Case Report

Surgical Treatment of Idiopathic Mesenteric Phlebosclerosis: A Case Report

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ABSTRACT

An 81-year-old woman who had been taking Chinese herbal medicines visited our hospital with the chief complaint of lower abdominal pain and constipation. She underwent colonoscopy because of the finding of fecal occult blood. Colonoscopy revealed mucosal edema along the entire circumference of the transverse colon to the descending colon with dark purplish and bluish discoloration and ulcerations. Computed tomography revealed thickening of the wall and linear and ramified calcifications on right side of the colon. These findings suggested idiopathic mesenteric phlebosclerosis. As conservative treatment did not improve symptoms, we performed subtotal colectomy. Histopathologic examination of the resected colon with Masson’s trichrome stain showed a significant increase in perivenous collagen fibers and occluded venous lumina, whereas Direct Fast Scarlet stain revealed no amyloid deposits. Therefore, a definitive diagnosis of idiopathic mesentery phlebosclerosis was made. The symptoms resolved after surgery, and the patient has had no recurrence of symptoms.

Key words: idiopathic mesenteric phlebosclerosis, mesenteric phlebosclerostic colitis, ischemic colitis

INTRODUCTION

Idiopathic mesenteric phlebosclerosis is a type of ischemic colitis that results from venous congestion due to phlebosclerosis of the right colon. It is a recently identified condition, and few cases have been reported. An association with the use of Chinese herbal medicines has been suggested. We report a case of idiopathic mesenteric phlebosclerosis that responded well to surgical treatment.

CASE REPORT

An 81-year-old woman, who had a history of hypertension and autonomic ataxia and a 15-year history of use of Chinese herbal medicines (kami shoyo san and tokaku jokito), had been having lower abdominal pain and constipation for 3 months. Because these symptoms did not improve, she visited our hospital’s department of gastroenterology. After fecal occult blood was found, she underwent colonoscopy, which revealed dark purplish to bluish discoloration, mucosal edema, and ulcerations. Because the lumen was occluded, endoscopic examination...
had to be discontinued at the midpoint of the transverse colon. She was admitted and treated with bowel rest and probiotics medication for two months.

However, because the symptoms of ileus did not improve with this conservative treatment, then she was referred to the department of surgery.

Results of physical examination at time of consultation were as follows: height, 140 cm; weight, 30 kg; temperature, 37.1°C; with a flat, soft abdomen but slight tenderness in the right lower quadrant.

Laboratory studies showed an elevated white blood cell count (11,700/µl), C-reactive protein level (8.65 mg/dl), and lactate dehydrogenase level (316 U/l).

Plain abdominal radiography showed punctiform calcifications along the intestinal wall between the cecum and the transverse colon (Fig. 1A).

Barium enema examination showed the disappearance of haustra coli, thumb-printing signs, mural sclerosis, and
narrowing of the lumen between the cecum and the splenic flexure (Fig. 1B).

Computed tomography (CT) of the abdomen showed fluid retention throughout the small intestine, edema and thickening of the intestinal wall, and calcification of the mesenteric veins between the ileocecum and the transverse colon (Fig. 1C, 1D).

Colonoscopy after conservative treatment revealed continued mucosal edema, dark purplish to bluish discoloration, and ulcerations throughout the circumference of the transverse colon and the descending colon (Fig. 2A, 2B). The narrow lumen forced us to abandon the endoscopic examination at the midpoint of the transverse colon. The mucosa from the transition between the sigmoid colon and the descending colon to the anus did not show bluish discoloration, and we applied clips as markers at the border of the bluish discoloration.

Conservative treatment with probiotics was not effective, and symptoms of paralytic ileus persisted. Therefore we performed subtotal colectomy. Laparotomy revealed dark purplish to bluish discoloration of the intestinal tract with calcifications from the cecum to nearly the midpoint of the transverse colon. The mesenteric veins of the colon showed an extremely hard, cord-like structure as a result of thrombi. The superior mesenteric artery pulse was palpated. The resection line had been marked with clips before surgery, and subtotal resection of the colon was performed at the junction between the sigmoid colon and the descending colon.

Gross examination of the resected specimen showed mucosal edema, dark purplish to bluish discoloration, and calcification of the intestinal wall and mesenteric veins were observed (Fig. 3A).

