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Abdominal Cystic Lymphangioma in Adults: Diagnostic Difficulties and Surgical Outcome

Ali M Turki

Department of Surgery, Faculty of Medicine, Tanta University, Tanta, Egypt

Corresponding author: Ali M Turki

turki5th@gmail.com

Department of Surgery, Faculty of Medicine, Tanta University, Tanta, Egypt.

Tel: 0020402080401

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Abstract

Background: Abdominal lymphangioma is a rare benign tumor can occur in children but its discovery in adults is very rare. This article was undertaken to study the presentations, diagnostic difficulties and surgical outcome of abdominal cystic lymphangioma in adult patients.

Materials and Methods: Clinical and imaging files of thirteen patients with abdominal cystic lymphangioma (ACL) treated in Tanta University hospital during a period of 15 years were reviewed for demographic data, presentations, radiological studies, histopathology and surgical outcome.

Results: The study included thirteen patients (8 males and 5 females) between 19 years and 52 years (mean 38.3 years). The main clinical presentation was abdominal mass or enlargement found in 12 patients, Abdominal US and CT highlighted a cystic mass in the mesentery of small bowel, large bowel, greater omentum and retroperitoneum in twelve patients, while the cyst discovered accidentally in one patient during laparoscopic appendectomy. Complete excision of the cyst could be done in eleven patients and incomplete resection in two with the result of recurrence of the cyst in one patient and intestinal obstruction in another patient during the follow up period.

Conclusion: Preoperative diagnosis of abdominal lymphangioma is usually difficult due to its variable misleading clinical presentations, rarity of the disease, and its misdiagnosis with other intra-abdominal cysts. Diagnostic imaging studies may help to suspect of the disease, but final diagnosis is confirmed after histopathological examination of excised cyst. The best therapeutic option of ACL is complete surgical excision to avoid the cyst complications and to reduce the risk of its recurrence.

Keywords: Abdominal cystic lymphangioma; Adults; Diagnostic imaging studies; Histopathology; Surgical outcome

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Introduction

Lymphangiomas are rare benign tumors of lymphatic origin. They are preferentially located in the head and neck (75%), and axilla (20%) in children [1]. However, lymphangiomas in the peritoneal cavity are extremely rare (5%), particularly in adults. In the abdomen, they are commonly of cystic type and occur in the mesentery, followed by the omentum, mesocolon, and retroperitoneum. The etiology is unclear, but they are considered primarily to be congenital in origin [2]. The clinical presentations of abdominal cystic lymphangiomas (ACL) are variable and nonspecific and usually not helpful in establishing the diagnosis [3]. Abdominal ultrasonography (US) and computed tomography (CT) are necessary in describing the cyst and providing important information regarding its location, size, and adjacent organs involvement but not sufficient to give an accurate preoperative diagnosis of lymphangioma [4]. Moreover, the differential diagnosis of intra-abdominal cystic lesions is broad and includes besides the cystic lymphangioma other benign and malignant cyst like lesions as pancreatic pseudocyst, cystic teratoma, ovarian cyst, duplication cyst, cystic mesothelioma, malignant mesenchymoma, undifferentiated sarcoma, and adnexal torsion [5]. All of the above present a great preoperative diagnostic difficulty of ACL. This study was done to report the various clinical presentations, diagnostic difficulties, and surgical outcome of abdominal cystic lymphangioma (ACL) in adults.

Patients and Methods

The medical records of thirteen adult patients with abdominal cystic lymphangioma (ACL) managed at the department of surgery in Tanta University Hospital during the period from 1998 to 2013 (15 years) were reviewed for age, sex, clinical presentations, diagnostic imaging studies, surgical interventions, histopathological features, follow up and outcome. Abdominal US and CT were done for 12 patients, while the cyst was discovered accidentally during appendectomy in one patient. All the patients proved to have ACL after surgery and histopathological examination of the excised specimens.

