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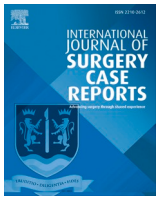

Case report

Mesenteric cystic lymphangioma misdiagnosed as ovarian cyst in a 63-year-old female: A case report and review of literature

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ABSTRACT

Introduction: Cystic lymphangioma is rare benign tumor that results from a lymphatic system malformation. The mesenteric location is even more uncommon.
Case report: We report the case of a menopausal 63-year-old woman who presented with a persistent painful well-defined mass of the pelvis. On ultrasound and computed tomography, the mass appeared as thick-walled unilocular homogenous cyst in favor of an ovarian cystadenoma. During laparotomy, the misdiagnosis was confirmed as the tumor was found to be embedded in the mesentery of the ileum. Subsequent histopathological examination confirmed the benign cystic lymphangioma diagnosis.
Discussion: Mesenteric cystic lymphangioma is rare peritoneal tumor of the adult. Clinically, it often masquerades as other abdominopelvic masses like ovarian cysts. Differential diagnosis is often challenging because of the overlapping clinical abdominal presentation and radiological features. Histopathological is the gold standard in diagnosing mesenteric cystic lymphangioma. Surgery is the mainstay treatment, and the recurrence rate is low if negative surgical margins are achieved.
Conclusion: Mesenteric cystic lymphangioma often mimics more frequent and potentially malignant lesions. It is essential for surgeons to remain vigilant for the possibility of this diagnosis when evaluating abdominopelvic cystic masses.

1. Introduction

Cystic lymphangioma is a benign peritoneal tumor, primarily observed in children and rarely encountered in adults [1]. It results from a developmental malformation of the lymphatic system. It predominantly manifests in the facial and axillary regions, however its occurrence in the mesentery is rare, constituting less than 1 % of all abdominal cases. Usually asymptomatic, its diagnosis is challenging due to its diverse clinical presentations, as it often mimics other abdominopelvic masses [2]. Imaging techniques like ultrasound, CT scan, and MRI are essential in localizing abdominal cystic lymphangioma and analyzing its features and extent. Nonetheless, final diagnosis relies on intraoperative findings and histopathological examination. Management options range from non-surgical approaches like sclerotherapy to surgical intervention, involving a large yet conservative resection [3].

This is the case of a mesenteric cystic lymphangioma diagnosed in a 63-year-old woman. The patient was indicated for laparotomy after misleading imaging diagnosis of serous cystadenoma. The diagnosis of mesenteric cystic lymphangioma was incidentally made intra-operatively.

This study aims to draw attention to this rare histological entity and emphasize the role of thorough radiological assessment of all abdominopelvic masses.

2. Case report

A 63-year-old patient, with no history of comorbidities, G5P5 (5 vaginal deliveries), menopausal for 15 years, presented to our department with severe intermittent pelvic pain evolving over the past 2 months. She had no metrorrhagia or digestive symptoms. Abdominal

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palpation coupled with vaginal examination revealed a well-defined, mobile, tender, right suprapubic mass. Transvaginal ultrasound showed a normal uterus with a purely anechoic, thickened-walled cystic lesion in the left parametrium space, measuring 59 × 53 mm without Doppler flow, suggestive of an ovarian cyst. The right ovary had a small anechoic cyst measuring 25 × 22 mm (Fig. 1).

Abdominal pelvic CT Scan revealed cystic formation above and left to the uterus measuring 66 × 50 mm, rounded with a thick and regular wall, and slightly dense liquid content, but homogeneous, without septations or vegetations, showing mild enhancement after contrast injection (Fig. 2).

Biologically, tumor markers were negative, including CA 19-9 at 2.06 IU/ml and CA 125 at 7.6 IU/ml. Given the age of the patient, the radiological findings and the biological markers levels, the diagnosis of ovarian serous cystadenoma was considered, and the patient was planned for surgery.

Laparotomy using Pfannenstiel incision was performed. During surgery, a large yellowish cystic mass of 70 × 60 mm was found arising from the mesentery of the ileum, 70 cm from the last ileal loop. The cyst was close to the ileal wall and was therefore resected with 25 cm of ileum (Fig. 3). A termino-terminal anastomosis was performed. Additionally, the left ovary and the uterus appeared normal with a small serous cyst in the right ovary, so a right adnexectomy was also performed.

