International Journal of Surgery Case Reports 98 (2022) 107606

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

A rare incidence of retroperitoneal Cystic Lymphangioma in a 45-year-old female - A Case Report

Jack Shaheen, MD [a], Omar Al Laham, MBBS [c,*], Fareed Atia, MD [a], Dana Ibrahim, MD [b], Bashir Hokouk, MD [c,d]

[a] Department of Surgery, Al Assad University Hospital, Damascus, Syria
[b] Faculty of Medicine, Tishreen University, Lattakia, Syria
[c] Faculty of Medicine, Damascus University, Damascus, Syria
[d] Department of Surgery, Al_Abbasieen Hospital, Damascus, Syria

ARTICLE INFO

Keywords:
Case Report
Cystic Lymphangioma
Cystic Hygroma
Retroperitoneal Mass
Vascular Oncology
Incidental Diagnosis

ABSTRACT

Introduction and importance: Cystic lymphangioma is an extremely rare benign vascular neoplasm of mesodermal origin, arising from lymphatic vessels and occurring principally in male children. The retroperitoneum is the rarest site, accounting for <1 % of all types of lymphangioma. The incidence of cystic lymphangioma is unknown due to the scarcity of published data.

Case presentation: A 45-year-old female presented to our hospital complaining of vague abdominal pain and intermittent episodes of vomiting over four months. Preoperative imaging via ultrasound and Computed Tomography revealed a well-demarcated retroperitoneal cystic mass between the right kidney and the liver.

Clinical discussion: The mass was excised laparoscopically. Histopathological examination confirmed a cystic hygroma (a subtype of cystic lymphangioma).

Conclusion: Cystic lymphangioma is rare, often misdiagnosed and may present with vague symptoms. Complete resection may be feasible and can be curative.

1. Introduction

Cystic lymphangioma is a benign and extremely rare vascular neoplasia. It is of mesodermal origin and is evident to arise from the body's lymphatic vessel system. They occur most predominantly in the neck region followed by the axillae with 75 % and 20 % respectively. Furthermore, merely 5 % of them occur in different sites inside the abdomen. These sites include the gastrointestinal tract, liver, mesentery, and spleen. However, the rarest of those sites is the retroperitoneum as retroperitoneal lymphangiomas comprise <1 % of all types of lymphangiomas [1,2].

With regards to clinical presentation, cystic lymphangiomas mostly occur without any or pathognomonic symptoms. These lesions are chiefly discovered incidentally either during surgery or while undergoing radiological assessment for a different clinical complaint [3].

In the rare instances where they cause symptoms, these complaints vary across the spectrum (i.e., general weakness, abdominal discomfort or pain, weight changes, episodes of fever, frank hematuria, infection, hemorrhage, or rupture) [4,5].

Preoperative clinical radiological assessments are largely inefficient to definitively differentiate cystic lymphangiomas from other different classes of cystic masses. Surgical intervention or diagnostic laparoscopy is the most likely required methods used to establish a firm diagnosis and to properly treat the condition [6].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [7].

2. Presentation of case

2.1. Patient information

We hereby demonstrate the case of a 45-year-old previously healthy Middle Eastern female patient who presented to the general surgery clinic at our tertiary hospital with the chief complaint of vague

Abbreviations: BMI, Body Mass Index; CT, Computed Tomography; IV, Intravenous; H&E, Hematoxylin and Eosin.

* Corresponding author at: Faculty of Medicine, Damascus University, Damascus, Syria.
E-mail addresses: jsbayern212@gmail.com (J. Shaheen), 3omar92@gmail.com (O. Al Laham), dr.fareedatia@gmail.com (F. Atia), danaibrahim.ej@gmail.com (D. Ibrahim), Mhmoodhokok@hotmail.com (B. Hokouk).

https://doi.org/10.1016/j.ijscr.2022.107606
Received 7 August 2022; Received in revised form 29 August 2022; Accepted 3 September 2022
Available online 7 September 2022

abdominal pain. The pain started 4 months prior to admission. It was intermittent, ill-defined, estimated to measure 06/10 on the pain severity scale according to the patient, was unrelated to food, was not aggravated by any specific factors, and was mildly relieved by analgesics. Her symptoms were accompanied by intermittent bouts of non-biliary vomitus. It was also unrelated to food and did not include any visible blood, pus or mucous.