Results

This retrospective study included thirteen patients (8 males and 5 females) with abdominal cystic lymphangioma. Their ages ranged between 19 years and 52 years (mean 38.3 years). The main presenting features were, abdominal mass found in nine patients, marked abdominal enlargement due to huge cyst in three patients, abdominal pain in four patients, and the cyst was discovered accidentally during laparoscopic appendectomy for acute appendicitis in one patient. Other associated manifestations were pallor, anorexia, fever and constipation. Plain x-ray of the abdomen showed a gasless space-occupying lesion displacing the bowel loops in seven patients. Abdominopelvic ultrasound and contrast CT scan were done for twelve patients. US showed hypoechoic cystic mass with fine septa but the origin was obscured in most of the cases, while contrast CT, revealed homogenous unilocular or multilocular cystic mass with enhanced septa. It defines also the size of the mass, its anatomical location and the adjacent organs involved **(Figure 1)**. Preoperative diagnosis of ACL was suspected in two patients, misdiagnosed as ovarian cyst and adnexal malignancy in two patients and initially diagnosed as ascites by the internist in one patient, to whom diagnostic paracentesis was done before surgical consultation.

On exploration, the cyst was found arise in the mesentery of small bowel in six patients (ileum n=4, jejunum n=2) **(Figure 2)**, root of mesentery (n=2), greater omentum (n=2), transverse mesocolon (n=1) **(Figure 3)**, sigmoid mesocolon (n=1), and the retroperitoneum (n=1). Intraoperative provisional diagnosis of ACL was made in three patients only. The size of cysts was variable **(Table 1)**, and the contained fluids ranged from 150 cc to 5 liters.

Complete surgical excision of the cyst (cystectomy) could be done in seven patients (53.8%), excision of the cyst with the adjacent part of intestine that is intimately adherent to it in four patients (30.8%), partial excision of a huge cyst with intraperitoneal marsupialization in one patient (7.7%) and incomplete excision of the cyst leaving a small part of its posterior wall adherent to superior mesenteric vessels done in one patient **(Table 1)**. After surgery, the excised specimens sent for histopathological examination and proved to be cystic lymphangioma by its characteristic histological criteria of lymphatic spaces lined by flat endothelial cells, lymphoid aggregates and foam cells containing lipoid material in stroma and smooth muscle fibers in their walls **(Figure 4)**. The cyst lumen contains chylous or serous fluid. The patients' follow up period ranged between two and six years with no mortality or morbidity except for recurrence of the cyst in one patient (7.7%), three years after operation, and adhesive intestinal obstruction required adhesolysis, two years after surgery, in another patient.

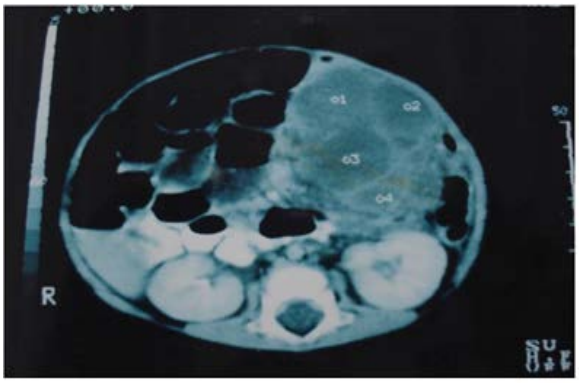

Figure 1 Contrast CT of abdominal cystic lymphangioma showing hypoechoic multi-locular cystic mass with multiple enhanced septa arises in the sigmoid mesocolon.

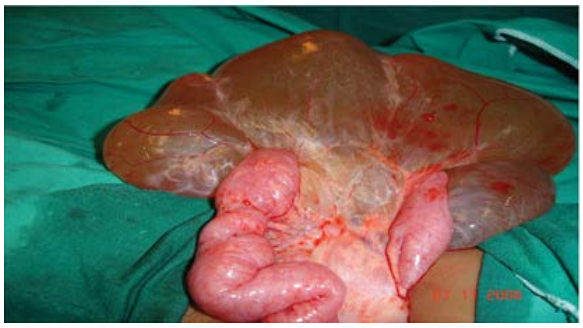

Figure 2 Cystic lymphangioma of ileal mesentery.

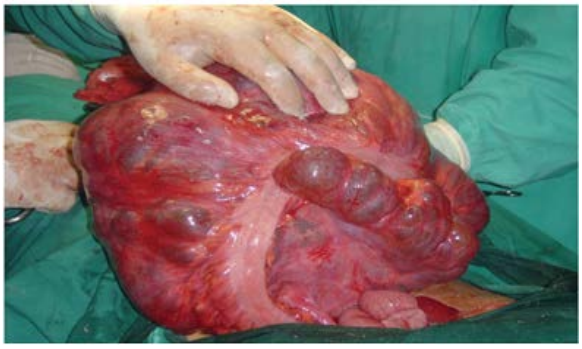

Figure 3 Cystic lymphangioma of transverse mesocolon.

