Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/12907
DOI URL: http://dx.doi.org/10.21474/IJAR01/12907

RESEARCH ARTICLE

CYSTIC LYMPHANGIOMA A CASE REPORT

Etoka E.C, MA Bicane, Rabbani K, Lammat H, Louzi A
Department of General Surgery, University Hospital Center Mohamed VI, Marrakech, Morroco.

Manuscript Info

Manuscript History
Received: 25 March 2021
Final Accepted: 29 April 2021
Published: May 2021

Abstract

Cystic lymphangioma of the pancreas is a rare benign vascular tumor. Its histogenesis is still hypothetical and its preoperative diagnosis difficult to establish. We report a case in a 26-year-old boy who presented with abdominal pain with the notion of a constipation-type transit disorder. Radiological investigations showed an intraperitoneal mesenteric cystic tumor. A complete resection of the mass was performed. Pathological examination of the operative specimen confirmed the presence of cystic formations, the wall of no need keep it.to the end which was lined by a squamous, endothelial epithelium with a fibrous wall dotted with a few lymphoid clusters in favor of cystic lymphangioma.

Introduction:-

Mesenteric cystic lymphangioma is a benign condition of probable vascular malformative origin, frequently appearing in infancy. The majority of cystic lymphangiomas appear at the cervico-axillary level. However, intra-abdominal locations such as the mesentery, retro peritoneum, omentum, etc. are possible, but less common, often benign, but many complications can occur, related to its location and volume. It is therefore essential to diagnose this mass in order to avoid the occurrence of an abdominal complication (transit disorder mesenteric ischemia, volvulus, intestinal distress, etc.)

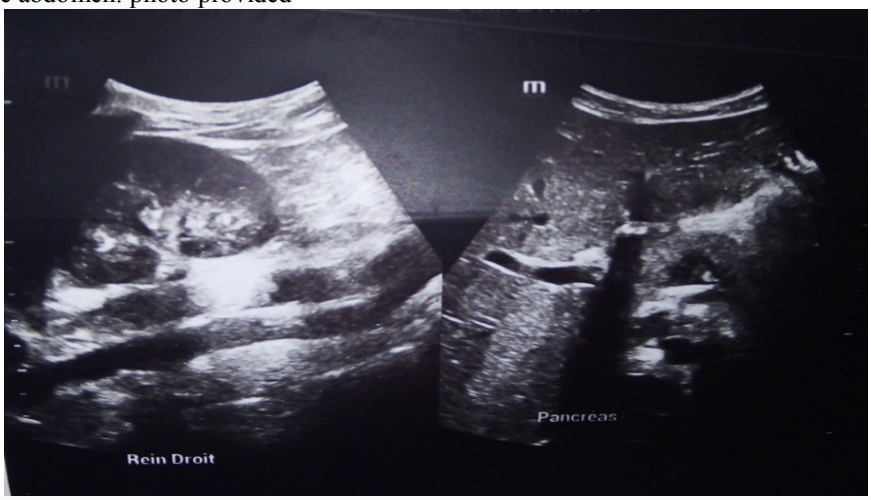
Clinical Case

A 26-year-old patient of Moroccan origin has been hospitalized for the development of an evolving abdominal pelvic mass for 2 months with paroxysmal intermittent abdominal pain, with the notion of a constipation-type transit disorder, which appeared 24 hours ago. The pain is crampoid, localized throughout the abdomen, without radiation and not relieved by taking an antispasmodic. The appetite is preserved without losing weight. This is the first painful episode of this type, the patient has been followed for a congenital pathology made up of harmonious growth retardation since the age of 12 years with an old face apparence. The abdominal examination revealed a voluminous abdominal mass, mobile without modification of the abdominal skin with a diffuse minimal sensitivity, and no sign of peritoneal irritation. Umbilicus reverse and transit is present. The patient has no fever and the remainder of the examination is unremarkable. The blood biology finds an inflammatory syndrome with a CRP at 10.3 mg / l without hyperleukocytosis; liver, kidney and pancreatic tests are normal. Abdominal ultrasound reveals a bulky bilateral intraperitoneal and para median abdomino pelvic cystic mass content 22 cm containing echogenic septa, mesuring 22 cm heights. . Echinococcus serology is negative. The more detailed development shows the presence of the tumor markers CEA and C19.9 in the upper limits of the normal, the abdominal CT- scann confirms the presence of a voluminous intraperitoneal unilocular mesenteric cystic formation with roughly rounded shape with thin wall of 22, 4 * 12.7 extended over 21 cm, at the top in contact with the inferior face of the liver, of the pancreas thus with a

