Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

A rare occurrence of an incidental primary intra-abdominal Cystic Lymphangioma in a Middle Eastern adult female: A case report

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Introduction and Importance: Cystic Lymphangiomas (CLs) are rare vascular anomalies of benign nature. The etiology remains a subject of controversy, but they are considered to arise due to anomalies that occur during the normal embryogenic developmental process of lymphatic vessels. They have an estimated incidence rate of merely 1 in every 20,000–250,000 individuals. Since they are mainly known as a disease of the pediatric population, accurate epidemiological rates of CLs, especially in adults, are still unidentified due to the scarcity of published data. Documentation is pivotal to collect further information about them to establish timely diagnoses and minimize the potentially high patient morbidity.

Case Presentation: The authors hereby demonstrate the case of a 46-year-old lady who presented to the outpatient general surgery clinic at our university hospital complaining of chronic right hypochondriac abdominal pain. Investigative radiological imaging marked a cystic formation with well-defined borders and homogenous content extending from the lower pole of the right kidney to the lower hepatic border.

Clinical Discussion: Surgical intervention was performed by completely resecting the lesion in question. Directly afterward, histopathological analysis was achieved and established the diagnosis of a CL. Conclusion: CL is a rare neoplasm with ill-defined symptoms and ambiguous clinical presentations. They remain poorly studied due to their rarity and lack of sufficient data in the published literature. This magnifies the importance of clinical awareness and time-efficient surgical intervention. Documenting these cases aids in identifying their subsequent etiological origins, disease-specific risk factors, clinical course, and yields proposals of novel therapeutic approaches.

Keywords: Case Report, Abdominal Mass, Abdominal Surgery, Cystic Lymphangioma, Intra-abdominal Lymphangioma, Vascular Neoplasia

Introduction

Cystic lymphangiomas (CLs) are benign lymphangiomas and are defined in the literature as aberrant benign maldevelopment of lymphatic vessels that classically arise in the anatomical distribution of the head and neck of neonates and grown-ups[1]. The main pathophysiological description behind their development is via an anomaly during the embryonic synthesis phase of lymphatic vessels, where said vessels do not ultimately succeed in forming the chief body lymphatic system[2,3]. Regarding CLs, they could potentially originate in multiple distinct anatomical sites. However, the predominant portion of them arises in the axial and cervical sites of the body. Moreover, CL that chooses the abdomen as a site of origin are rare and comprise even less than 5% of all known cases of CLs[4]. While looking out for symptoms that patients suffering from abdominal CL complain from, unfortunately, they are almost always nonexistent. In the rare cases in which symptoms do occur, they vary from non-specific abdominal pain, nausea, and episodes of vomitus[5]. Diagnosis of CL, especially abdominal CL, could be a difficult endeavor with classical methods like bedside ultrasound. However, the continuous evolution of complex imaging

HIGHLIGHTS

- Cystic Lymphangiomas are immensely rare and nonmalignant vascular anomalies of the lymphatic system.
- They are mainly thin-walled cystic formations, and merely 5% of them originate from sites within the abdomen.
- They are mostly asymptomatic but when symptoms occur, they are ill-defined and lead to misdiagnoses.
- A definitive diagnosis is reached because of meticulous histopathological analysis of the resected specimen.
- The cornerstone for treatment is utter surgical excision which leaves a minimal potential for lesion recurrence.

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Annals of Medicine & Surgery (2023) 85:231–235

Received 29 October 2022; Accepted 25 December 2022

Published online 18 January 2023

http://dx.doi.org/10.1097/MS9.0000000000000200

modalities, such as computed tomography (CT) and magnetic resonance imaging (MRI), has made diagnosing them easier[6,7]. Therefore, timely detection and diagnosis of CL are crucial to enable physicians to perform time-efficient and definitive surgical interventions to resect this vascular neoplasia[8]. The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines[9].