Histopathological examination of the mesenteric cystic tumor revealed fibroadipose tissue with numerous lymphatic vascular formations, consistent with a benign lymphangioma. The ileum was normal. The ovarian cyst was a simple serous cyst.

The postoperative course was uneventful with no signs of intestinal anastomotic failure. At the 1-month follow-up, the patient was completely relieved of pain. After 1 year, there were no signs of recurrence.

3. Discussion

Cystic lymphangioma is rare benign tumor that is observed mostly in children, rarely in adult patients. It occurs most commonly in the cervical (75 %) and axial (20 %) region, with less than 5 % in the abdominal region [4]. The mesenteric localization is even more uncommon, accounting for less than 1 % [5].

Cystic lymphangioma (CL) represents a complex malformation of the lymphatic system, the precise pathophysiology of which remains unclear. The prevailing hypothesis suggests an embryological anomaly characterized by aberrant connections between lymphatic channels and the venous system. This anomaly likely arises during the developmental stages, leading to the formation of cystic structures within the lymphatic vessels. Despite ongoing research, the exact mechanisms of this malformation remain incompletely understood [6].

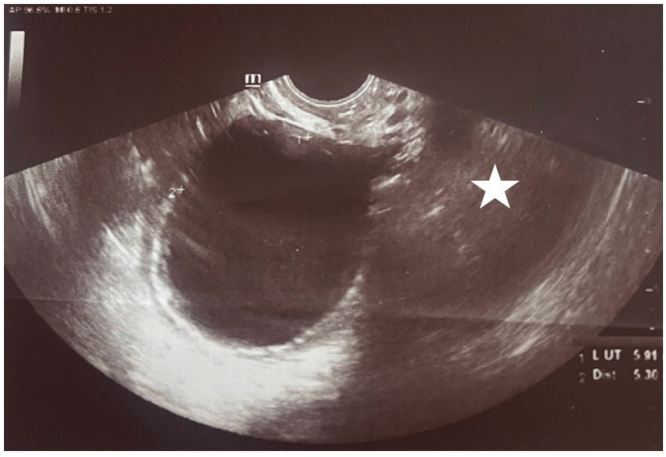

Fig. 1. Ultrasound image showing a purely anechoic, thickened-walled cyst in the left parametrium space, lateral to the uterus (white star), measuring 59x53mm.

Clinically, mesenteric CL is generally asymptomatic in adults, often diagnosed incidentally in radiological investigations for other abdominal affections or during surgery [7]. Mesenteric CL becomes symptomatic upon size expansion, presenting with intermittent pelvic pain and palpable mass [6]. It can be revealed by complications, notably bowel obstruction due to compression, volvulus, hemorrhage, infection and peritonitis if ruptured [8].

The differential diagnosis arises with any intra-abdominal cystic mass ranging from gynecological to nongynecological and from benign to malignant [7]. This includes abscesses, hematomas, digestive tumors, hydatic cysts, abdominal lymphomas, ovarian cysts and other mesenteric tumors [3,9].

The radiological diagnosis of mesenteric CL relies primarily on ultrasound (US), computed tomography (CT) scan, and magnetic resonance imaging (MRI). On ultrasound, CL typically presents as a purely anechoic cystic mass with a thick wall, often exhibiting internal septa forming a honeycomb pattern, and minimal to no flow on color Doppler imaging [7,10]. Occasionally, echogenicity may be observed within the cyst due to intracystic hemorrhage [6]. However, considering that CL can involve intraperitoneal as well as retroperitoneal organs, and ultrasound may not accurately determine its exact localization, CT scan is the exam of choice for diagnosis [7]. CT imaging typically reveals a unilocular or multilocular cyst with homogenous hypodense content, which is usually poorly enhanced by contrast administration [2]. Hyperdensity within the cyst may indicate the presence of intracystic hemorrhage [8].

However, it is worth noting that while CT may suffice as the primary diagnostic tool for CL, MRI can provide valuable information regarding the potential origin of the tumor and its relationship with adjacent organs. Furthermore, MRI consistently offers superior capabilities for comprehensive evaluation of pelvic cystic masses, particularly in menopausal women [10]. The case of our patient, along with other cases of CL misdiagnosed as ovarian cysts reported in the literature, highlights the importance of MRI in accurately assessing pelvic cystic masses [5,10,11]. Nonetheless, due to the rarity of CL, its diverse range of possible locations, and the potential for other abdominopelvic cystic masses to exhibit similar radiological features, even MRI may prove to be inadequate [12]. Therefore, the definitive diagnosis relies on intraoperative findings and histopathological examination of the cyst [11].