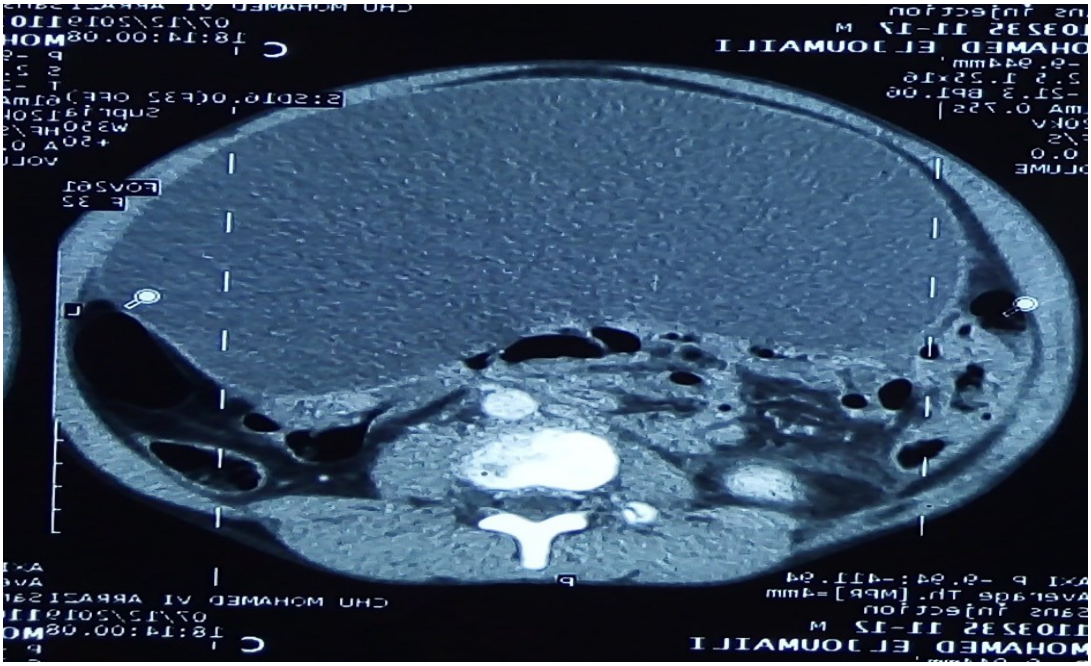
Corresponding Author:- Etoka E.C
Address:- Department of General Surgery, University Hospital Center Mohamed VI, Marrakech, Morroco.

well defined separation borders, pushing back the adjacent organs and comes into contact with the large retro-peritoneal vessels (Fig. 3), forward in intimate contact with the anterior abdominal wall and push back the transverse colon. The chest and cervical CT did not reveal any other mass, the diagnosis of benign cystic formation which may be compatible with a cystic lymphangioma, is retained. Surgical excision was the recourse to treatment. Thus, surgical excision (as complete and as conservative as possible) of this tumor is performed (figure). Pathological results reveal a 6 x 4cm fibro-membranous fragment with several limited cavity formations of a partially muscular endothelium confirming the diagnosis of cystic lymphangioma. The patient's progress is completely favorable one month after the operation: good general condition, disappearance of pain with an unremarkable abdominal ultrasound. The patient is informed of the risk of recurrence if the excision is not complete and must resume contact with his surgeon if symptoms recur.

