Radiology Case Reports 19 (2024) 3732–3739

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Case Report

Isolated unilateral ovarian cystic lymphangioma: A case report ☆,☆☆

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ARTICLE INFO

Article history:
Received 16 March 2024
Revised 8 May 2024
Accepted 17 May 2024

Keywords:
Ovary
Lymphangioma
Lymphatic vessels
Cell proliferation
Computed tomography
Magnetic resonance imaging

ABSTRACT

Ovarian lymphangiomas are rare benign neoplasms characterized by the proliferation of lymphatic vessels within the ovarian tissue. While lymphangiomas can manifest in various anatomical locations, their occurrence within the ovaries is exceptionally uncommon, posing diagnostic and therapeutic challenges for clinicians. The aetiology of ovarian lymphangiomas remains elusive, with theories suggesting congenital malformations, lymphatic obstruction, or acquired lymphatic proliferation as potential contributing factors. The clinical presentation of ovarian lymphangiomas often includes nonspecific symptoms such as abdominal pain, swelling, or discomfort, leading to difficulties in early detection and diagnosis. Radiological imaging, particularly Ultrasound, CT (computed tomography) and MRI (magnetic resonance imaging), plays a crucial role in identifying these lesions and guiding subsequent management strategies. Despite their generally benign nature, ovarian lymphangiomas can attain significant sizes, causing complications such as torsion, rupture, or compression of adjacent structures. Surgical intervention, typically in cystectomy or oophorectomy, is frequently pursued to alleviate symptoms and prevent potential complications. This paper aims to comprehensively review the existing literature on ovarian lymphangiomas, addressing their clinical presentation, diagnostic challenges, and management strategies. By synthesizing available data, we seek to enhance our understanding of this rare entity, providing valuable insights for clinicians encountering similar cases. Improved awareness and knowledge of ovarian lymphangiomas are essential for timely diagnosis and optimal patient outcomes.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

☆☆ Acknowledgments: The authors acknowledge the patient as a great source of learning. The authors declare that they have no financial support, and sponsorship was obtained for this research.

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https://doi.org/10.1016/j.radcr.2024.05.042

Introduction

Lymphangiomas are benign malformations involving the lymphatic system that occur due to obstruction of the local lymphatics. These lesions typically affect youngsters and are present in the head, neck, and axillary areas. Less than 1% of individuals developed retroperitoneal cystic lymphangiomas, an enlarged omentum, and mesentery [1]. These slow-growing tumors rarely cause symptoms and are typically discovered by accident. In some cases, they present with a palpable abdominal mass. The nonspecific clinical and radiological features are frequently mistaken for a malignant ovarian tumor. Thus, a definite diagnosis can only be made after a histological evaluation.

Case presentation

A 40-year-old woman complained of bloating and lower abdomen pain that had been worse during the previous 6 months. She reported irregular menstrual cycles but denied any significant medical history or familial predispositions to gynecological disorders. After the initial physical examination, a palpable lump in the lower abdomen prompted additional research.

Computed tomography (CT) abdomen (Plain) and contrast-enhanced computed tomography (CECT) Abdomen (Arterial, Venous, and Delayed Phases) show a bulky uterus with focal exophytic mild heterogenous enhancing isodense to the hypodense mass lesion (iso to myometrium) in the anterosuperior myometrial wall of the uterus (fundus)—Sub-serosal fibroid uterus with necrotic degeneration. A focal, non-enhancing multiloculated cystic lesion measuring 15.3 × 1.8 × 1.9 cm (CC x TR x AP) in the right adnexa (epicentre in the ovary) of serpiginous appearance extending superiorly along the right ovarian artery, right ovarian vein, and right ureter, along the para-caval region till the infra-renal segment of the pre-caval region—Ovarian lymphangioma (Figs. 1A-F, 2 A-F, and 3A-F).

Magnetic resonance imaging (MRI)—abdomen shows a bulky uterus with *T1W—focal* isointense (iso to myometrium) and *T2W*—heterogenous hypointense-hyperintense mass lesion in the antero-superior wall uterus (fundus) (*SPIR*: heterogeneous hypointense-hyperintense)—sub-*serosal fibroid with necrotic degeneration*. *T1W*: focal multiloculated hypointense, and *T2W*: hyperintense cystic lesion in the right adnexa (epicentre in the ovary) of serpiginous appearance extending superiorly along the para-caval region till the infra-renal segment of the pre-caval region (*SPIR*: hyperintense)—*Ovarian lymphangioma* **(**Figs. 4**A-L).**

Serum tumour markers CA-125 and CA-19.9 were within normal limits.