Systematic review for her was negative. She didn't report any cardinal or B-Symptoms. She also denied any presence of genitourinary symptoms.

The patient negated any history of trauma, infections, or radio-/chemotherapy. Her surgical history involved 5 previous cesarean sections. Her family, drug, allergic, and psychosocial histories were negative. Her Body Mass Index (BMI) was 26 kg/m^2.

2.2. Clinical findings

Physical examination was unrevealing. No pain was elicited, and no palpable masses were felt. Furthermore, there weren't any palpable lymph nodes. *Laboratory investigations* results were all normal.

2.3. Diagnostic assessment

At ultrasonography, we demonstrated an anechoic cystic formation with well-defined borders. It measured about (8.5 × 5.2 cm) and was situated between the right kidney and the liver. The remaining ultrasonographic findings of the abdomen were normal. For further clarification, we opted to perform a Computed Tomography (CT) scan. It showed a well-demarcated isointense cystic formation. It measured approximately (4 × 3.5 cm) and seemed to be attached to the right adrenal gland. There was no lymphadenopathy (Fig. 1A-B). The rest of the vital organs and structures were radiologically free of neoplastic involvement. Attempts to either biopsy the lesion or aspirate it via fine needle were out of the question because of the highly cystic appearance of the mass. Based on the radiological study, our differential diagnoses included renal cyst, adrenal mass, pancreatic pseudocyst, abscess, ovarian cyst, and cystic teratoma. Preoperatively, the patient had an Intravenous (IV) access installed. In addition, she was administered suitable IV preoperative antibiotics in the form of a single-dose Cefazolin injection. Blood sampling for crossmatch was done as part of the routine preoperative protocol. Fortunately, no remarkable challenges or obstacles were faced.

2.4. Therapeutic intervention

A surgical intervention was decided. The surgery was successfully achieved at the same tertiary hospital. It was performed by a general surgery consultant with 20 years of experience and was done under general anesthesia without any perioperative complications. At laparoscopy, it was achieved via 4 trocar incisions. Intraoperatively, the cystic mass was found between the right kidney and the liver. However, it was unattached to any surrounding structures. It had well-defined borders and its contents appeared to be chylous fluid. It was completely resected after meticulous isolation from its surroundings (Fig. 2A-B-C). The resected lesion was immediately sent for histopathological analysis. Postoperative histopathological analysis via Hematoxylin and Eosin (H&E) revealed a benign vascular neoplasm conformant with a cystic hygroma which is a subtype of cystic lymphangiomas. No atypical changes were seen (Fig. 3A-B-C).The patient underwent full postoperative recovery, and she was discharged from the hospital within 3 days of her operation. Regular wound dressings by a physician were applied and proper postoperative antibiotics were administered.

She has been surveilled for two months thus far. Clinical assessments at the general surgery included physical examination and radiological evaluation via ultrasound. All of which were yielded normal results.

