—Report on Experiments and Clinical Cases—

A Clinical Evaluation of Lymphangioma of the Large Intestine A Case Presentation of Lymphangioma of the Descending Colon and a Review of 279 Japanese Cases

Takeshi Matsuda[1], Takeshi Matsutani[1], Yoshikazu Tsuchiya[1], Yuji Okihama[1], Kaku Egami[2], Masatomo Yoshioka[2], Shotaro Maeda[3] and Masahiko Onda[4]

Departments of [1]Gastroenterology, [2]Surgery, and [3]Pathology, Tama-Nagayama Hospital, Nippon Medical School
[4]Department of Surgery (I), Nippon Medical School

Abstract

With the development and widespread use of colonoscopy, lymphangioma of the large intestine has recently been reported frequently. This paper presents some findings from a review of 279 cases of this disease in Japan, including a typical case that we encountered. A 69-year-old female was diagnosed as having lymphangioma of the descending colon based on the findings of a barium enema and a colonoscopy, and the lesion was successfully removed by an endoscopic resection. In the published reports, the etiology of this disease is not clear yet but the age at onset range shows a tendency toward a higher incidence in comparatively older patients and the male-to-female ratio indicates a higher incidence in males. If there is no complication, endoscopic treatment seems to be the preferable procedure for this disease. (J Nippon Med Sch 2001; 68: 262—265)

Key words: lymphangioma, large intestine, endoscopic treatment

Introduction

Colorectal lymphangioma used to be considered an extremely rare disease, but recently, along with the increasing prevalence of endoscopy, it has become more commonly encountered in general medical practice and has been reported more frequently. Until 2000, 279 cases have been reported in the Japanese literature, including our case. We encountered a typical case with lymphangioma of the descending colon, which was successfully treated by endoscopic resection.

Case Report

A 69-year-old Japanese woman with no significant past medical history consulted our hospital with a chief complaint of constipation and lower abdominal discomfort in July 1996. On physical examination of the abdomen, there was no remarkable abnormality. Laboratory studies were within normal limits. She underwent a barium enema study and a colonoscopy for close examination. The barium enema demonstrated an elevated lesion with a gently rising margin and smooth surface in the descending colon (**Fig. 1**), and the endoscopic examination revealed a glossy, pellucid, bluish, and soft submucosal tumor with a so-called

Correspondence to Takeshi Matsuda, MD, PhD, Department of Gastroenterology, Nippon Medical School Tama-Nagayama Hospital, 1–7–1 Nagayama, Tama-shi, Tokyo 206–8512, Japan
E-mail: matsuda takeshi/surg 1@nms. ac. jp
Journal Website (http://www.nms.ac.jp/jnms/)

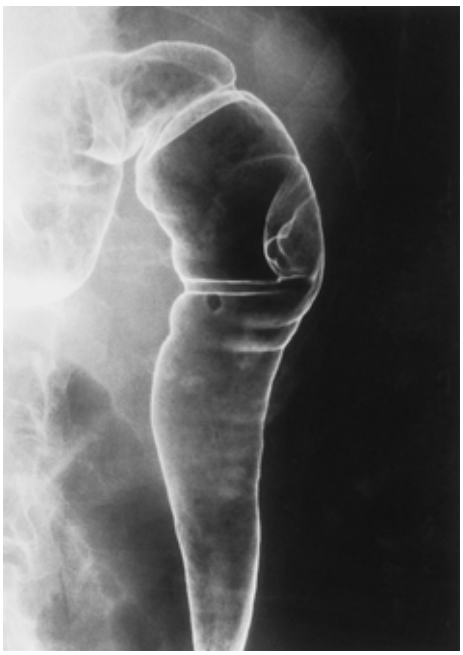

Fig. 1 Barium enema examination showed a tumorous lesion in the descending colon.

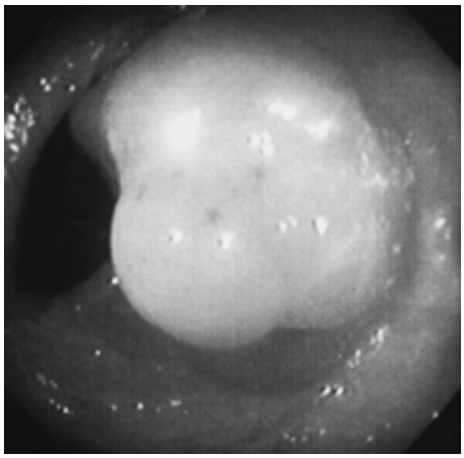

Fig. 2 Colonoscopy showed a protrusion, which was a slightly bluish and soft submucosal tumor with a smooth surface, located in the descending colon.

positive cushion sign in the same area (**Fig. 2**). These findings were suggestive of lymphangioma, and the entire tumor was removed by an endoscopic resection using a snare. The resected mass measured 0.8 × 0.7 × 0.5 cm, and was composed of several cysts filled with lemon-yellow serous content. Histopathologically, cysts whose lumen was covered with unilaminar endothelial cells were observed below the normal mucosa, leading to a diagnosis of cystic lymphangioma (**Fig. 3**). The patient is showing an uneventful course without any sign of recurrence.

