

Laparoscopic Management of retroperitoneal cystic lymphangiomas in children: a systematic review and an additional case report

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Table of Contents

1. Introduction	2
2. Case presentation	2
2.1. Technical details	3
3. Systematic review	4
4. Discussion	5
5. References	6

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Abstract. AIM: Retroperitoneal cystic lymphangiomas (RCLs) are rare vascular malformations which account for approximately 1% of all lymphangiomas in infancy. Surgical treatment of RCLs may be particularly challenging, due to their possible connection with vital structures. Laparoscopic excision has been recently described for RCLs also in pediatric patients, with only 14 published cases. METHODS: Authors provide a detailed systematic review of the pediatric literature according to PRISMA guidelines, focusing on laparoscopic treatment of RCLs in children. An additional case of a large RCL in a 4-year-old girl is also presented. RESULTS: The PubMed search yielded six relevant studies, encompassing a total of 14 children with RCLs; analysis was extended to our additional patient. Mean age at surgery was 6.6 years. Laparoscopic aspiration of the cysts before their removal was the common approach, used in 9 patients (64.3%). Conversion to open surgery was necessary in 3 cases (21.4%). Recurrence of the disease was reported only in one patient (7%). No mortality was reported. CONCLUSIONS: According to the limited available experience, accurate preoperative assessment and adequate training and skills make the laparoscopic resection of retroperitoneal lymphangioma a safe and feasible procedure, including within a pediatric context.

Keywords. Cystic lymphangioma; children; ultrasound; retroperitoneum; laparoscopy

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1. Introduction

Retroperitoneal cystic lymphangiomas (RCLs) are rare vascular, lymphatic malformations usually occurring during infancy (1). Since spontaneous resolution is uncommon, operative treatment is usually indicated (1-2). RCLs surgical excision with a laparotomy approach has been always considered the first therapeutic choice. Because of their anatomical location and relationship with vital organs and blood vessels, the sclerotherapy is considered as an alternative treatment in unresectable lesions (1-3). Laparoscopic treatment for RCLs has been recently reported in pediatric literature but only in few cases. In this study, we report our experience with a 4-year-old girl with a large RCL managed with primary laparoscopic excision, and provide a detailed systematic review of laparoscopically treated RCLs in childhood.

2. Case presentation

A 4-year-old otherwise healthy girl was admitted to our Department because of mild abdominal pain associated with fever and non-bilious vomiting. Physical examination on admission revealed tenderness with guarding in the left abdominal quadrants. Laboratory tests showed raised levels of C-reactive-protein and leukocytosis. Tumor markers (carcinoembryonic antigen, alpha-fetoprotein, CA 19.9, CA 125) were negative. Ultrasonography (US) and computed tomography (CT) showed a large (8 x 5 x 4 cm) infected retroperitoneal cystic mass extending from the lower left kidney pole to the ureter intersection with iliac vessels, strictly adhering to the left ureter and spermatic vessels (Figure 1 A-B). Abdominal MRI confirmed the suspicion of infected cystic lymphangioma (Figure 1 C). Antibiotic therapy was started with rapid clinical improvement and normalization of blood inflammatory tests. The patient was discharged and elective laparoscopic excision was scheduled one month later. Full informant consent was obtained from parents before all stages of the procedure.