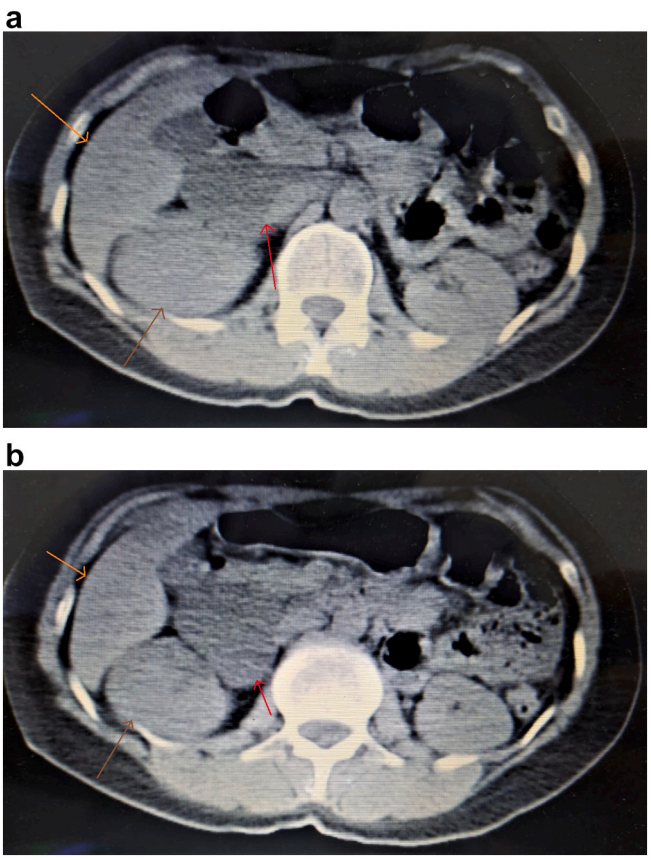

Fig. 1. (A-B): Preoperative CT It showed a well-demarcated isointense cystic formation. It measured approximately (4 × 3.5 cm) and seemed to be attached to the right adrenal gland. Red Arrow identifies the cystic mass. Brown Arrow identifies the right kidney. Orange Arrow identifies the liver. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3. Discussion

Koch was the scientist who was the first to demarcate the term "Lymphangioma" in 1913 [8,9]. The origin of the formation of a lymphangioma could be traced back to the period between 14 and 20 weeks of gestation. This is when it was perceived that the lymphatic vessels, which do not successfully develop to connect with the normal lymphatic drainage system, are the progenitors that later form a lymphangioma. We can therefore define a lymphangioma as a benign abnormal proliferation of the previously mentioned lymphatic tissue [10]. Therefore, we consider a lymphangioma to be a congenital neoplasm. Postpartum, these abnormal lymphatic tissues can get irreversibly dilated due to the collection of undrained lymphatic fluid [11].

There other etiological theories behind the development of a lymphangioma. These include inflammation, lymph node degeneration or obstruction, trauma, or fibrosis [12].

Wegner was the first to histologically classify lymphangiomas into 3 main types in 1877. The first type was lymphangioma simplex. These do not usually occur within the abdomen. The second type was cavernous lymphangioma. It is more common than the first type, but it is still rare to arise within the abdomen. However, it has the potential to undergo malignant transformation at a certain point. The third and final type was cystic lymphangioma. This one is almost always benign. With regards to retroperitoneal lymphangiomas, most of the documented cases are of the cystic type [13].

Retroperitoneal lymphangiomas are profoundly rare as their precise incidence rate isn't known to this day. They are found to occur largely in the pediatric population, specifically in male children where the male to