Based on imaging findings and clinical presentation, an *Ovarian lymphangioma* is considered. The decision for surgical intervention was influenced by the size of the ovarian lymphangioma, warranting consideration of hysterectomy. However, it is notable that the patient expressed a preference against hysterectomy, emphasizing the importance of shared decision-making in the treatment process.

Consequently, the patient underwent laparotomy, during which a myomectomy was performed to address the enlarged uterus, alongside a right ovarian cystectomy to manage the ovarian lymphangioma. Notably, the surgical approach employed meticulous dissection techniques aimed at preserving surrounding ovarian tissue, with a particular focus on conserving fertility. This nuanced surgical strategy underscores the commitment to individualized patient care and the prioritization of patient preferences in the management of complex gynecological conditions. Intraoperatively, the cystic mass was noted to be densely adherent to adjacent structures, necessitating careful separation to avoid intraoperative complications. The patient was discharged from the hospital on the third day after surgery after fully recovering. ***The preoperative image*** shows a cystic lesion in the right adnexa (epicentre in the ovary) of a serpiginous appearance — *Ovarian lymphangioma* (Fig. 5).

A detailed examination of the histopathological specimen (HPE) provided crucial insights into the nature of the cystic lesion. ***Photomicrograph analysis revealed the presence of multiple cystic spaces (black asterisk)*** within the stroma, characterized by a lumen filled with lymph fluid. Additionally, the examination identified foci of lymphocyte clusters, delineated by thin-walled lymphatic vessels forming a single layer of squamous endothelium. These characteristic features were consistent with the diagnosis of ovarian lymphangioma, and notably, no evidence of malignant transformation was observed (Fig. 6).

Follow-up appointments were scheduled to monitor for recurrence or complications, with an emphasis on maintaining regular gynaecological examinations and imaging surveillance.

Discussion

The complex network of one-way lymphatic veins that comprise the lymphatic system is intended to carry lymph toward the heart. It has been documented that congenital lymphatic system abnormalities can result in childhood lymphangiomas. Visceral lymphangiomas are very rare and primarily present in the mesentery and intestine. Ovarian lymphangioma is a rare condition.

Since the mesentery is home to the majority of lymphatic channels, it is the origin of most lymphangiomas in the abdominal cavity. Therefore, intraabdominal lymphangiomas were previously named ' mesenteric cystic lymphangiomas' [2].

Based on the extent of the lymphatic gaps, lymphangiomas can be categorized into 3 types: simple, cavernous, and cystic [3].

Lymphangiomas consist of multiple densely packed, thin-walled vascular spaces. Widely spaced flattened endothelial cells line these spaces, and the lumens of these spaces are filled with lymphocytes and pale eosinophilic material called lymph. The stroma of these structures also harbors aggregates of lymphocytes [4].

Typically, on imaging, lymphangiomas appear as distinct, multilocular cystic masses that either contain echogenic de-

