Intra-Abdominal Cystic Lymphangiomas in Infancy And Childhood

Chih-Cheng Luo, MD; Chen-Sheng Huang, MD; Hsun-Chin Chao[1], MD; Shih-Ming Chu[1], MD; Chuen Hsueh[2], MD

Background: Cystic lymphangiomas (CL) rarely present as intra-abdominal masses. Abdominal CL is often discussed in conjunction with mesenteric cysts; however, their histology, location and age of presentation differ significantly. In an attempt to establish a best diagnostic and treatment modality, we report our experience dealing with intra-abdominal CL during a 5-year period.

Methods: Between January 1998 and December 2003, 12 patients, 7 boys and 5 girls, with a diagnosis of CL were reviewed. Modes of clinical presentation, location of CL, methods of diagnosis, surgical intervention and histological examination were all analyzed.

Results: The ages of the 12 patients ranged from 8 days to 6 years. Eleven of the 12 patients were symptomatic with abdominal pain, abdominal distention or palpable mass , dysuria and severe acute abdominal pain mimicking appendicitis. Abdominal ultrasound was done preoperatively in all patients. At laparotomy, 5 CL were located in the omentum, 5 in the mesentery, and another 2 in the retroperitoneum. All omental CL were completely excised without difficulty. CL removal required resection of both the cyst and intestine in 2 patients. One of 2 retropritoneal CL was removed with small areas of the posterior wall of the cyst remaining on the inferior vena cava (IVC). There were no major postoperative complications, deaths, or recurrences in this series.

Conclusions: Intra-abdominal CL are usually involved in young children and are usually symptomatic. A preoperative diagnosis is possible with ultrasound study. Complete excision of the cysts with or without intestinal resection is mandatory to prevent recurrence. The long-term prognosis is excellent.
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Key words: **cystic lymphangioma, ultrasound, excision.**

Lymphangiomas that are localized or generalized are regarded as malformations that arise from sequestration of lymphatic tissue failing to communicate normally with the lymphatic system.(1) Intra-abdominal cystic lymphangiomas (CL) are uncommon benign tumors of congenital origin, and are often discussed in conjunction with mesenteric cysts.(2,3) However, their histology, location, and age

From the Department of Pediatric Surgery, [1]Divisions of Pediatric Gastroenterology and Neonatology, Department of Pediatrics; [2]Department of Pathology, Chang Gung Children's Memorial Hospital, Taipei; Chang Gung University College of Medicine, Taoyuan.

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Address for reprints: Dr. Chih-Cheng Luo, Department of Pediatric Surgery, Chang Gung Children's Hospital. 5-7, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext.8227; Fax: 886-3-3287261; E-mail: lifen@cgmh.org.tw

of presentation differ significantly.[4,5] The purpose of this study was to report our experience with abdominal CL in order to define its clinical presentation, diagnosis, and treatment in more detail.

METHODS

The medical records of patients with intra-abdominal CL at Chang Gung Children's Hospital during a 5-year period between January 1998 and December 2003 were reviewed. A total of 12 patients with abdominal CL were included in this study. There were 5 girls and 7 boys who ranged in age from 8 days to 6 years. All patients had surgery and had histopathologic confirmation of the diagnosis. Modes of presentation, location of the CL, methods of diagnosis, surgical intervention, histological examination and outcomes of these patients were reviewed and analyzed.

RESULTS

The clinical details of the patients are summarised in the Table 1. The most common initial

Table 1. Summary of the Clinical Information of Twelve Patients with Intra-Abdominal Cystic Lymphangiomas

