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Case Report

Cystic Lymphangiomyoma of the Uterus Mimicking an Ovarian Cyst: A Case Report

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Abstract

Background: Cystic lesions of the uterus are rare and are frequently misdiagnosed as adnexal pathology on imaging. Cystic lymphangiomyoma is an uncommon benign lesion characterized by dilated lymphatic channels within the uterine wall. Because imaging features are nonspecific, diagnosis generally requires histopathological confirmation supported by immunohistochemistry.

Case presentation: We report the case of a 34-year-old woman who presented with a pelvic cystic mass initially presumed to be of ovarian origin. Pelvic ultrasound and MRI suggested a paratubal cyst. Laparoscopy revealed a 6-cm intramural uterine cyst arising from the uterine wall. Laparoscopic cystectomy was performed with meticulous dissection to preserve the myometrium. Histopathological examination demonstrated multiple cystic spaces lined by flattened endothelial cells, consistent with uterine cystic lymphangiomyoma.

Conclusion: The postoperative course was uneventful. This rare case highlights the diagnostic challenge of distinguishing uterine cystic lesions from adnexal pathology based solely on imaging. Accurate diagnosis relies on histopathological and immunohistochemical evaluation, while complete surgical excision ensures definitive cure.

Keywords: Cystic lymphangiomyoma; Uterus; Uterine cyst; Lymphangiectatic leiomyoma; Adenomyotic cyst.

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1. Introduction

Cystic lesions of the uterus are rare. Among them, cystic lymphangiomyoma is an uncommon entity that typically affects young women and may be radiologically misinterpreted as adnexal disease. The main differential diagnoses include unusual variants of leiomyoma, as well as ovarian or paratubal cysts [1,2]. Because imaging findings are nonspecific, the diagnosis is usually established on the excised specimen, often with immunohistochemical (IHC) support [2]. This case report aims to highlight this diagnostic pitfall, describe the clinical and pathological pathway leading to the correct diagnosis, and review the relevant literature to improve preoperative recognition.

2. Case Presentation

A 34-year-old nulliparous woman was referred for evaluation and management of a left adnexal cyst identified on ultrasound (Figure 1) and MRI, described as a 7-cm paratubal cyst without suspicious features for malignancy. Based on these imaging findings, a benign paratubal cyst was initially suspected.

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The patient underwent diagnostic laparoscopy. Intraoperatively, the adnexa appeared normal. A 6-cm cystic myometrial mass was noted on the posterior-fundal surface of the uterus, without breach of the endometrial cavity. Initial aspiration yielded clear yellow serous fluid. A cystectomy was performed using divergent traction. The postoperative course was uncomplicated uneventful.

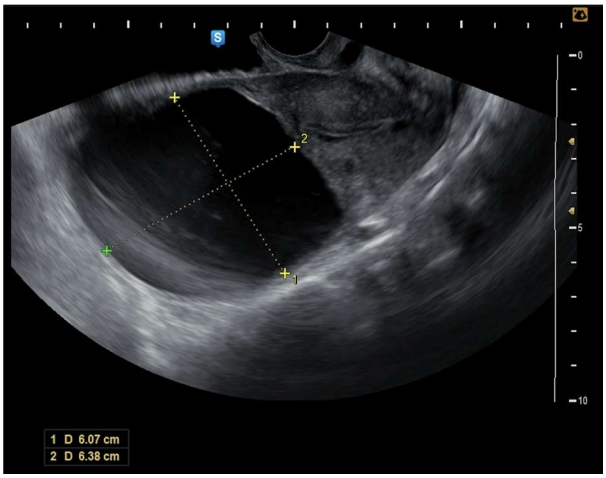

Fig. 1: Ultrasound image of a left paratubal cyst.