Microscopic examination showed significant transmural hyperplasia of collagen fibers in the resected colon and infiltration of slightly inflamed cells into the mucosa (Fig. 3B). In addition, Masson’s trichrome stain showed narrowing of the venous lumina (Fig. 3C). Direct Fast Scarlet stain did not show amyloid deposits (Fig. 3D).

The patient showed a satisfactory postoperative course with complete resolution of symptoms. After discharge, food intake was also satisfactory and allowed an 8-kg weight gain after 6 months. The patient is now undergoing follow-up observation as an outpatient and has had no recurrence of symptoms.

**Discussion**

Idiopathic mesenteric phlebosclerosis was first reported in 1991 by Koyama et al in a patient with right-sided ischemic colitis. Thereafter, an increasing number of cases were reported, leading to the establishment of a peculiar disease entity, which differs from previously known ischemic colitides due to impeded arterial blood flow that is
common in the left colon. In 2000, Yao et al., first used the term “phlebosclerotic colitis,” and in 2003 Iwashita et al., used the term “idiopathic mesenteric phlebosclerosis” to describe a condition caused by phlebosclerosis rather than inflammatory lesions.

A search of PubMed from the publication of the 1991 report by Koyama et al. through August 2013, excluding the minutes of meetings, and using the search terms “idiopathic mesenteric phlebosclerosis,” “phlebosclerotic colitis,” and “ischemic colitis” yielded 57 articles. Although most cases were reported in Japan, a few cases have been reported in other Asian countries. One case was reported in the European/American region, but the patient was a Taiwanese, which suggests the effect of race and environment. The number of reports has increased as idiopathic mesentery phlebosclerosis has become more widely known.

The observed characteristics of idiopathic mesentery phlebosclerosis are as follows: calcification from the site of lesions in the wall of the large intestine to the mesenteric veins, mainly in the right colon, and functional disorder caused by venous congestion. Although lesions during the early period are in the cecum or ascending colon, they are believed to gradually advance toward the anus. Clinical symptoms are localized to mainly the right lower quadrant.

Fig. 3A. Postoperative macroscopic examinations findings
The mesentery from the cecum to the splenic flexure showed dark purplish discoloration, and the intestinal wall showed thickening (arrowed area).

Fig. 3B/3C/3D. Findings of postoperative pathological examinations
3B: The mesenteric submucosa and vein walls in the ascending colon showed abundant deposition of collagen fibers (oval circled area) (Masson trichrome stain, ×200).
3C: Most of the blood vessels showing hyperplasia of collagen fibers were veins, and the lumina of most of these veins were occluded (arrowed areas).
3D: No amyloid deposits were observed (oval circled area) (Direct Fast Scarlet stain, ×200).
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to the right flank and include chronic pain, nausea, vomiting, diarrhea, bloody stool discharge, constipation, and ileus. Tenderness of the right lower quadrant and, sometimes, induration are also found. The disease progresses slowly and has no symptoms during the onset phase. Thus, some cases are identified through screening and other means. Blood tests indicate nonspecific inflammation, anemia, and malnutrition.

The diagnosis idiopathic mesenteric phlebosclerosis is based on characteristic imaging findings. On abdominal simple X-ray films, punctiform and ramified calcifications are observed along the vasa recta of the marginal artery. Barium enema examination of the affected colon shows thumb-printing signs, edematous lesions, narrowing, serration, calcification, the disappearance of semilunar folds, and longitudinal ulcers. Abdominal CT shows thickening of the intestinal wall and both punctiform and ramified calcifications along the veins on the inner wall of the digestive tract and along the vasa recta of the marginal artery. Colonoscopy reveals bluish-white to dark purplish discoloration of the mucosa of the lesions with edema and disappearance of the visible vascular pattern. The lumen is narrowed, hardened, and inelastic, and ulceration and ulcers (irregularly-shaped and longitudinal ulcers) are sometimes present. In the onset phase, colonoscopy shows dark bluish discoloration, but CT does not show characteristic calcification. Thus, the characteristic calcification is believed to develop as the disease progresses2.

The characteristics histological findings are significant fibrous thickening and calcification of the venous walls in the area of the lesion, a high degree of fibrosis in the submucosa, and significant perivascular deposition of collagen fibers in the lamina propria. Amyloidosis and other types of amyloid deposits are not observed.