2.1. Technical details

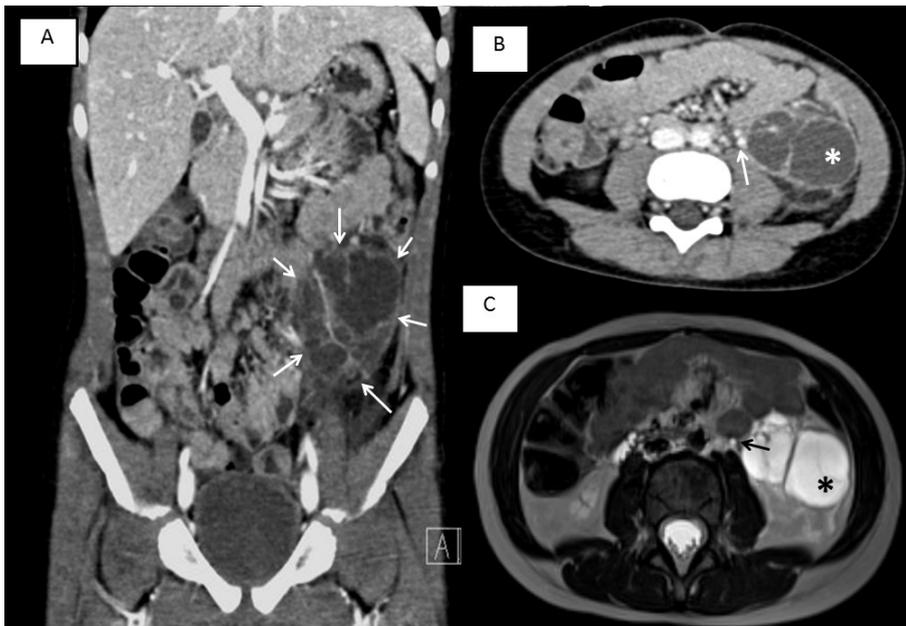


Fig. 1 A: Abdominal CT demonstrates a large (8 x 5 x 4 cm) retroperitoneal cystic mass (white arrows) extending from the lower left kidney pole to the ureter intersection with iliac vessels, tightly adhered to the left ureter. B: Relationship between the lymphangioma (asterisk) and spermatic vessels (arrow). C: T2-weighted MRI showing the close relationship between the mass (asterisk) and the left ureter (arrow).

At the time of surgery a double JJ catheter was inserted in the left ureter in order to facilitate the identification of the ureter itself. The patient was placed in 35° right lateral decubitus with a roll pad under the right back. An optical 10-mm Xcel trocar (Ethicon Endo-Surgery, Somerville, New Jersey) was inserted into the umbilicus using an open technique and pneumoperitoneum was created with CO₂ at a pressure of 10 mmHg and a flow of 1 L/min. Two additional 5-mm Xcel trocar were placed in the midline at the subxiphoid space and in the left iliac fossa, half-way between the umbilicus and the anterior-superior iliac spine. The left colon, together with the splenic flexure, was mobilized medially and the lower part of the mass exposed. The lienophrenic ligament was divided and Gerota fascia partially opened: tight adhesions between the cystic mass, the lower pole of the left kidney and the ureter were found and carefully divided with Ultracision (Ethicon Endo-Surgery) (Figure 2A-B). At the end of the dissection, the mass was completely removed and retrieved using an endobag. Pathological examination confirmed the diagnosis of cystic lymphangioma. The postoperative care was uneventful and the patient was discharged on the postoperative day two. At a 20-month follow-up she is doing well and abdominal US shows no recurrence of disease.

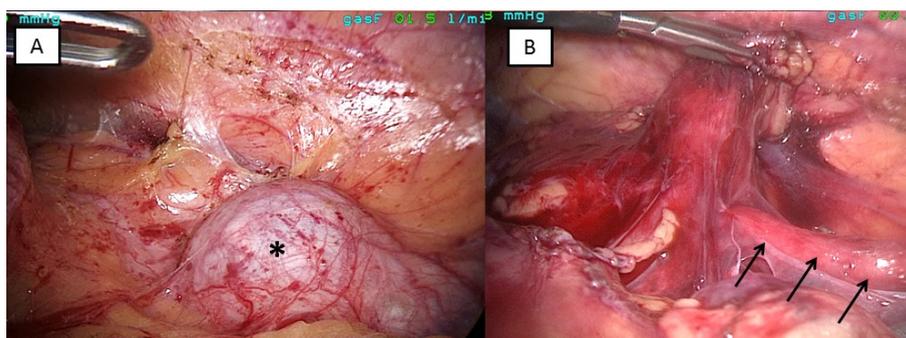


Fig. 2 A: Intraoperative picture showing the cystic mass (asterisk) after left colon mobilization. **B:** The mass strictly adheres to the left ureter (arrows).

3. Systematic review

For the review of the literature, the PubMed database (<http://www.ncbi.nlm.nih.gov/pubmed>) was searched for studies published since 2000 regarding the laparoscopic treatment of retroperitoneal lymphangiomas, in patients younger than 18 years old. The date of the last search was October 2015. The PubMed search yielded six relevant studies (5-10), encompassing a total of 14 children with RCLs (Table 1). All the studies were retrospective case reports or small case series (class of evidence III and rating scale of evidence E) (11). In two studies, additional data were included after personal communications with the Authors (6, 10). The analysis was extended with our additional patient. Patient mean and median age at presentation was 6.6 and 4.15 (SD 5.23, range 0-14) years, respectively. RCLs affect both sexes equally (5-10). When reported, abdominal pain was the commonest symptom at presentation. In terms of operation details, laparoscopic aspiration of the cysts before their removal was the common approach, used in 9 patients of this series. In one patient, as the mass was too large to be extracted via the umbilical port, it was sectioned and removed from the abdominal cavity through a mini-laparotomy (5). Conversion to open surgery was necessary in 3 cases (21.4%). One study did not report technical operation details (6). No other complications were reported. Recurrence of the disease was reported only in one patient (7%). No mortality was reported. Characteristics of the patients of the series were summarised in Table 1.

