Laparoscopic surgery of intra-abdominal lymphatic malformation in children

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Abstract. Lymphatic malformation (LM) in the abdomen is uncommon in children, and the standard treatment is surgical excision. The present study aimed to evaluate the safety and efficacy of laparoscopic surgery of intra-abdominal LM in a pediatric population. The medical records of 10 children with intra-abdominal LM treated by laparoscopic resection from March 2017 to June 2021 in The Second Affiliated Hospital of Xi'an Jiaotong University (Xi'an, China) were retrospectively reviewed. Equal numbers of female and male patients were included and underwent surgery at the median age of 55 months (range, 40 days-94 months). Abdominal pain was the most frequent symptom presenting in eight patients (80%). All children were referral patients, and half of them presented to the emergency department with acute abdominal pain. All 10 of the patients benefited from laparoscopic treatment; three patients underwent extraperitoneal segmental intestinal resection and anastomosis via an enlarged umbilical incision, and two patients were converted to laparotomy because of a large retroperitoneal LM with involvement of the colon. No recurrence or complications occurred in the patients during the follow-up period with mean of 35 months (range, 11-60 months) after surgery. Overall, laparoscopic resection of intra-abdominal LM was an effective, minimally invasive therapy in the pediatric population. Segmental intestinal resection is usually required to achieve the complete removal of lesions to lessen the risk for recurrence.

Introduction

Lymphatic malformation (LM) is one of the slow-flow vascular anomalies caused by abnormal embryologic development of the lymphatic system. LM manifests before the age of 2 years in >90% of patients, and has been called 'lymphangioma' or 'cystic hydroma' in the past (1-3). It most commonly occurs in the neck, followed by the trunk and extremities. However, intra-abdominal LM is rare, only accounting for 5-10% of cases (4-6). Abdominal pain and other gastrointestinal symptoms such as abdominal distension, vomiting and constipation are present in children with abdominal LM (4,6,7), which is different from the painless LM mass typical in the more common sites (2,8). LMs are classified by size as macrocystic, microcystic or mixed patterns, which is of therapeutic importance (2). In the abdomen, the majority of LM are classified as a macrocystic lesion (6-8), arising from mesentery, retroperitoneum or solid organs (4,8,9).

Historically, surgical resection has been the standard treatment regardless of location (1,2,7). Percutaneous sclerotherapy as a minimally invasive method by image-guidance has been preferred treatment applied for LM with good outcomes over the past years (1,2,6-8). However, surgical resection for intra-abdominal LM, especially with laparoscopy, has some advantages. In The Second Affiliated Hospital Of Xi'an Jiaotong University (Xi'an, China), laparoscopic surgery has been widely used in the pediatric population, and so the present retrospective analysis of intra-abdominal LM treated with laparoscopy was undertaken to provide pediatric surgeons with more experience in diagnosis and treatment of this lesion.

Materials and methods

Study population. A total of 13 patients with intra-abdominal LM were collected retrospectively from electronic medical records of The Second Affiliated Hospital Of Xi'an Jiaotong University (Xi'an, China) of patients from March 2017 to June 2021. Overall, 10 children were included in the study; one child who was simply monitored and two children who underwent sclerotherapy were excluded. Clinical data were reviewed including present and past history of illness, demographic characteristics, imaging choice for diagnosis, operative data and postoperative follow-up. Written informed consent was obtained from parents of children. The study was approved by the Ethics Committee of The Second Affiliated Hospital Of Xi'an Jiaotong University (approval number: 2022205).

