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Intra-Abdominal Cystic Lymphangioma: Report of 21 Cases

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Abstract

Purpose: Studying in a retrospective review of 21 cases, diagnostic, therapeutic and evolutionary aspects of intra-abdominal cystic lymphangioma (CL). Methods: Between 1992 and 2014, 21 patients were operated at our institution for a CL. Clinical presentation, location, surgical management and outcome were studied. Results: There were 14 women and 7 men. All CL were diagnosed by abdominal ultrasound and/or abdominal CT scan. The most common site was the retroperitoneum (24%) followed by equally by the mesentery, the mesocolon and abdominal wall. Surgical treatment consisted of a complete resection of cyst in 20 patients. This resection required a splenectomy in one case for a splenic location and digestive resection in 2 cases. Two cases of recurrence of CL were revealed. The first case was a result of partial resection, but the second case occurred in a patient who underwent a total cystectomy. These patients were asymptomatic, so we decided to monitor them. Conclusion: CL in adult is a rare disease. The preoperative diagnostic has benefited from the contribution of imaging mainly ultrasound and CT scan, treatment consisted of surgical complete excision to prevent recurrences.

Keywords

Cystic Lymphangioma, Intra-Abdominal, Diagnosis, Surgical Treatment, Evolution

1. Introduction

Cystic lymphangioma (CL) is a rare malformative benign tumor of the lymphatic vessels, preferentially located in the head, neck, and axilla and usually discovered in childhood.

However, the abdominal location is extremely rare and the clinical presentation is highly polymorphic.

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In this report, we present a retrospective study of patients operated on for abdominal CL in our hospital.

2. Materials and Methods

A retrospective review of cases of cystic lymphangioma was conducted.

Between 1992 and 2014, 21 patients were operated at our institution for a CL.

All these patients underwent radiological imaging, surgical excision and histological confirmation of diagnosis.

Clinical presentation, location, surgical management and outcome were studied.

3. Results

A total of 21 patients who underwent surgical removal of intra-abdominal CL were included.

Median age at diagnosis was 45 (range 15 - 65) years.

There were 14 women and 7 men.

Clinically, the main symptom was an abdominal pain found in 21 patients (100%).

Physical examination revealed an abdominal mass only in 4 patients (19%).

The CL was unique in 20 patients (95%), whereas only one patient had more than one CL.

All CL were diagnosed preoperatively by abdominal ultrasound and/or abdominal CT scan (**Figure 1**).

These intra-abdominal cystic lesions were unilocular in 15 cases (71%) and multilocular in 6 cases. Other radiological signs such as peripheral calcifications were found in 2 cases (9%).

The initial diagnosis varied from a CL in 13 cases, a hydatid cyst in 6 cases, a cyst of the mesentery in 1 case and mesenteric tumor in 1 case.

Clinical characteristics of patients and diagnosis tools are summarized in **Table 1**.

As related to the anatomical features of the cysts, their size ranged from 4 to 15 cm with a median size of 8 cm.

The most common site was the retroperitoneum (24%) followed by equally by the mesentery, the mesocolon and abdominal wall with 3 cases in each location (14%).

More rarely the CL was found in the posterior cavity of the omentum and the adrenal gland (9% each one).

The other locations were liver, spleen and diaphragm.

Surgical treatment consisted of a complete resection of cyst in 20 patients.

This resection required a splenectomy in one case for a splenic location and digestive resection in 2 cases.

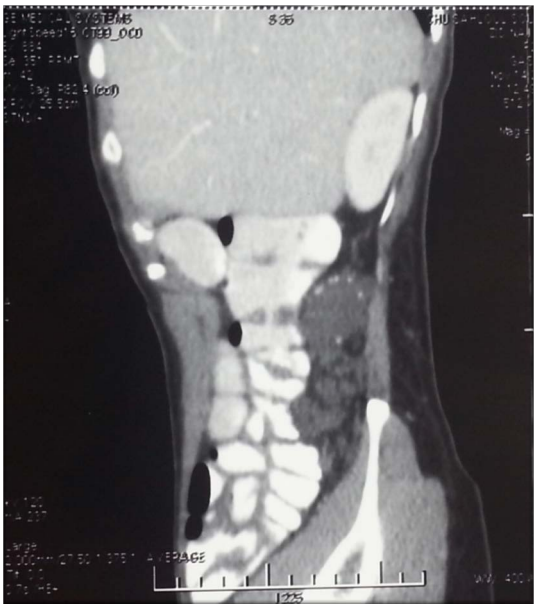

Figure 1. Abdominal CT scan shows unilocular cystic mass with calcifications.

