Pediatric cystic lymphangioma of the retroperitoneum: A case report and review of the literature

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Abstract
Rationale: Cystic lymphangioma (CL) is a rare benign tumor resulting from a failure of the lymphatic system development. It may occur at any age but it is more frequent during childhood. Its clinical presentation and location are various but abdominal CL are uncommon. Among those, mesenteric presentation is the most frequent form whereas CL of the retroperitoneum are particularly rare.

Patient concerns: Herein, we report the case of a 17-years-old patient with no medical history who presented with right-upper quadrant (RUQ) pain, but no other symptom. Physical examination showed tenderness of the RUQ without distension. Lab tests were unremarkable.

Diagnosis: Abdominal computed tomography (CT) highlighted a retroperitoneal cystic mass potentially infiltrating the mesenterium, raising suspicion of a CL of the retroperitoneum. Diagnosis of CL was confirmed by histological analyses.

Intervention: Patient underwent an exploratory laparoscopy that confirmed infiltration of the mesenterium and allowed for resection.

Outcomes: Postoperative course was uneventful and there is no evidence of recurrence after 14 months of follow-up.

Lessons: Although CL essentially occur in children, pediatric retroperitoneal CL is a rare finding, with only 21 cases identified in the literature.

In summary, CL are benign tumors rarely located in the retroperitoneum. Despite performant imaging technologies, preoperative diagnosis is challenging. Whenever possible, laparoscopic resection should be the treatment of choice. Herein, we report the largest CL pediatric case laparoscopically resected, and the first review of the literature on the topic.

Abbreviations: CL = cystic lymphangioma, CT = computed tomography, MRI = magnetic resonance imaging, RUQ = right-upper quadrant, US = ultrasound.

Keywords: benign tumor, pediatric surgery, retroperitoneal, surgical resection

1. Introduction
Cystic lymphangioma (CL) is a rare benign tumor resulting from a failure in the development of the lymphatic system that can occur at any age but more typically during childhood.[1] Craniofacial, cervical or axillary localisation are the most common locations. Intra-abdominal forms are rare. Retroperitoneal localization of the CL is particularly uncommon.[2] The clinical presentation of CL is various, ranging from incidental finding of abdominal cyst to acute abdominal presentation.[1,4] Preoperative diagnosis is challenging and differential diagnosis is extensive.[3] The diagnosis of CL mainly relies on imaging with either ultrasound (US), computed tomodiography (CT) or magnetic resonance imaging (MRI). It should thereafter be confirmed by histology.[6,7] Whenever possible, complete resection should be attempted. The risk of recurrence primarily depends on margins status.[4] Herein, we report the largest CL pediatric case laparoscopically resected, and the first review of the literature on the topic.

2. Case presentation
A 17-year-old boy with no medical history presented with right-upper quadrant (RUQ) pain, but no other symptom. Physical examination showed tenderness of the RUQ but no rebound. Lab
tests were unremarkable. An abdominal ultrasound revealed a right flank fluid collection of unknown etiology. An abdominal CT showed a retroperitoneal cystic mass infiltrating the mesentery near the right colic angle and right Morrison’s pouch. The lesion measured 14 cm and raised the suspicion of a CL (Fig. 1A).

Patient underwent an exploratory laparoscopy that showed a large cystic mass close to the cecum (Fig. 1B). Surgical exploration revealed a multiloculated cyst encapsulating other cysts of smaller sizes (Fig. 1C), and extending to the hepatic colonic angle without infiltrating the mesentery. The main cyst was filled with clear fluid which was aspirated and sent for cytological analysis. The tumor was resected almost entirely, only leaving a small portion of the capsule which was strongly adherent to the duodenum, precluding a safe complete resection. Postoperative course was uneventful and the patient was discharged on postoperative day 2. No sign of recurrence has been reported after 14 months of follow-up.

Macroscopically, specimens showed a bilobar cystic lesion containing hemorrhagic material measuring $3.0 \times 1.2 \times 1.0$ cm and a thin fibrotic fragment of $23.0 \times 2.0 \times 1.0$ cm.

Histological analyses of the resected cysts showed macrocystic cavities, containing thin fibrous tissue and lymphocytic infiltrate (Fig. 1D and E), boarded by a single layer of endothelial cells expressing D2-40 (Fig. 1F). These findings confirmed the diagnosis of CL. No malignant cells were detected by cytological analyses in the cystic fluid.

3. Discussion
Herein, we reported a rare case of symptomatic CL located in the retroperitoneum of a young patient successfully treated by surgery with laparoscopic resection.

