distress or fistulae. In some cases, the lesion is first noted as an incidental finding on ultrasonography or X-rays of the chest or abdomen.

Duplications of the foregut and hindgut are associated with additional congenital abnormalities of the spinal cord, vertebral column or intestine. Diagnosis relies on computerized tomography, ultrasonography [1] or radioisotopic scans, the latter being particularly useful in cases of bleeding. Neoplasia, including carcinoid tumors, may develop in duplications later in life [2,3].

Since duplications vary considerably in location and presentation, a wide variety of surgical techniques are advocated. Cystic lesions can generally be excised completely if surgery does not compromise blood supply to the normal segment. In such cases the duplication can be marsupialized or partially resected, and remaining mucosa stripped from the cyst wall. Tubular duplications should be totally excised if possible; however in some cases, the lesion can be employed as additional 'lumen' when re-anastomosing normal bowel.

Materials and Methods

The Tel Aviv Sourasky Medical Center is the principal tertiary hospital serving metropolitan Tel Aviv. The records of all patients treated for gastrointestinal duplications during 1987 through 1996 were collected, and the relevant published literature reviewed.

Results

During the past ten years we treated nine patients for gastrointestinal duplications: six patients had duplications in the small bowel, and one each in the cecum, colon and esophagus. None of the patients was found to have vertebral anomalies. The esophageal duplication was diagnosed only at surgery in a baby diagnosed as having hiatal hernia on prenatal ultrasound examination. Immediately following birth he was vomiting and could not be fed. The initial impression was of massive reflux due to the "hiatal hernia." At surgery a large cystic duplication was found, starting at the level of the upper esophagus and extending below the diaphragm and behind the gastroesophageal junction. The duplication had enlarged the hiatus and did not communicate with the gastrointestinal tract. Since the surgical approach was abdominal, the cyst was opened and the internal mucosal coat entirely removed [Figure 1]. At the end of the surgical procedure a contrast study demonstrated an intact esophagus. A Nissen fundoplication was performed. At present, the patient is 2 years postsurgery and thriving.

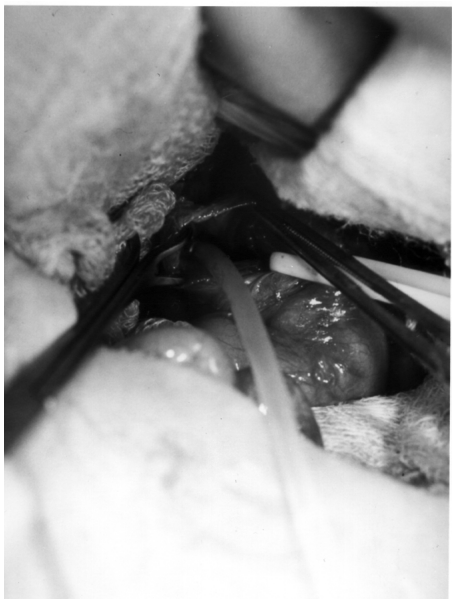

Figure 1. Intraoperative view looking cephalad from the abdominal cavity, showing a catheter inserted into the duplicated esophagus and a white band retracting the normal esophagus forward.

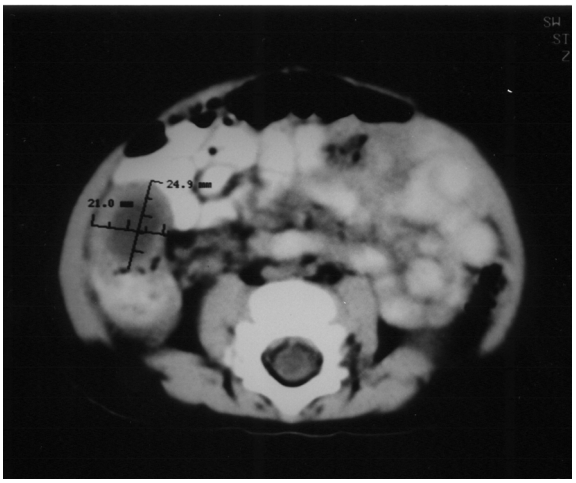

Figure 2. Axial CT scan of the abdomen, demonstrating a well-circumscribed oval fluid lesion adjacent to the cecum and measuring 2.1x2.5 cm.

All six small bowel duplications occurred in boys and were cystic in nature. One patient presented with intestinal obstruction at age 3 weeks. Another was diagnosed on prenatal ultrasound examination, and CT at birth confirmed the diagnosis. This infant underwent an elective resection and anastomosis at age 3 months. Two additional patients presented as intussusception at age 3 months and underwent successful surgery. One patient, a premature infant who weighed 800 g at birth and presented with free intestinal perforation, was one of two patients found to have ectopic gastric mucosa in the resected bowel. At surgery, we encountered three adjacent duplications, one of which had perforated. The sixth patient was a 3-year-old boy who presented with rectal bleeding. A technetium scan was

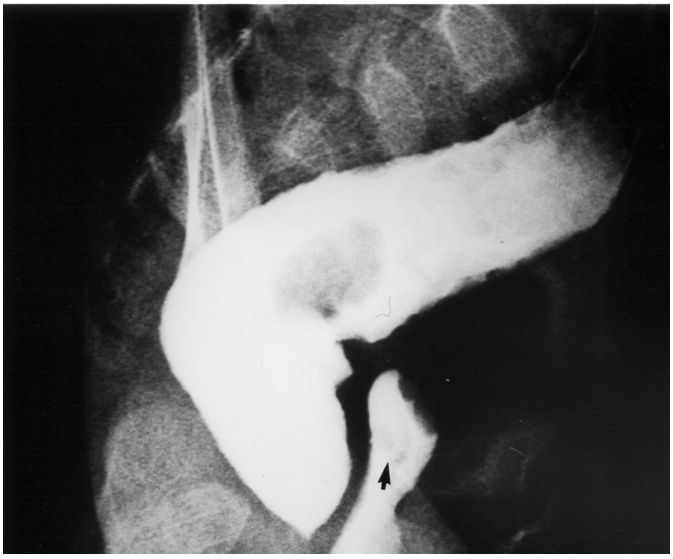

Figure 3. Barium cologram (via colostomy) demonstrating the duplicated colon with a fistula into the vagina (arrow).

