Intra-Abdominal Cystic Lymphangiomas in Infancy And Childhood

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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 retroperitoneal 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. (Chang Gung Med J 2004;27:509-14)

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.¹⁰ Intra-abdominal cystic lymphangiomas (CL) are uncommon benign tumors of congenital origin, and are often discussed in conjunction with mesenteric cysts.²³ However, their histology, location, and age

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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.

**RESULTS**

The clinical details of the patients are summarised in the Table 1. The most common initial presentation of abdominal CL in children is abdominal pain, followed by palpable mass.

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

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

**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 anastomosing lymphatic channels with occasional lymphoid aggregates in the wall. (H&E, ×20)
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. The most common sites involved are the neck and axilla, but they also occur in the mouth, arm, mediastinum, lung, abdomen and viscera. Intra-abdominal CL are rare. However, some recent reports indicated that CL present more commonly in children under 10 years of age. 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. 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. 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.

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. 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.

REFERENCES


腹内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 朱世明 薛 純

背景：囊狀淋巴管瘤很少以腹內腫瘤表現，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，為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：於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，皆被诊斷為腹內囊狀淋巴管瘤)。臨床表現，發生的位置，診斷的方式，手術的機會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、腹部、觸摸到腫塊，小便疼痛，甚至有以類似闖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。所有病人術後都併發症，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：手術前的臨床表現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。

(長庚醫誌 2004;27:509-14)

關鍵字：囊狀淋巴管瘤，腹部超音波，切除。