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CASE REPORT

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Large leiomyomatosis peritonealis disseminata with ascites and pleural effusion eight years after laparoscopic supracervical hysterectomy: an extremely rare case and review of literature

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Abstract

Leiomyomatosis peritonealis disseminata is a rare benign disease characterized by the appearance of multiple pelvic and abdominal nodules. Its pathogenesis remains unclear and preoperative diagnosis is difficult. Here, we report a case of Leiomyomatosis peritonealis disseminata in a 48-year-old woman with post-polio paralysis and complaints of lower abdominal distending pain. Eight years ago, she underwent a laparoscopic supracervical hysterectomy with electronic power morcellator without an in-bag containment system. Medical imaging showed unilateral pleural effusion, a small amount of ascites and a large cystic-solid mass that filled the pelvis and abdomen, measuring 25 cm in size, following which she underwent bilateral adnexectomy, omentectomy, residual cervical resection and removal of all visible lesions. Histopathology confirmed the diagnosis of Leiomyomatosis peritonealis disseminata. To our best knowledge, large Leiomyomatosis peritonealis disseminata with ascites and pleural effusion has not been previously reported in literature. Our study showed that the patient was disease-free without any evidence of recurrence for over half a year after surgery, suggesting that the surgical resection of the tumors and bilateral adnexal was an effective and reasonable treatment. These observations suggest that more consideration should be given to Leiomyomatosis peritonealis disseminata after laparoscopic myomectomy or supracervical hysterectomy with power morcellation.

Keywords

Leiomyomatosis peritonealis disseminata; Power morcellation; Leiomyoma; Ascites; Pleural effusion

1. Introduction

Leiomyomatosis peritonealis disseminata (LPD), also called disseminated peritoneal leiomyomatosis (DPL), is a rare benign disease of smooth muscle tissue characterized by the formation of multiple nodules spread throughout the peritoneum. It was first described in 1952 and named after Taubert in 1965 [1]. Currently, the pathogenesis of LPD remains unknown, but iatrogenic implantation after morcellation of myoma during laparoscopic surgery has been conclusively established as a risk factor for LPD [2, 3]. Minimally invasive laparoscopic surgery, including myomectomy, supracervical hysterectomy and hysterectomy, is widely used in China. The use of electric power morcellator to remove a large uterus or tumors from a small abdominal incision may result in myoma fragments in the abdominal cavity during laparoscopic myomectomy or supracervical hysterectomy with electrical morcellation, which may progress to iatrogenic LPD even after many years following the sruegry [4]. However, previous studies reported that most iatrogenic tumors were numerous small nodules. Large LPD with ascites and pleural effusion remains extremely rare. Here, to our best knowledge, we present the first case of LPD larger than 25 cm in China appearing after laparoscopic supracervical hysterectomy. The case report was in line with the surgical case report (SCARE) criteria [5].

2. Case report

A multiparous 48-year-old woman with post-polio paralysis was admitted to our hospital with complaints of vague abdominal distending pain in September 2021. She had undergone laparoscopic supracervical hysterectomy at our hospital 8 years earlier because of menorrhagia and multiple uterine fibroids. The uterus and tumors were removed using a power morcellator without an in-bag containment system. Histopathological examination revealed multiple uterine leiomyomas without hypercellularity and mitotic activity. She had no history of prolonged medication use and denied any family cancer history. She started to experience persistent lower abdominal distending pain and slight dyspnea when lying down approximately

3 months ago. Physical examination revealed a firm abdominal mass extending above the umbilicus. Chest computed tomography (CT) showed right-sided pleural effusion (Fig. 1), and abdominal sonography demonstrated that the large tumor was hypervascularized (Fig. 2). Magnetic resonance image (MRI) revealed a small amount of ascites but a large cystic-solid mass that filled the pelvis and abdomen, measuring 25 cm in size (Fig. 3). MRI also detected tumors attached to the bowel. The imaging examination findings aroused the suspicion of a malignant ovarian tumor, metastatic leiomyosarcoma or intestinal tumor with enlarged lymph nodes. However, tumor markers, including carcinoembryonic antigen (CEA), carbohydrate antigen199 (CA199), alpha-fetoprotein (AFP) and human epididymis protein4 (HE4), were within normal range except for cancer antigen 125 (CA125), which showed an elevated level of 175.60 U/mL.