Histopathologically, macroscopic examination reveals a well-defined single or polycystic lesion, exhibiting a whitish or translucent appearance [3]. Microscopically, CL features thin walled dilated lymphatic vessels lined with endothelial cells, loose connective tissue with abundant lymphoid aggregates tissues and smooth muscle [2,13]. Various immunohistochemical studies, including CD31, CD34, CD45, factor VIII-related antigen, HMB-45, D2–40, and calretinin, aid in detailed histopathological examination [13].

Therapeutic options of CL are both surgical and non- surgical. Non-surgical options include abstention for asymptomatic lesions, as well as per-cutaneous aspiration with or without injection of sclerosing agents like Bleomycin, acetic acid, OK 432 and alcohol (Ethibloc ®) [6]. This technique may be used for paucicystic and accessible CLs when surgery is difficult or refused by the patient, with variable results and a high risk of recurrence [2]. Therefore, the treatment of choice of CL is surgery and consists of a large surgical resection in order to avoid recurrence [14]. Due to the benign nature of the tumor, the resection should be as conservative as possible. However, depending on the localization and nature of CL, the necessity for partial resection of the neighboring organs like the spleen, the omentum and the bowel might arise [10]. Such is the case of our patient, whose tumor was embedded in the mesentery and required a partial resection of the ileum. Laparoscopic approach is nowadays preferred to laparotomy, even for sizable cysts which are made easier to resect after aspiration [11,15].

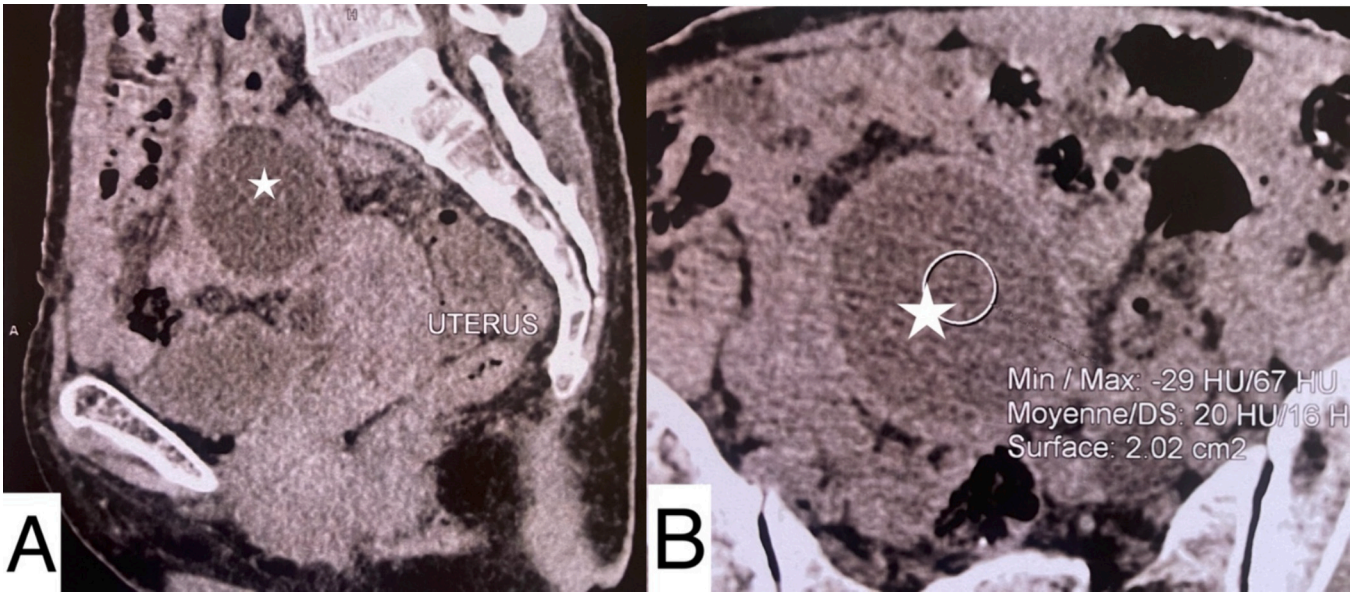

Fig. 2. CT scan images at the sagittal (A) and axial (B) planes showing a unilocular cystic formation (white star) above and left to the uterus, rounded with a thick and regular wall, homogeneous.

