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Leiomyomatosis peritonealis disseminate associated with recurrence: A case report

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Summary

Purpose of Investigation: The present study reviewed and analyzed a case of LPD, including the medical history, diagnostic process, and treatment strategy, and it will improve our understanding of the disease. *Material and Methods:* A 25-year-old female was referred to the present gynaecology unit for abdominal pain. A following up investigation was conducted until a gynecological ultrasound revealed a myoma in the lower abdomen. Then surgery was conducted to remove the myoma. *Results:* The ultrasound revealed a mass in the left ovary. Furthermore, the patient was diagnosed with ovarian tumor pedicle torsion and was hospitalized. During the laparoscopic surgery, a 10×7.0-cm myoma in the left posterior uterine wall which was removed using a morcellator was identified. The post-operative pathology report determined that the uterus was rich in leiomyoma cells and bleeding. *Conclusion:* As a rare benign tumor, LPD is commonly seen in female of reproductive years, but its pathogenesis has not been identified. Lack of specific clinical manifestations and signs, LPD is difficult in the preoperative diagnosis.

Key words: Leiomyomatosis peritonealis disseminate (LPD); Myoma of uterus; Laparoscopy; Immunohistochemistry.

Introduction

A 25-year-old female was referred to the present gynaecology unit for abdominal pain. The ultrasound revealed a mass in the left ovary (Figure 1). Furthermore, she was diagnosed with ovarian tumor pedicle torsion and was hospitalized. As the patient presented with no specific symptoms, the patient decided to continue with follow-up examinations only. In July 2009, the ultrasound revealed the other mass in the left ovary, which was very small but continued to grow. In May 2011, a gynecological ultrasound revealed a myoma in the lower abdomen. During the surgery, the laparoscopic identified a 10×7.0-cm myoma in the left posterior uterine wall which was removed using a morcellator. The postoperative pathology report determined that the uterus was rich in leiomyoma cells and bleeding (Figure 2).

Case Report

The female was referred to the present gynaecology unit again for abdominal pain when she was 25-years-old. A gynecological ultrasound revealed a 5.2×3.0-cm mass in pelvic cavity (Figure 3). Three days after the conservative therapy, a large mass with a size of 10.6 cm in diameter was identified at the uterine bladder peritoneal reflection, impossible at the left ovary (Figure 4). Furthermore, she was diagnosed with ovarian tumor pedicle torsion and was hospitalized. The patient underwent an exploratory laparotomy. During the surgery, a myoma was identified at the left uterine wall. Numerous unequally sized leiomyoma tubercles were identified on the uterine surface and a leiomyoma tubercle with size of 2 cm in diameter was identified on the surface of the right fallopian tube (Figure 5). The mass was removed from the uterine bladder peritoneal reflection and frozen section analysis determined a uterine leiomyoma, therefore, a myomectomy was performed. Post-operative pathology determined that the lesion was a leiomyoma (Figure 6).

In September, 2015, the female was referred to the present gynaecology unit again because "physical examination revealed pelvic masses for ten days" when she was 29-years-old. A gynecological ultrasound revealed a 15.2×4.8-cm abnormal echo mass under the right side of diaphragm, while the solid mass on the left side of the attachment was about the size of 8.3×3.3 cm. There was also a 1.2×1.0-cm intrahepatic strengthened echo mass, therefore a hepatic hemangioma was suspected. In October 2015, the giant ovarian cyst and the right side of the diaphragmatic peritoneum (from right to the top of liver and peripheral area of peritoneal attachments, local S7 segment of hepatic parenchymal involvement, but without the clear boundary between ascending colon), and the uterine rectum concave cystic focal were identified by the CT. Adnexal tumor derived from cystadenoma/carcinoma with metastasis. Adnexa of abdominal cavity or the digestive tract derived from a malignant tumor or with metastasis, uterine myoma, pelvic effusion, or pelvic fasciitis (|Figure 7). In October 2015, PET-CT performed in Shanghai Fudan University Affiliated Tumor Hospital revealed a cystic low density in the hepatic surrounding areas, the right lower quadrant mesentery and bilat-