Presentation of case

Patient information

We hereby demonstrate the rare case of a 46-year-old Middle Eastern female patient who has been a known case of hypertension for 14 years before admission. She presented to the outpatient clinic at our university hospital with the chief complaint of recently aggravated symptoms of right hypochondriac vague abdominal pain. Her complaint began 3 years before admission. It was sudden, intermittent, poorly defined, limited to her right hypochondriac region, had no radiation to any other site, and was valued to equal 04/10 on her pain severity scale. The patient elaborated that it was unrelated to the ingestion of solids or liquids, did not get provoked by any triggers, and was somewhat relieved by over-the-counter analgesic medications. The pain was not accompanied by any other symptoms, as her systematic review was negative. Furthermore, symptoms that constituted red flags for tumor formation were all denied by the patient. These included unintentional loss of weight, poor appetite, night sweats, and general fatigue. Since her symptoms persisted and did not subside, but rather increased over the course of the few days prior to her visit, she decided to visit our clinic. Her past medical history only includes well-controlled hypertension and Meniere's disease for 14 and 8 years, respectively. Her surgical history included two previous operations for bilateral ovarian cysts removal via open surgery 18 years ago, a cesarian section 28 years ago, an ectopic pregnancy 28 years ago, and an open appendectomy. Her drug history includes cetirizine and bisoprolol to control her dizziness and elevated blood pressure, respectively. She is an active smoker with a 20-pack-year smoking history but does not consume alcohol. Her family history for a previous similar incidence and neoplastic occurrences is negative. Lastly, her allergic history was negative, and her body mass index was 27 kg/m^2.

Clinical findings

Physical examination yielded negative findings as no skin changes or bulging across the overlying skin of the abdomen were noted. Moreover, the lesion was not palpated in either superficial or deep palpation. In addition, no pain was elicited during the examination. Furthermore, no abnormal sounds or bruits were heard during auscultation. Preoperative laboratory investigations results were within normal.

Diagnostic assessment

We began our preoperative radiological assessment with an abdominal ultrasound, and it revealed a well-demarcated anechoic cystic lesion with well-defined borders. It measured ~4 × 11 cm and extended from the lower pole of the right kidney to the lower hepatic border. It did not appear to be attached to any surrounding organs. The remaining ultrasonographic findings of the abdomen and pelvis were all normal. Finally, no regional lymphadenopathy was marked. We needed further insight into the lesion, hence a contrast-enhanced CT scan was performed. It revealed a demarcated isointense cystic lesion with homogenous content which was measured (4 × 6 × 14 cm) and extended from the lower pole of the right kidney until the lower hepatic border. There was no surrounding lymphadenopathy (Fig. 1a, b). The remaining fields, like the chest, abdomen, and pelvis were seen to be radiologically free of neoplastic involvement (Fig. 2a–c). Based on the previous results of our radiological assessment, our

