Case Report
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Incidental peritoneal lymphangiomatosis associated with a giant uterine leiomyoma: a diagnostic challenge

Abstract

Introduction: Peritoneal lymphangiomatosis is an exceptionally rare lymphatic malformation in adults that may closely mimic malignant peritoneal conditions, including peritoneal carcinomatosis and pseudomyxoma peritonei. According to the International Society for the Study of Vascular Anomalies (ISSVA), peritoneal lymphangiomatosis is classified as a complex lymphatic anomaly within the spectrum of Generalized Lymphatic Anomaly (GLA).

Materials and methods: We present the case of a female patient who was evaluated for large adnexal cystic masses and uterine fibroids at the Instituto de Cancerología y Dr. Bernardo del Valle S Hospital (INCAN), Guatemala City, Guatemala.

Results: A 43-year-old woman presented with a two-year history of continuous abdominal pain and progressive abdominal enlargement. Physical examination revealed a well-defined, non-tender abdominopelvic mass extending from the pelvis to the mesogastrium. Computed tomography demonstrated a giant multilobulated abdominopelvic tumor with cystic components, internal calcifications, and necrotic areas, without ascites or distant metastasis. Tumor markers were negative.

Exploratory laparotomy was performed, including total hysterectomy with bilateral salpingo-oophorectomy, omentectomy, appendectomy, and peritoneal lavage. Intra-operative findings revealed a giant uterine leiomyoma and multiple cystic peritoneal implants involving the root of mesentry, omentum, appendix, and rectosigmoid region.

Definitive histopathological and immunohistochemical analysis confirmed the diagnosis of peritoneal lymphangiomatosis.

Conclusion: This case highlights the importance of recognizing peritoneal lymphangiomatosis as a benign but deceptive entity that can simulate advanced peritoneal malignancy.

Keywords: peritoneal lymphangiomatosis, giant uterine leiomyoma, complex lymphatic anomaly

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Introduction

Lymphangiomatosis is a rare disorder in adults. It is characterized by diffuse or multifocal proliferation and dilation of lymphatic vessels. The International Society for the Study of Vascular Anomalies (ISSVA) classifies lymphangiomatosis within complex lymphatic anomalies, specifically under the spectrum of Generalized Lymphatic Anomaly (GLA).[1,2]

Lymphangiomas are extremely rare in the abdominopelvic region. They are described as benign fluid-filled cystic lesions that may cause compression of adjacent organs. Approximately 1% of lymphangiomas occur in the retroperitoneum. Most cases are diagnosed incidentally or postmortem.[3]

Reactive lymphatic ectasia may occur secondary to obstruction or inflammation and generally follows established lymphatic drainage pathways. In contrast, true lymphangiomatosis represents a primary lymphatic malformation and may affect anatomical sites unrelated to the drainage of the primary organ.[4,5]

Abdominal lymphatic malformations account for approximately 5% of cases; however, retroperitoneal involvement remains particularly rare. Anatomically, lymphangiomatosis most commonly affects the mesentery, omentum, and mesocolon.[3]

In exceptional cases, these lesions may reach large sizes, causing compression of adjacent structures and progressive abdominal distension. Clinical manifestations vary according to the structures involved and may include intestinal obstruction (constipation), gynecological symptoms such as infertility or pelvic heaviness, adnexal mass-like presentation, and, in rare cases, bleeding or inflammatory symptoms mimicking appendicitis. Peritoneal involvement is exceedingly rare and represents a diagnostic challenge due to its close resemblance to malignant peritoneal diseases such as peritoneal carcinomatosis and pseudomyxoma peritonei.[6,7]

Ultrasound and computed tomography (CT) are frequently used imaging modalities. CT typically reveals multiple thin-walled cystic structures filled with fluid resulting from lymphatic obstruction. Magnetic resonance imaging (MRI) provides additional information regarding disease extent and shows similar findings.[3,8]

Differential diagnoses include mucinous cystadenoma, cystic teratoma, cystic mesothelioma, pseudomyxoma retroperitoneal with cystic changes, mucinous perianal carcinoma with cystic changes,

liquefied retroperitoneal hematoma, retroperitoneal hydatid cyst, and retroperitoneal bronchogenic cyst.[3]

Treatment is primarily surgical, consisting of excision of large cystic lesions to relieve symptoms. Minimally invasive approaches such as laparoscopy are preferred; however, when masses exceed 10 cm, exploratory laparotomy is appropriate. In complex, unresectable, or diffuse cases, alternative treatments include sclerotherapy (OK-432, bleomycin) or targeted therapy with sirolimus (mTOR inhibitor).[9–11]

Case presentation

A 43-year-old woman from Esquipulas, Chiquimula, Guatemala, with no significant personal or family history, presented with progressive abdominal pain of two years' duration and increasing abdominal mass. She denied constitutional symptoms and reported allergy to acetaminophen.