Laparoscopic technique. Laparoscopy was performed with patients under general anesthesia in the supine position. At

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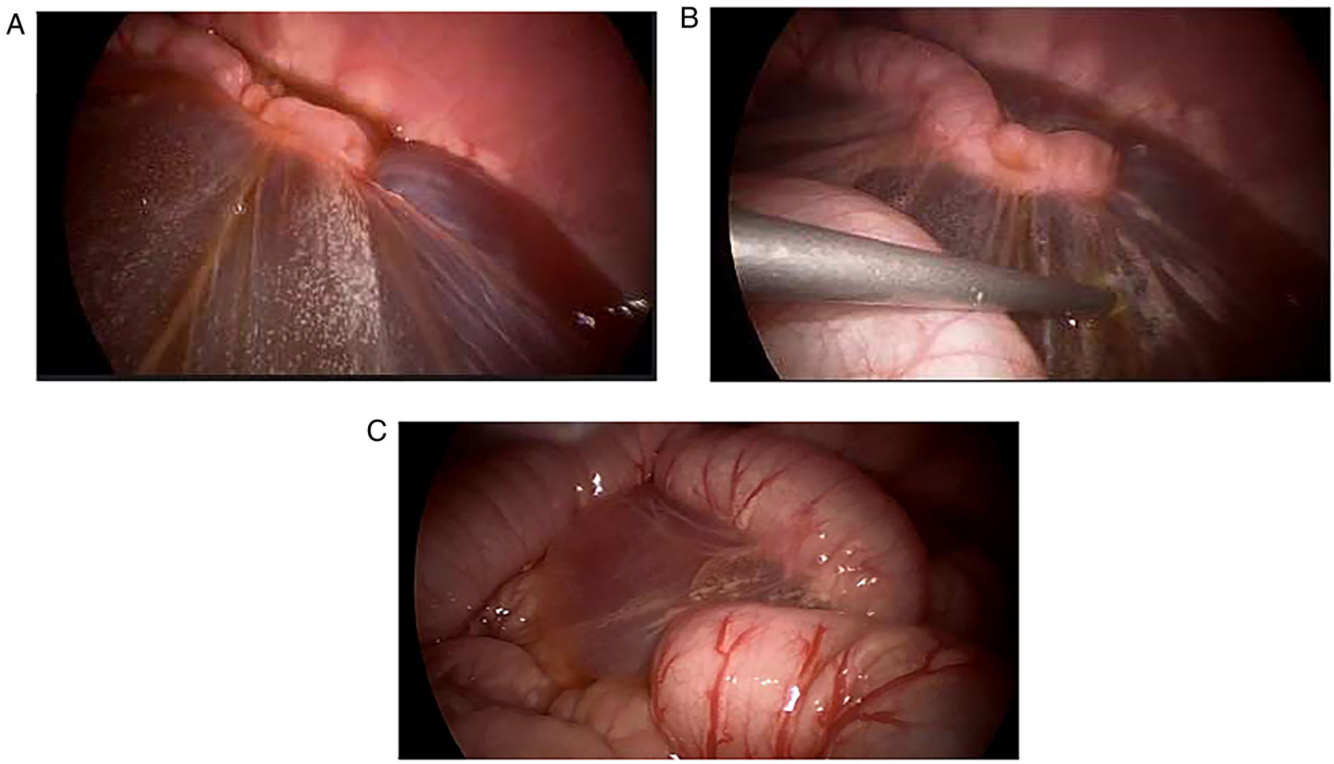

Figure 1. Intraoperative findings of intra-abdominal lymphatic malformation. (A) The appearance of intra-abdominal lymphatic malformation and intestine surrounded before suction. (B) Suctioning by suction apparatus. (C) Condition after suction.

first, a 5-mm umbilical trocar was inserted with the open method (10). Pneumoperitoneum was established and maintained at 8-12 mmHg by CO_2 gas according to the child's age and weight. Subsequently, the next two 5-mm (or 3-mm) trocars were inserted according to lesion location.

After confirmation of the origin and involvement of the lymphatic cysts, the surgeon decided how to finish the resection. LMs can contain huge cysts that block sight and be difficult to extract via the small incisions created by the trocars. Therefore, large cysts were punctured with Veress needle guided by laparoscopy and lymphatic fluid was suctioned clear (Fig. 1) to extracorporeal container. The lymphatic cysts, mesentery or adjacent tissues were then resected. Complete resection of the LM and intestinal anastomosis after partial resection of the small intestine were performed by exteriorization of the small bowel via the enlarged umbilical trocar port. Nearly complete resection was made in instances of severe adhesion to important tissues or organs, and the cyst wall was cauterized by electrocoagulation.