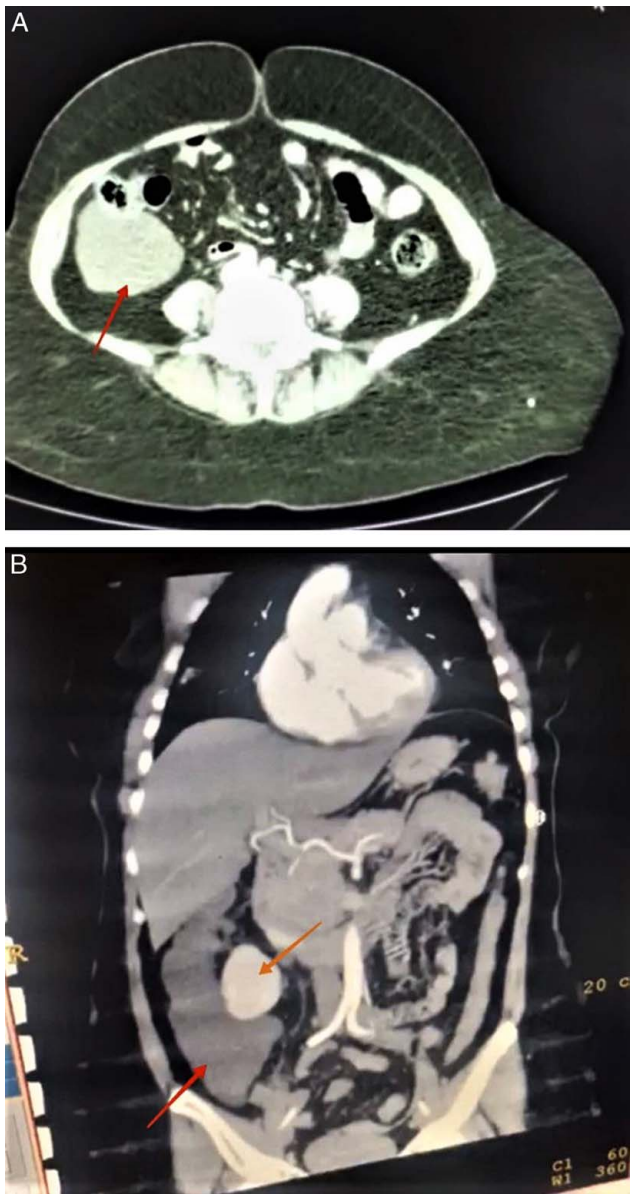

Figure 1. (a) Preoperative computed tomography image with contrast in cross-sectional view revealing a well-demarcated cystic formation with homogenous content overlapping the ascending colon. It is labeled by the red arrow and measures 4 × 6 × 14 cm. It extends upward toward the lower hepatic border and the lateral border of the right kidney. (b) Preoperative computed tomography image with contrast in coronal view revealing a well-demarcated cystic formation (brown arrow) with homogenous content overlapping the ascending colon (red arrow). It measures 4 × 6 × 14 cm. It extends upward toward the lower hepatic border and the lateral border of the right kidney.

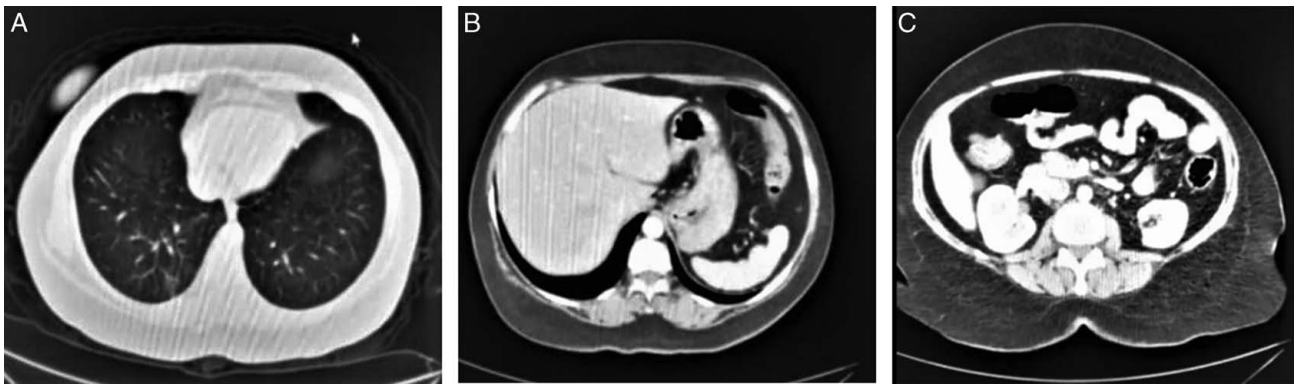

Figure 2. (a–c) Preoperative computed tomography image in cross-sectional view revealing clear lung, abdominal, and pelvic regions. No metastasis is visualized.

differential diagnoses revolved around a variety of possibilities, such as renal cysts, an ovarian mass, a lipoma, CL, a teratoma, and a fibroma. We opted to perform a complete surgical resection of the lesion. Therefore, to prepare the patient for surgery, she had an intravenous access line fixed and was administered intravenous prophylactic antibiotics. Finally, blood sampling for grouping and crossmatch was performed. Fortunately, no remarkable challenges or obstacles were faced during the perioperative phase.

Therapeutic intervention

Surgical excision of the lesion via midline abdominal incision was the intervention of choice. The surgery had successfully taken place at our university hospital. It was achieved by a general surgery professor and a general surgery first assistant with 35 and 5 years of experience, respectively. The therapeutic intervention was performed under general anesthesia with no perioperative complications. Intraoperatively, the lesion was found overlapping the ascending colon. However, it did not appear to originate from any surrounding organs. Several aggregating cystic formations with well-defined borders with serous contents were visualized (Fig. 3) and were utterly excised after meticulous isolation from their soft tissue surroundings (Fig. 4). The resected specimens were directly sent to our specialized pathology laboratory for complete histopathological analysis. Postoperative final histopathological analysis through hematoxylin and eosin staining demonstrated a type of benign vascular neoplasia corresponding to a CL. No atypical variations nor malignant transformations were noted (Fig. 5a, b). The patient underwent a complete and uneventful postsurgical recovery, and she was discharged to the outpatient setting within 4 days of her surgery. Consistent wound dressings by a medical professional were applied, and suitable postoperative antibiotics were prescribed accordingly. She is still

