Characteristics of Adult Abdominal Cystic Lymphangioma: a single center Chinese cohort of 12 cases

CURRENT STATUS: UNDER REVIEW

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DOI:
10.21203/rs.2.11209/v1

SUBJECT AREAS
General Surgery

KEYWORDS
Cystic lymphangioma, Abdominal cystic lymphangioma, Surgery, Adult
Abstract

Background

Cystic lymphangioma is a kind of rare benign cystic developmental disease entity, mostly affected cervical and axial regions. Clinical features of abdominal cystic lymphangiomas (ACL) was reported among childhood but unclear for adult ACL. The purpose of this study is to demonstrate the clinical characteristics of Chinese adult ACL patients and introduce our experience of treating this disease.

Methods

We conducted a single-centered non-intervention retrospective study of 12 adult ACL patients who admitted to Peking Union Medical College Hospital (PUMCH, Beijing, China) from November 1984 to August 2017. The demographic, clinical, laboratory, imaging, histologic and therapeutic data were collected.

Results

Detailed information of 7 male and 5 female was available. The mean age at diagnosis was 39.1 (17.3) years. The mean duration of follow-up was 6.9 years. Six (50%) patients were asymptomatic and abdominal pain was the leading symptom for three (25%) patients. The cysts were evaluated by ultrasound (n=8, 67%), CT (n=10, 83%) and MRI (n=4, 33%). Therapeutic modalities include laparotomy (n=6, 50%), laparoscopy (n=4, 33%) and aspiration (n=2, 17%), with a variable response. Mean postoperation hospital stay was 6.8 days. Complete excision was accomplished among 8 patients and only one relapse was observed during follow-up. One partial resection patients reported disease reccurrence.

Conclusions

This is the first Asian cohort of adult ACL patients. Typical imaging features could lead to timely diagnosis and treatment. Radical resection is recommended for ACL with a longer period of follow-up. The analysis of this cohort deepens our understanding of adult ACL.
Introduction

Cystic lymphangioma (CL) is a kind of rare benign cystic developmental disease entity. It has been suggested to result from an embryological incorrect connection of the lymphatics when primary lymphatic cysts fail to converge into the main lymphatic system (1, 2). They may occur in a variety of anatomic locations, while in most cases the lesions are located in the cervical and axial regions (3). Abdominal cystic lymphangiomas (ACL) are a rare occurrence accounting for less than 5% of all CL cases (4). Even though ACL is a rare disease, there have already been several reports published to illustrate its clinical characteristics mainly about children (5, 6).

However, the clinical features of adult ACL remain unclear. The real prevalence of adult ACL may be vastly underestimated because of lack of recognition or an atypical clinical characteristic. It may be wrongly diagnosed as pancreatic pseudocyst, ovarian cyst, kidney cyst or other diseases according to its location. Most literatures about adult ACL patients are case reports and limited cohorts were reported (7, 8). The purpose of this study is to demonstrate the clinical characteristics of Chinese adult ACL patients and introduce our experience of treating this disease.

Methods

We retrospectively reviewed the medical records of the adult ACL patients recruited between November 1984 and August 2017 at Peking Union Medical College Hospital (PUMCH), Beijing, China. Informed consent was given from each patient, and this study was approved by the Institutional Review Board of PUMCH.

Detailed demographic and clinical data including locations, symptoms, physical signs, were collected. Ultrasound, computed tomography (CT) and Magnetic Resonance Imaging (MRI) results were all reviewed, as available. Therapeutic choices, scope of surgery, length
of incision, amount of bleeding and hospital stay were detailed recorded. Follow-up of these patients were performed via telephone, e-mail or last outpatient checkup in June 2019.

All data were recorded and analyzed in SPSS statistics software version 22. Descriptive data was expressed in numbers (%) for categorical variables and mean (SD) for continuous variables, as appropriate. T-test and Fisher's exact test was used for continuous and categorical variables respectively. All data tests were two-sided and P<0.05 was regarded to be statistically significant.

Results

A sum of 12 adult ACL patients were recruited, including 7 male and 5 female patients (male: female=7:5), all of Chinese origin. The mean age at treatment was 39.1 (17.3) years. The duration from initial symptom and first imaging evaluation to treatment was 17.8 (12.2) and 4.8 (3.9) months, respectively. The follow-up time of this cohort was 6.9 (4.5) years till June 2019 (Table 1).

