

Abdominal Cystic Lymphangioma-
about Seven CasesRachid Khmekhem^{1*}, Houda Rahay¹, Sofiene Ghorbel¹, Said Jlidi¹, Wiem Douira²
and Ibtisem Bellagha²¹Department of pediatric surgery, University Tunis El Manar, Hospital of children of Tunis, Tunisia²Department of pediatric Radiology, University Tunis El Manar, Hospital of children of Tunis, Tunisia

Article Information

Received date: May 13, 2016

Accepted date: Oct 24, 2016

Published date: Oct 26, 2016

*Corresponding author

Rachid Khemakhem, Department of
pediatric surgery, Children Hospital of
Tunis, University Tunis El Manar, Tunisia,
Email: kakoum_rochdi@yahoo.frDistributed under Creative Commons
CC-BY 4.0Keywords Cystic lymphangioma;
Abdomen; Children; Surgery

Abstract

Cystic Lymphangioma (CL) is a rare benign tumor of malformative etiology. It is seen mainly in children and sits at the neck and axilla. Abdominal localization is rare and representing only 10% of cases.

Aim: To study the clinical, radiological and therapeutic management of abdominal cystic lymphangioma.

Patients and methods: This is a retrospective study of seven cases of abdominal CL collected over a period of 11 years in the department of pediatric surgery, of the children's hospital of Tunis. These are four boys and three girls with a mean age of 2 years 7 months.

The diagnosis was made prenatally in only one case and in postnatal period in the remaining cases due to abdominal pain in 3 cases, an abdominal mass in 4 cases and an acute intestinal obstruction in 2 cases.

The radiological diagnosis was made by abdominal ultrasound in 6 cases, and during surgical exploration in emergency in case of acute intestinal obstruction.

All patients were operated. The localization of CL was the mesentery in four cases, the left meso-colon in one case, the spleen lodge in one case and the retro-peritoneum in a case. The lesion was totally removed in five cases and partially in 2 cases.

The outcome was favorable in all cases outside of flange on occlusion occurred late postoperatively in one case and who has advanced medical treatment.

Conclusion: Abdominal cystic lymphangioma is a benign tumor malformation, preferably sitting in the mesentery. Surgical resection should be as complete as possible but without sacrifice of body, putting the patient free from complications and recurrences.

Introduction

The Cystic Lymphangiomas (CL) are connective vascular malformations lesions corresponding to a sequestration of the lymphatic tissue secondary to an abnormal embryological development of the lymphatic system whose origin remains uncertain [1,2]. Described for the first time by Koch in 1913, this is a rare benign anomaly occurring primarily in children with nearly 60% of cases are diagnosed before the age of 5 years [3].

The cystic lymphangiomas essentially sit at the subcutaneous tissue of the cervical region (nearly 70%), extremities (15%) and in the armpits (near 15%). Abdominal localization is rare and represents only 10% of cases with a preference for the mesentery and omentum. This location is problematic specific in diagnostic and therapeutic approach.

Patients and Methods

This is a retrospective descriptive study that interested 7 patients with an abdominal cystic lymphangioma collected between January 2002 and December 2012 in the Department of Pediatric Surgery "B" of the children's hospital of Tunis (Table 1).

There are four boys and three girls whose age at diagnosis ranged from 1 day to 6 years and 3 months with an average of 2 years 7 months.

The diagnosis was made prenatally in one case, due to the existence of a cystic intra-abdominal lesion at the obstetrical ultrasound of the third trimester of pregnancy.

For the six remaining children, diagnosis was made in postnatal period with symptomatic patients: abdominal pain in 3 cases, mass and/ or bulging of the abdominal wall in 4 cases and bowel obstruction in 2 cases.

Abdominal ultrasound is the most widely used radiological examination done in all cases it shows a well defined multicystic mass. Doppler Ultrasound has made the diagnosis of small bowel volvulus in one case in presence of whirlpool sign. Further exploration was deemed necessary and directed by CTScan in 4 cases and abdominal MRI in 1 case that confirms the ultrasound diagnostic

Table 1: Summary of observations of abdominal CL.

