Y-Shaped Colonic Duplication: Report of A Case and Literature Review

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Colonic duplication is a very rare congenital anomaly that is usually detected in infancy and early childhood. In the English literature, 6 cases of Y-shaped colonic duplication have been reported since 1953. We conducted a review of the reported cases, and we present a new case of a Y-shaped duplication of the sigmoid colon manifesting as long-term abdominal pain and constipation. *(Chang Gung Med J 2011;34(6 Suppl):43-7)*

Key words: colon, duplication, pediatric

Intestinal duplications are rare congenital anomalies that show an incidence rate of 1 in 4000–5000 births.[1] The underlying causes remain poorly understood and speculative. They can occur anywhere in the alimentary tract and are typically located on the mesenteric side of the native bowel. Intestinal duplications can be cystic or tubular, with the former being more common. This condition is usually detected in infancy or early childhood when the child complains of pain and shows signs of obstructive disease. Surgical resection remains the treatment of choice in most cases. The ileum is most frequently involved, and colonic duplications are among the rarest, representing less than 10–15% of all intestinal duplications.[1,2] A Y-shaped colonic duplication, a variant of the tubular type, is extremely rare. Only 6 cases have been documented in the English literature since 1953. In this article, we report a Y-shaped colonic duplication in a teenage girl who experienced long-standing abdominal pain and constipation.

CASE REPORT

A 12-year-old girl visited our hospital because of long-standing abdominal pain and constipation. She was diagnosed with gastric volvulus 3 years previously, and had undergone a laparoscopic gastropexy; however, the symptoms persisted, and only symptomatic treatment was given thereafter. On examination at our hospital, a painful ovoid mass approximately 10 cm in diameter was found in the mid-abdominal region. Blood test results were within normal limits. Plain radiography of the abdomen revealed dilation of the intestine with abundant fecal content. A barium enema examination revealed a Y-shaped colonic duplication originating from the sigmoid colon (Fig. 1).

During the operation, a 21.5 x 5 x 5 cm tubular bowel segment originating from the mesenteric side of the sigmoid colon was identified. The tubular segment, which was distended with fecal material, extended along the retroperitoneum beneath the descending colon and terminated at the level of the

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left kidney. This tubular structure shared a wall with the native sigmoid colon for about 3 cm, and a small, 1 cm. orifice was noted at this junction. The duplicated colon and the involved sigmoid segment were resected after complete mobilization of the left side of the colon (Fig. 2). An end-to-end anastomosis was achieved without any complications. Histological examination revealed well-developed smooth muscle layers on both the original and the duplicated colon. The mucosa and submucosa were flattened and atrophic without gastric or pancreatic ectopic tissue. The patient tested negative for vertebral and genitourinary malformations.

DISCUSSION

Colonic duplications are rare congenital anomalies, and more than 85% of cases are cystic.[1] Tubular duplications can be further categorized as double-barreled and Y-shaped types,[3] of which the latter is rarer. The tubular duplicated colon shares enteral continuity with the native colon, forming a Y-shaped structure; thus, the entity is called a Y-shaped colonic duplication. The 6 previously reported cases and the present case of Y-shaped colonic duplication are summarized in the Table.

Colonic duplications are mainly diagnosed and treated in infancy and early childhood. This anomaly is predominant in females with an incidence ratio of around 2:1; no familial or racial predilection has been reported.[4] Most patients present with a combination of pain and obstruction symptoms, whereas a few cases show abdominal masses or complications such as bleeding, perforation, volvulus, and intussusceptions.[1,3] Colonic duplications can be completely asymptomatic throughout the course of the disorder, as reported in about 10% of patients.[5] Nearly 80% of the patients with colonic duplications are reported to have at least 1 extragastrointestinal anomaly, mostly in the genital and urinary systems, and diverse degrees of genitourinary or enterourinary fistulous communication are found in half of patients.[6,7] Less frequently, colonic duplication may manifest in the form of skeletal anomalies such as a duplicated vertebral column, hemivertebrae, or separated pubic symphysis.[8,9] More than half of patients with double-barreled lesions have distal anomalies,