Results

Clinical characteristics of patients. Overall, 10 patients with intra-abdominal LM underwent laparoscopic surgery, and their characteristics are presented in Table I. The male to female ratio was 1:1, and the average age at operation was 55 months (range, 1-94 months). All of them were referral patients with 20% (2/10) diagnosed with LM, half of whom presented to the emergency department because of acute abdomen. Subsequently, one patient was diagnosed on prenatal ultrasound, one infant presented with restlessness and the other eight patients suffered from abdominal pain accompanied by vomiting or fever. The referral diagnosis was obtained from the medical records or description by the parents. The interval from primary diagnosis to surgery ranged from 5 days to 21 months (median, 24 days).

Ultrasound as the first choice was performed in all of the patients to identify the lesion. Computed tomography (Fig. 2) and magnetic resonance imaging were performed for further information in four and five cases, respectively. Overall, 40% (4/10) of the patients were diagnosed with LM preoperatively without any biopsies; three mesenteric cysts, two intra-abdominal cysts and one intestinal duplication were suspected in the remaining six patients preoperatively. By imaging, LM size ranged from 2.8-13.8 cm (mean, 9.5 cm).

Surgical outcome. The final diagnosis was confirmed by intraoperative findings and postoperative histopathology. As presented in Table II, LMs were located in the mesentery in 70% (7/10) patients, other locations included retroperitoneum and greater omentum. Macrocystic LM was diagnosed in nine patients by intraoperative and imaging findings after final diagnosis.

Laparoscopic resection of LMs was completed in eight patients; however, two patients were converted to laparotomy because of the retroperitoneal origin of huge LMs that involved the colon. In three of the eight patients who underwent complete laparoscopic resection, an assisting and enlarged incision via umbilical trocar site to finish the extraperitoneal intestinal anastomosis. Incomplete excision of the LM was performed in patient 7 because of severe adhesion to the inferior vena cava. The residual cyst wall was

Table I. Patient characteristics and preoperative assessment.

Patient no.	Sex	Age, months	Weight, kg	Emergency experience	Referral diagnosis	Symptoms	Duration, days	Preoperative imaging	Size, cm
1	Male	37	16.5	Yes	Peritoneal effusion	Abdominal pain, fever	7	US + CT	12
2	Male	77	19	Yes	Lymphangioma	Abdominal pain	639	US + CT	7.8
3	Male	54	18	Yes	Intestinal duplication	Abdominal pain, vomiting	9	US	2.8
4	Female	94	20	No	Omental cyst	Abdominal pain	93	US + MRI	4.6
5	Female	53	16	Yes	Mesenteric cyst	Abdominal pain, fever	5	US + CT	9.9
6	Male	1	5	No	Lymphatic malformation	Prenatal examination	45	US + MRI	10
7	Female	74	19	No	Giant mass	Abdominal pain	36	US + MRI	12.7
8	Female	78	24	Yes	Intra-abdominal cyst	Abdominal pain, vomiting	29	US + MRI	11.8
9	Female	73	21	No	Intra-abdominal cyst	Abdominal pain	19	US + CT	9.6
10	Male	9	10	No	Intra-abdominal cyst	Restless	6	US + MRI	13.8

CT, computed tomography; MRI, magnetic resonance imaging; US, ultrasound.

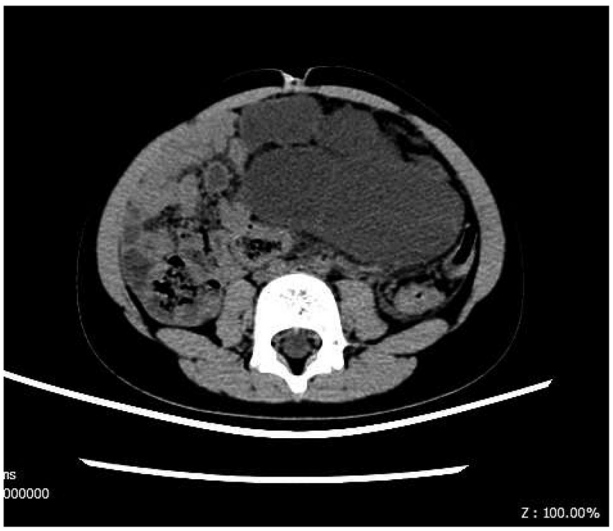

Figure 2. Computed tomography imaging of intra-abdominal lymphatic malformation.