For 12 ACL patients, retroperitoneum was the most commonly affected location. Four (33%) patients had ACL at left retroperitoneum and 2 (17%) patients at right. Moreover, 2 (17%) patients reported ACL in posterior cavity of omentum and 2 (17%) in mesentery. Rare locations of hepatoduodenal ligament and spleen were reported in two different patients, respectively. None of these 12 ACL patients reported a family history of ACL. Primary relatives of only two of them suffered from abdominal tumors, bladder tumor and gastric cancer (Table 1).

Six (50%) patients were asymptomatic and ACL was incidentally detected during routine medical examination. For the other six patients having symptoms, abdominal pain was the leading symptom (33%). Abdominal distention, fatigue and waist circumference increase were reported by 2 patients each. No patient suffered from nausea, vomiting or acute
abdomen, such as bowel obstruction, volvulus and hemorrhage. Appendicitis, uterus myoma, tuberculosis infection, hypertension and other sporadic complications were observed in different patients, seeming to have no relationship with ACL (Table 1).

A detailed physical examination were performed to each patient. Nine (75%) of 12 did not have obvious physical signs. Abdominal mass could be touched only in 2 patients. Moreover, one patient was initially misdiagnosed as ovary cyst and treated in Department of Gynecology in our center. A 10cm × 10cm mass was palpable under bimanual genecological examination (Table 1).

Diagnosis imaging procedures included ultrasound (67%), CT scan (83%) and MRI (33%). Only two cases did not perform CT scan. One patient was diagnosed in 1980s when CT was not introduced to our hospital. The other one was misdiagnosed as ovary cyst before surgery and CT scan was not commenly performed for ovary cyst patients. The longist diameter of each imaging methods was 12.1 (6.2), 9.7 (4.3) and 11.3 (3.7) centimeters, respectively. A paired t-test of the longist diameter was calculated for patients who performed more than one imaging procedures. P values were 0.473, 0.440, 0.180 for comparision of ultrasound and CT, CT and MRI, ultrasound and MRI, respectively, all non-significant (Table 1, Figure 1 & 2).

At therapeutc aspect, ten patients were treated by surgery, 6 (50%) open surgery and 4 (33%) laparoscopic surgery. None of them required urgent surgical intervention.

Aspiration under CT guidance was performed in 2 patients and no sclerosing agents were injected into the cyst. Histopathologic study matched the diagnosis of CL in 10 surgical patients. For 2 aspiration patients, a diagnosis was made by typical imaging features and laboratory-confirmed chylous fluid of the drainage. For ten surgical patients, complete excision was accomplished among 8 patients while partial excisions in 2 patients, one with ACL in mesentery while the other one in spleen. Surgical bleeding volum was less than
50 milliliters among eight patients. Two patients had relatively larger amount, 200ml and 1600ml, both under open surgery. Besides, for six open surgery patients, the mean length of the incision was 16 (7.3) centimeters and 8.3 (6.6) centimeters longer than the longest diameter of imaging procedures, including ultrasound, CT and MRI (Table 2 & Figure 3). Median hospital stay was 12.8 (6.9) days (6.0 days before treatment and 6.8 days after treatment). Patients with laparotomy and patients with ACL at mesentery and retroperitoneum had relatively longer hospital stay after treatment (8.0 vs 5.7, p=0.255; 7.1 vs 6.3, p=0.583). Only one patient presented hypokalemia in postoperation day 1. No other severe complications were reported. With an average 6.9 years follow-up, no recurrence was noticed among 8 patients. One patient with partial cystectomy of spleen ACL reported slowly enlarging remained tumor. The other one reported disease relapse one year after radical excision of retroperitoneum cyst. Two were lost of follow-up, one with partial resection of mesentery ACL, while the other one with aspiration of retroperitoneum ACL (Table 2).

Discussion

This is the first and largest Chinese adult ACL cohort described in the literature to date. We summarized detailed demographic, clinical, imaging, histologic data and treatment choices for all adult ACL patients admitted to our hospital. Up to our knowledge, there have only been two adult ACL cohorts published in English literature (7, 8). By comparing our results with other cohorts, the similarities and differences between these cohorts could be drawn (Table 3).