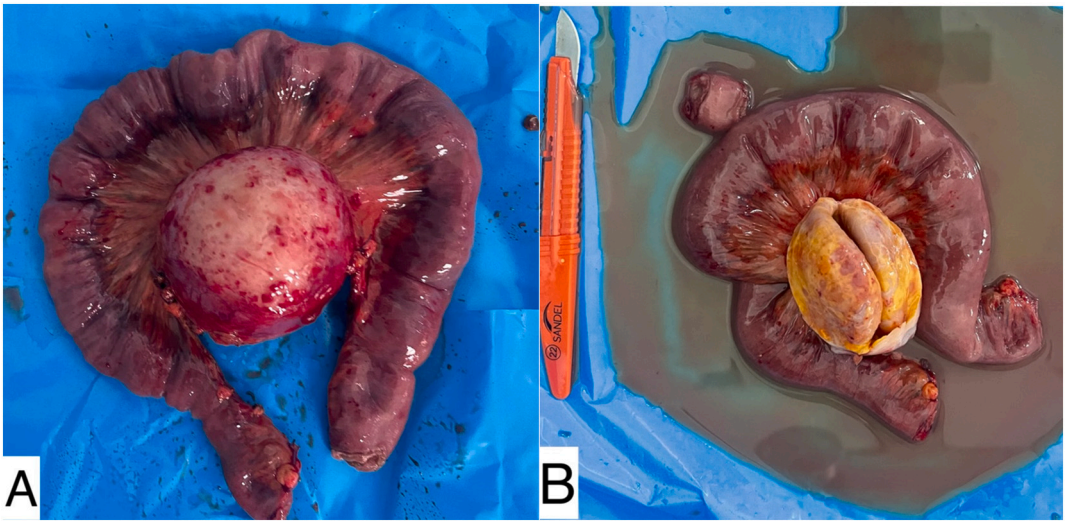

Fig. 3. Image of the resected mesenteric cystic lymphangioma with the ileum (A), opened (B).

Recurrence of CL is up to 40 % for incomplete resection and 17 % for complete resection [6]. If untreated, complications such as rupture, compression and infection may occur [9]. Spontaneous regression was noted in 1,6–16 % of cases [8]. Post-operative complications include lymphocele and chylous ascites, which can be avoided with careful lymphostasis [6]. A novel approach to treating CL involves targeted therapy, which selectively targets tumor cells while preserving normal cells. This treatment strategy focuses on specific lymphatic markers like PRox-1, VEGFR-3, PDGFR-b, and D2–40, particularly beneficial for cases exhibiting aggressive or recurring behavior [16].

In our patient's case, a laparotomy via a Pfannenstiel incision was conducted initially, intending to perform a total hysterectomy for the treatment of serous cystadenoma in a post-menopausal woman. However, intraoperatively, the diagnosis of mesenteric cystic lymphangioma (CL) was established, requiring the assistance of a gastrointestinal surgeon to achieve complete resection of the mass.

Our work has been reported in line with the SCARE Guidelines [17].

4. Conclusion

Mesenteric CL is a rare type of abdominal masses whose diagnosis is challenging, sometimes presenting as more frequent or potentially malignant lesions like ovarian cysts. An exhaustive radiological assessment including computed tomography and magnetic resonance imaging is essential to prevent misdiagnosis, however the final diagnosis is often made intra-operatively. Finally, it is safe to say that a surgeon ought to always be prepared to perform a different surgery than planned.

Consent

Written informed consent was obtained from the patient to publish this case report and accompanying images. On request, a copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethical approval

This case report is exempt from ethical approval in our institute.

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Author contribution

Hounaida Mahfoud, Sarah Benammi, Amina Etber: performed surgery.

Hounaida Mahfoud, Amina Etber: literature review, paper writing and editing.

Amina Etber, Aziz Baidada: Supervision.

Hounaida Mahfoud, Farah Flissate, Samia Tligui: Manuscript editing, picture editing,

Guarantor

Hounaida Mahfoud.

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Not applicable.

Conflict of interest statement

The authors declare that they have no competing interests relevant to the content of this article.

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