Table 1. Clinical characteristics of patients and diagnostic tools.

Patient	Age	Sex	Symptomatology	Physicalsigns	Diagnosis
1	45	M	Abdominal pain	Absent	US, CT scan, MRI
2	27	F	Abdominal pain	Absent	US, CT scan, MRI
3	51	F	Abdominal pain	Absent	US, CT scan
4	65	M	Abdominal pain	Absent	US, CT scan
5	15	M	Abdominal pain + fever	Mass	US, CT scan
6	61	F	Abdominal pain	Absent	US, CT scan
7	52	F	Abdominal pain	Absent	US, CT scan
8	49	F	Abdominal pain	Absent	US, CT scan
9	49	F	Adbominal pain	Absent	US, CT scan
10	38	F	Abdominal pain	Absent	US, CT scan
11	63	F	Abdominal pain	Absent	US, CT scan
12	56	F	Abdominal pain	Absent	US, CT scan
13	24	F	Abdominal pain	Absent	US, CT scan
14	28	M	Abdominal pain	Mass	US, CT scan
15	51	M	Abdominal pain	Absent	US, CT scan
16	37	F	Abdominal pain	Mass	US, CT scan, MRI
17	36	F	Abdominal pain	Mass	US, CT scan
18	13	M	Abdominal pain + fever	Absent	US
19	4	M	Abdominal pain	Absent	US
20	75	F	Abdominal pain	Absent	US, CT scan
21	24	F	Abdominal pain	Absent	US, CT scan

20 patients underwent open laparotomy, whereas the remaining patient was treated by laparoscopic approach.

All patients had an uneventful postoperative recovery.

After a median follow up of 16 months (1 - 72 months) with physical examination and an abdominal ultrasound, two cases of recurrence of CL were revealed. The first case was a result of partial resection, but the second case occurred in a patient who underwent a total cystectomy (**Figure 2**).

In both cases, patients were asymptomatic, so we decided to monitor them.

The pathological characteristics and management of patients are summarized in **Table 2**.

4. Discussion

Cystic lymphangioma is a rare malformation described for the first time by Koch in 1913 [1] considered to be congenital and resulting in sequestration of lymphatic tissue failing to communicate normally with the lymphatic system [2].

However, it has been suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery, and/or radiation therapy may lead to the secondary formation of this benign tumor [3].

50% of these lesions would be present at birth, and 90% of cystic lymphangiomas would grow until the age of 2 years [4].

They are usually found in the neck (75%, also called cystic hygromas) and axilla (20%) [5] and up to 95% of cystic lymphangiomas are reported in combination [6].

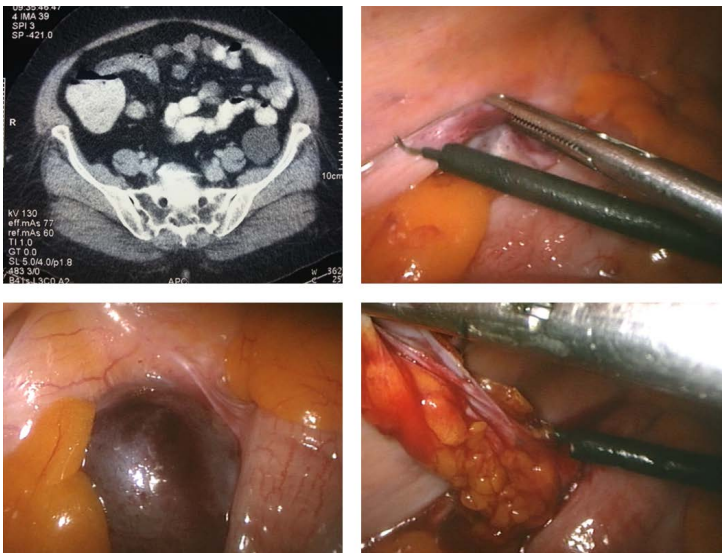

Figure 2. Introperative view of total cystectomy after puncture and evacuation of cyst.

Table 2. Pathological characteristics and management of patients.