cauterized carefully to avoid recurrence. Complete resection was performed in the other nine patients. The mean duration of operative time was 106 min (range, 65-165 min) in eight patients with complete laparoscopic resection. Total blood loss during the operation was very small. Intracystic hemorrhage and infection was confirmed by intraoperative findings and histopathology in 60% (6/10) patients. No complications or mortality occurred after surgery. The mean follow-up period was 35 months (range, 11-61 months). Patients were asymptomatic and no recurrence was identified by ultrasound from 1 to 12 months after surgery except for missing one.

Discussion

Intra-abdominal LM is rare in children and challenging for surgeons to differentiate from other cystic lesions (8). Surgical excision is indicated for patients with LM who have complications such as infection or hemorrhage (2). Laparoscopic surgery is a minimally invasive method for treating intra-abdominal LM, and the rapid development of laparoscopic technique has benefitted patients with LM (11). Because of less intraoperative blood loss, postoperative pain and scarring, laparoscopic surgery has more advantages compared with conventional open surgery (11).

The majority of LMs are noted in the first few years of life, and grow proportionately with the growth of children (2). Intra-abdominal LMs are usually diagnosed with acute gastrointestinal symptoms because of complications of large LMs at the mean age of 5 years (4), which is similar to the results of the present study. Female patients outnumbered male in some previous studies (4,6); male predominance was reported in other studies (7,12,13), but there was no sex predilection in the present cases, which may be because of the small number of patients included.

The majority of patients of intra-abdominal LM are reported to have acute gastrointestinal symptoms (4,6,13), which differed from the majority of LMs presenting as a painless mass (1,2,8) diagnosed clinically and radiologically (7,14). Although varied symptoms were present in 80-96% of patients, abdominal pain was the most common symptom of intra-abdominal LMs (4,6,7,12) as in the present study. Macrocystic LMs were the most common pattern of intra-abdominal LMs although combined and microcystic were also identified in the abdomen (4,6,7,9,13). Except for symptoms from mass effect and volvulus, complications

Table II. Intraoperative and postoperative data.

Patient no.	Location	Type	Laparoscopic approach	Mode of operation	Operative duration, min	Intraoperative bleeding, ml	Final diagnosis	Follow-up, months
1	Greater omentum	Macrocystic	Yes	Lesion with partial greater omentum	165	80	Omental LM with hemorrhage	60
2	Mesocolon	Macrocystic	Yes	Lesion with segmental colon resection	135	<5	Mesenteric LM with infection	Missing visit
3	Mesoileum	Mixed	Yes	Lesion excision only	85	<5	Mesenteric LM with hemorrhage	50
4	Mesocolon	Macrocystic	Yes	Lesion excision only	65	<5	Mesenteric LM	46
5	Mesocolon	Macrocystic	Yes	Lesion excision only	80	<5	Mesenteric LM with infection	36
6	Mesocolon	Macrocystic	Yes	Lesion excision only	95	<5	Mesenteric LM	17
7	Retroperitoneum	Macrocystic	Converting to laparotomy	Lesion with segmental colon resection	310	20	Retroperitoneal LM with hemorrhage	11
8	Mesoileum	Macrocystic	Yes	Lesion with segmental ileum resection	100	<5	Mesenteric LM	48
9	Jejunum-ileum mesentery	Macrocystic	Yes	Lesion with segmental jejunum-ileum resection	120	<5	Mesenteric LM	30
10	Retroperitoneum	Macrocystic	Converting to laparotomy	Lesion with segmental colon to ileum resection	260	10	Retroperitoneal LM with infection	19

LM, lymphatic malformation.

including intracystic hemorrhage and infection occurred in 60% of the present patients and may be the main cause of acute abdomen (3). Therefore, the diagnosis was challenging preoperatively and misdiagnosis occurred in ~1/3 of patients with intra-abdominal LM (4).

