Int Surg 2021;**105:**784–787
DOI: 10.9738/INTSURG-D-20-00018.1

Case Report

Laparoscopic Resection for Lymphangioma Originating From Small Intestine: A Case Report

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Background: Lymphangioma originating from the small intestinal wall is a very rare finding. There have been only 15 case reports on the disease. Here, we report a case of lymphangioma in the small intestine that was resected laparoscopically. We also provide a literature review.

Case Presentation: A 37-year-old man came to our hospital with a chief complaint of abdominal pain. Physical examination showed mild tenderness in the lower abdomen, but rebound tenderness or guarding was absent. A contrast-enhanced computed tomography scan showed local poor enhancement in distal ileum, suggesting the possibility of mucous necrosis. Conservative therapy was started. On the eighth hospital day, although his abdominal symptoms improved, a follow-up computed tomography scan showed no improvement in the distal ileum lesion. Laparoscopic tumor resection was performed. The patient's postoperative course was unremarkable, and he was discharged on postoperative day 7. The pathologic result of the surgical specimen was lymphangioma of the ileum. The patient is still alive with no sign of recurrence 19 months after surgery.

Conclusion: Lymphangioma originating in the small intestinal wall is a very rare disease. Laparoscopic resection seems to be a preferable method for surgery.

Key words: Laparoscopy – Lymphangioma – Small intestine

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Lymphangioma (LA) is more commonly seen in children than in adults,[1] and it is usually found in the head, neck, and axillary region. The reports on LA in the small intestine often describe LA originating from small intestinal mesentery, whose frequency is reported to be less than 1% of LA.[2] Moreover, the finding of LA originating from small intestinal wall is even rarer, and its frequency is unknown, with only 15 case reports available. Despite its benign nature, LA sometimes requires surgical resection when it causes severe symptoms that must be corrected. Herein we report a case of LA of the ileum that was surgically resected.

Case Presentation

A 37-year-old Japanese man came to our hospital with the chief complaint of abdominal pain and a high fever of 38.3°C. Physical examination showed a flat abdomen with mild tenderness in the lower abdomen, but rebound tenderness and guarding were absent. Laboratory data showed an exacerbated inflammatory response (leukocyte: 18,000/μL; C-reactive protein: 1.02 mg/dL) and elevated lactate dehydrogenase level (273 U/L). Contrast-enhanced computed tomography (CE-CT) scan showed a thickened edematous wall of distal ileum with poor enhancement of mucous membrane (Fig. 1), suggesting ileitis with mucous necrosis. Both the superior mesenteric artery and superior mesenteric vein were enhanced as well. Magnetic resonance imaging (MRI) was performed, but it yielded no additional diagnostic clues. Differential diagnosis included infectious ileitis, ectopic pancreas, gastrointestinal stromal tumor, and small intestinal carcinoma, but a series of investigations did not help us reach a diagnosis. Parasitic enteritis including Anisakis and Spiruroid nematodes were unlikely from his food history. Although a definite diagnosis was not made from a series of examinations, conservative therapy with abstinence from food was started.

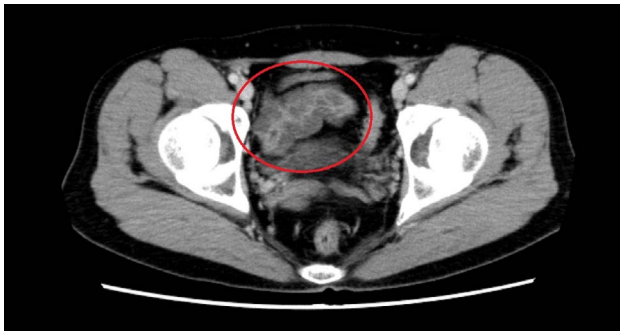

Fig. 1 CE-CT scan at presentation showed a thickened edematous wall of distal ileum with poor enhancement of the mucous membrane (a red circle).

