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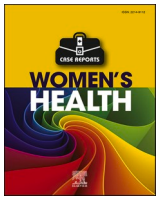

Ovarian lymphangioma resected during abdominal hysterectomy: A case report

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ABSTRACT

Ovarian lymphangioma, a rare pathologic finding, is an ovarian mass characterized by lymphatic tissue lined with endothelial cells. It is normally asymptomatic and may be found incidentally during abdominal surgery for other pathologies.

This report describes a case of a 49-year-old woman presenting to her primary care physician for three months of abdominal bloating and irregular menses. Magnetic resonance imaging revealed a 31 × 23 × 20 cm uterine mass suspected to be the cause of her symptoms. Total abdominal hysterectomy and bilateral salpingectomy were performed. During surgery, the right ovary was flattened and densely adhered to the body of the uterus, necessitating right oophorectomy. Pathology of the right ovary revealed flattened endothelial cells lining cystic spaces, consistent with the diagnosis of ovarian lymphangioma. Taken together, this case and the literature suggest that ovarian lymphangioma should be considered in the differential of ovarian masses, and their management shared more widely to help encourage the development of standard practice guidelines. There are no clear guidelines for when, and how often, to monitor these lesions after resection. In this case, the patient was seen at two-week and six-week follow-up visits with no new symptoms. Given that some case reports describe malignant transformation, patients should be followed this closely in the post-surgical period, and the best cadence for follow-up should be determined to improve outcomes.

1. Introduction

A lymphangioma is an abnormal proliferation of lymphatic tissue characterized histologically by cystic spaces lined by endothelial cells [1,2]. While they may arise anywhere in the body, the most common locations in childhood are in the head, neck, and axilla, while superficial cutaneous or intra-abdominal lesions may be found in adulthood [3,4]. Ovarian lymphangiomas are difficult to distinguish from malignant ovarian lesions on imaging alone, and therefore pathologic evaluation is needed for definitive diagnosis. Given the rarity of this condition, little is known about its rate of recurrence or capability for malignant transformation.

This report describes the case of a woman presenting with abdominal bloating and abnormal uterine bleeding who was found to have an enlarged, fibroid uterus requiring total abdominal hysterectomy with bilateral salpingectomy. An abnormal ovarian mass incidentally found adhered to the uterus was removed, and pathologic examination of the mass revealed characteristics consistent with a benign ovarian lymphangioma. A brief literature review of other cases of ovarian lymphangioma is presented, as well as suggested options for follow-up and surveillance of these lesions.

2. Case Presentation

A 49-year-old, premenopausal woman initially presented to her primary care physician with a three-month history of progressively worsening abdominal bloating and irregular menses. Initial pelvic ultrasound revealed a 25 × 19 × 20 cm multi-fibroid uterus and could not rule out leiomyosarcoma. Magnetic resonance imaging (MRI) of the uterus was done for further characterization of the pelvic masses, and the patient was referred to an obstetrician-gynecologist for further work-up of the irregular bleeding and enlarged uterus. An MRI scan two

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months later showed a 31 × 23 × 20 cm uterine mass, arising from the right lower segment of the uterus and extending to the right upper quadrant of the abdomen. The deviated uterus contained other smaller intramural and subserosal fibroids, most between one and seven millimeters in size, with a few significantly larger fibroids. Additionally, the MRI noted mild right hydronephrosis, the absence of free fluid and the absence of pelvic lymphadenopathy. The patient presented for gynecologic care three months after initial presentation, with persistent abdominal bloating and irregular vaginal bleeding. She had no previous surgery. Her medical history was notable only for diabetes controlled with metformin, and subclinical hypothyroidism not requiring treatment. She had no significant family or social history. After counseling on various treatment options, the patient elected to undergo a total abdominal hysterectomy with bilateral salpingectomy to resolve her symptoms.