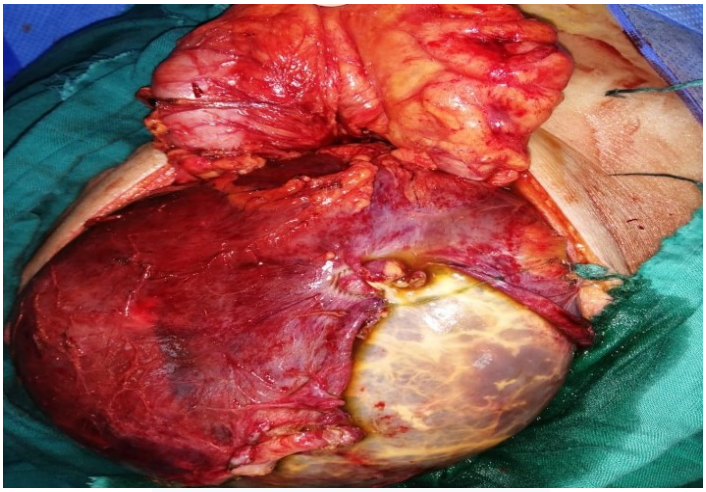
Iconography of the abdomen: photo provided



Iconography1:- Abdominal ultrasound reveals a voluminous abdominopelvic mass intraperitoneal median and para median bilateral with cystic content containing echogenic partitions of 22 cm, heights 10 * 10 cm



Iconography 2:- abdominal CT-scan presence of a voluminous intra-unilocular mesenteric cystic formation of roughly rounded shape with a thin wall of 22.4 * 12.7 cm extended over 21 cm.



Iconography 3:- of the preoperative.

Discussion:-

Cystic lymphangioma is a benign and rare tumor of the lymphatic vessels: its frequency in the mesentery is estimated at 1 / 100,000 in adults and 1 / 20,000 in children (1-2). Depending on how fast it grows (3), cystic lymphangioma can develop at any age, but it is seen in 60% of cases before the age of 5 (1, 3, 4). The physio pathogenesis of cystic lymphangioma remains unclear, but can however 2 theories:

Malformative theories of the lymphatic system

Cystic lymphangioma is thought to result from a disorder of embryogenesis with the appearance of a connection defect between a group of lymphatic chains and the venous system resulting in the isolation of lymphatic capillaries, causing dilation and the formation of multiple cysts (3,4).

Theory acquired

Cystic lymphangioma is thought to exit as a result of obstruction of the lymphatic vessels linked to a secondary cause (inflammation, trauma or degeneration) (3,5 and 6). This theory is increasingly abandoned (7). The usual locations are cervical or axillary (95% of cases), more rarely at the mediastinal or abdominal level (5-10% of cases). Regarding the abdominal region, the cystic lymphangioma accounts about 7% of cysts in adults. It preferentially affects the mesentery and the retro peritoneum, due to the great richness of the lymphatic structures (8). However, it can affect other intra-abdominal organs (spleen, pancreas, kidney, etc.). The non-specific and polymorphic clinical presentation of cystic lymphangioma is linked to the tumor volume, the variable location and the types of complications that it causes (mechanical / infectious / hemorrhagic) (1,2,4). Cystic lymphangioma can be revealed by the perception of an abdominal mass, abdominal pain (38% of cases), febrile state, hematemesis, volvulus, etc. Finally, there is a particularly rare clinical form, the cystic spread type mimicking peritoneal carcinoma, called peritoneal cystic lymphangiomatosis (3). According to Losanoff and Kjossev, a classification based on the morphotype of the lesion is used to optimize the surgical approach (5). The "type 1" is pedicled with a risk of torsion, volvulus. Its resection is easy. “Type 2” is sessile, less mobile and may require sacrifice. "Type 3" involves retro-peritoneal extension (sometimes affecting vital structures) making total removal of the LK impossible. "Type 4" corresponds to extensive multi-organ disease.

The discovery of an intra-abdominal cystic lesion requires careful investigation to rule out a malignant process. In many cases, non-invasive focusing is not sufficient to give an accurate diagnosis. Indeed, the differential diagnoses of intra-abdominal cysts to consider are numerous. Usually, the definitive diagnosis is made by histological analysis of the total resection specimen. In children, the diagnosis can be made on the basis of clinical practice and radiology in the event of cervico-facial involvement, which is characteristic (3). In other circumstances (rare localization), the diagnosis is not always obvious, but ultrasound is the useful examination initially and for follow-up. It is not irradiating for the child and sometimes allows antenatal screening (3). Cystic lymphangioma is characterized on ultrasound by a hollowed-out mass of cystic cavities with hypoechoic fluid content, of varying sizes and thin walls (hemodynamically inactive on Doppler). The CT scan is an excellent complementary diagnostic tool in adults. It shows a homogeneous tumor, hypodense, with fine partitions, not enhanced by contrast (3). The density of the