Due to the possibility of malignancy, we performed an exploratory laparotomy and found three mass lesions scattered on the surface of the pelvic peritoneum and omentum. All the mass lesions varied in size from 3.5 to 26 cm, totally weighing 4600 g. The maximum diameter of the largest tumor fed by large vessels from the omentum was 25 cm and weighed 4400 g (Fig. 4). The patient underwent bilateral salpingo-oophorectomy, omentectomy, residual cervical resection and removal of all the lesions. The surgery was completed in 240 min, and the total amount of blood loss was 300 mL. Intraoperative frozen section revealed that the largest tumor was leiomyoma with cystic degeneration and that some tumor regions had active cell proliferation. Histopathological examination showed three discrete masses, two consistent with leiomyomas (Fig. 5A) and one with cystic degeneration of leiomyoma (Fig. 5B). Immunohistochemistry showed that the spindle cells were positive for Desmin (Fig. 5C), smooth muscles actin (SMA) (Fig. 5D) and Ki-67 (2%+) but negative for S-100, CD-34, CD-117 and Dog-1.

Based on the immunohistochemistry results and tumor morphology, the patient was diagnosed with LPD. She was discharged 4 days after the surgery without any postoperative complications and did not undergo any adjuvant therapy. The patient was followed up to 9 months after surgery and remained asymptomatic, without anomalies on imaging.

3. Discussion

LPD is a rare disease characterized by the formation of multiple nodules spread throughout the peritoneum, greater omentum, intestinal mesentery and surface of abdominal organs. LPD is often asymptomatic or presents with nonspecific symptoms. It can mimic ovarian carcinoma, metastatic leiomyosarcoma and peritoneal carcinomatosis. Hence, the preoperative diagnosis of LPD remains quite challenging for clinicians. In some cases, iatrogenic LPD is expected to have hypercellularity and mitotic activity, which requires a differential diagnosis with uterine smooth muscletumor of uncertain malignant potential (STUMP). Howerever, postoperative pathology didn't reveal active cell proliferation and atypia mitosis in our case. Some studies have reported that Ki-67 expression is different in STUMP and leiomyoma (LM), with significantly higher expression in STUMP compared with LM [6, 7]. Our case showed low Ki-67 index (2%). The patient is asymptomatic, and no abnormal imaging results are found one year after surgery. A close and long-term follow-up for this patient will be performed in the further study.

Although LPD is a benign disease, the risk of malignant degeneration reported in literature is 2% to 5% [8, 9]. The etiology and pathophysiology of LPD remain unclear, but some possible causes have been proposed, including hormonal issues, subperitoneal mesenchymal stem cells, metaplasia and genetic or iatrogenic causes following morcellation of the myoma during laparoscopic surgery [2]. The hormonal theory suggests that LPD results from the metaplastic change of subcelomic mesenchymal stem cells with exposure to high endogenous or exogenous sex hormone levels [10, 11]. This hypothesis is supported by cases found in women with tumors secreting estrogens or in reproductive age, especially those who are pregnant, have prolonged use of contraceptive pills or have received hormone replacement therapy [12, 13]. Moreover, tumors in reproductive-aged women can demonstrate spontaneous regression after delivery, oophorectomy or cessation of oral contraceptive use. However, LPD has also been found in menopausal women and even in a man [14, 15]. The patient in our study was not pregnant nor had a history of oral contraceptive use. Therefore, factors other than hormones contributed to her LPD.