N°	Sex	Age	Clinic	Diagnosis	localization	Treatment	Evolution
1	M	1 Y	Intestinal obstruction	X-ray Laparotomy	Mesentery 5 cm	Limited Resection of intestine	Good FUp 2 years
2	M	2Y 3M	Abdominal pain and Mass	Abdominal Ultra Sound CT Scan	Retro-peritoneum 15 cm	Partial resection marsupiation	Adhesive obstruction FUp 4 years
3	M	5 M	Intestinal obstruction	Doppler US(volvulus)	Mesentery 7 cm	Limited Resection of intestine	Favorable 6 M
4	F	5Y 4M	Abdominal pain and Mass	Abdominal Ultra Sound CT Scan	Splenic loge 13 cm	Subtotal excision	Favorable FUp 6 years small stable residue
5	F	7 M	Abdominal Mass	Ultrasound RMI	Transverse Mesocolon 10 cm	Total removal	Favorable FUP 2 years
6	M	1 D	Antenatal diagnostic	Abdominal Ultra Sound CT Scan	Mesentery 4 cm	Limited Resection of intestine	favorable Fup 2Y1/2
7	F	6Y 3M	Abdominal pain and Mass	Abdominal Ultra Sound CT Scan	Meso-jejunum 6 cm	Jejunal resection and excision	favorable Fup 2 Y

and give specified the location of the lesion. One patient was operated in emergency after abdominal X-ray showing an intestinal obstruction with air-fluid levels type small bowel.

Two patients underwent emergency surgery in presence of acute intestinal obstruction, then for the remaining five patients, surgical exploration was performed routinely (with one by laparoscopy).

Intraoperatively, there was a cystic lymphangioma of the mesentery in 4 cases, in the Mesocolon in a case of retro-peritoneal region in 1 case and near the spleen in one case.

The operation consisted of resection of the lymphangioma with limited resection to adjacent intestinal loop hail in 4 cases of lymphangioma of the mesentery (Figure 1), total excision of the lymphangioma only in one case and sub-total in the remaining two cases with minimal residue which was cauterized.

The outcome was favorable in all cases even those with residue which remained stable lack of long-term recurrence for the remaining patients. One patient presented a bowel obstruction one year later that was managed conservatively with good result.

Discussion

The cystic lymphangiomas are rare benign lesions of congenital origin. The abdominal localization is rare, accounting for only about 10% of cases (6/50 cases or 12% in our series). Lesions are then essentially localised in the mesentery, but can involve the

retroperitoneum, omentum, spleen, kidney, liver and pancreas [1-3,5]. In this location has a male predominance with a sex ratio of 2 to 3 [1,5,6].

Lymphangioma may look multilocular and sometimes it is unicystic.

The pathological lymphangiomas are divided into three dominant groups: capillary, cavernous and cystic. The first two are predominantly skin, while cystic lymphangioma is predominantly intra-abdominal and retro peritoneal.

The cyst wall is surrounded by an epithelium composed of a single row of endothelial cells and has small lymphoid aggregates whose existence helps make difference with the simple cyst of the mesentery. In case of secondary bleeding into the cystic cavity, C L can be difficult to distinguish from hemangioma which the diagnosis can be made on the immunohistochemical data [3,6].

the content of CL can be, serous or sero-sanguineous. These different aspects can be explained by different degrees of lymphatic stasis, a variable number of connections with the lymphatic system and the protein content of the cyst contained. The sero-hematic cyst appearance is secondary to intracystic hemorrhage. Rarely CL can be purulent by infection [5,6].

it is accepted that, CL is the result of a lack of connection between nodes and the venous system during embryonic period resulting in

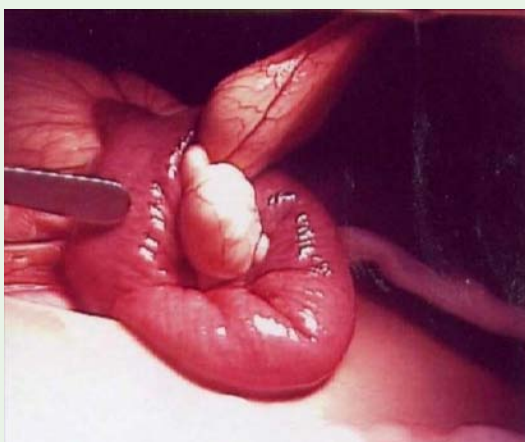


Figure 1: Intraoperative appearance of chylous cystic lymphangioma of the mesentery.