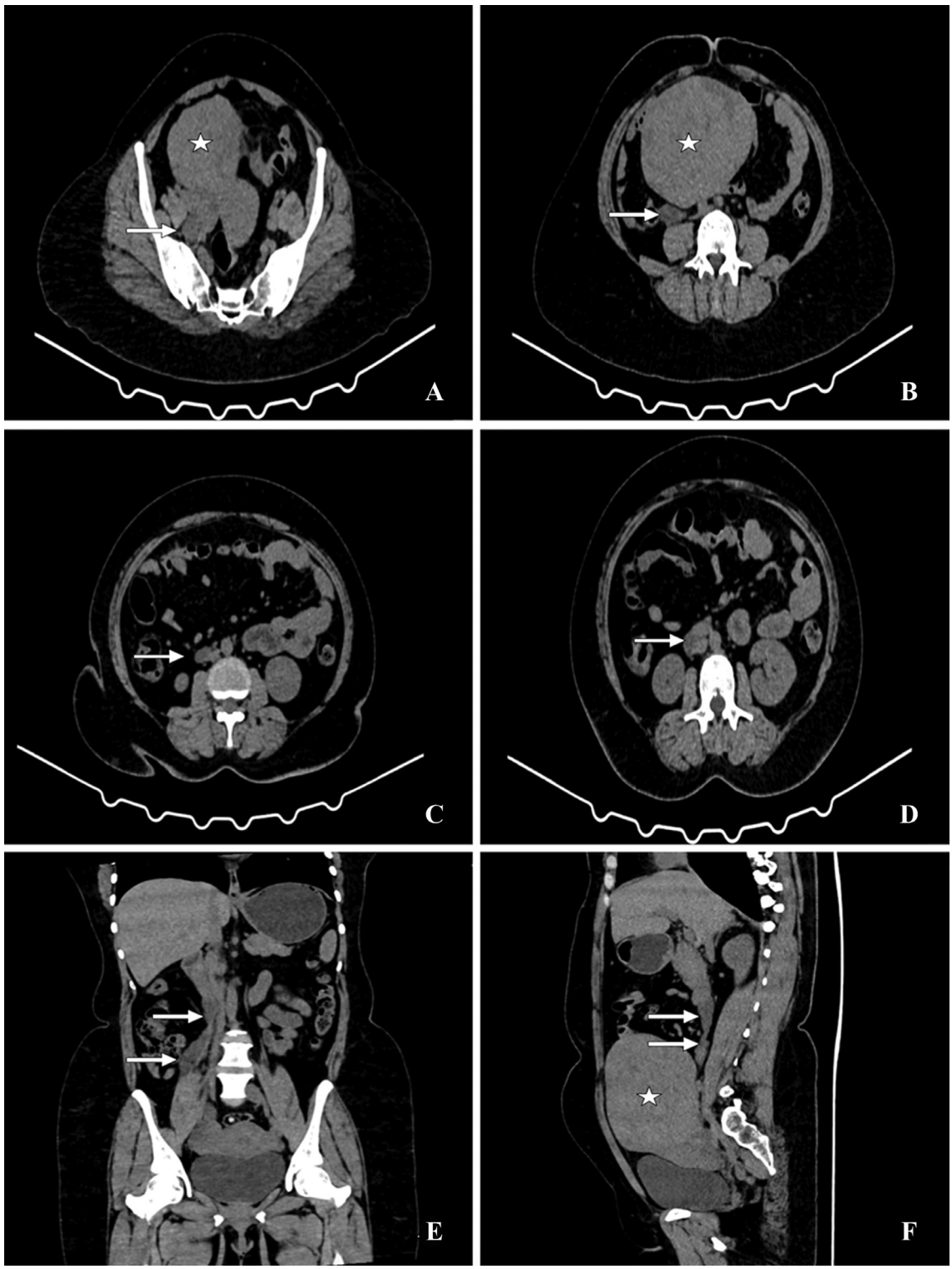

Fig. 1 – (A-F): A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. Pre-operative Computed Tomography (CT) Abdomen (Plain): Multi-Planar Reformatted (MPR) images: (A, B, C, D - Axial, E - Coronal, F - Sagittal sections) show a bulky uterus with focal exophytic mild heterogeneous isodense to hypodense mass lesion (iso to myometrium) in the antero-superior wall myometrium of the uterus (fundus)—Sub-serosal fibroid uterus with necrotic degeneration (white asterisk). A focal multiloculated cystic lesion in the right adnexa (epicenter in the ovary) of serpiginous appearance extending superiorly along the para-caval region till the infra-renal segment of the pre-caval region—Ovarian lymphangioma (short white arrows).