Laparoscopy is widely used for myomectomy and subtotal hysterectomy in China. Myomectomy or subtotal hysterectomy is often performed with the use of a morcellator. Recently, iatrogenic tumor implantation following laparoscopic myomectomy or hysterectomy has aroused widespread attention. There has been an increasing number of reports on LPD after laparoscopic myomectomy with power morcellation [16–18], during which small fibroid fragments might be missed and left in the abdominal cavity. They may implant into the peritoneum, mesentery, intestines or omental tissues. Further, they can receive abundant blood supply from new blood vessels in the surrounding tissues, contributing to the development of large LPD. Some literature reviews showed that iatrogenic LPD or parasitic myoma (PM) could be mainly caused by previous laparoscopic surgery with power morcellation [14, 19]. The incidence of LPD or PM after laparoscopic morcellation is reported to range between 0.12% and 0.95% [19]. In April 2014, the FDA recommended manufacturers to incorporate information regarding the risk of malignant and benign uterine tissue spread in the labeling of laparoscopic power morcellators [20], which has since discouraged the use of power morcellator. The patient reported in this present study underwent laparoscopic subtotal hysterectomy using power morcellation, which might have led to the possibility of fibroid fragments into the abdominal cavity during surgery. Thus, to reduce the risks of tissue dissemination, novel methods, including laparoscopic in-bag morcellation techniques [21, 22], the incision of posterior vaginal fornix or mini-laparotomy for tumor retrieval [23, 24] and irrigation after laparoscopic power morcellation [25], have been implemented in the surgery of such cases.

In most case reports, the size of PM or LPD after laparoscopic morcellation was reported to be $\leq$15 cm [26]. Only two cases, one by Yasunori *et al.* [27] and the other by