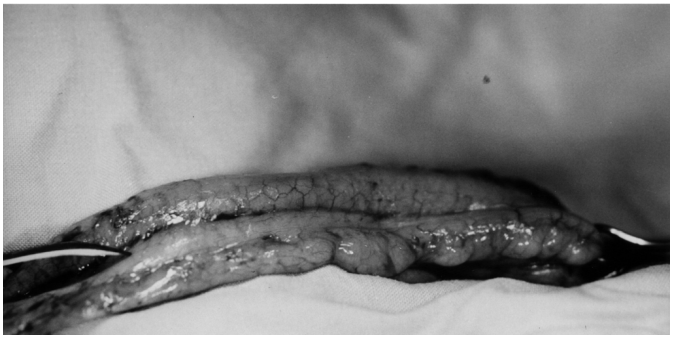

Figure 4. Intraoperative photograph of colonic duplication.

performed and interpreted as Meckel's diverticulum. Surgery revealed the cause of bleeding to be a duplication of the ileum.

The cecal duplication was diagnosed prenatally on maternal ultrasonography, and was confirmed postpartum using ultrasound and CT examination [Figure 2]. Elective surgery was performed at age 3 months. The last patient, a girl with tubular duplication of the colon, was referred to us at age 18 months. She was the product of a normal pregnancy and delivery, but at age 3 months the mother noted that stools were being passed both through the vagina and a normal anus. A right diverting colostomy identified a duplication extending from the distal ascending colon to the upper rectum and an associated rectovaginal fistula. Each arm of the colostomy contained two openings. A subsequent contrast study demonstrated an extremely redundant sigmoid and transverse colon ending at the anus, and a similar colonic duplication [Figure 3] at the rectovaginal fistula. The urogenital system was normal on ultrasonography. At surgery, the entire duplicated colon [Figure 4] was excised and the fistula was closed. The colostomy was closed 6 weeks later, and the patient is doing well one year after surgery.

Discussion

Alimentary tract duplication is a rare anomaly. Such anomalies are more common in boys than girls, and most cases are identified before the age of 2 years [4]. Foregut lesions in particular are associated with vertebral anomalies, most commonly cleft vertebrae [5].

Thoracic duplications are found in the posterior mediastinum and are generally cystic [6]. They may or may not contain gastric or squamous epithelium, may communicate with the duodenum or stomach, or may protrude into the right thoracic cavity. Patients may present with upper respiratory distress due to airway compression or vomiting. Diagnostic modalities include barium swallow and CT. Our case was quite unusual in that we were initially misled by a prenatal ultrasound examination, and a compressing mass was only noted on retrospective review of the upper gastrointestinal series.

Gastric duplications usually appear along the greater curvature and are generally cystic. Resection is the treatment of choice. Duodenal duplications are also cystic, but usually involve the second part of the duodenum and may communicate with the pancreatic ductal system. In this case, complete resection is also preferred, but marsupialization with internal drainage may be necessary for lesions in close proximity to the biliary system or pancreas. Intraoperative cholangiography is always indicated [4].

The jejunum and ileum are the most common sites for gastrointestinal duplication. Most lesions are cystic and are easily resected with the adjacent small bowel. Occasionally small bowel duplications are associated with intestinal atresia, suggesting that both anomalies may be related to a common vascular accident [4,7]. As in one of our cases, patients may present with intestinal obstruction of unknown cause, in which duplication is discovered only at surgery. In two additional patients, the anomaly presented as recurrent and irreducible intussusception. Other findings may include acute abdomen due to free perforation. Bleeding may occur in cases of communication with adjacent bowel lumen, and if gastric mucosa is present in the duplication, pertechnetate 99m Tc scintigraphy may be useful in locating the lesion [8,9].

Today, with ubiquitous use of perinatal ultrasound, cystic masses such as duplications within the fetal abdominal cavity should be suspected, especially in boys. Following delivery, close neonatal follow-up and a planned approach to surgical intervention will prevent unnecessary complications [10].

Large bowel duplications may be cystic or tubular, and are associated with anorectal or genitourinary malformations. Such associations may be related to the close anatomic proximity of the hindgut and genitourinary sinus [11]. Evaluation of the genitourinary tract is obligatory and therapy should be planned accordingly. Rectal duplications are rare and usually present as masses of variable shape, often distended with mucus lying within the presacral space [12,13]. Drainage of pus or mucus from the perineum may be an initial presenting feature.

In summary, alimentary tract duplications are uncommon. Investigations should be directed toward appropriate and planned surgery. The surgeon must be familiar with the anatomy and clinical characteristics of this entity, which should be included in the differential diagnosis of thoracic mass, gastrointestinal bleeding and rectal masses. The treatment of choice is resection.

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Correspondence: Dr. H. Nagar, Director of Pediatric Surgery, Tel Aviv Medical Center, 6 Weizmann St., Tel Aviv 64239, Israel. Tel: (972-3) 697 4538; Fax: (972-3) 613 2892; email: hnagar@post.tau.ac.il

Tell me and I know. Show me and I remember. Let me do it and I understand. To know is to know that you know nothing. That is the meaning of true knowledge.

Confucius