Case No.	Sex	Age	Clinical manifestations	Imaging study	Location	Diameter (cm)	Treatment	Pathologic diagnosis	Follow-up	Remarks
1	M	2.7 years	Abdominal pain	Ultrasound CT	Sigmoid mesocolon	7	Complete excision	CL	Recovery, well 5 yr after operation	
2	M	3.5 years	Abdominal distention, palpable mass	Ultrasound CT	Omentum	20	Complete excision	CL	Recovery, well 4.5 yr after operation	
3	F	8 days	Palpable mass	Ultrasound CT	Omentum	8	Complete excision	CL	Recovery, well 4 yr after operation	
4	M	5.4 years	Abdominal pain	Ultrasound CT	Retroperitoneum	16	Complete excision	CL	Recovery, well 3.6 yr after operation	
5	F	3 months	Palpable mass vomiting	Ultrasound CT	Mesentery of jejunum	9	Combined cyst and intestinal resection	CL	Recovery, well 5 yr after operation	Partial intestinal obstruction
6	F	4 years	Abdominal pain	Ultrasound CT	Omentum	7	Complete excision	CL	Recovery, well 2.2 yr after operation	
7	M	6 years	Abdominal pain	Ultrasound CT	Mesentery of jejunum	6	Partial excision	CL	Recovery, well 2.6 yr after operation	Posterior cyst wall left on SMA & SMV
8	F	2 months	Vomiting	Ultrasound CT	Omentum	8	Complete excision	CL	Recovery, well 2.2 yr after operation	
9	F	4 years	Mimicking appendicitis	Ultrasound CT	Ascending mesocolon	10	Complete excision	CL	Recovery, well 2 yr after operation	
10	M	3.6 years	Dysuria	Ultrasound CT	Retroperitoneum	10	Partial excision	CL	Recovery, well 1.6 yr after operation	Posterior cyst wall left on IVC
11	M	3.2 years	Incidently found	Ultrasound CT	Transverse mesocolon	8	Combined cyst and intestinal resection	CL	Recovery, well 1 yr after operation	
12	M	5 years	Abdominal pain	Ultrasound CT	Omentum	11	Complete excision	CL	Recovery, well 8 mo after operation	

Abbreviations: CL: cystic lymphangioma; CT: computed tomography; SMA: superior mesenteric artery; SMV: superior mesenteric vein; IVC: inferior vena cava.

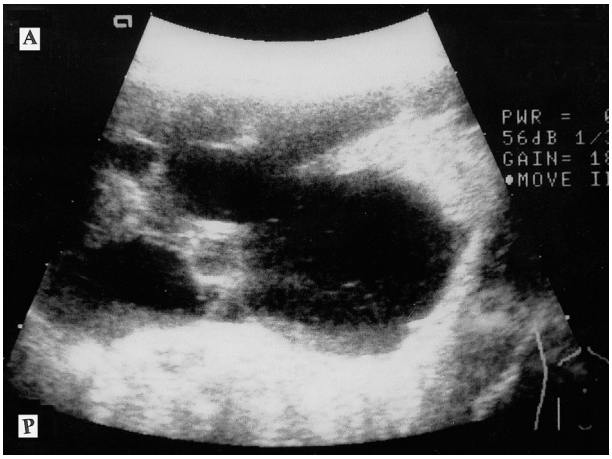

Fig. 1 Abdominal ultrasound study showing a portion of a large cystic lobulated mass located in the right lower abdomen. A, anterior, P, posterior.

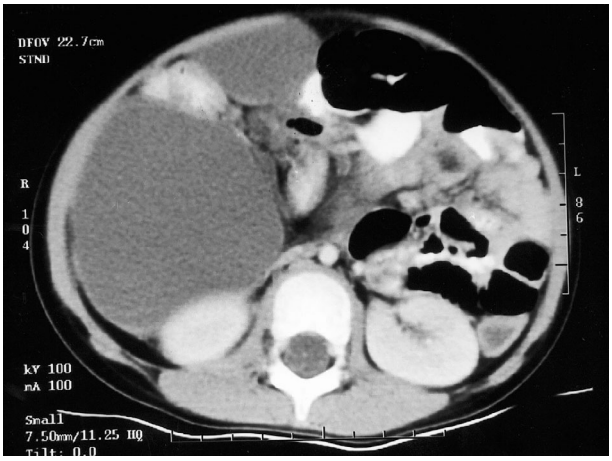

Fig. 2 Abdominal CT scan showing a large cystic mass with septa displacing the bowel loops.