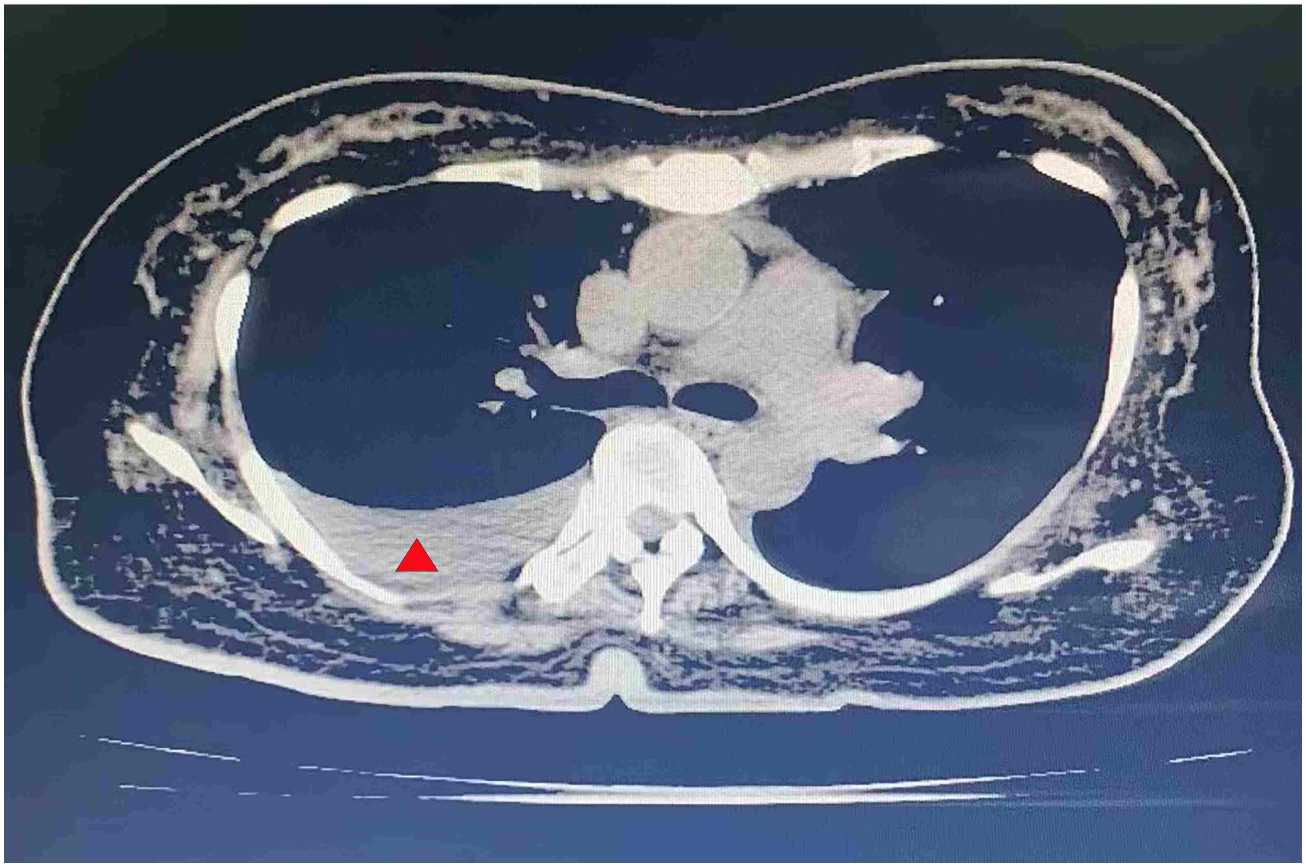

FIGURE 1. Chest computed tomography (CT) showing a relatively small amount of right-sided pleural effusion.