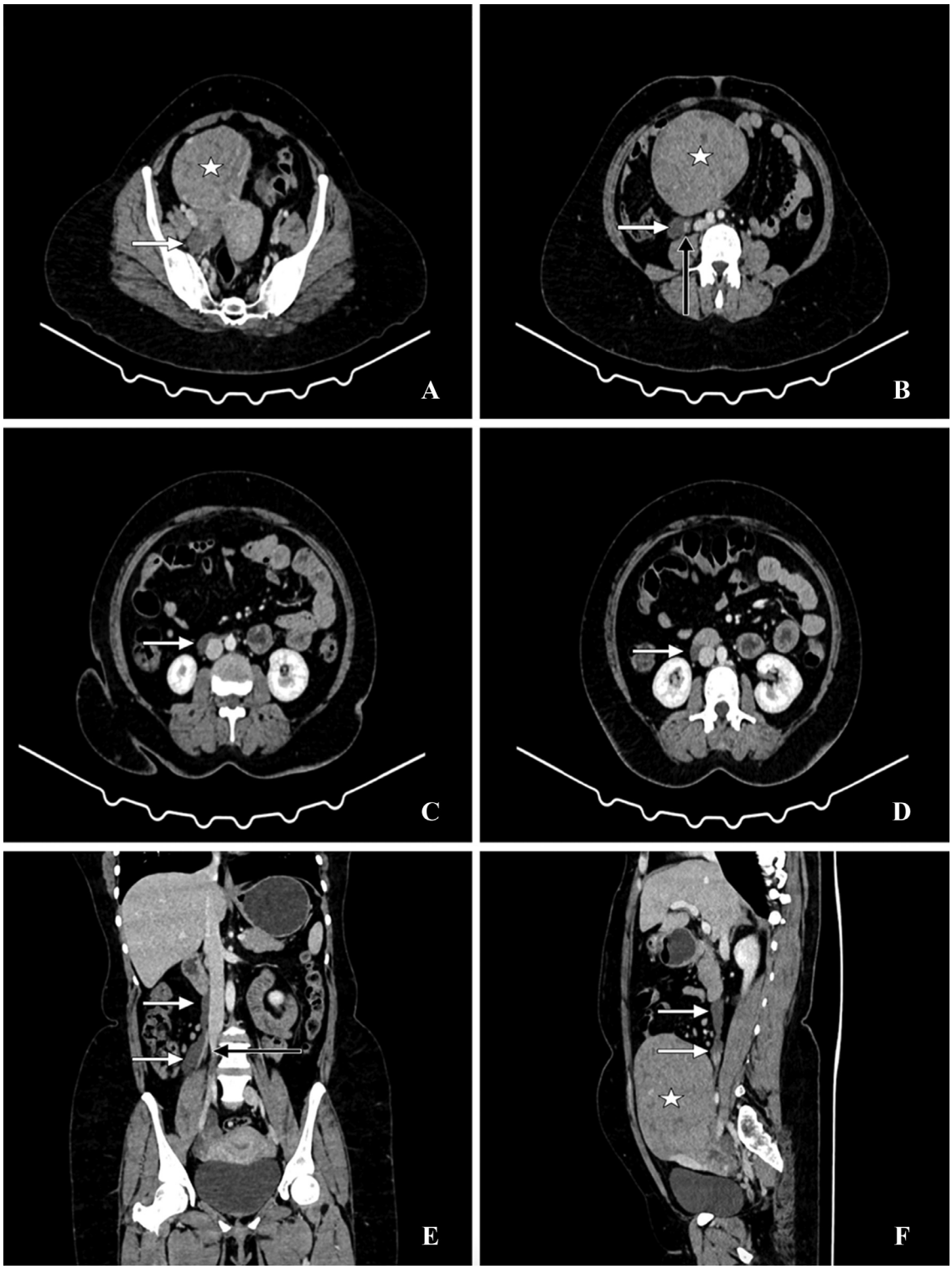

Fig. 2 – (A-F): A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. Pre-operative contrast-enhanced computed tomography (CECT) Abdomen (Venous phase): Multi-Planar Reformatted (MPR) images (A, B, C, D - Axial, E - Coronal, F - Sagittal sections) show a bulky uterus with a focal exophytic mild heterogenous enhancing isodense to hypodense mass lesion (iso to myometrium) in the antero-superior wall myometrium of the uterus (fundus)—Sub-serosal fibroid uterus with necrotic degeneration (white asterisk). A focal, non-enhancing multiloculated cystic lesion in the right adnexa (epicenter in the ovary) of serpiginous appearance extending superiorly along the right ovarian vein (large black arrows), along the para-caval region till the infra-renal segment of the pre-caval region—Ovarian lymphangioma (short white arrows).