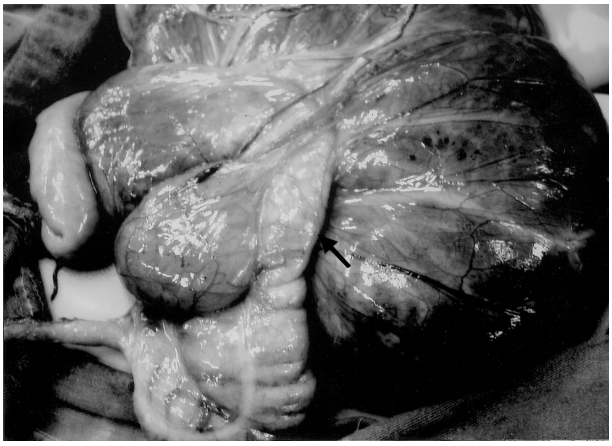

Fig. 3 Operative photograph demonstrating multiple cysts involving the ascending mesocolon (arrow). Total excision of the lesion required a segmental bowel resection.

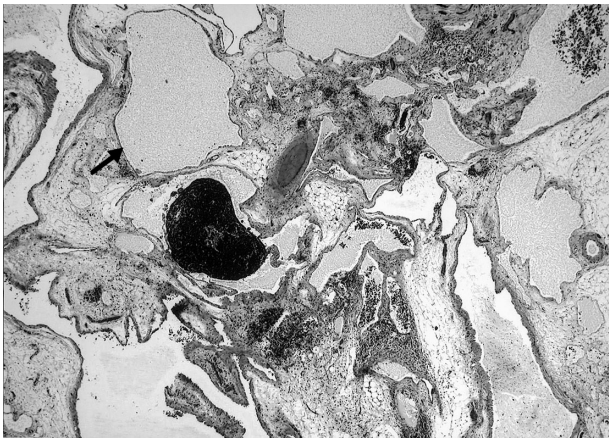

Fig. 4 Light microscopy revealing cystically dilated (arrow) and anstomosing lymphatic channels with occasional lymphoid aggregates in the wall. (H&E, ×20)

complaint was abdominal pain associated with vomiting. A palpable abdominal mass was detected in three patients. One patient with acute abdominal pain was thought to have appendicitis and one presented with urinary symptoms. Another patient was diagnosed incidentally by routine ultrasound study. Abdominal ultrasound study was performed preoperatively in all patients and invariably showed a cystic mass with septa (Fig. 1). Computed tomography (CT) showed the extent of the cyst (Fig. 2).

All patients underwent laparotomy, and the CL which were located in the omentum in four patients were completely excised easily. Locations of the mesenteric CL included the jejunal mesentery in two patients, trasverse mesocolon in one, ascending mesocolon in one, and sigmoid mesocolon in one. CL removal with intestinal resection was performed in 2 of these 5 patients (Fig. 3). One CL was firmly adherent to the base of the small bowel mesentery, involving the superior mesenteric artery (SMA) and vein (SMV) and was peeled off these vessels without damage, but small areas of the posterior wall of the cyst had to be left behind. Two CL were located in the retroperitoneum. Total excision was performed in one, and in the other, partial excision was performed with small areas of the posterior wall of the cyst

remaining on the inferior vena cava (IVC). Histological studies confirmed the diagnosis of cystic lymphangioma (Fig. 4). There were no operative deaths or major postoperative complications. The follow-up period ranged from 8 months to 5 years. There was no recurrence during follow-up in these patients.

DISCUSSION

Lymphangiomas constitute about 5 % of all benign tumors in infants and children.[6,7] The most common sites involved are the neck and axilla, but they also occur in the mouth, arm, mediastinum, lung, abdomen and viscera.[7-9] Intra-abdominal CL are rare. However, some recent reports indicated that CL present more commonly in children under 10 years of age.[10,11] In accordance with these reports, the age of the CL patients in this series ranged from 8 days to 6 years. Abdominal CL occur most commonly in the mesentery of the small bowel, with the retroperitoneum being the second most frequent site.[12-14] Indeed, in this series, 5 of 12 lymphangiomas were located in the mesentery, 5 in the omentum and 2 in the retroperitoneum.