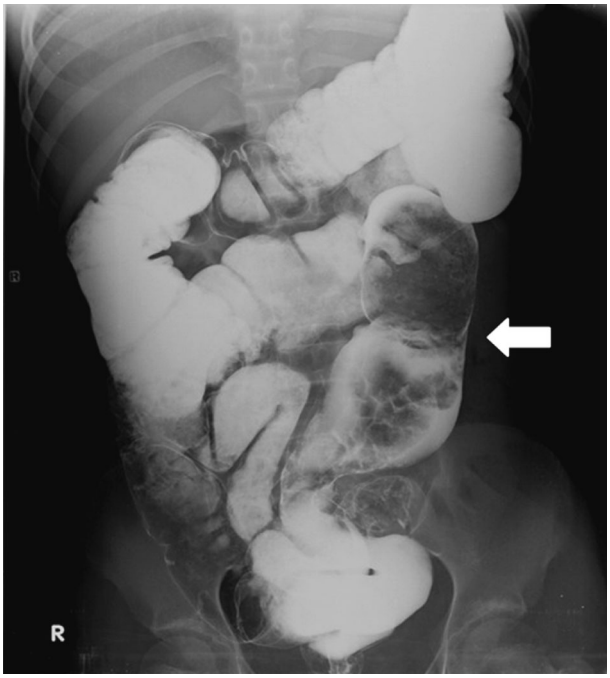

Fig. 1 Barium enema examination reveals a Y-shaped structure formed by the sigmoid and duplicated colonic segment (white arrow). The sac of the duplicated colon segment is distended with abundant fecal material.

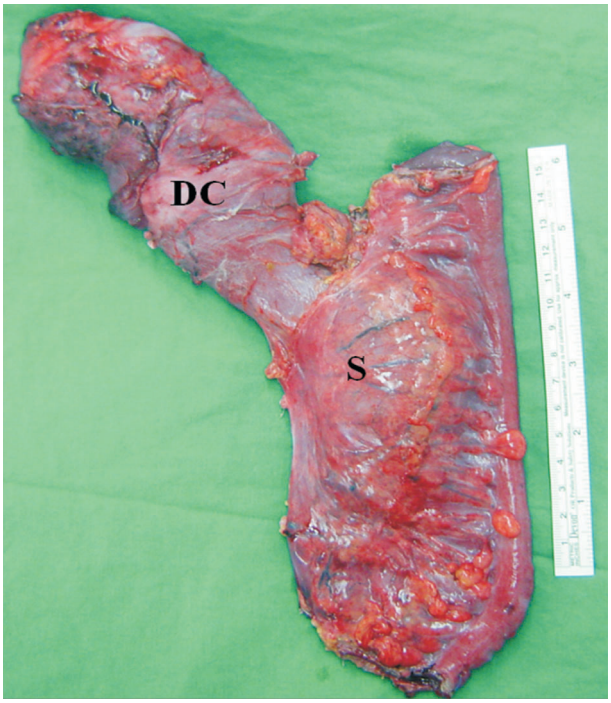

Fig. 2 The Y-shaped duplicated colon (DC) and the native sigmoid colon (S).

Table Reported Cases of Y-shaped Colonic Duplication

Reference	Age / Sex	Origin / Location	Length	Gastric (cm)	Symptoms mucosa	Anomaly	Diagnosis	Treatment
3	8d/F	Sigmoid / Intraperitoneum	5	nil	Peritonitis due to perforation	nil	Intra-operative	Resection†
14	12y/M	Transverse / Intraperitoneum	12	nil	Pain, abdominal mass, obstruction	nil	LGI series	Resection
15	15y/F	Right / Intraperitoneum	35	nil	Incidental detection when appendicitis was diagnosed	nil	Intra-operative	Resection†
13	6y/F	Transverse / Intraperitoneum	24	nil	Pain, abdominal mass, obstruction	nil	Intra-operative	Resection
16	7y/F	Descending / Retroperitoneum	31	nil	Pain, abdominal mass	nil	CT, LGI series	2-stage operation (including temporal loop colostomy)*
17	35y/F	Sigmoid / Both	12.5	nil	Pain, abdominal mass	nil	CT, LGI series, colonoscopy	Resection*
Present case	12y/F	Sigmoid / Both	21.5	nil	Pain, abdominal mass, obstruction	nil	LGI series	Resection