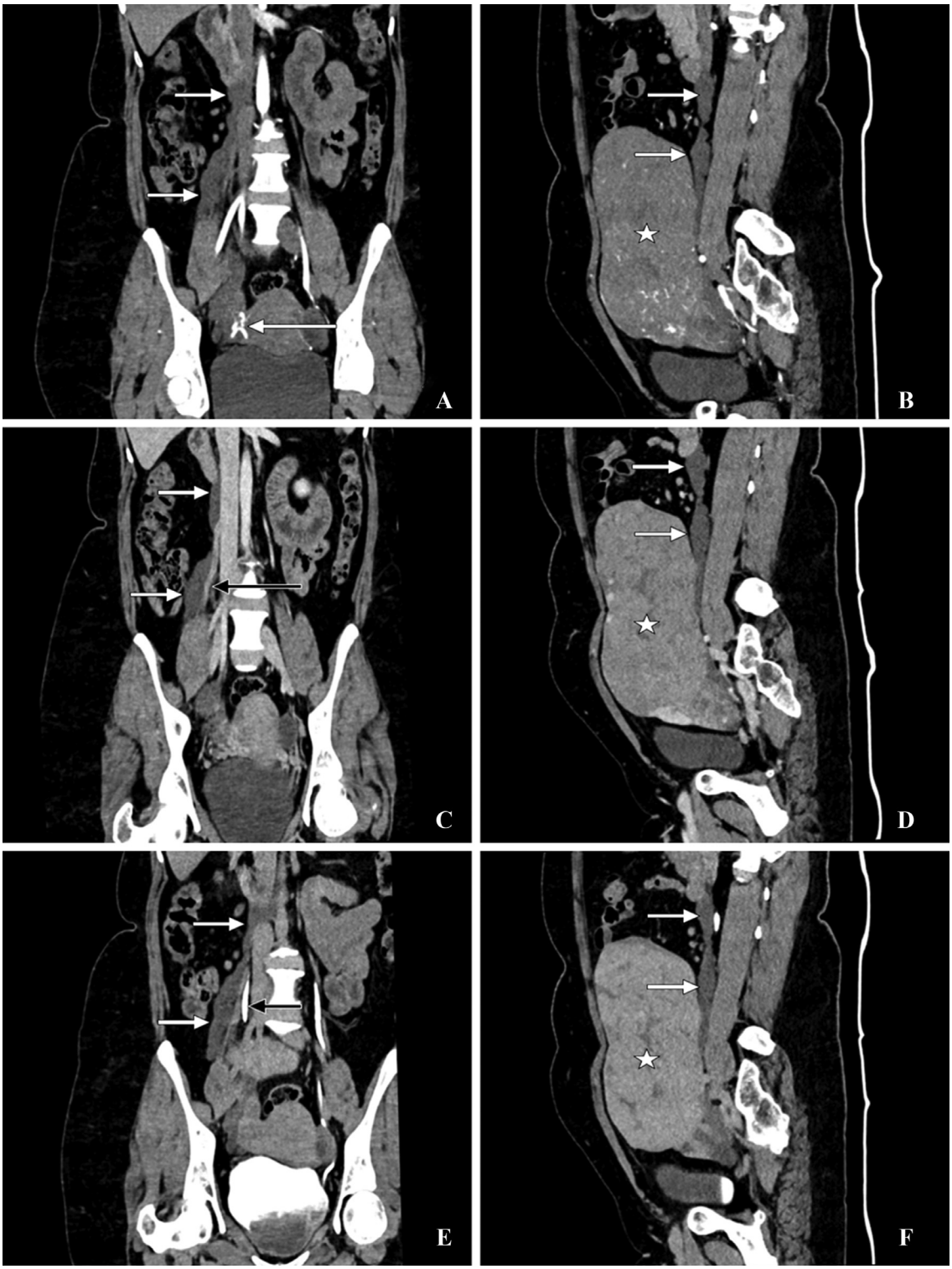

Fig. 3 – (A-F): A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. Pre-operative Contrast-enhanced Computed Tomography (CECT) Abdomen: Multi-Planar Reformatted (MPR) images: (A - Coronal, B - Sagittal in Arterial Phase, C - Coronal, D - Sagittal in Venous Phase, E - Coronal, F - Sagittal in Delayed Phase) shows a bulky uterus with focal exophytic mild heterogenous enhancing isodense to hypodense mass lesion (iso to myometrium) in the anterosuperior wall myometrium of uterus (fundus)—Sub-serosal fibroid uterus with necrotic degeneration (white asterisk). A focal non-enhancing multiloculated cystic lesion in the right adnexa (epicenter in the ovary) of serpiginous appearance extending superiorly along the right ovarian artery (large white arrow), right ovarian vein (large black arrow), right ureter (short black arrow), along the para-caval region till the infra-renal segment of the pre-caval region—Ovarian lymphangioma (short white arrows).