Patient	Suspecteddiagnosis	Incision	Number	Seat	Size (cm)	Gesture
1	CL	Laparotomy	1	Retroperitoneum	6	Total cystectomy
2	CL	Laparotomy	1	Retroperitoneum	4	Total cystectomy
3	Hydatidcyst	Laparotomy	1	Liver	5	Total cystectomy
4	CL	Laparotomy	1	Abdominal wall	5	Total cystectomy
5	CL	Laparotomy	1	Mesocolon	15	Total cystectomy
6	CL	Laparotomy	1	Spleen	5	Splenectomy
7	Hydatidcyst	Laparotomy	1	Diaphragm	5	Total cystectomy
8	CL	Laparotomy	1	Mesentery	8	Digestive resection
9	CL	Laparotomy	1	Mesocolon	13	Total cystectomy
10	CL	Laparotomy	1	Mesocolon	6	Total cystectomy
11	CL	Laparoscopy	1	Abdominal wall	5	Total cystectomy
12	Mesentericcyst	Laparotomy	1	Abdominal wall	14	Total cystectomy
13	Hydatidcyst	Laparotomy	1	Adrenal gland	7	Total cystectomy
14	CL	Laparotomy	1	Retroperitoneum	15	Total cystectomy
15	Mesenterictumor	Laparotomy	1	Mesentery	10	Partial cystectomy
16	CL	Laparotomy	1	Retroperitoneum	10	Total cystectomy
17	Hydatidcyst	Laprotomy	1	BCO	5	Total cystectomy
18	Hydatidcyst	Laparotomy	2	BCO	10	Total cystectomy
19	CL	Laprotomy	1	Mesentery	10	Digestive resection
20	CL	Laparotomy	1	Retroperitoneum	5	Total cystectomy
21	Hydatidcyst	Laparotomy	1	Adrenal gland	10	Total cystectomy

Intra-abdominal cystic lymphangiomas are rare and comprise less than 5% of all cystic lymphangiomas [7] and cystic lymphangioma represents 7% of abdominal cystic lesions in adults [8].

Cystic lymphangiomas are usually benign but can be locally invasive [9] and malignant transformation is possible [10].

Approximately 60% of lymphangiomas are diagnosed before the age of 5 years [11].

Abdominal cystic lymphangiomas occur most commonly in the small bowel mesentery and the retroperitoneum [12] [13].

In the literature, men and women are affected similarly in adulthood [14].

The clinical manifestations of abdominal cystic lymphangioma are highly polymorphic depending on size and location and including abdominal pain, an increase in waist circumference, and palpable mass.

Other complications can cause acute clinical presentations, such as a hemorrhage cyst or a secondary infection, intestinal obstruction, volvulus, rupture, cystic torsion, and obstruction of the urinary and biliary tract [15] [16].

Preoperative diagnosis of intra-abdominal lymphangioma is difficult, and the main mode of investigation is radiological imaging.

Ultrasound examination is useful initially and typically shows a unilocular cystic mass, thin wall, containing hypoechoic fluid. Sometimes the cyst may be complicated by an intracystic hemorrhage, causing a partially echogenic content [17].

On CT imaging, they appear as a uni- or multilocular mass with enhancement of the wall and septum by contrast medium [18] and calcification may occur but is uncommon [19].

The fluid component is typically homogeneous with low attenuation values but the density is lower in case of chylous content and denser if lesion is complicated by hemorrhage [20].

Magnetic resonance imaging is the most useful preoperative radiological tool for diagnosis and in surgical planning [21] [22].

Other radiological investigations were reported in the literature, including lymphoscintigraphy to albumin does not appear to provide additional diagnostic element, the lymphangioma not communicating with the peripheral lymphatic system [23].

The definitive treatment for abdominal CL is radical excision, even when asymptomatic [24].

But with increasing tumor size, radical resection becomes more difficult and local recurrence more probable [25].

For all sites combined, a recurrence rate of 40% after incomplete resection and 17% after macroscopically complete resection was shown [26].

Other modalities for treatment for intraabdominal lymphangioma including percutaneous injection of sclerosing agent (doxycycline, alcohol, acetic acid…) appear to be a very safe and effective procedure and have long-term effects, such as frequent recurrences, up to 100% in some series [27]-[32].

5. Conclusions

The abdominal CL is a benign tumor of the lymphatic system, affecting mostly young children.

The diagnosis is established by ultrasound or CT scan.

To prevent recurrence, complete excision seems to be the best option.

Conflict of Interest Statement

Ben Mabrouk Mohamed and other co-authors have no conflict of interest.

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