Abbreviations: LGI series: lower gastrointestinal series; CT: computed tomography; *: Laparoscopic treatment; †: Resection of the lesion alone; others: resection of the lesion and the involved native bowel segment; d: day; y: year

terminal fistulae, or an imperforate anus, which prevent adequate drainage. However, lesions in patients with inherently adequate drainage through distal communication with the colon or a second anus are usually asymptomatic and the condition may remain unrecognized unless it is associated with other congenital anomalies that necessitate clinical attention. In these cases, the patient is usually diagnosed in the perinatal period or during infancy. Non-communicating Y-shaped lesions, such as double-barreled lesions with inadequate drainage, are often enlarged due to internal secretion, and at times, impaction of bowel contents; these lesions progressively compress the adjacent organs. Since these duplications communicate proximally and not distally, they become distended with fecal material and cause obstruction of the adjacent bowel. Furthermore, pain may result from overdistension of the duplication, inflammation caused by secretions containing proteolytic enzymes, or mass effect on the blood supply associated with the lesion. Although gastrointestinal bleeding is rarely severe, it may result from ulceration and eventual erosion of the adjacent organs and/or vessels because of enzymatic secretions or ischemia caused by stretching of the mesenteric vessels around the duplications. None of the reviewed patients with Y-shaped colonic duplications had related anomalies; this finding is attributable to the small number of reviewed cases. A delayed diagnosis of isolated colonic duplication may be related to the equivocal and non-specific symptoms.[7] Four of the 7 isolated Y-shaped colonic duplications reviewed here were diagnosed when the patients were almost teenagers or later, rather than in infancy or early childhood.

The challenge for clinicians is not the treatment of colonic duplication, especially for Y-shaped colonic duplication, which is simple, but its clinical diagnosis because the condition is rarely diagnosed accurately before surgery. The detection of an additional intestinal loop originating from the native colon or bifurcation of the intestinal lumen on computed tomography or magnetic resonance imaging after barium enema administration, or that of bifurcation of the colonic lumen on colonoscopy can help diagnose colonic duplication. The treatment for colonic duplication is surgical resection, not only to relieve symptoms, but also to eliminate the risks of complications caused by ectopic gastric mucosa and mucoviscidosis. Furthermore, surgery can help eliminate the risk of adenocarcinoma since the rate of

occurrence of adenocarcinoma in the colon is higher than at other locations.[10-12] Y-shaped colonic duplications are different from double-barreled duplications in that they can usually be resected completely without compromising the original bowel. Moreover, the treatment for double-barreled duplications is complicated because they are strongly associated with distal anomalies, terminal fistulae, or an imperforate anus, whereas, the treatment for Y-shaped duplications is quite simple. Complete surgical resection of the involved intestine is possible because of the common blood supply between the duplicated and the native colon. However, an extended excision for at least 2 cm from the junction is necessary because pathologic fibrosis may occur near the opening of the duplication.[13]

In conclusion, Y-shaped colonic duplications are rare congenital anomalies that tend to affect the left-side of the colon predominantly in female patients; however, the exact cause is unknown. Pain and obstruction with evidence of a segmental bowel loop containing gas or fecal material are commonly evaluated by radiography. Y-shaped colonic duplications should be included in the differential diagnosis of abdominal pain in children. The diagnosis of colonic duplication needs special attention in clinical practice, and surgical resection should be indicated to avoid complications and a tendency for malignant degeneration.

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Y 型結腸複製畸形：案例報導與文獻回顧

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結腸複製囊腫是一極罕見，通常是被診斷於嬰兒與幼兒的先天畸形。其中，Y 型結腸複製畸形更是少見，自 1953 年起於英文文獻僅有 6 案例被報導。本文章報導一以長期腹痛與便秘作表現的新診斷個案與過往案例的回顧。(長庚醫誌 2011;34(6 Suppl):43-7)

關鍵詞：大腸，複製囊腫，小兒科的

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