Discussion

Although abdominal lymphangioma is rare, it presents a diagnostic challenge to physicians and surgeons. In this series, ACL was suspected preoperatively in two patients only and misdiagnosed as ovarian cyst, adnexal pathology and ascites in three patients,

Table 1 Demography, presentations, imaging studies, surgical intervention and outcome of thirteen patients with abdominal cystic lymphangioma.

Case No.	Age (Y)	Sex	Clinical Presentations	Imaging Studies	Cyst Location	Size of Cyst	Surgery	Outcome	Remarks
1	38	F	Mobile abdominal mass	US, CT	Greater omentum	18 cm × 10 cm × 6 cm	Cystectomy	Alive & well	Misdiagnosed as ovarian cyst
2	41	M	Abdominal mass	AXR, US, CT	Jejunal mesentery	9 cm × 6 cm × 4 cm	Excision of the cyst with a part of intestine	Alive & well	
3	43	M	Abdominal enlargement & discomfort	AXR, US, CT	Transverse mesocolon	20 cm × 22 cm × 9 cm	Cystectomy	Alive & well	
4	36	F	Abdominal mass, vague abdominal pain and constipation	AXR, US, CT	Root of mesentery	17 cm × 12 cm × 10 cm	Cystectomy	Alive & well	
5	47	F	Left pelviabdominal mass & abdominal pain	US, CT	Sigmoid mesocolon	10 cm × 9 cm × 4 cm	Excision of the cyst with a part of intestine	Alive & well	Misdiagnosed as adnexal mass
6	19	M	Right lower abdominal pain, nausea, vomiting, fever	Discovered during laparoscopy	Ileal mesentery	6 cm × 5 cm × 3 cm	Laparoscopic cyst excision & appendectomy	Alive & well	Accidentally discovered
7	27	F	Abdominal mass & recurrent abdominal pain	US, CT	Ileal mesentery	10 cm in diameter	Excision of the cyst with a part of intestine	Alive & well	
8	45	M	Abdominal mass	AXR, US, CT	Retro-peritoneum	27 cm × 22 cm × 15 cm	Partial cyst excision & marsupialization	Recurrence	
9	31	M	Diffuse abdominal enlargement & ballottement	AXR, US, CT	Greater omentum	26 cm in diameter	Cystectomy	Alive & well	Misdiagnosed as ascites
10	52	F	Abdominal mass, epigastria pain	US, CT	Jejunal mesentery	17 cm × 11 cm × 6 cm	Excision of the cyst with a part of intestine	Alive & well	
11	34	M	Abdominal mass, pallor	US, CT	Ileal mesentery	11 cm × 7 cm × 5 cm	Cystectomy	Alive & well	
12	46	M	Abdominal mass	AXR, US, CT	Root of mesentery	21 cm × 14 cm × 7 cm	Incomplete cyst excision	Adhesive IO	
13	39	M	Abdominal mass	US, CT	Ileal mesentery	13 cm × 8 cm × 7 cm	Cystectomy	Alive & well	

US=Ultrasound; CT=Computed Tomography; AXR=Abdominal X-Ray; IO=Intestinal Obstruction; Cystectomy=Complete Excision of the cyst only

however, the definitive diagnosis is judged in all patients after histopathological examination of excised specimens.

Within the abdomen, the most common site of lymphangioma is the mesentery (mesenteric lymphangioma) in which most lymphatic channels are included [6]. Lymphangioma of small bowel mesentery is the commonest (70%) of intraperitoneal sites, with 50% to 60% of all cysts located in the ileal mesentery [7]. Although mesenteric cysts and mesenteric cystic lymphangiomas are uncommon and clinically confusing lesions, histological and ultrastructural evidence suggests that they are pathologically distinct and should be differentiated because lymphangioma behaves differently [8]. Other possible locations include the omentum (omental cyst), retroperitoneum, mesocolon, pancreas, spleen and adrenal gland [9]. The most frequent classification system used for lymphangiomas is still that of Wegner who categorized them as simple, cystic or cavernous however, Losanoff et al. [10] reported another classification into pedicled, sessile, retroperitoneal extended or multicentric that may help in their treatment strategy. All the cases in this study were of cystic type and 66.7% of intraperitoneal types arise in the small bowel mesentery, 33.3% in ileal mesentery, 16.7% in jejunal mesentery and 16.7% in the root of mesentery.