Figure 2: Abdominal ultrasound showing a multicystic intra abdominal dotted thin wall suggestive of CL.

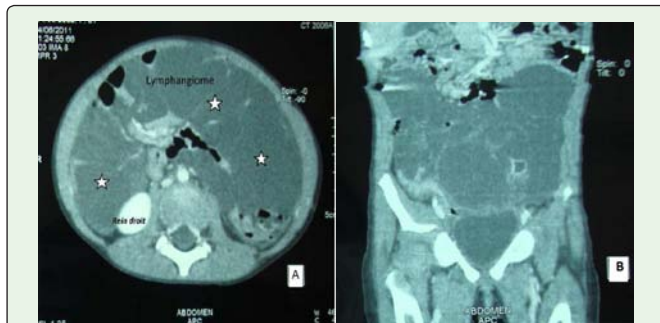


Figure 3: Abdominal CT Scan with injection showing a large intra-abdominal multi celled cystic lesion with thin walls backing the handles periphery.
3a: axial section of the floor in meso-colic.
3b: reconstruction showing the extent of lesions in height.

isolation of a bud lymphatic evolving on its own behalf to provide a CL [1-2, 7]. The persistence of abnormal retroperitoneal bag would be at the origin of retroperitoneal, mesenteric or mesocolic locations after his attraction to forward in the development of meso [1]. This theory is reinforced by congenital CL observation in the neonatal period.

Also a chronic intermittent volvulus may compromise the adequate lymphatic drainage and lead to venous stasis and congestion of the lymphatic system leading to the formation of a mesenteric lymphangioma or chylous cyst. Bleeding, inflammations, trauma leading to lymph obstruction have also been proposed as causes of acquired lesions [5].

Most of the patients with CL are initially asymptomatic or presenting vague symptoms that can change rapidly or later depending on the size and the location of the cysts [6]. As in our patients, abdominal pain with or without abdominal mass is the most common diagnosis circumstance, found in 58.4% of cases in one serie, this is rarely an occlusive syndrome revealing 30 to 40% of cases, 28.7% in our series) [1,2,4,6,7]. The diagnosis can also be done during complications with the most frequent are bowel volvulus, traumatic rupture, intra-abdominal hemorrhage or intra-cavitary sur-infection [2,6,8]. These are the CL which are relatively mobile that can be subject to complications, while those localised in retroperitoneal region are few symptoms and therefore often discovered late. Currently, and with the increase in prenatal ultrasound, the CL is increasingly diagnosed in prenatal, (1 case in our series).

The abdomen X-ray has no place in the diagnosis of cystic lymphangioma, it only shows the shadow of the lesion as a water tone opacity relatively well limited backing the digestive gas in periphery (6).

Abdominal ultrasound is the modality of choice for the investigation of intra-abdominal masses with high sensitivity and specificity. It was used in 83.3% of our patients and was able to make the diagnosis in all cases. These liquid formations appear as hypochoic well circumscribed uni or multi-celled lesions with intracystic septa (Figure 2). these lesions can contain dispersed internal echoes in case of infection. Ultrasound is also an examination of choice in postoperative, especially in children [1,2,6].

CT and MRI can provide important preoperative information regarding the location of lesions, the invasion of adjacent organs, the size of the cysts and the possible complications. Measuring the

density of the intracystic liquid can distinguish serous, hematic and chylous content (Figure 3a, 3b). These tests are important for clarify the relationship between the lesion and the superior mesenteric vessels [3,6].