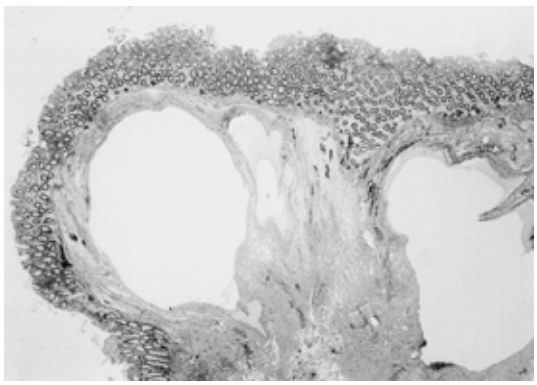

Fig. 3 Microscopic findings of the resected specimen revealed that the tumor was a lymphatic cyst covered with normal colonic mucosa, and that the lumen was lined by monolayered endothelial cells (H & E).

Discussion

Lymphangiomas arising in the abdominal organs are rare, and the majority arise in the mesentery, omentum, mesocolon, and retroperitoneum[1]. Those a rising in the wall of the intestine are considered to be even more rare. Colorectal lymphangioma was initially described by Chisholm et al. (1932)[2]. In Japan, since a report by Yoshitoshi et al. (1965)[3], this disease has been regarded as an exceptionally rare illness. However, with the development and widespread use of colonoscopy, it has recently been reported more frequently. Until 2000, 278 cases had been reported in the Japanese literature, and 279 cases, including our case, were invastigated for the following items.

1. Age at onset and gender

The age at onset ranged from one to 83 years (mean: 55.2 ± 14.1 years), and the male-to-female ratio was 150: 92, indicating a higher incidence in males (**Table 1**). It is generally agreed that lymphangiomas are a kind of hamartomatous or malformative lesions, and most of them are thought to be present at birth[1]. However, almost ail of the reported cases of lymphangioma of the large intestine had been found in adults, and patients aged 60 years or older comprised 40.4% (92 cases) of the patients whose ages were defined, showing a tendency toward a higher incidence in

Table 1 Frequency of colorectal lymphangioma by age and sex

Age (years)	Sex			Total
	Male	Female	Unknown	
0～10	0	1	0	1
11～20	1	1	0	2
21～30	5	3	0	8
31～40	10	9	0	19
41～50	40	18	0	58
51～60	24	25	0	49
61～70	33	24	0	57
71～80	24	9	0	33
81～90	1	0	0	1
Uncertain	12	2	37	51
Total	150	92	37	279

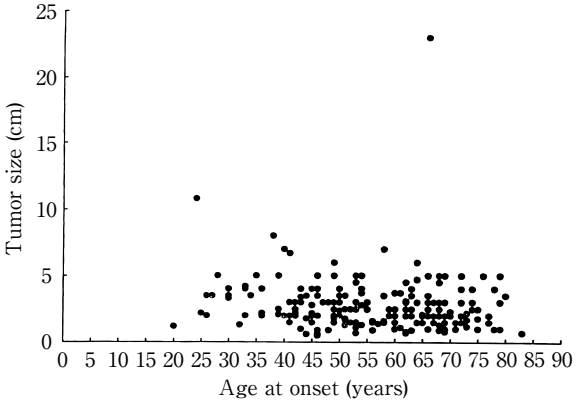

Fig. 4 Correlation between the size of the colorectal lymphangioma and the age at onset.

comparatively older patients. In cases of lymphangioma appearing late in adulthood, the possibility that the lesions have developed secondarily from local disturbance of the lymphatic circulation is not excluded.

2. The site of origin

In the 252 cases in which the site of origin was identified, it was the cecum: ascending colon: hepatic flexure: transverse colon: splenic flexure: descending colon: sigmoid colon: rectum=34: 72: 8: 87: 8: 26: 16: 7 (the overlapping cases were included in every count). Thus, lesions can develop at any site in the large bowel, but occurrence in the transverse colon and ascending colon tended to be comparatively frequent, and many of them were single lesions.

3. Tumor size

The tumor size was identified in 210 cases, with the maximal diameter ranging from 0.5 to 23.0 cm (mean: 2.8±2.1 cm), showing no particularly close correlation between the maximal diameter of the tumor and age ($r = -0.096$, **Fig. 4**). The lesion site was not significantly correlated with the maximal diameter of the tumor, either.

4. Subjective symptoms and complications

Symptoms of abdominal pain, bloody stool, and diarrhea were observed and our patient often complained of constipation with lower abdominal discomfort, but there were no characteristic symptoms of this disease. All patients demonstrating intussusception[4], massive bleeding[5] or protein-losing enteropathy[6] had larger tumors, while many patients with smaller tumors appeared to be asymtomatic. There have been no reported cases in which this tumor underwent malignant transformation, but colonic cancer accompanied it in some cases of lymphangioma[7].