On physical examination, ECOG performance status was 0. Abdominal palpation revealed a large, firm, non-tender mass involving the right upper quadrant and mesogastrium with limited mobility. No ascites or lymphadenopathy were detected.

CT imaging showed a giant multilobulated pelvic-abdominal mass measuring approximately 24 × 14 × 25 cm, with cystic components, internal calcifications, and areas of necrosis, without ascites or distant metastasis. Tumor markers were within normal limits. Exploratory laparotomy was performed with total hysterectomy, bilateral salpingo-oophorectomy, omentectomy, appendectomy, and peritoneal lavage (Figure 1).

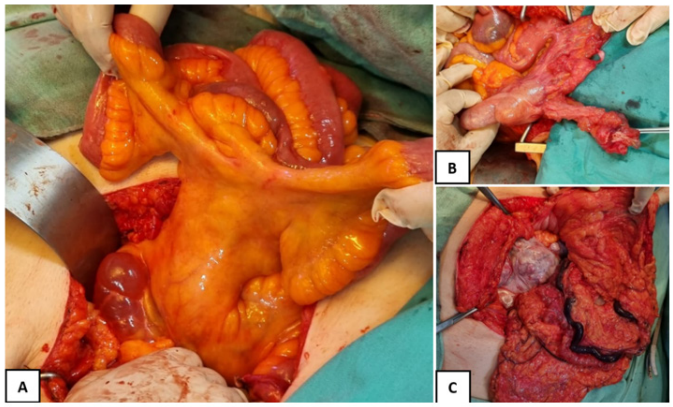

Figure 1 Exploratory laparotomy findings. A: Multiple cystic peritoneal implants affecting the root of mesentery. B: Cystic formations in the appendix and rectosigmoid junction. C: Giant uterine leiomyoma.

Intra-operative findings: The uterus was enlarged with multiple leiomyomas. A cystic implant measuring approximately 15 × 10 cm was identified at the root of the mesentery. Additional uterine leiomyomas measured 29 × 17 × 16 cm and 7 × 4 × 3 cm. A cystic lesion was found 15 cm from the ileocecal valve adherent to the larger mesenteric lesion. Another 5 cm cystic implant was located in the rectosigmoid region. The omentum was diffusely involved with multiple small cystic lesions (~1 cm). The appendix appeared edematous and surrounded by mucinous-appearing material. No other intraabdominal abnormalities were observed (Figure 2).

Surgical treatment: Peritoneal lavage, total hysterectomy with bilateral salpingo-oophorectomy, omentectomy, and appendectomy were performed. Intra-operative frozen section confirmed benign leiomyomas. Cytological analysis of the peritoneal lavage was negative for malignant cells.

Histopathological and Immunohistochemical Findings: Histology demonstrated dilated vascular spaces lined by flattened endothelial cells without atypia and absence of erythrocytes within these spaces, suggesting lymphatic origin.

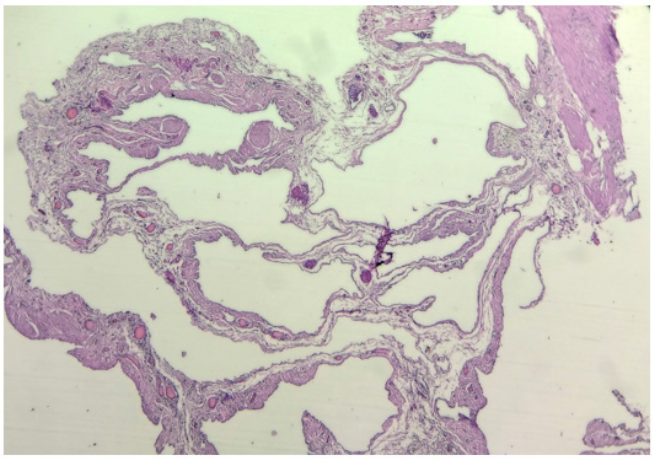

Figure 2 Dilated spaces are observed in the appendiceal wall. These spaces are lined by flat endothelial cells without atypia. No erythrocytes are observed within the dilated spaces, suggesting a lymphatic origin of these structures.

Immunohistochemistry showed strong positivity for podoplanin (D2-40) in the endothelial lining cells of most dilated vascular channels, confirming lymphatic origin. CD31 was positive in endothelial cells of all observed vessels. CD34 was positive in a single mildly dilated vessel consistent with a blood vessel. Overall findings confirmed peritoneal lymphangiomatosis (Figure 3).