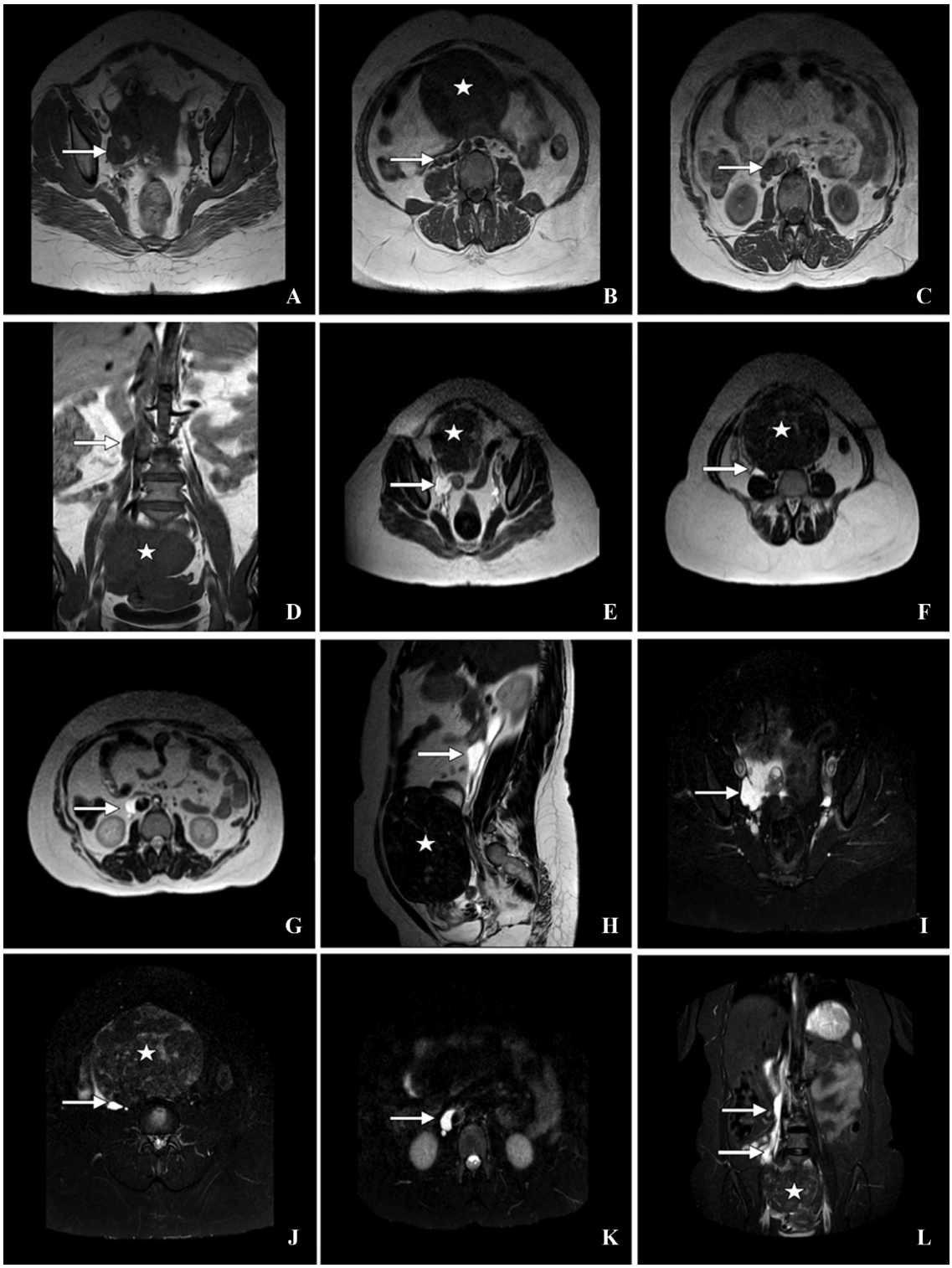

Fig. 4 – (A-L): A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. Magnetic Resonance Imaging (MRI)—Abdomen: (A, B, C) T1W Axial, (D) T1W Coronal, (E, F, G) T2 Axial, (H) T2 Sagittal, (I, J, K) SPIR Axial, (L) SPIR Coronal shows a bulky uterus with T1W: focal isointense (iso to myometrium) and T2W: heterogenous hypointense-hyperintense mass lesion in the antero-superior wall uterus (fundus) (SPIR: heterogeneous hypointense-hyperintense)—Sub-serosal fibroid with necrotic degeneration (white asterisk). T1W: A focal multiloculated hypointense, and T2W: hyperintense cystic lesion in the right adnexa (epicenter in the ovary) of serpiginous appearance extending superiorly along the para-caval region till the infra-renal segment of the pre-caval region (SPIR: hyperintense)—Ovarian lymphangioma (short white arrows).