Table 1. Review of laparoscopic excision resection of retroperitoneal lymphangioma in childhood

Author	Patients (n)	Age	Clinical Presentation	Technical details	Complications	Outcome
Wildhaber BE et al, 2006	2	1.5 y	Chronic vomiting, abdominal pain	Aspiration of the cyst before excision with low-current electrocautery	None	No recurrence of disease at 18-month follow-up
		4 y	Acute abdominal pain	Division the cyst in portion with		

				low-current electrocautery; removal with endobag		
De Lagausie P et al, 2007 *	2	8 y, 14 y	Nor reported	Not reported	1 conversion to open surgery	No recurrence of disease
Singh R et al, 2009	1	15 y	Recurrent urinary infections, neurofibromatosis	Double J catheter; Aspiration of the cyst before excision; excision with SonoSurg	None	Not reported
Solari V et al, 2011	1	2 w	Antenatally identification of intraabdominal cystic mass	Aspiration of the cyst before excision	None	No recurrence of disease at 24-month follow-up
Padilla BE et al, 2011	1	9 y	Not reported	Magnet-assisted laparoscopy	None	Not reported
Son TM et al, 2012 *	7	4.3 (mean) y	Not reported	Aspiration of the cyst before excision	2 conversion to open surgery	1 recurrence
Our case, 2015	1	4 y	Acute abdominal pain, fever, vomiting	Excision with Ultracision; removal with endobag	None	No recurrence of disease at 20-month follow-up

Legend: * additional data derived from personal communication with Authors, y: years, w: weeks

4. Discussion

Lymphangiomas are vascular malformations arising from sequestration of lymphatic tissue that fails to communicate with the lymphatic system (1-2). They can be very wide-ranging, from localized masses, to areas of diffuse infiltration, to chylous fluid accumulations in various body cavities. RCLs are extremely rare, accounting for less than 1% of all lymphatic malformations (1). These malformations are usually diagnosed in infancy and are histologically classified as cavernous or cystic. Infections located elsewhere in the body

or viral illnesses can cause increased size and tension of abdominal LMs, leading to acute symptoms of abdominal pain, tenderness, fever, leukocytosis, peritonitis and dysuria (5-6). In these cases, the diagnosis must differentiate it from other fluid-filled lesions as pseudocysts, dermoid cysts, cysts of urothelial and enteric origin, hydatid cyst or neoplasms like sarcomas, mesotheliomas, pancreatic tumours, teratomas. In addition, lymphangiomas should be differentiated from hemangioma with secondary hemorrhage (5-7). Well-localized and cystic RCLs are easily characterized by ultrasonography, while MRI is better at documenting the full extent of more complex malformation as well as their macrocystic and microcystic components (7). Increasing evidence suggests that an alternative treatment is the image-guided percutaneous catheter drainage followed by Sclerotherapy, especially for RCLs with macrocystic components (4, 12-15). This noninvasive approach sometimes requires multiple sessions based on response to therapy and recurrence of symptoms, and is been also adopted as a “rescue” treatment after partial surgical excision (3, 13). Other treatment options, like aspiration, drainage and irradiation have given poor results (3). Nevertheless, the effectiveness of sclerotherapy in microcystic disease is limited (15). Surgery with complete excision may be important for patients with a life-threatening problem such as compartmental syndrome, rupture, superinfection and bleeding, and also after failed sclerotherapy and those who have the microcystic disease (14-15).

The traditional treatment of choice for RCLs is complete primary excision, providing the definitive histological diagnosis and avoiding complications such as enlargement, superinfection, rupture or bleeding and recurrence (1-3). Even if conservative management in asymptomatic patients has been described (1), most of the studies of this series agree that the removal of the tumor as soon as possible is essential, either in order to provide definitive pathological diagnosis, to avoid the above mentioned complications or because growth of the mass might prevent complete removal at a later date (2,6-7, 10). Malignant degeneration to lymphangiosarcoma has been reported rarely in children and adolescents (1). RCLs in childhood have a high recurrence rate of up to 53%, mostly in cases of subtotal excision that generally occur in cases of large and complicated lesions (2-3).