On imaging, cystic lesions such as teratoma, enteric duplication cyst and ovarian cyst can be confused with intra-abdominal LMs (8). To avoid radiation, ultrasound is the first choice to evaluate intra-abdominal LMs in children, which typically shows multilocular cystic masses. Further evaluation with cross-sectional imaging provides additional information on diagnosis and location of the lesion. However, although imaging findings play an important role in the diagnosis of intra-abdominal LMs, there is no highly specific radiological presentation that makes the definitive diagnosis (4,8).

Percutaneous sclerotherapy by image-guidance has been preferred as primary treatment modality for LM over the past years (1,2,15). However, repeated sclerotherapy with residual lesions (6,7,13) cannot replace surgery as the first treatment modality because of frequent recurrence (3,16-18). The final diagnosis was usually corrected by intraoperative findings and the histopathology of lesions (8,19), which makes resection important (4,11,12,16,17,20). Surgical resection is a potential curative procedure for LM (19), and the laparoscopic approach

can be performed to find the location and dissect primarily without large scars (4,11).

In fact, it is also challenging to remove intra-abdominal LMs completely because of the involvement of neighboring structures such as blood vessels and the alimentary tract (4,17), and recurrence may result after incomplete resection (3). Therefore, segmental resection of the intestine is usually necessary to achieve complete the resection of and decrease the potential recurrence of LMs (3,4,20). A total of five patients in the present study underwent intestinal anastomosis after segmental intestinal resection, which was common (84% of patients) in a previous study (4) and in case reports (11,16-18). In some circumstances, LM cannot be resected completely because of the involvement of vital structures, and percutaneous sclerotherapy may be suitable for recurrence after surgical resection (6).

Ileus and recurrence are the frequent postoperative complications of surgical resection (3,4). With the exception of residual lesion in one patient who underwent incomplete resection, there were no instances of recurrence or ileus in our patients during the mean follow-up period of 35 months. Conversion to laparotomy occurred in patients from the current study who had large retroperitoneal LM, suggesting that these larger-volume lesions had more involvement of adjacent organs and tissues (9). It is worthwhile to perform laparoscopic exploration to confirm the origin and extent of the lesion as well as the possibility of laparoscopic resection. Furthermore, the classification of mesenteric LM by radiological information would give surgeons guidance in preoperative planning (4).

Compared with adults, pediatric laparoscopic surgery of intra-abdominal LM is challenging because of the smaller space in children. Puncturation and aspiration guided by laparoscopy is recommended to minimize the lesion and to provide more space for laparoscopic resection because of benign cysts (3). After the removal of lesions, segmental intestinal resection and anastomosis of can be finished by expanding the umbilical trocar incision in some cases. Complete resection can be performed by laparoscopy without relapse during the follow-up. This approach was proved to be effective for treating intra-abdominal LM in the present study.

To the best of our knowledge, this is the largest study of laparoscopic resection of intra-abdominal LM in the pediatric population. However, there were limitations to the current study, as in the majority of case series: Small number of patients, retrospective review and no controls because of the rarity of intra-abdominal LMs.

In summary, intra-abdominal LM in pediatric population of the present study was diagnosed by initial acute gastrointestinal symptoms, mainly onset of abdominal pain at the mean age of 5 years. Macrocytic LM was the most frequent type of intra-abdominal LM and was effectively resected laparoscopically. Complete resection was associated with a lower recurrence, which was usually accompanied by segmental intestinal resection.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

QL and JF participated in study design and data collection, carried out the initial analysis and drafted the article. QY and WG analyzed and interpreted the patient data regarding the technique and outcome of laparoscopic surgery, and revised the manuscript. PL and XG conceptualized and designed the study, and performed critical revisions of the article. QL and JF confirm the authenticity of all the raw data. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Written informed consent was obtained from parents of children. The study was approved by the Ethics Committee of The Second Affiliated Hospital Of Xi'an Jiaotong University (approval no. 2022205).

Patient consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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