The clinical symptoms of adult ACL are highly polymorphic. A huge tumor may induce abdominal pain and compression symptoms. Infection, hydatid cyst, intracystic hemorrhage, weight loss and other rare symptoms were also reported (7, 9). In Chinese cohort, half of the patients did not complain about any symptom before incidental
diagnosis of the cyst during annual medical examination. This ratio is slightly but not significantly higher than other cohorts maybe because of socio-economic development and increased awareness of health promotion (50% vs 21%, p=0.128). Vague presentations of adult ACL may lead to misdiagnosis of masquerading diseases (10). In Chinese cohort, only two patients were diagnosed as ACL before surgery due to typical imaging features. For diagnostic imaging methods, ultrasound was regarded as first-line examination and suitable for disease screening. Thin Clear boundary with strong echo, hypoechoic fluid in the center, no blood flow signal in Color Doppler Flow Imaging (CDFI) are all typical features of ACL. Noticeably, when the cyst were complicated by internal bleeding, it may also have echogenic content, which is not observed in Chinese cohort. Whether the cyst has septum is not deemed to have diagnostic value. In Chinese cohort, 5 cysts were multilobular under ultrasound and were verified pathologically. CT scan and MRI were also excellent diagnostic tools. Mostly, typical ACL under CT is a low-density cyst, with an average CT value of 19.7 Hounsfield unit in Chinese cohort. The shell of the cyst is normally glossy and regularly shaped, while the homogeneous content never take the contrast. For MRI, ACL was all presented as low signal in T2WI sequence and high signal in T1WI sequence. Similarly, no contrast could enter the cyst in MRI either. In Chinese cohort, MRI was normally used as an assistant tool for distinguish the possible origin and positional relationship with adjacent organs. The differences of longest diameter of each imaging method are not significant, which indicates the accuracy and stability of these procedures for evaluating ACL. An accurate diagnosis of ACL may be established by combination of these imaging examinations, but final confirmation should be done by pathologists. Other radiological investigations, such as lymphoscintigraphy, were not performed in Chinese cohort, because of poor potential value (11).

The size of adult ACL varied from 5×4 to 23×17 centimeters in Chinese cohort, which was
mainly within the range of 3-20 centimeters reported in literatures, except for the largest one (7, 9). The growth rate could be estimated by 3 patients with detailed records before admitting to our center. An average growth of 1.6 centimeters was observed. In the absence of prospective controlled studies, the indication of adult ACL is not standard. We recommend 5 centimeters as the smallest size for interventions. Both too small and too large ACLs may increase the difficulty of surgical procedures. But since ACL may also develop acute abdomen such as internal bleeding or volvulus, timely diagnosis and treatment is still needed especially for patients with intense anxiety (12).

Both laparotomy and laparoscopic surgery were reported to treat ACL with acceptable outcome (13, 14). In Chinese cohort, open surgery was preferred due to the complexity and uncertainty of ACL surgery. The length of incision should be a little bit longer than the largest diameter of imaging results. A longer duration of hospital stay and larger amount of operation bleeding may be disadvantages of laparotomy according to our data, but more studies are still demanded to confirm this assumption because of small sample size.

Both retroperitoneoscopic and anterior approach for laparoscopic surgery were performed in Chinese cohort. A retroperitoneoscopic approach was more suitable for ACL that was closed to urinary systems. Two aspiration procedures under CT guidance were performed. Unfortunately, one case were lost of follow-up, while the other one did not show any recurrence tendency with a regular follow-up of 10 years. Percutaneous interventions are controversial as therapeutic modality for ACL (15). More studies argued that aspiration, no matter with or without injection of sclerosing substances, had higher relapse rate in ACL (16, 17). Our data supported that aspiration and drainage may also be considered as an adjunctive therapy in the treatment plan especially for widespread and difficult-to-operate lymphangiomas.