In MRI, the content of the lesion may be explained more specifically. In case of fluid content, there is a hypointense signal on T1, hyperintense on T2, which reinforces the late echoes of T2. Walls and partitions are usually hypointense on both sequences T1 and T2. Gadolinium-taking by the wall and the septa can however be observed [1].

The treatment of abdominal cystic lymphangioma is surgical excision often associated with limited bowel resection because of the intimate relationships between cysts and intestinal wall. Omental resection is easy in this case, it can be done by laparotomy or laparoscopy which is increasingly performed and offers better postoperatively comfort. After all, this surgery should remain conservative due to the benign nature the lesion. No sacrifice of vital organ is tolerated [2,5,6,8,9]. Complete resection was possible in 66% of cases in our serie with a limited intestinal resection in 75% of cases of the mesenteric CL.

In case of invasion of vital structures surgical excision becomes hazardous and incomplete resection or marsupialization is performed. These techniques expose to high rate of complications such as bleeding, fistula and risk of recurrence in nearly of 10 to 30% of cases [2,6]. Performed in two of our patients, this technique was sufficient and effective since no recurrence was recorded.

This treatment should be offered as early as possible because of the risk of complication [4]. However, small asymptomatic lesions may be monitored by repeated imaging, indeed, possible spontaneous regression were reported in approximately 10% of cases [2,3,10].

Aspiration of the cystic content with or without injection of a sclerosing substance has variables results with high recurrence rate reaching 100% in some series [2]. Bleomycin is one of the first sclerosing products used to reduce the lesion volume but has low cure rates. More recently, has been proposed injection of the Tissucol, Ethibloc which would of greater efficiency in the range of 40 to 50%. But these treatments are aimed at macro-micro-lesions cystic and easily therefore accessible locations: subcutaneous cervicofacial and axillaries [2,6,9].

Conclusion

The final proof of the diagnosis of CL is provided by the pathological examination. Microscopically, the lesions appear as dilated lymphatics lined by endothelial cells without atypia, with abundant lymphoid tissue [2].

References

1. Mabrut JY, Grandjean JP, Henry L, Chappuis JP, Partensky C, Barth X, et al. Les lymphangiomes kystiques du mésentère et du méso-colon. Prise en charge diagnostique et thérapeutique. *Ann Chir.* 2002; 127: 343-349.
2. Bezzola T, Buhler L, Chardot C, Morel P. le traitement chirurgical du lymphangiome kystique abdominal chez l'adulte et chez l'enfant. *J. Chir.* 2008; 145: 238-243.
3. Ghatak S, Ray S, Sanyal S, Sonar PK, Khamrui S, Basu K, et al. An Unusual Cause of Acute Abdomen in Adults: Giant Cystic Lymphangioma of the Pancreatic Head. A Clinical Case and Literature Review. *JOP.* 2011; 12: 266-270.

4. Fayad Ali H. Abdominal Cystic Lymphangioma in Children. *Annals of Pediatric Surgery*. 2009; 5: 132-136.
5. Weeda VB, Boojij KAC, Aronson DC. Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature. *J Pediatr Surg*. 2008; 43: 1206-1208.
6. Losanoff JE, Richman BW, El-Sherif A, Rider KD, Jones JW. Mesenteric Cystic Lymphangioma. *J Am Coll Surg*. 2003; 196: 598-603.
7. Prabhakaran K, Patankar J Z, Loh D L S K, Ahamed Faiz Ali M A. Cystic lymphangioma of the mesentery causing intestinal obstruction. *Singapore Med J*. 2007; 48: e265-e267.
8. Day W, Kan D. A small bowel lymphangioma presenting as a volvulus. *Hong Kong Med J*. 2010; 16: 233-234.
9. Luo CC, Huang CS, Chao HC, Chu SM, Hsueh C. Intra-Abdominal Cystic Lymphangiomas in Infancy and Childhood. *Chang Gung Med J*. 2004; 27: 509-514.
10. Méndez-Gallart R, Solar-Boga A, Gómez-Tellado M, Somoza-Argibay I. Giant mesenteric cystic lymphangioma in an infant presenting with acute bowel obstruction. *Can J Surg*. 2009; 52: E42-E43.