CL is a rare benign tumor resulting from a failure in the development of the lymphatic system.[3] Two theories exist about
its origin. The first one relies on a malformation due to a lack of connection between abdominal lymphatic chains and venous system.[4] The second one suggests an origin acquired due to inflammation, trauma or degeneration.[5] The histological types of lymphangiomas are divided into cystic, capillary, and cavernous patterns. Retroperitoneal lymphangioma mostly shows cystic type.[5]

CL is most frequently located in the subcutaneous area of the cervix (~7.5%) or axilla (~20%).[6] Intra-abdominal forms are observed in only 5% of cases.[2] Retroperitoneal localization of the CL is particularly rare. Abdominal CLs are most common in children and 90% of the cases are diagnosed before the end of second decade of life, which is consistent with our 17-years-old patient.[2]

The clinical manifestations of abdominal CL are various.[3] It varies from incidental discovery of an abdominal cyst to acute abdominal presentation. The most common symptom is abdominal pain often related to tumor volume, which can be associated with a palpable mass.[4] Complications such as intestinal obstruction, intracystic hemorrhage, infection, torsion, spontaneous rupture of the cyst or digestive hemorrhage can cause acute abdomen.[4]

Differential diagnoses of cystic retroperitoneal lymphangioma include retroperitoneal hematoma, abscess, duplication cysts, ovarian cysts, microcystic pancreatic adenoma, pancreatic pseudocysts, mucinous pancreatic neoplasms, branch-type intraductal papillary mucinous neoplasia, lymphangiosarcoma, cystic metastases (gastric/ovarian), undifferentiated sarcoma, cystic teratoma, cystic mesothelioma, and malignant mesenchymoma.[10]

There is no specific sign and diagnosis is usually guided by imaging. To establish the diagnosis, ultrasound is the typical initial exam. CL appears as a sharply marginated, unilocular or multilocular liquid tumor, often with scattered echoes.[8] In our case, ultrasound detected an anechoic fluid collection, well-delineated, without calcification and without any sign of complications. Considering the non-specific ultrasound aspect of these lesions, CT provides additional information on the size, extent of the lesion and its relation with adjacent structures. CL typically shows a well-circumscribed homogeneous cyst with contrast-enhancing walls and septa.[8] MRI helps to better define the nature of the cyst, exhibiting hypointensity in T1 images and increased intensity in T2 images.[9]

The diagnosis of CL can only be confirmed by histological analyses and is based on well-established criteria.[6] Those include a well circumscribed cystic lesion with or without endothelial lining, a stroma characterized by meshwork of collagen and fibrous tissue and a wall containing focal aggregates of lymphoid tissue.[10] Lymphatic vessel endothelial receptor-1, vascular endothelial growth factor-3, monoclonal antibody D2-40 and prox-1 are used as immunohistochemical markers in the diagnosis of lymphangioma.[11]

Surgery is the cornerstone of CL treatment. Decision-making must take into account the benign nature of the tumor, potential complications, the infrequent spontaneous regression of the cyst and the need for definitive diagnosis.[6] Surgical excision, open or laparoscopic, is the gold-standard for abdominal CL. The excision must be complete to reduce the risk of recurrences, which varies from 7% after complete resection, to 50% after incomplete resection.[4]

Our review of the literature only identified 21 cases of pediatric CL of the retroperitoneum.[12–26] Details of the reported cases are provided in Supplementary Table 1, http://links.lww.com/MD/E532. Characteristics of these cases are summarized in Table 1. Briefly, a PubMed search was performed using the following terms “Retroperitoneal”, “cystic” and “lymphangioma” (between January 1970 and January 2019). Each abstract was carefully reviewed by two separate authors (FP and IL). Inclusion criteria were (I) retroperitoneal CL, (II) age under 18 years, (III) full-text available, (IV) manuscript written in English.

Overall, a majority of patients were male with an average age of 6 years. CL showed a mean size of 12 cm and were essentially available, (IV) manuscript written in English.