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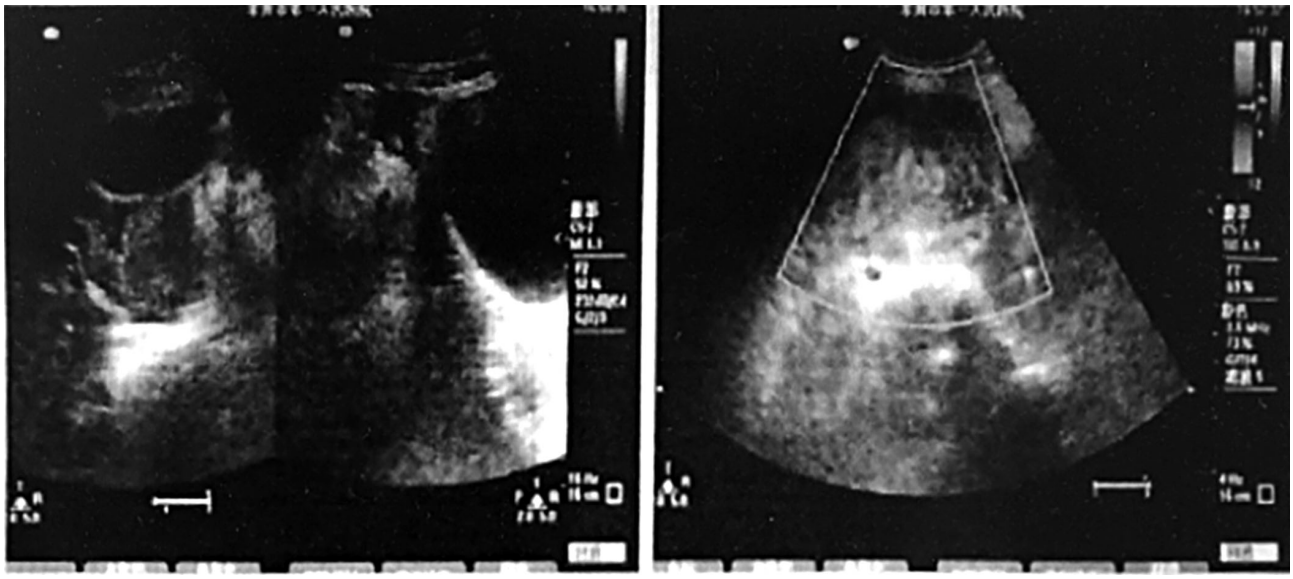

Figure 1. — Ultrasound views of the first surgery.

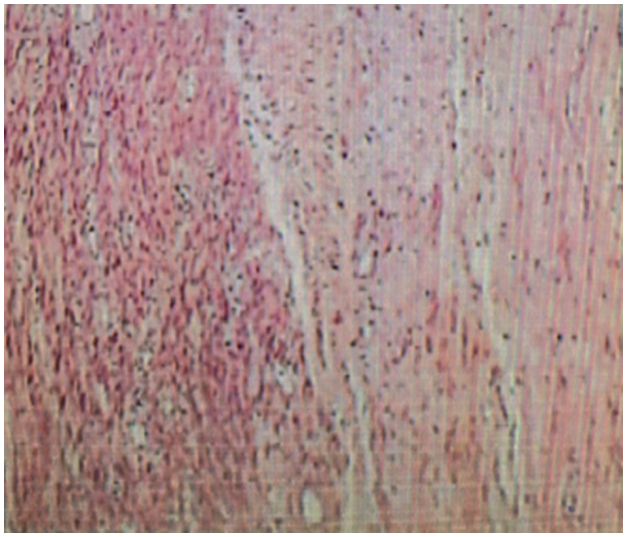

Figure 2. — Histological examination of biopsy. Magnification of nodule showing the transition between stromal endometrial and smooth muscle component (original magnification ×400).