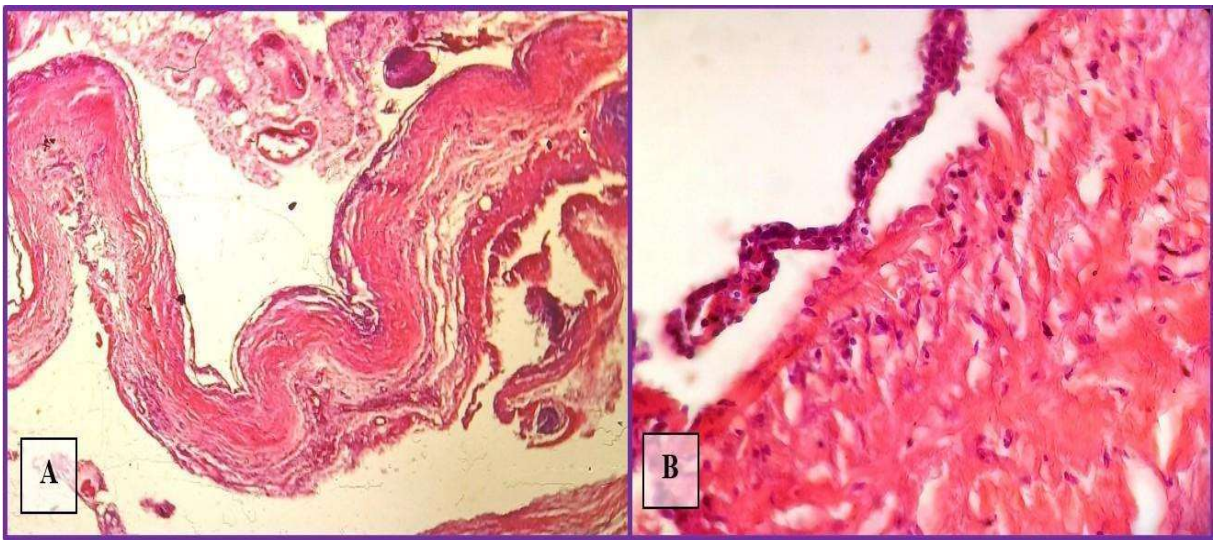

Fig. 2. Histopathology of cystic lymphangiomyoma. (A) Cystic wall containing smooth muscle fibers. (B) Lymphatic-type epithelial lining.

3. Pathology

Gross examination revealed a thick-walled, opened cystic lesion measuring 6 × 3 cm. Microscopically, the cyst wall was lined by a single layer of flattened to cuboidal, bland endothelial-type cells, and the supporting wall contained intersecting bundles of smooth muscle. Immunohistochemistry was strongly positive for D2-40 and CD31 in the endothelial lining, and for smooth muscle actin (SMA) and Desmin in the underlying spindle cells, confirming the diagnosis of cystic lymphangiomyoma.

4. Discussion

Cystic lymphangiomyoma of the uterus is an exceptionally rare entity, with fewer than 10 cases reported in the world literature to date [6,11]. Clinically, these lesions are almost invariably misdiagnosed as ovarian or paratubal cysts due to their location and cystic appearance on ultrasound and MRI, as in our patient [3,10].

Histologically, the differential diagnosis includes lymphangiectatic leiomyoma and adenomatoid tumor [4,7]. Unlike lymphangiectatic leiomyomas, which represent degenerative variants of common fibroids, cystic lymphangiomyomas are true vascular malformations [8]. The distinction is crucial and is reliant on immunohistochemistry: lymphangiomyomas show diffuse positivity for lymphatic markers D2-40 and CD31 in the endothelial lining, whereas lymphangiectatic spaces in leiomyomas typically lack this specific staining pattern [7,11].

Our case mirrors previously reported instances in which conservative, uterus-sparing cystectomy was curative, indicating that radical surgery (hysterectomy) is not required when the diagnosis is correctly established [1,6].

5. Conclusion

Uterine cystic lymphangiomyoma should be considered in the differential diagnosis of pelvic cystic masses in reproductive-age women, even when imaging strongly suggests an adnexal origin. This case demonstrates that laparoscopic cystectomy is a safe and effective fertility-preserving treatment. Pathologists must recognize this entity to ensure appropriate immunohistochemical evaluation and to avoid misdiagnosis as a malignant or degenerative lesion.

Declaration

Ethics approval and consent to participate

The management of this case was conducted in accordance with the ethical standards of the institution.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

RB and DB drafted the manuscript. DB, AH, SBS, and AL contributed to the histopathological diagnosis. All authors approved the final manuscript.

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