Author contributions

Acquisition of data: FP, SAB, IL, LDM
Analysis and interpretation of data: FP, DP, SAB, IL, LDM
Critical revision of the manuscript for important intellectual content: FP, DP, SAB, IL, LDM
Drafting of the manuscript: FP, IL
Study concept and design: FP, IL, LDM

Table 1

| Characteristic                                      | Cases (n = 21) |
|----------------------------------------------------|----------------|
| Gender (male)                                      | 12 (57)        |
| Age                                                 | 6 (±4)         |
| Tumor size (cm)                                    | 12 (±7)        |
| Imaging techniques                                 |                |
| US                                                  | 15 (71)        |
| Abdominal CT                                        | 14 (67)        |
| MRI                                                 | 4 (19)         |
| Other imaging techniques†                          | 10 (48)        |
| Treatment                                           |                |
| Conservative                                        | 1 (5)          |
| Surgery                                             | 19 (90)        |
| Open                                                | 9 (43)         |
| Laparoscopy                                         | 3 (14)         |
| Other treatment                                     | 1 (5)          |
| Unknown                                             | 7 (33)         |
| Postoperative complications†                        | 0 (0)          |
| Recurrence                                          | 0 (0)          |
| Follow-up (mo)                                      | 13 (±6)        |

Results are provided as number of cases (%), or mean values (±standard deviations).

CL = cystic lymphangioma, CT = computed tomography, MRI = magnetic resonance imaging, US = ultrasound.

† Other imaging techniques: X-ray, dimercaptosuccinic acid technetium-99m scan, intravenous pyelography, excretory urogram, total body opacification, abdominal angiogram, selective celiac arteriogram, percutaneous retrograde abdominal angiography.

§ Postoperative complications: minimal paresthesia along the distribution of the genitofemoral nerve, chronic lymphatic fistulas, death.

Table 1: Characteristics of pediatric cases of retroperitoneal CL reported in the literature.
References

[1] Chaker K, Sellami A, Ouans E, et al. Retroperitoneal cystic lymphangioma in an adult: a case report. Urol Case Rep 2018;18:33–4.
[2] Fattahi AS, Maddah G, Motamedolshariati M, et al. Chronic low back pain due to retroperitoneal cystic lymphangioma. Arch Bone Jt Surg 2014;2:72–4.
[3] Celia A, Breca G. Laparoscopic Excision of a Retroperitoneal Cystic Lymphangioma: An Insidious Case [Internet]; 2007. Available at: https://home.liebertpub.com/lap [cité 26 févr 2019]. Disponible sur: https://www.liebertpub.com/doi/abs/10.1089/lap.2006.0167.
[4] Surlin V, Georgescu E, Dumitrescu C, et al. Retropancreatic cystic lymphangioma – considerations upon a case. Rom J Morphol Embryol Rev Roum Morphol Embryol 2011;52(Suppl):493–6.
[5] Gachabayov M, Kubachev K, Abdullaev E, et al. A huge cystic retroperitoneal lymphangioma presenting with back pain. Case Rep Med [Internet] 2016;2016:1–3. [cité 16 mai 2019]. Disponible sur: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5097799/.
[13] Leonidas JC, Brill PW, Bhan I, et al. Cystic retroperitoneal lymphangioma in infants and children. Radiology 1978;127:203–8.
[14] Iyer R, Eftekhar F, Varma D, et al. Cystic retroperitoneal lymphangioma: CT, ultrasound and MR findings. Pediatr Radiol 1995;23:305–6.
[15] Meyer T, Stöhr G, Post S, et al. Retroperitoneal lymphangioma presenting as a mesenteric cyst. Eur J Radiol 1995;21:143–4.
[16] Irvine AD, Sweeney L, Crobert JR. Lymphangioma circumscriptum associated with paravesical cystic retroperitoneal lymphangioma. Br J Dermatol 1996;134:1135–7.
[17] Waldhausen JH, Holterman MJ, Tapper D. Identification and surgical management of cystic retroperitoneal lymphangioma in children. Pediatr Surg Int 1996;11:283–5.
[18] Freund E, Farkash U, Cassella R, et al. Childhood retroperitoneal lymphangioma presenting following minor trauma. Injury 1999;30:380–3.
[19] Khetarpal R, Halwai G, Marwaha RK, et al. Retro-peritoneal cystic lymphangioma in association with fetal hydantoin syndrome. Indian J Pediatr 1999;66:294–7.
[20] Shankar KR, Roche CJ, Carby HM, et al. Cystic retroperitoneal lymphangioma: treatment by image-guided percutaneous catheter drainage and sclerotherapy. Eur Radiol 2001;11:1021–3.
[21] Wilson SR, Bohrer S, Losada R, et al. Retroperitoneal lymphangioma: an unusual location and presentation. J Pediatr Surg 2006;41:603–5.
[22] Pratap A, Tiwari A, Sah BP, et al. Infected retroperitoneal cystic lymphangioma masquerading as psoas abscess. Urol Int 2008;80:325–7. discussion 328.