eral accessories, FDG metabolism was slightly higher, the gallbladder was chronically inflamed, right kidney had a calculus, and FDG metabolism of pelvic small intestine increased. The patient gave a birth to a girl by cesarean in April 2015 and no abnormality was seen during the operation. No obvious abnormality was seen in ultrasound examination during postpartum examination on July 19, 2015. In October 2015, metrectomy + uterine bilateral adnexectomy + pelvic exenteration + omentum majus resection + intestinal adhesion separation + appendicectomy were all performed. No obvious myoma bulge was seen during operation. There was poor activity and close adhesion between the bladder and cesarean section scar; no abnormality was seen in bilateral annex. Part of the intestine had close adhesion with the right side of the abdominal wall and the left oviduct ovarian. There was a solid mass of approximately 7.0×8.0 ×8.0 cm in the pelvic cavity and pedicle attached to the omentum majus with an abundant blood supply. A solid mass of about 20.0×10.0×6.0 cm was seen in the right pelvic cavity towards the lower edge of the liver, with no adhesion to the surrounding tissue. The pedicle was at the omentum minus (Figure 8). There was adhesion between appendix and pelvic wall; solid nodules of size range from about 0.5-0.3cm were scattered in the Intestinal surface and mesentery. Neither ascites nor abnormality seen in the surface of liver, stomach, and diaphragm (Figure 8). The postoperative pathological diagnosis after the consultation between Fan Qin and Professors from Jiangsu Province People Hospital was the following: (omentum majus routine examination): (omentum majus): OTI: immunohistochemistry I2015-1853: Desmin (+), Ki67 (about 5%), S100 (-), SMA (+), Vimentin (+), CD117 -), DOG1 (-), and CD34 (vascular +). Combined with HE morphology, it was inferred that this case was smooth muscle tumor. Cells were rich; oncocytes had light to moderate atypia, there was interstitial mucinous degeneration of 8×7×3 cm, The features conformed with potential malignancy, but appeared with malignant behavior in clinic practice. Appendix, entire uterus + bilateral adnexa included proliferative phase endometrium, chronic cervicitis, (left) ovarian graafian follicles; (Right) ovarian corpus luteum hemorrhage and graafian follicles, fallopian tube chronic inflammation, the left broad ligament was wrapped by nodules along with multinucleated giant cell reaction, the right broad ligament had small smooth muscle tumors of about 0.7×0.7 cm size. There was chronic appendicitis, and no obvious change in omentum majus. Intestinal surface and mesenteric nodules inspection revealed multiple smooth muscle tumors (abdominal tumor): smooth muscle neoplasms along with mucous degeneration, 20×16×6 cm in size, and tumor cells had certain atypia - features conformed with the potential malignancy, but appeared malignant in clinic practice. Immunohistochemical analysis included I2015-1863: CD10 (focal+), Desmin (+), Ki67 (about + 5%), and SMA (+) (Figure 9). Postoperative diagnosisi included LPD (potential malignancy),