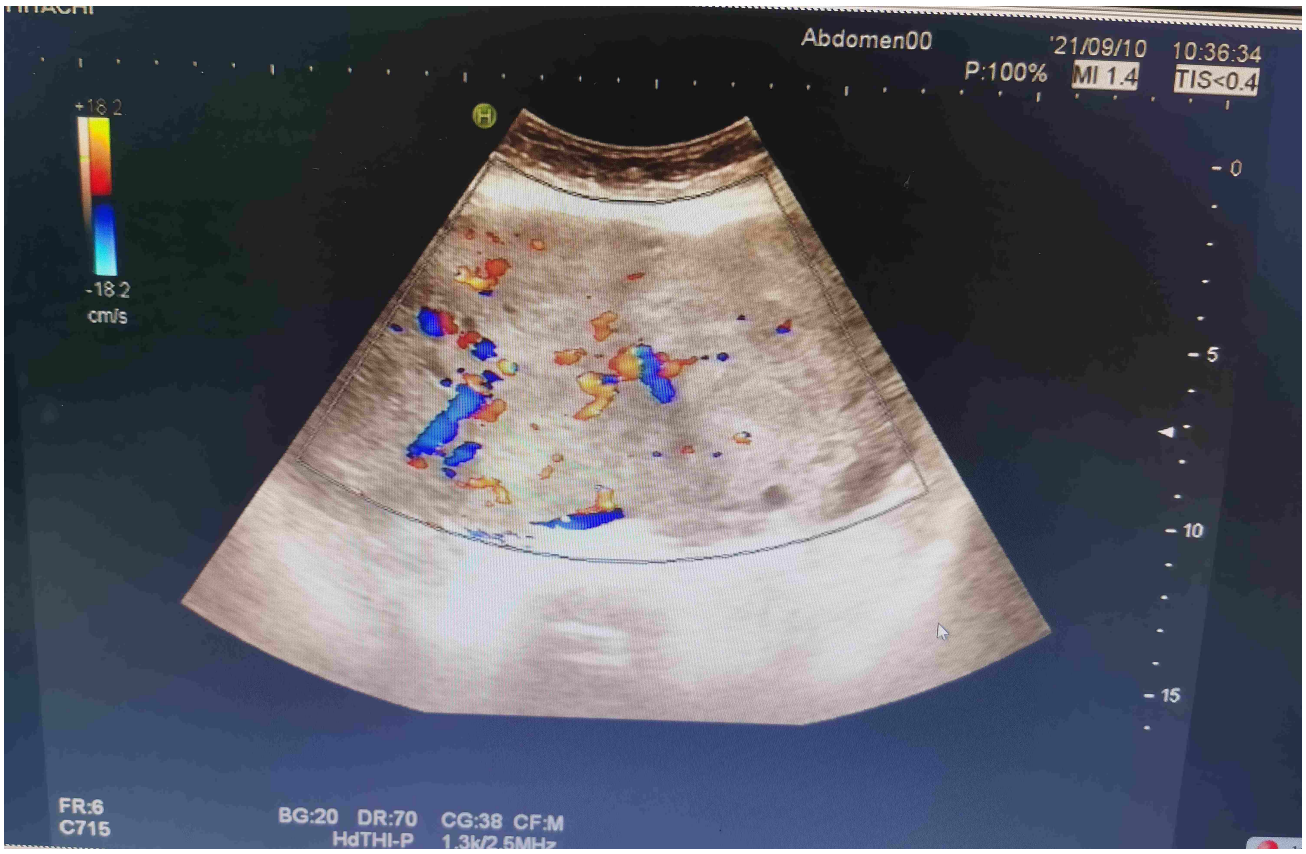

FIGURE 2. Abdominal sonography demonstrating a cystic-solid mixed mass in the abdomen and showing abundant color Doppler blood flow signal inside the solid part of the tumor.