5. Diagnosis and pathologic findings

For diagnosis of this disease, barium enema, colonoscopy, endoscopic ultrasonography are useful[8]. Endoscopic findings are characteristic, showing a translucent, rather bluish torous lesion with a smooth surface and a pedunculated or broad base. Cysts are multilocular, with septa being reflected on the surface in some cases. Concerning colonoscopic findings, Takahashi et al.[9] described how the tumor was characterized by (1) a steep rising margin and a somewhat narrow base, (2) the presence of the properties of submucosal tumors covered with normal colon mucosa, (3) a smoother, more glossy, and more translucent surface than that of adenoma, and (4) soft lesions that changed in shape on postural alterations or compression. The lesion is soft when pushed with a forceps, and shows a so-called positive cushion sign. Our case had all of these features. Cysts are filled with serous fluid, and drainage is sometimes observed on biopsy, which is helpful for diagnosis. In general, a diagnosis of lymphangioma is possible by means of careful colonoscopic examination.

Histologically, lymphangioma of the intestine is usually located in the submucosa with the overlying mucosa remaining intact, and this tumor is classified into

Table 2 Treatment for colorectal lymphangioma reported in Japan

Year	Procedure					Total
	Colectomy	Local excision	Endoscopic resection	Others	Unknown	
1965～1975	4	0	0	0	0	4
1976～1980	5	4	2	1	1	13
1981～1985	15	1	6	0	7	29
1986～1990	38	8	44	6	13	109
1991～1995	22	4	27	4	3	60
1996～2000	7	0	25	4	28	64
Total	91	17	104	15	52	279

(1) simple, (2) cavernous, and (3) cystic types in general[10]. Of the 279 subjects in the present study, the records of 148 patients were available and examined. Cystic tumor was noted in 104 cases, cavernous tumor in 43 cases, simple tumor in one case. Thus, cystic lymphangioma comprised the majority.

6. Treatment and prognosis

Although laparotomy was the main form of treatment for patients with this disease, endoscopic treatment has become increasingly indicated recently (**Table 2**). Because this is a benign disease, endoscopic resection is sufficient, and excessive invasive treatment should be avoided. The indications for endoscopic polypectomy are pedunculated or subpedunculated tumors measuring 2 cm or smaller in the maximal diameter. However, there are no problems in many cases if tumors even larger than these are left without complete resection, but with only segmental or partial resection, or puncture aspiration[11]. Because the lesion in this patient was less than 2 cm, we elected to resect the entire tumor endoscopically.

In the present study, we reported a patient with colonic lymphangioma, and reviewed 279 cases reported in Japan. Although there have been no reported cases in which this tumor underwent malignant change, the lesion rapidly grew, intussusception, ileus, or massive bleeding developed in some patients. Hence, this disease should not be ignored, and is considered to be a good indication for endoscopic treatment. If there is no complication, endoscopic treatment (resection, suction or incision) seems to be the preferable procedure for lymphangioma of the large intestine.

References

1. Enzinger FM, Weiss SW: Tumors of lymph vessels. "Soft Tissue Tumors. 3 rd ed." 1994; pp 679–700, Mosby, St. Louis.
2. Chisholm AJ, Hillkowitz P: Lymphangioma of the rectum. Am J Surg 1932; 178: 281–282.
3. Yoshitoshi Y, Oda T, Utsumi Y, Kaneko E, Yamashita K, Nakamura K: A case of lymphangioma of the ascending colon (in Japanese). Jpn J Clin Med 1965; 23: 2264–2267.
4. Matsui A, Okajima K, Ishii M, Namio H, Kawanishi M, Fujii Y, Arakaki Y, Toyoda H: Lymphangioma of the cecum with intussusception. J Jpn Surg Assoc 1986; 47: 234–239.
5. Sugiura H, Katoh H, Inomata H, Maeda Y, Saitoh M, Tanabe T: A case of bleeding lymphangioma of the large intestine. J Surg Hokkaido 1990; 35: 123–126.
6. Zilko PJ, Laurence BH, Sheiner H, Pollard J: Cystic lymphangioma of the colon causing protein-losing enteropathy. Am J Dig Dis 1975; 20: 1076–1080.
7. Morozumi K, Omoto T, Miyazaki H, Shibata S, Narai S, Yamauchi T, Kodaira K: Two cases of lymphangioma of the colon accompanied with colon cancer. Gastroenterol Endosc 1999; 41: 1323–1329.
8. Sato K, Maekawa T, Yabuki K, Tomita N, Eguchi M, Matsumoto M, Sugiyama N: Cystic lymphangioma of the colon. J Gastroenterol 1999; 34: 520–524.
9. Takahashi H, Ishida H, Togo S, Anazawa S, Sakurai K: A case of lymphangioma of the transverse colon treated with endoscopic polypectomy. Gastroenterol Endosc 1984; 26: 70–74.
10. Waston WL, McCarthy WD: Blood and vessel tumors. Surg Gynec & Obst 1940; 71: 569–588.
11. Matsuda T, Okihama Y, Kojima T, Hiroi M, Onda M: A case study of colonic lymphangioma treated by endoscopic resection and a review of Japanese 234 cases. Prog Dig Endosc 1999; 55: 92–93.

(Received, December 28, 2000)
(Accepted, January 11, 2001)