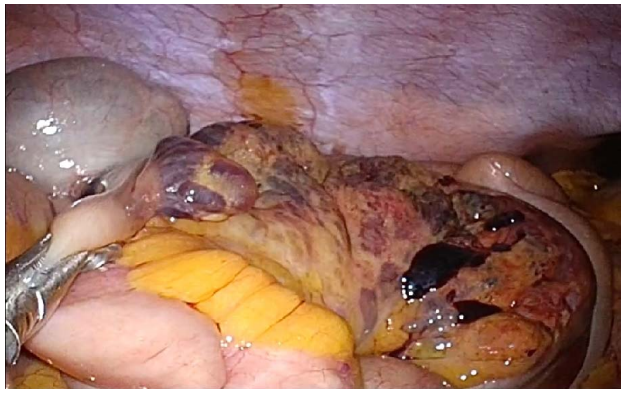

Fig. 2 An intraoperative picture through laparoscopy revealed edematous, discolored ileum located from 100 to 120 cm from the terminal ileum.

His high fever and inflammatory response gradually improved with time, but a follow-up CE-CT scan on hospital day 8 showed no improvement in the ileum lesion. The mucous membrane of the affected ileum was still poorly enhanced, suggesting necrotic change. Because enteritis showed no improvement despite an 8-day duration of abstinence, and because stricture of ileum was likely to occur even if the enteritis was treated with conservative therapy, we decided to perform surgery. Laparoscopic partial enterectomy was performed. The ileum lesion was located from 100 to 120 cm from the terminal ileum, and it looked like a tumor with many small black cysts and small hematomas (Fig. 2). The tumor was soft and showed no sign of malignancy. No adhesion, invasion, or ascites were present in the abdomen. The tumor was resected, and the small intestine was anastomosed with a functional end-to-end procedure. The operation time was 116 minutes, and the bleeding amount was 10 mL. The postoperative course was unremarkable, and he was discharged on postoperative day 8.

The pathologic result of the surgical specimen was LA of the ileum wall. Macroscopically, the tumor was 9.5 × 3.5 cm in size and consisted of yellow polyps (Fig. 3). Microscopically, the tumor was a collection of lumens with serous content. Immunostaining was positive with D2-40 and CD31.

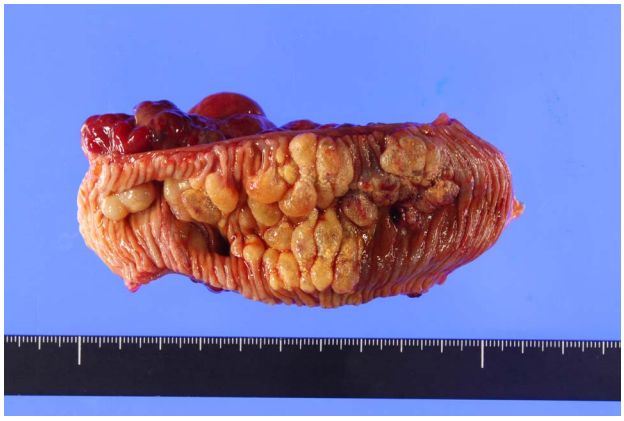

Fig. 3 Surgical specimen of the tumor consisted of yellow polyps.

The patient is still alive with no sign of recurrence 19 months after surgery.

Discussion

LA is one of the least frequently encountered tumors originating from small intestine. LA in the small intestine is reported to account for approximately 1.4% to 2.4 % of all small intestinal tumors by Worapop *et al*,[2] but considering that LA originating from the small intestinal mesentery is included in their report, LA originating from the small intestinal wall is an extremely rare finding. As far as we searched in PubMed, there has been only 16 reports,[3–16] including our case on lymphangiomas in the small intestinal wall (Table 1).

LA is a benign tumor, so basically resection of the tumor is not necessary. However, surgery is often required, especially when clinical symptoms caused by the small intestinal tumor are so severe that surgical resection is the only way to relieve them, or when preoperative diagnosis is unavailable and therefore the possibility of the tumor being malignant cannot be excluded. Symptomatic LA in the small intestine sometimes causes bleeding that results into anemia,[6] melena,[10] or positive fecal occult blood test,[5] whereas it sometimes causes digestive tract symptoms including intestinal obstruction[1] or volvulus.[8] Of the 16 cases in Table 1, 13 cases underwent surgery, all of which were resected without preoperative diagnosis of LA. Only 2 cases (cases 6[7] and 13[14]) were left unresected, because in