Radical resection was considered to have lower recurrence rate than partial excision (18,
In Chinese cohort, only one of eight patients with complete resection relapsed after 6.9 years follow-up (range from 2 to 16 years). The follow-up duration is longer than other adult ACL cohorts published (7, 8). Complete resection should be attempted initially if there is no obvious contraindication for surgical intervention. However, since adult ACL is a kind of benign tumor and no malignant tendency was observed. Resection of other normal organs should be avoided, such as small intestine and spleen. In Chinese cohort, associated bowel resection was only performed in one case. Notably, after a partial resection of the spleen ACL, it was reported that the diameter of remaining ACL had grewed 3 centimeters during past 3 years. So a balance should be achieved between minimizing the surgery invasion and possibility of relapse for ACL especially in solid organs.

Our study had some evident limitations. First, it was a retrospective study and this may induce recall bias and missing data, so we conducted different methods to gather these information. Secondly, the patient number in our cohort was relatively small, although given that adult ACL is a kind of rare disease around the world. Thirdly, our hopital is a national comprehensive medical center in China, and therefore some severe cases may have been over-represented.

Conclusions

In summary, our cohort is the first Chinese adult abdominal cystic lymphangioma cohort to the best of our knowledge. We have provided detailed demographic and clinical data of Chinese adult ACL patients and compared the differences between Chinese cohort and other cohorts. With the longest duration of follow-up, our findings supported that complete surgical excision of cyst is recommended in these lesions. We also provided evidence that adult ACL could be discovered and treated timely with typical imaging features. Chinese cohort enriches the disease spectrum of adult ACL and deepens understanding of the
disease.

List Of Abbreviations

CL: Cystic lymaphangioma
ACL: Adult cystic lymphangioma
PUMCH: Peking Union Medical College Hospital
CT: Computed tomography
MRI: Magnetic Resonance Imaging
ml: Milliliter
cm: Centimeter

Declarations

Ethics approval and consent to participate
Informed consent was given from each adult cystic lymphangioma patient, and this study was approved by the Institutional Review Board of Peking Union Medical College Hospital, Beijing, China

Consent for publication
Not applicable.

Availability of data and materials
The datasets used and analysed during the current study are available from the corresponding author on reasonable request.

Competing interests
The authors declare that they have no competing interests

Funding
...The role of the funding is in the design of the study and collection of data.

Authors' contributions
XJC, SYM and ZS contributed equally to the design of the work and acquisition, analysis and interpretation of data. SYM drafted the manuscript. XJC, ZS and HXD substantively revised it. All authors read and approved the final manuscript.

Acknowledgements

The authors thank all the adult abdominal cystic lymphangioma patients enrolled in this study.

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Tables

**Table 1. Demographic and clinical characteristics of adult ACL patients**
Demographic characteristics n=12

| Characteristic                                      | Value     |
|-----------------------------------------------------|-----------|
| Sex male/female                                     | 7/5       |
| Age at diagnosis, mean (S.D.), years                | 39.1 (17.3) |
| Duration from first imaging evaluation to diagnosis, mean (range), months | 21.9 (1, 96) |
| Duration of follow-up, mean (S.D.), years           | 6.9 (4.5) |

Clinical characteristics n=12

Location

| Location                                | Value (Percentage) |
|-----------------------------------------|--------------------|
| Left retroperitoneum                    | 4 (33%)            |
| Right retroperitoneum                   | 2 (17%)            |
| Posterior cavity of omentum             | 2 (17%)            |
| Mesentery                               | 2 (17%)            |
| Hepatoduodenal ligament                 | 1 (8%)             |
| Spleen                                  | 1 (8%)             |

Symptoms

| Symptom                                | Value (Percentage) |
|-----------------------------------------|--------------------|
| Asymptomatic                            | 6 (50%)            |
| Abdominal pain                          | 4 (33%)            |
| Abdominal distention                    | 2 (17%)            |
| Fatigue                                 | 2 (17%)            |
| Waist circumference increase            | 2 (17%)            |
| Acute abdomen                           | 0 (0%)             |

Physical signs

| Physical sign                           | Value (Percentage) |
|-----------------------------------------|--------------------|
| Absent                                  | 9 (75%)            |
| Mass touched on abdomen                 | 2 (17%)            |
| Mass touched via vagina                 | 1 (8%)             |

Number of cysts

| Number of cysts                         | Value (Percentage) |
|-----------------------------------------|--------------------|
| One                                     | 11 (92%)           |
| More than one                           | 1 (8%)             |