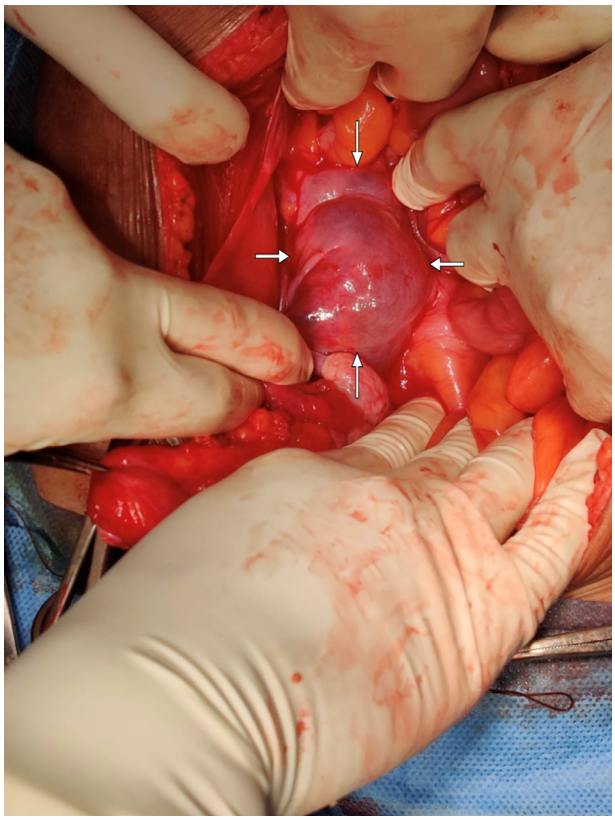

Fig. 5 – A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. The per-operative image shows a cystic lesion in the right adnexa (epicenter in the ovary) of a serpiginous appearance—Ovarian lymphangioma (short white arrows).

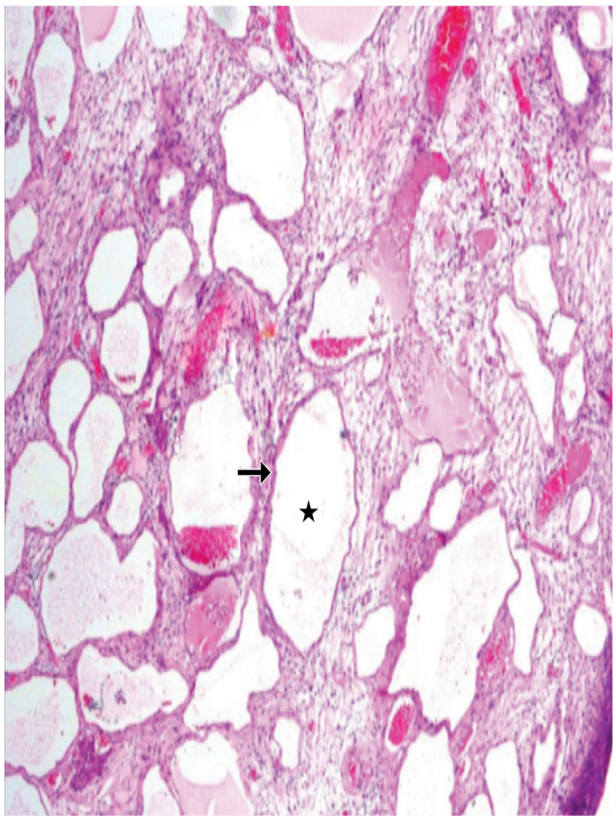

Fig. 6 – A 40-year-old woman complained of lower abdominal pain, distension, and bloating that had been worse during the previous 6 months. A photomicrograph of the histopathological specimen (HPE) examination of cystic lesion shows Multiple cystic spaces (black asterisk) within the stroma, a lumen filled with lymph fluid, and foci of lymphocyte clusters lined with thin-walled lymphatic vessels form a single layer of squamous endothelium (short thick black arrow)—Ovarian lymphangioma with no evidence of malignant transformation.

bris or are anechoic. IV contrast-enhanced CT may show the cyst wall and septa enhancement. Usually, the fluid component has low attenuation values and is homogenous. There are times when chyle causes negative attenuation values. Hemorrhaging and calcification are rare, but they can happen. Lymphangiomas are hypointense on T1-weighted magnetic resonance imaging images and hyperintense and CT attenuation may seem more solid if bleeding or infection occurs within the lesion [5]. Rare side effects of retroperitoneal lymphangiomas include ascites, bleeding, rupture, torsion, and volvulus [6].