In the last few years, laparoscopy has been described as the choice treatment for RCLs, especially in adults, with only 14 pediatric cases reported so far (1, 5-10). The advantages of laparoscopy compared to open surgery in regard of postoperative pain and adhesions, faster recovery and better cosmetic results are well known (8-10). Furthermore, laparoscopy may also be indicated as a better diagnostic tool for the characterization of the lesion and its relationship with the surrounding organs. If laparotomy or sclerotherapy are deemed necessary, they may be better targeted after a laparoscopic evaluation. The results of laparoscopic treatment of RCLs are good with a low complication rate (8). Infected lymphangiomas can be definitively reduced with a preoperative antibiotic therapy, allowing a safer excision. When a lymphangioma is tightly adhered to the ureter, a cystoscopic ureteral double JJ catheter inserted immediately before surgery could be very useful to avoid ureteral injuries during dissection (9).

Our limited experience suggests that when sclerotherapy is not indicated, laparoscopic resection is a safe and feasible procedure also in pediatric setting, including for large retroperitoneal lymphatic malformations.

5. References

1. Bhavsar T, Saeed-Vafa D, Harbison A, et al: Retroperitoneal cystic lymphangioma in an adult: a case report and review of the literature *World J Gastrointestinal Pathophysiol* 1(5): 171-6, 2010.

2. Steyaert H, Guitard J, Moscovivi J, et al: Abdominal cystic lymphangioma in children: benign lesion that can have a proliferative course. *J Pediatric Surg* 31: 677-80, 1996.
3. Nam SO, Kim DY, Kim SC, et al: The surgical experience for retroperitoneal, mesenteric and omental cyst in children. *J KorSurgSoc*; 83 (2): 102-6, 2012.
4. Guvenc B H, Ekingen G, Tuzlaci A, et al. Diffuse neonatal abdominal lymphangiomatosis: management by limited surgical excision and sclerotherapy *Pediatric SurgInt* 21: 595-8, 2005. .
5. Wildhaber BE, Chardot C, Le Coultre C, et al: Total laparoscopic excision of retroperitoneal cystic lymphangioma. *J LaparoscAdv Tech*; 16(5): 530-3, 2006.
6. De Lagausie P, Bonnard A, Berrebi D, et al: Abdominal lymphangioma in children: interest of the laparoscopic approach. *Surgical Endosc* 21: 1153-1157, 2007.
7. Singh R, Govindarayan K, Bowen C, et al: Retroperitoneal cystic lymphangioma: a rare presentation in childhood, treated laparoscopically. *J LaparoendoscopicAdvSurg Tech* 2:249-25, 2009.
8. Solari V, Mullassery D, Lansdale N, et al: Laparoscopic excision of a retroperitoneal lymphatic malformation in a newborn. *J Pediatric Surg* 46 (2): e15-e17, 2011.
9. Padilla BE, Dominguez G, Millan C, et al: The use of magnets with single-site umbilical laparoscopic surgery. *Semin Pediatric Surg*; 20: 224-31, 2011.
10. Son TM, Liem NT: Laparoscopic management of abdominal lymphatic cyst in children. *J LaparoendoscopicAdv Surgical Tech* 22(5): 505-7, 2012.
11. Moher D, Liberati A, Tetzlaff J, et al: PRISMA Group. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. *BMJ* 339: b2535, 2009.
12. Bagrodia N, Defnet AM, Kandel JJ. Management of lymphatic malformations in children. *Curr Opin Pediatr* 27(3):356-63, 2013.
13. Olímpio Hde O, Bustorff-Silva J2, Oliveira Filho AG2, Araujo KC. Cross-sectional study comparing different therapeutic modalities for cystic lymphangiomas in children. *Clinics (Sao Paulo)* 69(8):505-8, 2014.
14. Stillo F, Baraldini V, Dalmonte P, El Hachem M, Mattassi R, et al. *Vascular Anomalies Guidelines by the Italian Society for the study of Vascular Anomalies (SISAV)*. *IntAngiol* 34(2 Suppl 1):1-45, 2015.
15. Gaied F, Emil S. Laparoscopic excision of a gastric lymphatic malformation. *Am Surg* 78:E232–E234, 2012.