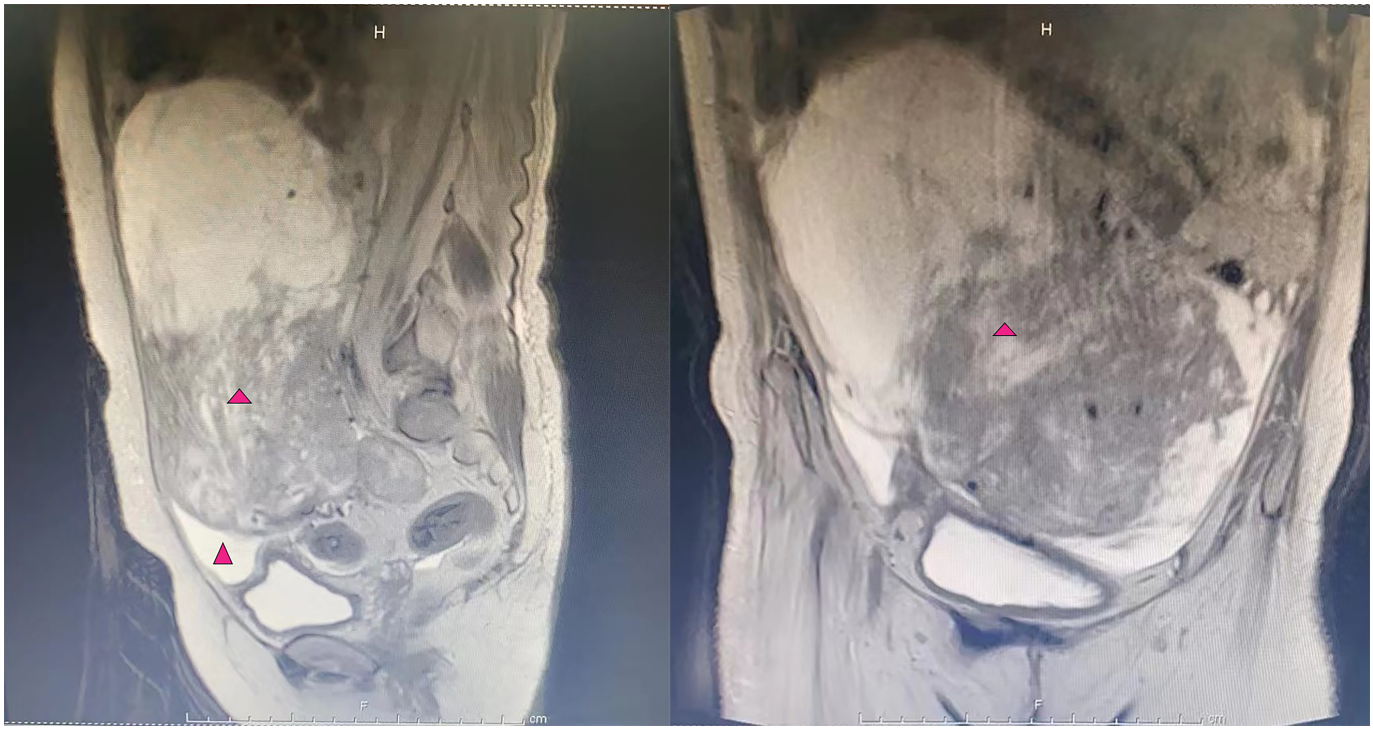

FIGURE 3. Abdominal magnetic resonance imaging showing a small amount of ascites and three masses with short T1 and long T2 signals.