Idiopathic mesenteric phlebosclerosis was first thought to be caused by ischemic colitis but is actually a disorder of mesenteric venous congestion accompanied by venous sclerosis. The mechanism is thought to involve the presence of intestinal stimulants, vasculitis accompanied by systemic collagen disease, thrombophlebitis, autoimmune disease, hypercoagulability, arteriosclerosis, and portal hypertension but has not been fully clarified. However, many patients are married couples who have a history of long-term use of Chinese herbal medicines4,9, which are suggested to be a possible cause.

Most of the effective components of Chinese herbal medicines are glycosides that are metabolized mainly by the bacterial flora and are absorbed in the intestine10. Therefore, as the glycosides are absorbed, toxic agents in the Chinese herbal medicine cause damage to the tunica intima of the superior mesenteric vein and its tributaries. This damage causes phlebostasis and progresses to phlebosclerosis. Lesion formation occurred from right colon which is a tributary of the ileocolic vein (which itself is a tributary of the superior mesenteric vein) toward the anal side.

Hiramatsu et al11 investigated 25 patients with idiopathic mesenteric phlebosclerosis who had used Chinese herbal medicines: 12 used kami shoyosan, 5 used shinisei haito, 4 used orenkei dokuto, and 1 each used kami kihito, bofutsu shosan, gorinsan, or inchinko to. The components these medicines share include Gardenia jasminoides (contained in 24 of the 25) and Scutellaria baicalensis root or ogon (contained in 11 of the 25), which suggests their involvement in the development of idiopathic mesenteric phlebosclerosis. G. jasminoides is an evergreen shrub (family Rubiaceae) that grows on mountainsides in warm climates and whose ripe fruit is dried and used as a medicine for the treatment of liver disease, hypertension, autonomic ataxia, dysmenorrhea, and other disorders due to its choleretic, antihypertensive, sedative, antipyretic, antiphlogistic, and hemostatic effects. Most of the cases suffered from IMP had been taking Chinese herbal medicine more than ten years. However there was a case for four years. Therefore if patients who have been taking Chinese herbal medicine for more than five years have a repeated symptom of abdominal pain, diarrhea, constipation, distension or nausea, stopping medications and checkups by CT or Endoscopy should be recommended. The present case was a long-time user of Kami Shoyosan for fifteen years, which suggests that it may have been one cause of the onset of the disease.

The clinical manifestations of idiopathic mesenteric phlebosclerosis include edema and peristaltic dysfunction due to venous congestion, which leads to abdominal pain, nausea, vomiting, diarrhea, bloody bowel discharge, constipation, and ileus. The symptoms often resolve with conservative treatment, including bowel rest and gastrointestinal prokinetic agents. However, if the symptoms continue to recur or if conservative treatment is not effective, surgical treatment is necessary.
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<th>Gender</th>
<th>Chief complaints</th>
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I: Ileum, C: Cecum, A: Ascending colon, T: Transverse colon, D: Descending colon, S: Sigmoid colon, R: Rectum
Our review of 58 previously reported cases of idiopathic mesenteric phlebosclerosis shows that surgical treatment was performed in 29 cases (Table 1). The patients were 12 men and 17 women with a mean age of 59.1 years (age range, 33–81 years). Surgical indications were as follows: symptoms of ileus, 13 cases; abdominal pain, 8 cases; perforation, 2 cases; surgery for other illnesses, 3 cases; and not documented, 3 cases. The affected areas were as follows: the ileum terminal or cecum to the ascending colon, 3 cases; cecum to the transverse colon, 16 cases; cecum to the descending colon, 4 cases; and cecum to the sigmoid colon, 6 cases. The surgical procedures selected were as follows: right hemicolectomy, 10 cases; extended right hemicolectomy, 2 cases; subtotal colectomy, 15 cases; and total colectomy, 2 cases. These cases indicate that the more advanced the disease, the more extensive the resection. Neither complications associated with the surgery nor recurrence has been reported.

Recently, idiopathic mesenteric phlebosclerosis has been treated with laparoscopic surgery, which provides good visibility with only a minimal skin incision and allows the color of the affected colon to be monitored. Because idiopathic mesenteric phlebosclerosis is not a malignant disease, laparoscopic surgery might reduce the incidence of surgical complications and be more satisfying cosmetically.

If identified at an early stage, idiopathic mesenteric phlebosclerosis shows improvement with conservative treatment. However, if such treatment is ineffective, surgery is necessary. Because the area of resection increases as the disease progresses, proactive approaches, such as early detection and treatment and the discontinuation of Chinese herbal medicine use, are necessary.

Authors have no conflict of interest.

References