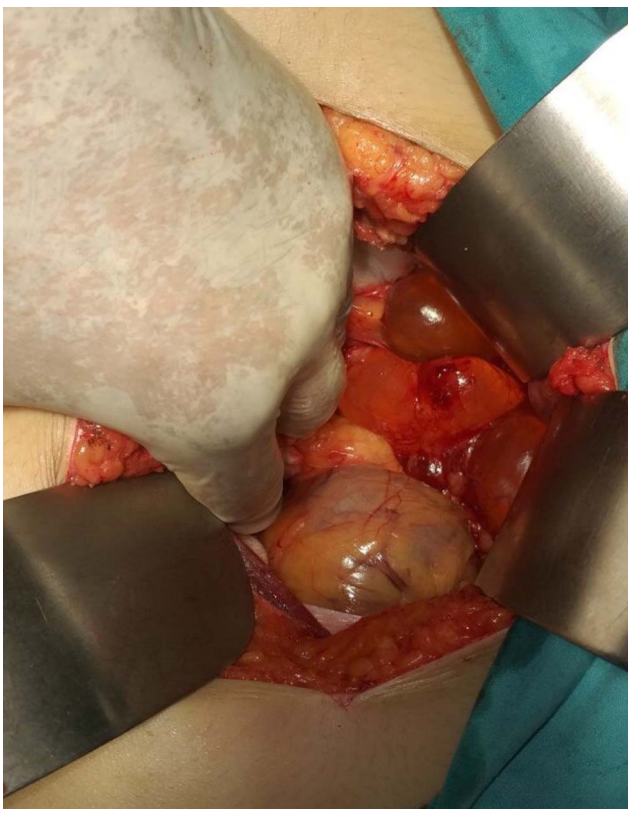

Figure 3. Intraoperative image revealing the cystic mass.

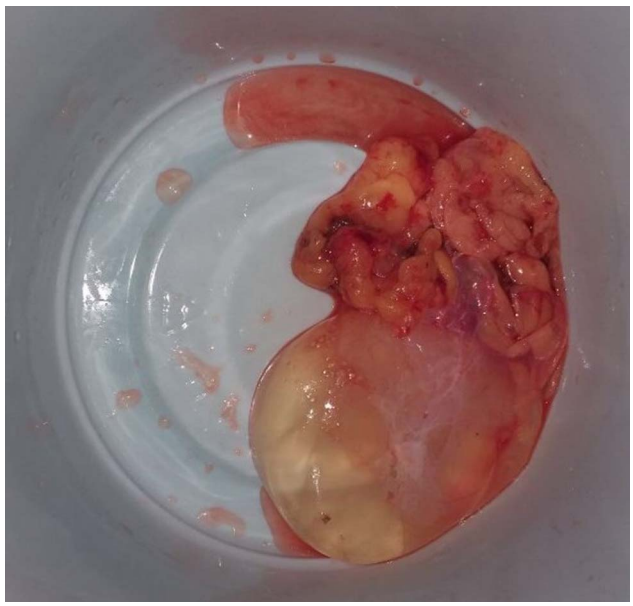

Figure 4. Postoperative image depicting the excised lesion.

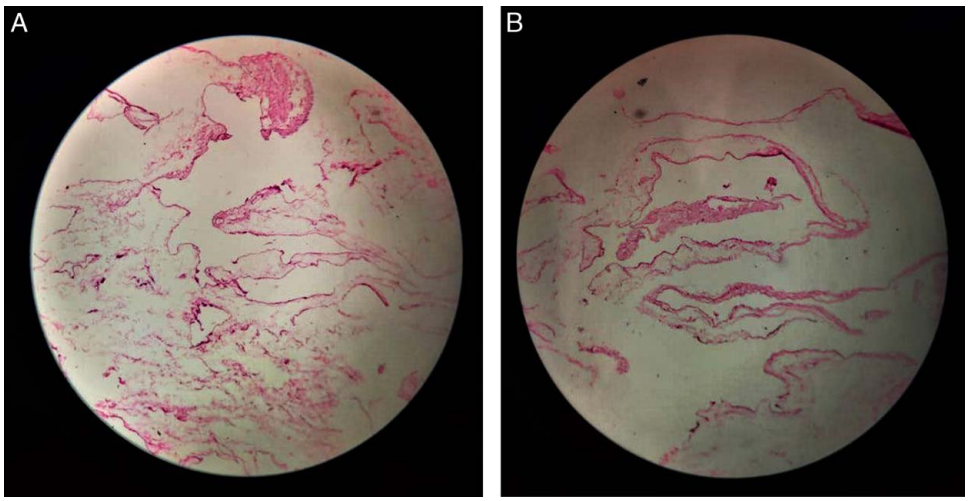

Figure 5. (a, b) Histopathological analysis via hematoxylin and eosin staining. They are marked by large lymphatic channels in loose connective tissue stroma. In addition to the presence of focally disorganized smooth muscle cells with peripheral lymphoid aggregates. No atypia is seen.

under postoperative patient surveillance for 7 months thus far. In addition, regular clinical evaluations at the general surgery clinic were done. These included proper physical examination and radiological assessment via ultrasound. Their subsequent results yielded normal results as no lesion recurrence was demonstrated.