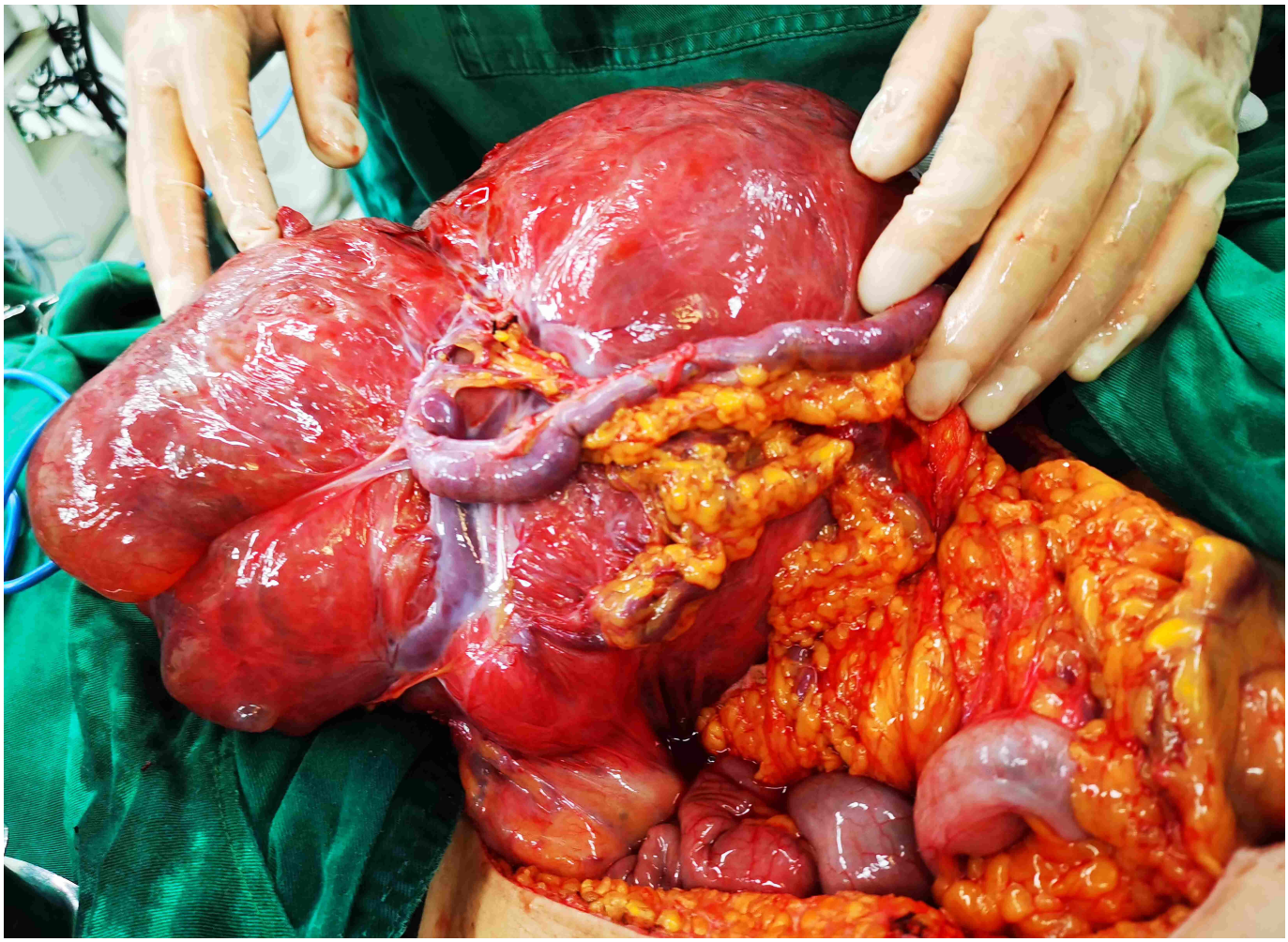

FIGURE 4. Intraoperative image showing a large tumor feeding by large vessels from the omentum.