Malignant transformation in a lymphangioma is extremely rare [7]. Past studies, such asthose by Rice et al. [8] and Aristizabal et al. [9], have described lymphoma with rare malignant counterparts. Rice et al. wrote about a benign ovarian lymphangioma that came back within 6 months and spread to the other ovary, the liver, and the peritoneum as a whole. A peritoneal lymphangioma that twice returned in less than 2 years was reported by Aristizabel et al. Thus, radiation treatment was used to manage the illness.

Spontaneous regression of the cyst is a rare occurrence, and due to the risk of recurrence, surgical intervention becomes necessary [10]. The preferred method is simple total excision. However, it's important to note that post-surgery complications such as bleeding, peritonitis, torsion, and abscesses may rarely occur. While the spread to the retroperitoneum is a highly uncommon event, it is considered a potentially fatal complication. Therefore, managing these components of the disease requires significant thought and attention.

Learning points

a. **Awareness of Rare Entities:** Isolated unilateral ovarian cystic lymphangiomas are sporadic benign tumors. Clinicians should maintain awareness of the possibility of encountering such rare entities in clinical practice, particularly when evaluating patients with nonspecific symptoms or imaging findings suggestive of ovarian pathology.
b. **Diagnostic Challenges:** The diagnosis of isolated unilateral ovarian cystic lymphangioma can be challenging due to its rarity and nonspecific clinical presentation. Clinicians should be mindful of the potential for misdiagnosis or delayed diagnosis and consider this entity in the differential diagnosis of ovarian cystic lesions.

c. **Role of Imaging:** TVUS (Transvaginal ultrasound) and MRI are crucial in diagnosing and preoperatively evaluating ovarian cystic lymphangiomas. Familiarity with the characteristic imaging features, such as multicystic ovarian masses with variable echogenicity on TVUS and well-defined cystic structures on MRI, can aid in accurate diagnosis.
d. **Histopathological Confirmation:** The definitive diagnosis of isolated unilateral ovarian cystic lymphangiomas relies on the histopathological examination of surgical specimens. Pathologists should be aware of the histological features to confirm the diagnosis, including cystic spaces lined by flat endothelial cells and positive immunohistochemical staining for lymphatic markers.
e. **Surgical Management:** Surgical excision, either via laparoscopic or open approaches, remains the mainstay of treatment for isolated unilateral ovarian cystic lymphangiomas. With few recurrence rates documented in the literature, complete resection is frequently curative. Surgeons should aim for complete excision while preserving ovarian function whenever possible.
f. **Multidisciplinary Approach:** Given the rarity and complexity of isolated unilateral ovarian cystic lymphangiomas, an interdisciplinary approach involving gynecologists, radiologists, pathologists, and surgeons is essential for optimal patient care. Collaboration among different specialities ensures comprehensive evaluation, accurate diagnosis, and appropriate management of this rare ovarian tumor.
g. **Research and Reporting:** Continued reporting of cases and research efforts are crucial for enhancing our understanding of isolated unilateral ovarian cystic lymphangiomas. Clinicians and researchers should contribute to the literature by reporting cases, sharing experiences, and conducting further studies to improve diagnostic and therapeutic strategies for this rare entity.

Conclusion

Lymphangiomas, typically observed in the head and neck, are infrequently found in pelvic regions, with reported cases in the ovaries being limited. Though multidetector CT and MRI play a crucial role in diagnosis and surgical planning, definitive confirmation still relies on histopathological examination for this condition. It's important to remember that lymphangiomas can be found in the para-ovarian region, emphasizing the need to consider them as a differential in pelvic lesions.

Patient consent

The authors certify that they have obtained all appropriate patients consent.

Declaration of generative AI and AI-assisted technologies in the writing process

During the preparation of this work, OpenAI was used in order to improve language and readability. After using this tool/service, the author(s) reviewed and edited the content as needed and take full responsibility for the content of the publication.

Authors contributions

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