Discussion

Koch coined the scientific term 'Lymphangioma' during his research in 1913[10,11]. Lymphangiomas are explicitly defined in the published literature as rare lymphatic vessel developmental maldevelopments that are considered congenital in origin[12]. Furthermore, lymphangiomas which tend to originate in various sites within the abdomen are rare occurrences. They have a plummeting incidence rate of merely 1 in every 20,000–250,000 individuals[13,14]. Furthermore, CL which arise from within the abdomen are extremely rare and comprise even less than 5% of all known cases of CLs[4]. Researchers have not yet precisely identified the responsible etiology for CL. Several possible factors have been proposed, such as occlusion of lymphatic vessels, various infections, and surgical interventions[15,16]. In 1877, Wegner first set the now-known classification system for lymphangiomas. He divided them into three distinct categories. CLs are one of those three subtypes. As a differentiating quality from the other two types, CL – according to Wegner – is the one that could originate from within the abdomen and is almost always benign[17]. Patients suffering from a lymphangioma originating in the abdomen have vastly variable clinical presentations. This is also highly reliant on the precise anatomic site from which the CL has grown. Nonetheless, those patients mostly have an uneventful and otherwise silent clinical course. In the instances where symptoms manifest, they mainly include episodes of vomitus, intestinal obstruction, nausea, and loss of body weight[18–20]. Preoperative diagnostic modalities for intra-abdominal CL are mainly comprised of ultrasound, CT, and MRI. Nevertheless, CT and MRI hold higher regard in being able to preoperatively diagnose CL or otherwise guide physicians to suspect its occurrence. In addition, they can also provide more key information about the lesion's site, size, components, and relationship to nearby vital structures. These advantages enable medical care providers to effectively opt for a timely surgical intervention[12]. On the other hand, preoperatively diagnosing CL, especially abdominal ones, remain a difficult endeavor to reach. The definitive diagnosis is often only reached after the final histopathological analysis of the surgically removed lesion has been made. This is mostly done via hematoxylin and eosin staining[21]. Treatment options for this rare anomaly vary. Invasive methods naturally include complete surgical resection of the lesion, whereas minimally invasive methods include radiotherapy/chemotherapy and the use of sclerosing agents[22,23]. On the other hand, utter surgical excision of the lesion remains the modality of choice for treating CL originating from within the abdomen. This, in turn, results in outstanding prognoses for such patients[24]. However, we must not be negligible toward the devastating postsurgical clinical consequences of intra-abdominal CL. These involve hemorrhage, infections and sepsis, and vascular injury[25]. In conclusion, we must highlight that there is a 10% risk of recurrence for CL[26]. Therefore, we must adequately follow up with our patients to ensure no disease recurrence takes place.

Conclusion

It is rare to witness a case of intra-abdominal CL. It is even rarer for this pathology to present in this age group. We must remember that it is an illness with poorly understood etiology, vague symptoms, and misleading clinical courses. Our patient kept suffering from vague symptoms for 3 years before she visited our clinic. Without clinical awareness and consideration of this disease, timely interventions will not be taken, and the resulting patient-related morbidity inevitably rises. We must document these rare incidences to aid in the conduction of informative epidemiological research which identifies disease-specific statistical information. This improves physicians' capacities to diagnose and manage this rare vascular anomaly appropriately and preemptively.

Ethics approval

This study is exempt from ethical approval in our institution.

Patient consent

Written informed consent was obtained from the patient for the 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.

Sources of funding

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

Author contributions

O.A.: Conceptualization, resources, who wrote, originally drafted, edited, visualized, validated, literature reviewed the manuscript, and the corresponding author who submitted the paper for publication.

G.A., M.A., and E.A.: Supervision, visualization, validation, resources, literature review, and review of the manuscript.

O.B.: First surgical assistant in the operation, supervision, project administration, validation, resources, and review of the manuscript.

A.A.: General surgery specialist who performed and supervised the operation, in addition to supervision, project administration, and review of the manuscript.

All authors read and approved the final manuscript.

Conflicts of interest disclosure

The authors declare that they have no competing interests

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

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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's computer-based in-house system. Data are available from the corresponding author upon reasonable request.

Acknowledgements

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Provenance and peer review

Not commissioned, externally peer-reviewed.

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