intracystic fluid may vary depending on the content, which may turn out to be serous, chylous or hemorrhagic (3-6). MRI is used only as a second intention, it allows a more precise study of the anatomical relationships of the lesion with the neighboring structures. More invasive techniques can also be used, such as a fine needle puncture which specifies the nature of the intracystic fluid and allows cytological examination to reveal the presence of lymphocytes. Nevertheless, diagnostic certainty is provided by the pathological analysis of the tumor.

Macroscopically, cystic lymphangioma can be single or polycystic, with oligomacrocystic, microcystic and mixed forms (3). Microscopically, three criteria are necessary for the diagnosis: 1) it is a cystic formation; 2) the septa are made up of a connective stroma with lymphoid tissue and smooth muscle; 3) the cyst is lined with a lymphatic-like endothelial coating (factor D2-40 positivity) demonstrating the vascular origin of the tumor (1,3,6,9). In case of accidental discovery, abstention from treatment with regular monitoring is recommended if the cystic lymphangioma is asymptomatic (1, 3). Spontaneous regression can be seen in 1.6 to 16% of cases (3). Surgical excision is the classic approach (3), because there is a high risk of the lesion progressing (increase in volume, appearance of complications) (7). It must be total to avoid recurrence as much as possible, by laparotomy or laparoscopy and the most conservative for the organs because of the benign nature of the lymphangioma (3). During the procedure, attention should be paid to lymphostasis in order to limit postoperative complications such as lymphocele or the development of chylous ascites (3). Despite this, there is a recurrence rate of 40% after incomplete resection and 17% after macroscopically complete resection (1, 3). An alternative treatment is aspiration of the contents of the cyst with or without injection of a sclerosing product (bleomycin, tissucol, OK-432 (picibanil), ethibloc (zeïne)). This treatment is interesting in the event of oligo-macro-cystic lesion and easily accessible. However, the recurrence rate is high and therefore, it is preferentially used symptomatically for unresectable lesions without extensive intestinal sacrifice (1).

In conclusion, Cystic lymphangioma is a benign tumor of lymphatic origin, the clinic of which is polymorphic. The diagnosis is suggested by imaging and confirmed by histology. Resection surgery is the only treatment to prevent local recurrence.

Bibliography:-

1. Mabrut JY, Grandjean JP, Henry L, et al.— Cystic lymphangiomas of the mesentery and mesocolon. Diagnostic and therapeutic care.. Ann Chir, 2002, 127, 343-349.
2. Prabhakaran K, Patankar JZ, Loh DLSK, et al.— Cystic lymphangioma of the mesentery causing intestinal obstruction. Singapore Med J, 2007, 48, 265-266.
3. Bezzola T, Bülher L, Chardot C, et al.— Surgical treatment of abdominal cystic lymphangioma in adults and children
4. . J Chir, 2008, 145, 238-243.
5. Rifki Jai S, Adraoui J, Khaiz D, et al.— Le lymphangiome kystique rétropéritonéale. Prog Urol, 2004, 14, 548-550.
6. Losanoff JE, Kjossev KT.— Mesenteric cystic lymphangioma : unusual cause of intraabdominal catastrophe in an adult. Int J Clin Pract, 2005, 59, 986-987.
7. Weeda VB, Booij 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.
8. Chegdali O, Guedira M, Benaddi L, et al.— Lymphangiome kystique abdominal de l'adulte. Acta Endosc, 2004, 34, 245-248.
9. Kably A, Moumen M, Raissouni N, et al.— Le lymphangiome kystique du mésentère et de l'épiploon. A propos de deux cas. Gynecol Obstet Fertil, 2003, 31,136-138.
10. Okamoto D, Ishigami K, Yoshimitsu K, et al.— Hemorrhagic mesenteric cystic lymphangioma presenting with acute lower abdominal pain : the diagnostic clues on MR imaging. Emerg Radiol, 2009, 16, 327-330.