Table 1 Cases of lymphangioma originating itself from the small intestinal wall

Case no.	First author	Year	Age	Sex	Main symptom	Procedure (finding)	Location	Intervention
1[3]	Walker-Smith	1969	5 wk	Female	Diarrhea	Autopsy	Jejunum and ileum	None
2[1]	Hanagiri	1992	53 yr	Male	Abdominal pain, nausea	Intestinal tube (obstruction)	Ileum (250 cm from Treitz ligament)	Surgery (open)
3[4]	Barquist	1997	33 yr	Female	Dyspnea	Enteroclysis	Middle jejunum	Surgery (Lap)
4[5]	Honda	2003	31 yr	Female	Positive fob	CT	Jejunum (60 cm from Treitz ligament)	Surgery (not available)
5[6]	Hsu	2007	75 yr	Female	Tarry stool	DBE	Jejunum (200 cm from ileocecal valve)	Surgery (Lap)
6[7]	Li	2009	69 yr	Male	Melena	DBE	210 cm from front teeth	Endoscopic hemostasis
7[8]	Day	2010	10 yr	Female	Abdominal pain, vomit	CT (volvulus)	Jejunum	Surgery (open)
8[9]	Morris-Stiff	2011	34 yr	Female	Dyspnea	CapE, CT	Proximal jejunum	Surgery (open)
9[10]	Fang	2012	57 yr	Female	Melena	Endoscopy	Jejunum (30 cm from Treitz ligament)	Surgery (not available)
10[11]	Al-Obeed	2014	56 yr	Male	Epigastic pain	CT	Ileocecal valve	Surgery (Lap)
11[12]	Bucciero	2015	28 yr	Male	Melena	Endoscopy, CT	Jejunum	Surgery (not available)
12[13]	Iwaya	2018	70 yr	Male	Tarry stool	DBE	Jejunum (120 cm from Treitz ligament)	Surgery (Lap)
13[14]	Costa	2019	56 yr	Female	None	DBE	Proximal jejunum	Biopsy
14[15]	Giuliani	2019	41 yr	Male	Abdominal pain	CT (perforation)	Ileum (80 cm from ileocecal valve)	Surgery (open)
15[16]	Teng	2020	55 yr	Female	RUQ abdominal discomfort	CT, DBE	Jejunum (60 cm from Treitz ligament)	Surgery (open)
Our case	Imamura	2020	37 yr	Male	Abdominal pain	CT	Ileum (100 cm from ileocecal valve)	Surgery (Lap)

CapE, capsule endoscopy; DBE, double-balloon endoscopy; FOB, fecal occult blood; Lap: laparoscopy; RUQ, right upper quadrant.

case 6, the patient was asymptomatic, and in case 13, hemostasis was successfully achieved with an endoscopic procedure instead of surgical resection. The location of tumors is also described in Table 1. In 10 of 16 cases (62.5%), LA was found in the jejunum, whereas in 4 cases (25%), LA was found in the ileum.

Lymphangiomas are classified into 3 types based on histologic findings: capillary (simple), cavernous, and cystic.[2] Diagnosis of LA in the small intestine is made histologically, but it is actually difficult to make a correct diagnosis because the small intestine is where endoscopy is least likely to reach a tumor. Radiologic examinations such as CT or MRI may provide us some information about LA. Al-obeed and Abdulla[11] reported that LA appears on CT scans as homogenous, nonenhancing lesions with variable attenuation values depending on whether the fluid is chylous or serous, which was also true of our patient. However, radiologic examinations still cannot be informative enough to exclude the possibility of malignancy. This is why surgical resection is sometimes necessary without making a preoperative diagnosis. Li *et al*[7] reported that endoscopic resection was a possible option for LA in the small intestine, but in our case, where necrotic change of intestinal wall was suspected, endoscopic procedure would have possibly caused perforation. That was why we chose surgical resection for our patient instead of an endoscopic procedure. Of the 13 cases in Table 1 that required surgery, 5 cases underwent open laparotomy and 5 underwent laparoscopic surgery. In the remaining 3 cases, detailed information about the surgical procedure was not available. Considering that the small intestine is a highly motile organ and that LA is less likely to cause adhesion or invasion because of its benign nature, laparoscopy seems to be a better procedure for resection than laparotomy. Of the 13 surgery cases, none refer to surgical difficulties such as adhesion or invasion. In our case, laparoscopy was an optimal choice because surgery was completed less invasively. Because laparoscopic surgery is now widespread, reports on laparoscopic resection for LA in the small intestine will increase in number.

Acknowledgments

The authors have no conflicts of interest to declare related to this manuscript. This manuscript does not require any ethical approval. The authors obtained a permission for publication from the patient.

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