The clinical presentations of ACL are usually variable and nonspecific. It may be clinically silent and discovered accidently during investigation or laparotomy for unrelated pathology or may mimic a variety of vague abdominal symptoms like abdominal discomfort, distension, diarrhea, anorexia, nausea and vomiting. In the symptomatic cases, the cystic mass may mimic other abdominal cysts like ovarian cyst, pancreatic cyst, duplication cyst or adnexal torsion [11-13]. Very few cases may present with

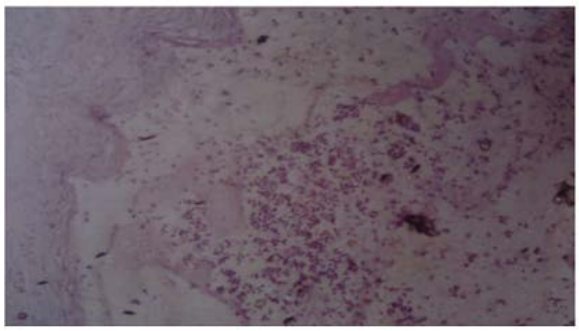

Figure 4 Microscopic photo of a case of abdominal cystic lymphangioma showing lymphatic spaces lined by flattened endothelial cells, lymphocyte aggregates in stroma and remnants of smooth muscle fibers in the walls.

acute abdomen due to cyst complications, which include rupture, hemorrhage, secondary infection, or intestinal obstruction due to torsion, volvulus or external pressure on nearby intestine [14-16]. ACL may also be misdiagnosed as appendicitis or Meckel's diverticulitis [17]. Most of the cases (92.3%) in our study were symptomatic, may be due to late presentation, except one case of ileal lymphangioma discovered accidentally during laparoscopic appendectomy.

Ultrasonography, Computed tomography and Magnetic resonance imaging studies are useful preoperative radiological tools for diagnosis and surgical planning of ACL. They can help to determine the cystic nature of the tumor, its anatomical location and its relation to surrounding structures, but they are not sufficient to establish a single preoperative diagnosis, and the final diagnosis is confirmed after histopathology and/or immunochemistry [18].

Although lymphangioma is a benign lesion, it has an aggressive invasive behavior; it may grow to an enormous size and develop life threatening cyst complications like, rupture, infection, acute abdomen due to cyst complications or intestinal obstruction, or invade adjacent vital structures and make complete resection of the tumor difficult. So, ACL should be excised completely as early as diagnosed, with or without the adjacent part of adherent bowel, to avoid the cyst complications and reduce the risk of recurrence. Prognosis is excellent if cyst resection is complete [19,20]. If complete cyst excision is not possible because of its huge size, deep location within the root of mesentery or it infiltrates main intra-abdominal artery or IVC, the other option is partial excision of the cyst with marsupialization of its remaining part into the abdominal cavity. Approximately 10% of patients require this form of therapy [21]. One (7.7%) of our patients required this option of treatment, with recurrence of the cyst in this patient.

The recurrence rate ranges from 0% to 13.6%, averaging about 6.1%. Most recurrences occur in patients with retroperitoneal cysts or those who had only a partial excision [22]. Location of the cyst does not influence the recurrence rate if it is resectable but surgery and hospital stay become more prolonged as in retroperitoneal and mesenteric root lymphangiomas [23]. The importance of surgical treatment of ACL is to avoid the potential cyst complications and reduce the incidence of recurrence, while, non-surgical treatments with bleomycin and steroids, or aspiration and injection of sclerosing agents as doxycycline has not been established to be superior to surgery, for the same reasons [19].

Conclusion

From the above we can conclude that, preoperative diagnosis of ACL is usually difficult due to its variable misleading clinical presentations, rarity of the disease, and its resemblance to many other intra-abdominal cysts. Although US, CT and MRI are helpful imaging tools, they can give only suggestive diagnosis of the disease; however, the definitive diagnosis is made only after histopathological examination of the excised specimen. Complete surgical excision is the treatment of choice of ACL to avoid cyst complications and reduce the incidence of recurrence.

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