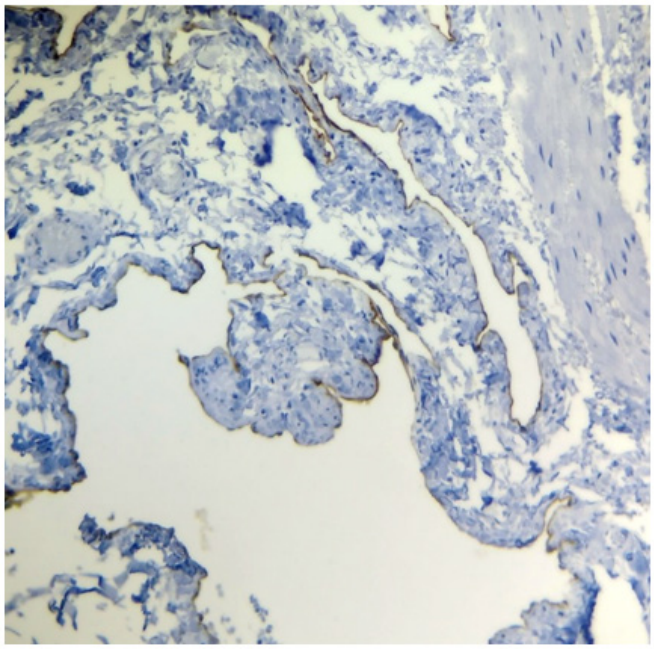

Figure 3 Immunohistochemistry with D2-40 (podoplanin) showed positivity in the endothelial cells lining the dilated spaces. D2-40 is a highly selective marker of the lymphatic endothelium.

Postoperative course: The patient had an uneventful postoperative recovery and was discharged 48 hours later in stable condition.

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Materials and methods

Case report of a female patient who was treated for adnexal masses and large fibroids at the Instituto del Cancer y Dr. Bernardo del Valle S Hospital (INCAN), Guatemala City – Guatemala.

General objective: To describe and analyze the presentation of a rare benign pathology.

Discussion

Peritoneal lymphangiomatosis is a rare manifestation of complex lymphatic anomalies and may closely resemble malignant peritoneal disease intra-operatively. In the present case, although a giant uterine leiomyoma was identified, the anatomical distribution of the cystic lesions involving the mesenteric root, omentum, appendix, and rectosigmoid region did not correspond to the known pathways of uterine lymphatic drainage. This finding supports the diagnosis of true peritoneal lymphangiomatosis rather than secondary reactive lymphatic ectasia.[7]

In the case presented, the involvement of adjacent structures and the presence of large cystic lesions resulted in progressive abdominal enlargement, as described in several case series, with minimal pain symptoms attributable to a mass effect and its association with uterine leiomyomatosis.[8,10,11]

Although ultrasound remains an accessible and convenient imaging modality, it does not adequately demonstrate the distribution of multiple cysts or their relationship to surrounding structures, thus playing a limited role in comprehensive evaluation. In contrast, computed tomography and magnetic resonance imaging provide superior anatomical detail, allowing for appropriate surgical planning and preoperative assessment.[9,10]

The coexistence of gynecological pathologies with peritoneal lymphangiomatosis has been described, particularly involving adnexal structures such as the ovary, where it may be misinterpreted as an adnexal mass due to non-specific symptomatology and imaging findings suggestive of multiple cystic lesions. Definitive diagnosis can only be established through histopathological examination, which demonstrates dilated lymphatic spaces lined by endothelial cells, with fibrous stroma, collagen deposition, and lymphoid aggregates. However, marked congestion and inflammation may pose a diagnostic challenge for pathologists. In this context, immunohistochemical techniques play a crucial role. The condition is characterized by endothelial cell positivity for CD31, CD34, and D2-40 (podoplanin).[11,12]

Limitations of this report include the absence of molecular genetic testing and limited long-term follow-up.

Clinical contribution

This report emphasizes the importance of histopathological and immunohistochemical confirmation to avoid overtreatment.

Recommendation

Lymphatic malformations should be considered in the differential diagnosis of diffuse cystic peritoneal lesions with negative tumor markers.

Conclusion

Peritoneal lymphangiomatosis is a benign yet deceptive lymphatic malformation that may simulate advanced peritoneal malignancy. Recognition of this entity according to ISSVA classification and confirmation through histopathology and immunohistochemistry are essential to guide appropriate management and prevent unnecessary aggressive treatment.

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Conflict of interest

The authors have no financial conflicts of interest, real or perceived, in the publication of this manuscript.

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