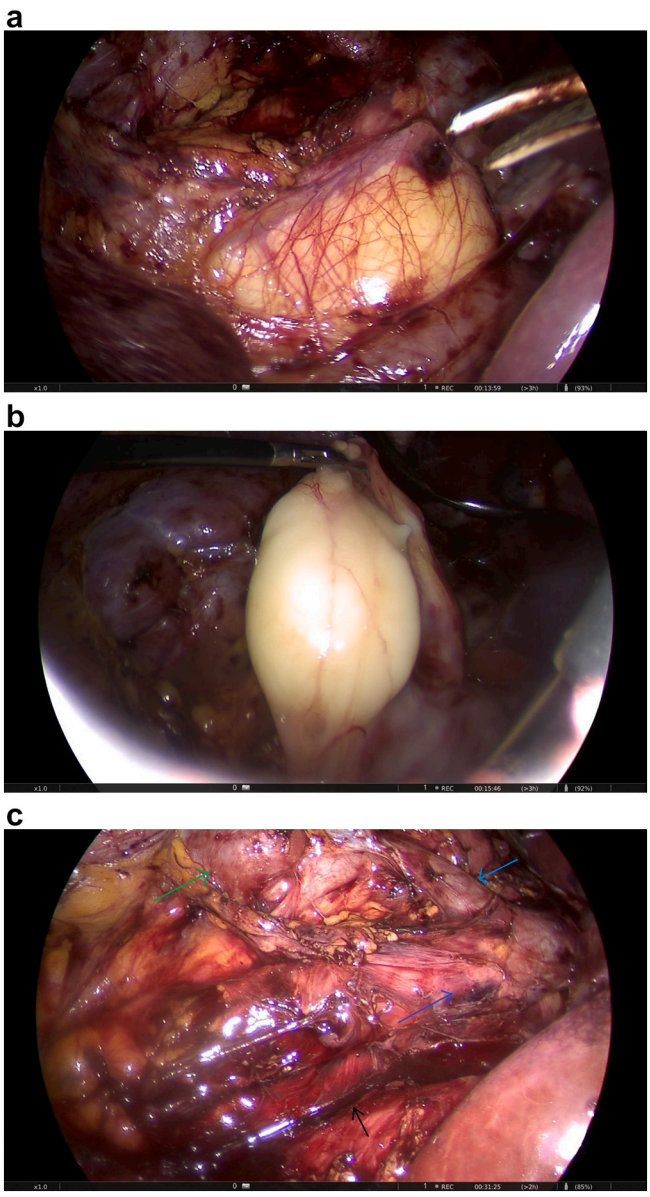

Fig. 2. A: Intraoperative image prior to resection revealing the cystic mass.
B: Intraoperative image revealing the cystic mass while isolating it from its surroundings.
C: Intraoperative image after resection of the mass has taken place. Black arrow points towards the Abdominal Aorta. Blue Arrow points towards the Inferior Vena Cava. Turquoise Arrow points towards the right renal vein. Green Arrow points towards the right kidney. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

female incidence ratio is 5:2 [14,15].

Possible etiologies behind their occurrence in the adult population remain ill-defined. Proposed factors include surgery, trauma to the abdomen, inflammations, and radiotherapy [5,16,17].

Retroperitoneal lymphangiomas have variable presentations and lack any specific or pathognomonic patient complaints [18].

The chief clinical course for retroperitoneal lymphangiomas is silent as they mainly lack any presenting symptoms. Nonetheless, in some cases they manifest as a complaint of a palpable abdominal bulging. This results in a great level of confusion for physicians as they cannot accurately specify whether such a mass is a retroperitoneal lymphangioma or a distinct mass arising from vital intraabdominal organs such as the kidneys, liver, or pancreas [1].

With regards to cystic retroperitoneal lymphangiomas, most cases occur as incidental clinical findings. Nevertheless, presenting symptoms arise when the cystic mass has proliferated and started to compress nearby structures such as the ureters or different small intestinal segments [3]. Furthermore, patients with retroperitoneal lymphangiomas could present with an array of nonspecific symptoms, such as weight changes, episodes of vague abdominal pain, general weakness, or fever. Other potentially fatal manifestations involve rupture of the cyst, sepsis, or bleeding [4,5].

With regards to preoperative diagnostic modalities, radiological assessment via ultrasound or CT could aid in guiding the surgeon towards the next step. By performing ultrasound, lymphangiomas appear as anechoic multilocular cystic masses. Whereas through CT scan with IV contrast material, the cyst may be better visualized and could reveal its septa and fluid contents. When the cystic contents are comprised of chyle, the fluid appears homogenous with low attenuation values on CT scan [19].

However, diagnosing retroperitoneal lymphangiomas remains a challenge. Such a diagnosis is sometimes solely made after performing laparoscopy or laparotomy and is definitively established after thorough histopathological analysis of the excised specimens [1].

Histologically, a lymphangioma is diagnosed relying on a set of preexisting conditions. These include a cystic lesion with or without an endothelial lining, a well-circumscribed wall having accumulations of lymphoid tissue, a stroma consistent of collagen and fibrous tissue [13].

With regards to possible differential diagnoses for cystic retroperitoneal lymphangioma, the possibilities include of abscesses, pancreatic adenoma, ovarian cysts, pancreatic pseudocysts, sarcomas, cystic teratoma, and pancreatic neoplasia [3,20,21].

Possible life-threatening complications of this lesion include volvulus, sepsis, cystic leaking or rupture, and hemorrhage within the cyst [22].