Imaging methods

| Imaging method                          | Value (Percentage) |
|-----------------------------------------|--------------------|
| B ultrasound                            | 8/12 (67%)         |
| Longist diameter, mean (S.D.), cm       | 12.1 (6.2)         |
| CT scan                                 | 10/12 (83%)        |
| Longist diameter, mean (S.D.), cm       | 9.7 (4.3)          |
| MRI                                     | 4/12 (33%)         |
| Longist diameter, mean (S.D.), cm       | 11.3 (3.7)         |

ACL: adult cystic lymphangioma; cm: centimeter
**Table 2. Treatment and outcome**

| Treatment                              | Count   |
|----------------------------------------|---------|
| Open surgery                           | 6/12 (50%) |
| Laparoscopic surgery                   | 4/12 (33%) |
| Aspiration under CT guidance           | 2/12 (17%) |

Incision of open surgery, n=6

| Description                              | Count   |
|------------------------------------------|---------|
| Length, mean (S.D.), cm                  | 16 (7.3) |
| Longer than longest diameter of imaging results*, mean (S.D.), cm | 8.3 (6.6) |

Surgery**

| Type                      | Count   |
|---------------------------|---------|
| Complete excision         | 8/10 (80%) |
| Partial excision          | 2/10 (20%) |
| Bleeding ≤50ml            | 8/10 (80%) |
| 50ml                      | 2/10 (20%) |

Hospital stay

| Type                          | Count   |
|-------------------------------|---------|
| Before treatment, mean (S.D.), days | 6.0 (4.2) |
| After treatment, mean (S.D.), days | 6.8 (3.4) |
| Total, mean (S.D.), days      | 12.8 (6.9) |

Follow-up

| Type                  | Count   |
|-----------------------|---------|
| Loss of follow-up     | 2/12 (17%) |
| No relapse            | 8/10 (80%) |
| Relapse               | 2/10 (20%) |

*Including ultrasound, CT scan and MRI.

**Including open surgery and laparoscopic surgery

cm: centimeter, ml: milliliter

**Table 3. Comparison of Chinese adult ACL cohort with other published cohorts**
| Cohorts            | Tunisian (7) | French (8) | Chinese |
|--------------------|--------------|------------|---------|
| Patients, n        | 20           | 9          | 12      |
| Male, %            | 40           | 10         | 58      |
| Age at treatment, years | 46          | 36         | 39      |
| Follow-up, months  | 5            | 28         | 83      |
| Locations          |              |            |         |
| Retroperitoneum, % | 23           | 66         | 50      |
| Omentum, %         | 14           | 11         | 17      |
| Mesentery, %       | 23           | 22         | 17      |
| Spleen, %          | 7            | 0          | 8       |
| Symptoms           |              |            |         |
| Asymptomatic, %    | 20           | 22         | 50      |
| Abdominal pain, %  | 75           | 88         | 33      |
| Acute abdomen, %   | 10           | 0          | 0       |
| Physical signs     |              |            |         |
| Absent, %          | 40           | 44         | 75      |
| Mass touched on abdomen, % | 60    | 55         | 17      |
| Imaging methods    |              |            |         |
| Ultrasound, %      | 90           | 100        | 67      |
| CT, %              | 85           | 77         | 83      |
| MRI, %             | 0            | 22         | 33      |
| Open surgery, %    | 65           | 0          | 50      |
| Laparoscopic surgery, % | 35      | 100        | 33      |
| Aspiration under CT guidance, % | 0    | 0          | 17      |
| Complete excision, % | 90          | 100        | 80      |
| Partial excision, % | 10          | 0          | 20      |
| Follow-up          |              |            |         |
| No relapse, %      | 90           | 100        | 80      |
| Relapse, %         | 10           | 0          | 20      |

**Figures**
Figure 1

A. ACL closed to left renal pelvis  
B. ACL between spleen and stomach  
C. ACL between aorta and left kidney of three-dimensional CT reconstruction
Figure 2

A. Low signal in T1WI sequence of MRI  
B. High signal in T2WI sequence of MRI
Figure 3

A. ACL in mesentery of three-dimensional CT reconstruction  B. ACL in mesentery  
C. Root of mesenteric ACL  D. Clear fluid in mesenteric ACL