She underwent surgery seven months after initial gynecological consultation. During the surgery, myomectomy of the largest 30 cm fibroid was initially performed to facilitate the subsequent hysterectomy. During this step of the procedure, the right ovary was noted to be flattened and adhered to the large fibroid. Because the fibroid could not be safely removed without avulsing the infundibulopelvic ligament of the ovary, a right salpingo-oophorectomy was performed. The surgery was complicated by an estimated blood loss of 2400 mL from the large fibroid, requiring transfusion of two units of packed red blood cells. The remainder of the surgery was uncomplicated, and the ovarian specimen was sent to pathology. The patient was discharged from hospital on post-operative day 3 and was recovering well at two-week and six-week, in-office post-operative follow-up.

Histopathological examination of the right ovarian cystic mass revealed numerous dilated vascular channels of varying size lined by a single layer of flattened endothelial cells (Fig. 1). The cysts were separated by loose fibrous stroma, and some cysts showed hypereosinophilic material consistent with lymph fluid (Fig. 2). No atypia, hyperchromasia, or pleomorphism were identified. There were no mitotic figures or necrosis. Immunohistochemical studies for vascular markers CD34 and CD31 were performed and were positive. D2–40, a more specific marker for lymphatic endothelial cells, was positive, consistent with the diagnosis of lymphangioma (Fig. 3). Additional pathologic findings in this specimen included a 31 cm lipoleiomyoma, multiple leiomyomas, secretory phase endometrium and bilateral fallopian tubes within normal limits.

3. Discussion

Ovarian lymphangioma is a rare pathologic finding, with approximately 20 case reports in the literature describing the phenomenon [1].

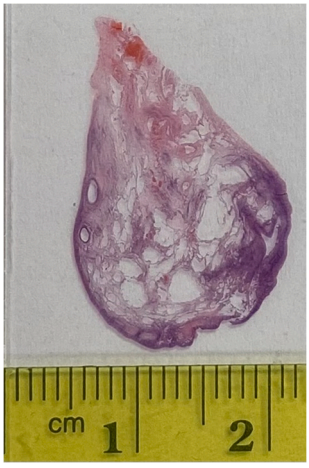

Fig. 1. Appearance of ovary on glass slide with multiple cystic spaces.

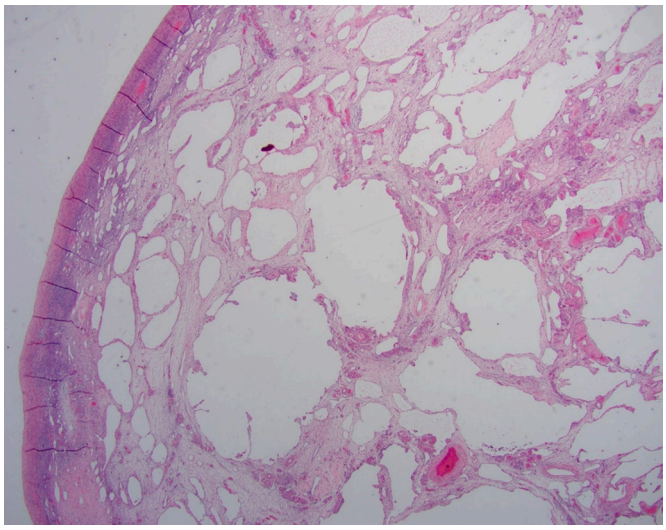

Fig. 2. H&E stain of cystic spaces lined by lymphatic endothelial cells.