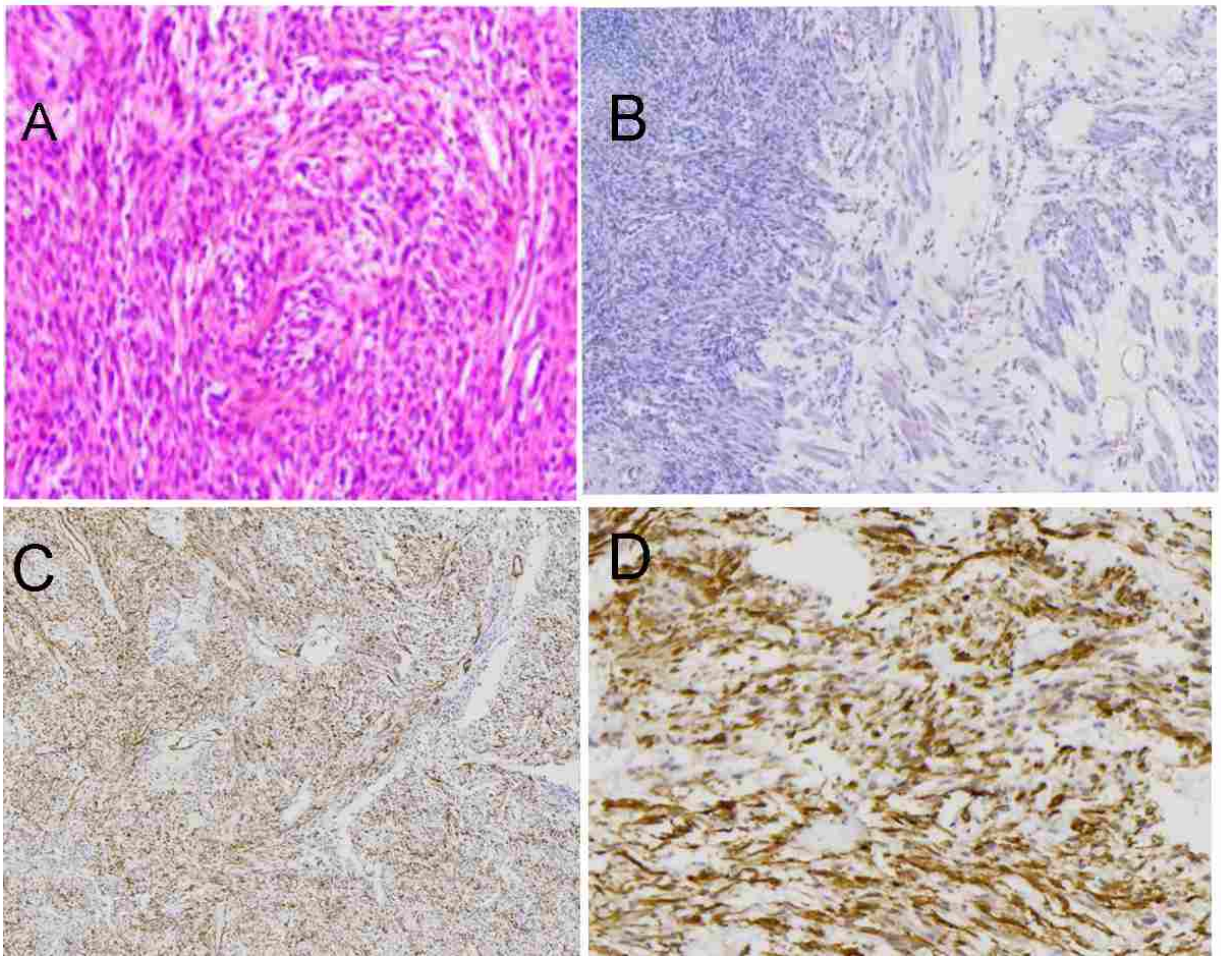

FIGURE 5. Microscopic examination showing three discrete masses. Two consistent with leiomyomas without malignancy (A) and the largest one with cystic degeneration of leiomyoma (B). Desmin (C) and SMA (D) expressions were strongly positive.

Kumar *et al.* [28], were large iatrogenic LPD of size >25 cm. LPD with ascites and pleural effusion measured 25 cm in our case. The reasons for such large tumors could be because they were fed by large vessels of the omental artery [27], yet our patient remained asymptomatic or had less obvious and atypical symptoms for a long period of time.

Until now, there are no standard treatment guidelines for the management of LPD. Soni *et al.* [8] thought that individualized treatment should be considered based on the patient's age, fertility requirements and symptoms, while treatment could be unnecessary in patients with asymptomatic LPD due to difficulties in removing all nodules and the clinically benign course of LPD [8]. Gonadotropin-releasing hormone agonists (GnRHa) and aromatase inhibitors therapy after local excision are feasible for patients with reproductive desires [2]. Aromatase inhibitors are effective for non-resectable tumors in post-menopausal women. However, since LPD has potential risks of malignant transformation, removal of symptomatic or suspected malignant LPD is necessary. Surgical resection of the uterus, bilateral appendage, abdominal mass and even the greater omentum should be considered for women without fertility requirements [14].

4. Conclusion

Large LPD, especially those with ascites and pleural effusion, is often mistaken for peritoneal carcinomatosis, leiomyosarcoma and gastrointestinal or ovarian tumors. The diagnosis of LPD is based on medical history, intraoperative observations and postoperative pathological findings. In this reported case, prior power morcellator use without an in-bag containment system was considered to have played a key role in the pathogenesis of LPD. This rare disease must be considered whenever a patient presents with abdominal masses following myomectomy or supracervical hysterectomy. Long-term follow-up might be necessary to detect LPD after laparoscopic myomectomy or supracervical hysterectomy with power morcellation.

AVAILABILITY OF DATA AND MATERIALS

The data used during the study are available from the corresponding author.

AUTHOR CONTRIBUTIONS

HYC—wrote the manuscript and collected the images. DLL and JLH—analyzed the data. All authors were involved in the management of the case. All authors read and approved the

final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

This case report was approved by the Ethics Committee of Hengzhou People's Hospital (LW2022-001). Written informed consent was obtained from the patient to publish this case report with the corresponding images.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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