The clinical presentation of an abdominal CL depends on its size and location. In this study, omental CL presented with a large, freely movable mass along with abdominal distention. Mesenteric CL usually presented with abdominal pain with or without vomiting. One of the 5 patients with acute abdominal pain had symptoms mimicking appendicitis. One small lymphangioma did not generate any symptoms. One retroperitoneal CL which compressed the ureter and bladder presented with urinary symptoms.

Abdominal ultrasound study is the diagnostic procedure of choice in cases of suspected abdominal CL. On ultrasound examination, a CL appears as a well-circumscribed cystic structure with thin walls often containing septa.[15,16] A higher index of suspicion and a simple ultrasonography may lead to an earlier correct diagnosis in many of these patients. A CT scan can show the extent of the abdominal CL and demonstrate that the cyst does not originate from solid organs such as the pancreas, kidney, or ovary.[17]

The definitive treatment for abdominal CL is complete surgical excision. During surgery, a bowel resection will often be performed because of the intimate relationship between the cyst and the intestine.[18-20] In this series, most CL were localized within the omentum and could be completely excised without difficulty. Four CL which occupied the bowel mesentery or much of the retroperitoneal space required a bowel resection for complete excision. Pathological examination with a light microscope confirmed the diagnosis of CL in all our patients. .

In conclusion, intra-abdominal CL are usually involved in young children and usually symptomatic. The diagnosis is established by ultrasound or CT scan. To prevent recurrence, complete excision of the CL with or without intestinal resection and near-total resection are mandatory.

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腹內囊狀淋巴管瘤在嬰兒及小孩子的表現

駱至誠 黃振盛 趙舜卿[1] 朱世明[1] 薛 純[2]

背 景： 囊狀淋巴管瘤很少以腹內腫瘤來表現，而時常和腸系膜囊腫一起被討論。不過它們之間有不同的年紀和發生位置的表現，爲了建立正確的診斷及治療的原則，我們報告本院5年間有關治療此病的經驗。

方 法： 從1998年1月至2003年12月，我們分析12位病人(包括7位男性5位女性，皆被診斷爲腹內囊狀淋巴管瘤)。臨床表現，發生的位置，診斷的方式，手術的方法及病理檢查的結果是本研究分析的重點。

結 果： 12位病人的年齡分布在8天至6歲，其中7位以腹痛、腹脹、觸摸到腫塊，小便疼痛，甚至有以類似闌尾炎的急性腹痛等症狀來表現者。所有病人在術前都接受腹部超音波的檢查。手術當中，我們發現有5位的囊狀淋巴管瘤長在大網膜，5位長在腸系膜，餘2位長在後腹腔。所有長在大網膜的淋巴管瘤都可完全切除。有2位長在腸系膜者要一併切除小腸，2位長在後腹腔的其中1位，有小部份的囊腫後壁要遺留在下腔靜脈上。所有病人無術後併發症，死亡或再發。

結 論： 腹內囊狀淋巴管瘤通常發生在年輕的小孩且有症狀；手術前的臨床表徵和腹部超音波檢查常可作爲診斷的依據；完全的切除囊腫有時需要一併切除小腸可防止再發，長期追蹤顯示病人的預後不錯。

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關鍵字： 囊狀淋巴管瘤，腹部超音波，切除。

長庚兒童醫院　台北院區　兒童外科，[1]兒童內科，[2]病理科
受文日期：民國93年2月19日；接受刊載：民國93年5月12日。
索取抽印本處：駱至誠醫師，長庚兒童醫院　兒童外科。桃園縣333龜山鄉復興街5-7號。Tel.: (03)3281200轉8227; Fax: (03)3287261; E-mail: lifen@cgmh.org.tw