Congenital Segmental Dilatation of the Colon

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Summary: Segmental dilatation of the sigmoid colon in adults and the surgical treatment are reported with a review of some of the literatures. 1) There have been 10 cases of congenital segmental dilatation of the sigmoid colon, including this case. 2) Hirschsprung's disease is an important disease for the differential diagnosis of segmental dilatation of the colon. Resection of the segmentally dilated portion of the colon and end-to-end anastomosis are the best treatments, and these procedures can produce good results. 3) The cause of segmental dilatation of the colon is unknown.

Key words: congenital—segmental dilatation of the colon—anorectal manometry—Hirschsprung's disease—Auerbach's plexus.

Introduction

Segmental dilatation of the colon is very rare. This disease was first reported by Swenson et al. (1959). However, the etiology is still unknown. Because there is no mechanical stenosis of the intestine or pathological abnormality of the ganglion cells, only local dilatation is found.

One female patient, 26 years of age, with congenital segmental dilatation of the sigmoid colon was treated in our department. The case history, clinical examination and histological details are presented together with a short review of the literature.

Case Report

A 26 year-old woman was admitted to our department. She complained of abdominal distension and constipation. The family and past histories were not unusual. In 1981, a laparotomy and artificial miscarriage were performed because of a volvulus of the sigmoid colon. The postoperative prognosis was good. In 1982, the operation was performed again for the same reason. After the operation, constipation continued to occur and laxatives usually had to be used. An abdominal X-ray examination showed a large loop in the colon.

The blood and chemical results were:

- WBC, 5,100/mm³; RBC, 440 × 10⁴/mm³; Hb, 9.5 g/dl; Ht, 31.3%; B Plat, 29.4 × 10⁴/mm³; Tbil, 6 mg/dl; GOT, 11.7 mg/dl; GPT, 6.0 mg/dl; LDH, 194.6 mg/dl; ALP, 6.5 mg/dl; TP, 7.8 g/dl; ALB, 3.6 g/dl; TTT, 1.8 g/dl; ZTT, 11.4 g/dl; Ch-E, 0.8 g/dl; T-cho, 145.5 g/dl; Amylase, 120 g/l; Na, 141 mEq/l; Cl, 105 mEq/l

Digestive hormone- Gastrin, 20 pg/ml; Glucagon, 72 pg/ml; PGE, 415 pg/ml; PGE2α, 570 pg/ml; Somatostatin, 1.0 pg/ml; Secretin, 67 pg/ml; VIP, 35 pg/ml.

Before the operation an abdominal X-ray (Fig. 1) showed a large loop and considerable gas at the left side of the colon with small quantities of gas in the small intestine. A barium enema (Fig. 1) demonstrated a distended loop from the de-
scending colon to the sigmoid colon. However the sigmoid colon and rectum were almost normal.

The results of an anorectal manometry of the rectum were normal (Fig. 2). The length of anus was also normal.

Fig. 1. Abdominal sample X-Ray (right) and Barium enema (left) showed a dilatation of sigmoid colon.

Fig. 2. Ano-rectal Resting Pressure Profile is normal.

Fig. 3. Condition of sigmoid colon in the operation.
Preoperative diagnosis: Segmental dilatation of the sigmoid colon.

Operative findings (Fig. 3, 4): It was observed during the median laparotomy of the lower abdomen that the sigmoid colon was widened, hypertrophic and distorted by 180 degrees with no adhesion by the peritoneum. Passage failure in this part was not present. The root of the mesentery and the rotatory of the colon were normal. The sigmoid colon including the dilated part of the colon was resected. An end-to-end anastomosis was performed.

Pathological examination (Fig. 5, 6): The resected colon had a normal Auerbach's Plexus between the inner circular muscle and outer longitudinal muscle layer. After staining for acetylcholinesterase by the method of Karunovsky or Root, the histological findings were also normal in the resected part.

Fig. 4. Operative finding showed a dilatated sigmoid colon.

Fig. 5. Pathological examination (H and E. ×200) is normal.
Fig. 6. Pathological examination (Karunoysky and Root ×200) is normal.
CONGENITAL SEGMENTAL DILATATION OF THE COLON

Postoperative anorectal manometry: Normal as before the operation.
Postoperative diagnosis: congenital segmental dilatation of the sigmoid colon.
Postoperative prognosis: there was considerable gas in the descending colon, but no constipation.

Discussion

In 1959, Swenson reported 3 cases of segmental dilatation of the colon as a new disease. In these cases the obstructive symptoms of the digestive tract were considered to be caused by movement disturbances of the dilated tract. Resection of the dilated tract was performed in these cases. The movement of the tract without the dilated region was normal. The pathological examination indicated that the muscle layer was normal, the number and condition of the nerve fibers and nerve plexuses were also normal. Thus a neural mechanism was suspected for the movement disturbance of the dilated segment of the tract. The differential diagnosis consisted of invagination, diverticulum, and Hirschsprung’s disease. Now 10 cases of segmental dilatation including the present case, have been reported. The ages of the patients were: 9 days after birth to 26 years. There were 5 males and 5 females. The dilated portion of the tract usually occurred in the small intestine and colon, and most frequently in the small intestine.

There are only 9 cases of the segmental dilatation of sigmoid colon, including this case, in the literature. The chief complaints are abdominal distension and constipation, it is very difficult to distinguish between segmental dilatation of the sigmoid colon and Hirschsprung’s disease; thus anorectal manometry and a biopsy of the rectum should be performed. In this case the barium enema did not demonstrate a narrow segment and a caliber change. The anorectal manometry was normal, so it was easy to distinguish from Hirschsprung’s disease.

The etiology remains unclear. In adults, the chronic constipation, malrotation and elongation of the sigmoid colon are considered the cause of the symptoms. But three papers which (Swenson et al. 1959; Aterman et al. 1967; De Lorimier et al. 1971) reported support a congenital cause, especially in infants with segmental dilatation of the colon which was reported by (De Lorimier et al. 1971; Kusaba et al. 1983); because 2 cases of segmental dilatation of the sigmoid colon were complicated with cystocele, atresia and abnormalities of the spine and ribs, chromosome aberrations, polydactyly and congenital heart disease. Furthermore, pathological thickening and atrophy of the muscle layer were found.

As to the treatment of this disease, segmental resection with end-to-end anastomosis is recommended. This is the procedure that was performed in all the reported cases.

References