Complete and thorough surgical resection of the cystic mass remains the treatment modality of choice for cystic lymphangiomas [23]. On the other hand, there are modalities of noninvasive therapeutic approaches, such as cystic contents aspiration and the injection of sclerosing material [24].

Finally, emphasis must be made that the surgical resection of the lesion is of utmost importance. 10 % of cystic lymphangiomas tend to recur as a direct result of inadequate surgical removal [25]. Therefore, meticulous surgical removal of the lesion is vital to avoid potential complications, such as infections, continued growth of the mass, hemorrhage, and cystic rupture [25,26].

4. Conclusion

Cystic lymphangioma is a rare but benign lymphatic vascular neoplasm, especially when retroperitoneal. It mostly presents with nonspecific symptoms. The diagnosis is sometimes only reached histologically, after utter resection is performed. This approach should be curative and in our case was possible laparoscopically.

Abbreviations

BMI	Body Mass Index
CT	Computed Tomography
IV	Intravenous
H&E	Hematoxylin and Eosin

Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the Data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

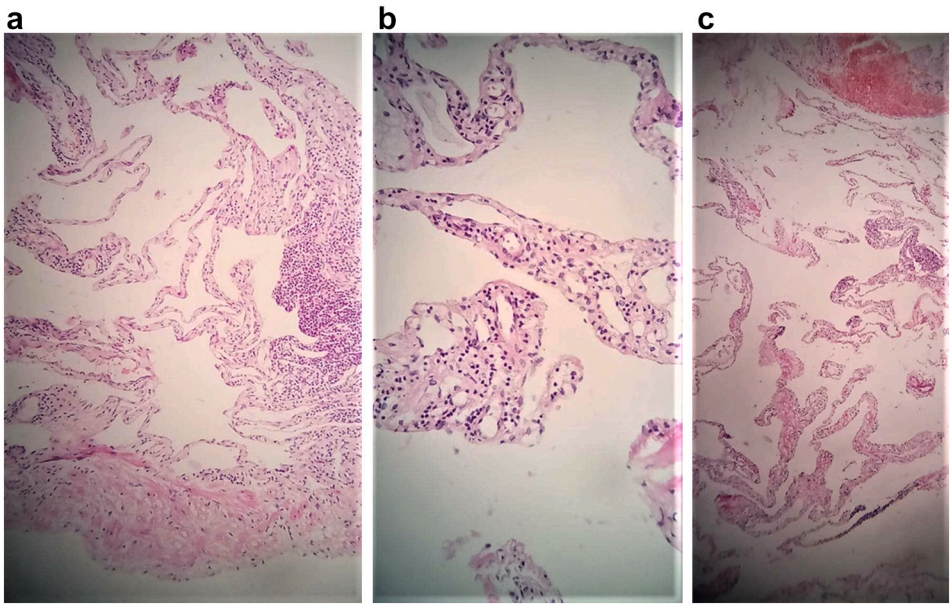

Fig. 3. (A-B-C): Postoperative histopathological analysis via H&E staining revealed well circumscribed thin-walled cystic cavities with variable sizes. It also shows lymphoid tissue and stroma consistent with a benign vascular neoplasm conformant with a Cystic Hygroma which is a subtype of Cystic Lymphangiomas. No atypical changes are evident.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

This study is exempt from ethical approval in our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT authorship contribution statement

OA: Conceptualization, resources, who wrote, original drafted, edited, visualized, validated, and literature reviewed the manuscript.

JS: Supervision, project administration, resources, and review of the manuscript.

FA, DI: Review of the manuscript and review of the literature.

BH: General Surgery consultant who performed and supervised the operations, supervision, project administration, and review of the manuscript.

OA: The corresponding author who submitted the paper for publication.

All authors read and approved the final manuscript.

Registration of research studies

Not applicable in our case.

Guarantor

Omar Al Laham.

Declaration of competing interest

None.

Acknowledgements

Maher Nassar's Histopathology laboratory, Damascus, Syria. For their role in the histopathological diagnosis of the excised specimens.

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