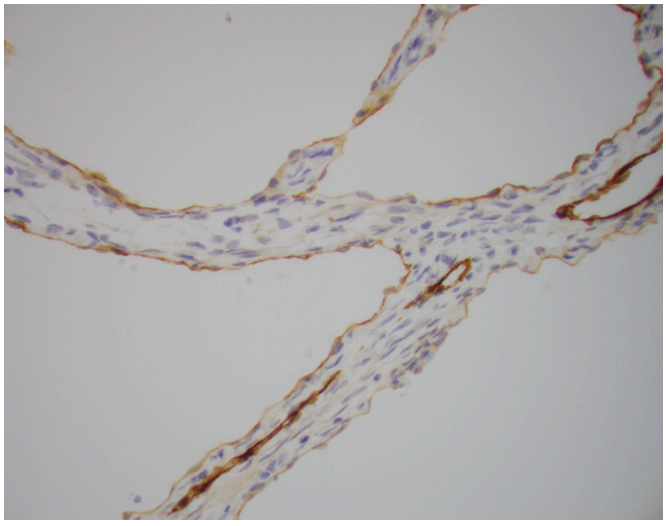

Fig. 3. Cystic spaces lined by lymphatic endothelial cells marked with D2–40 stain.

While the etiology of ovarian lymphangiomas is unclear, prevailing theories include embryonic malformations of lymphatic vessels, as well as fistulation of lymphatic ducts due to inflammation [5,6]. The vast majority of ovarian lymphangiomas are asymptomatic, though some may grow large enough to cause symptoms due to mass effect [1], or ascites [7,8]. Most cases are found in women of middle to late reproductive age, though cases of lymphangiomas have been described in pediatric patients, including in utero [1,9,10]. In addition, though most cases are unilateral, lymphangiomas of the bilateral ovaries have been described [2,8,11,12]. Many ovarian lymphangiomas are thus found incidentally during surgical procedures for other indications [2,8,13,14]. This case falls within this latter group, as the patient's symptoms were not thought to be due to her ovarian pathology.

Due to the overwhelming size and locations of the multiple fibroids in the uterus of the patient, and the inability of imaging to delineate an ovarian mass, a differential diagnosis for an adnexal mass was not initially considered. The patient described here did not experience any precipitating event – such as abdominal trauma, surgery, or radiation – that would explain the genesis of a lymphatic malformation. Furthermore, her medical history was negative for significant disease that would explain a lymphangioma resulting from chronic inflammation.

Ovarian lymphangiomas are thought to be largely benign; however, there have been occasional reports of metastatic neoplasms [13,15]. Similarly, ovarian lymphangiomas are not typically associated with elevated CA-125. However, one case report describes an ovarian lymphangioma causing pseudo-Meigs syndrome with an elevated CA-125 [16]. Despite the evidence that ovarian lymphangiomas may behave similarly to malignant neoplasms, there are no guidelines in place for follow-up imaging or screening for malignant conversion. Most case reports suggest follow up for two years, first suggested by Aristizabal [13] and Evans [2], two of the earliest case reports describing ovarian lymphangioma. Most do not describe the cadence of surveillance during these two years, though two case reports show good follow-up at six months [8,17], and one scheduled follow-up every three months for two years [18].

In the case presented here, there was no evidence of complications at two-week or six-week follow-up, nor any other indication for follow-up ultrasound examination. Because of this, further imaging was deferred. To date, the patient has had no further complications from the surgery or evidence of recurrence of the lymphangioma on the contralateral side and she reports feeling well. More research is needed to understand the frequency with which these neoplasms should be surveilled to prevent recurrence and malignant transformation.

4. Conclusion

Ovarian lymphangioma is a rare pathological diagnosis that is largely asymptomatic, though may present with symptoms and imaging similar to a malignant neoplasm. Ovarian lymphangioma should be considered in the differential diagnosis of an adnexal mass, both pre-operatively and intraoperatively, given its morphologic similarity to an ovarian malignancy. More research is needed to know if, and when, further surveillance after resection is needed.

Contributors

Brian Benitez undertook the literature review and revised the article critically for important intellectual content.

Eva Patalas contributed to patient care, and revising the article critically for important intellectual content.

Tara Singh contributed to patient care, conception of the case report, acquiring and interpreting the data, drafting the manuscript and revising the article critically for important intellectual content.

All authors approved the final submitted manuscript.

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Patient consent

Consent for publication of this case report was obtained from the patient.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflicts of interest statement

The authors report no conflicts of interest.

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