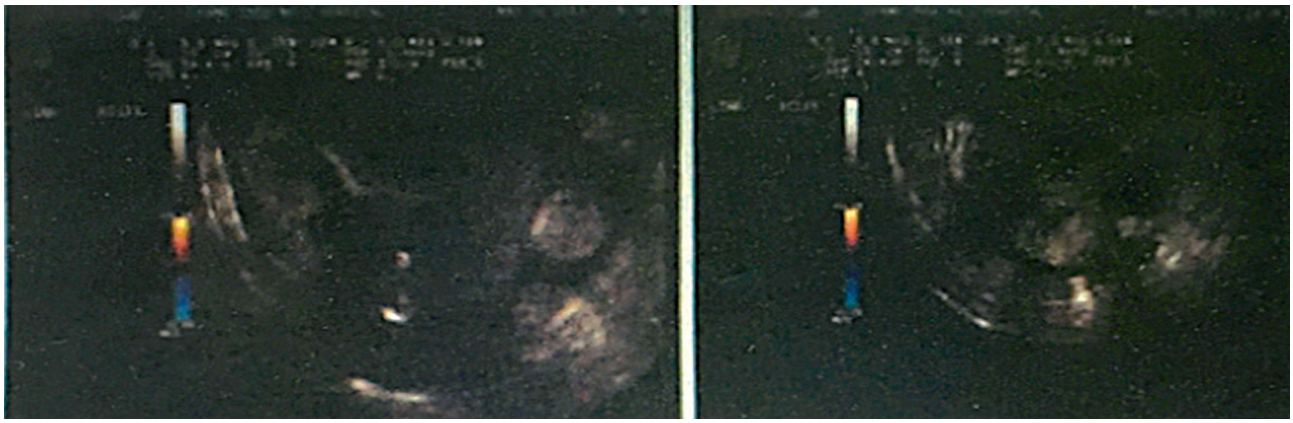

Figure 3. — Ultrasound views of the second surgery.

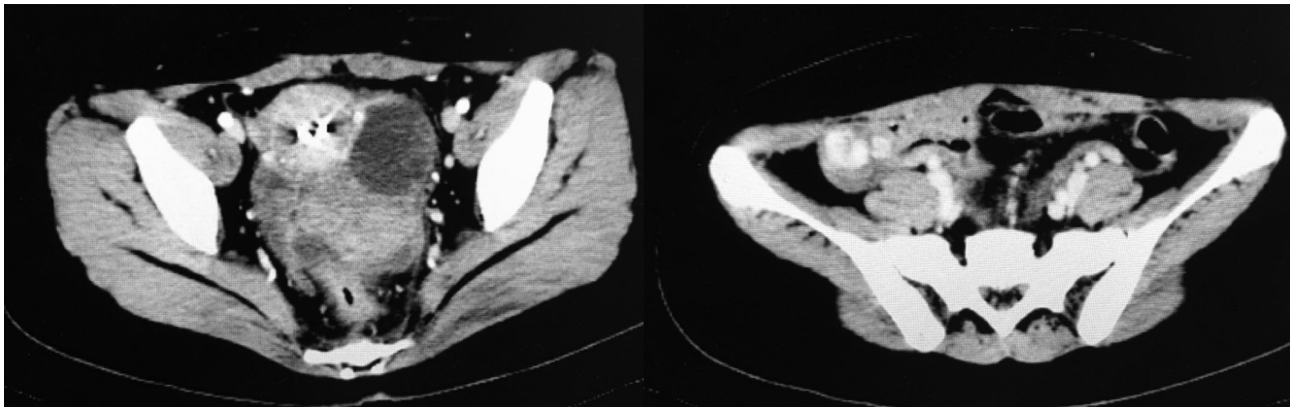

Figure 4. — CT views of the second surgery.

cholecystitis, right kidney calculi, and liver hemangioma. CA12527.49 U/ml. So far, abnormality was found during outpatient follow-up.

Discussion

As a rare benign tumor, LPD is commonly seen in female of reproductive years, but its pathogenesis has not been identified [1]. It may involve in many factors, sex hormones, iatrogenic, genetic, biochemistry of pluripotent stem cells on the peritoneum, endometriosis, etc [2].

The patient developed the first occurrence of LPD when she was 25-years-old, and the recurrence occurred two years later, which met the average onset age of LPD. Research has shown that estrogen levels are closely associated with the development of the disease. The factors of rising estrogen levels, including pregnancy, estrogen replacement therapy, oral contraceptives, and ovarian endocrine tumors, can induce the disease or cause its progression. The patient had three pregnancies: cesarean delivery in 2007, abortion in 2008, and another cesarean section in 2015. Incipient uterine fibroids were found in a medical examination in 2009, shortly after miscarriage but gradually increased. After half a year of the first occurrence of LPD, the patient had physical examinations with no abnormalities, later achieved another pregnancy, cesarean delivery operation without abnormal mass in April 2015, normal postpartum ultrasound in July, and a giant mass found during a physical examination in September, which indicated that the LPD relapsed. On this basis, considering that pregnancy results in higher levels of estrogen, stimulating growth of cell differentiation is the important inducing factor of LPD's occurrence and recurrence. In addition, in 2011 and 2013, the patient has undergone laparoscopic myomectomy in the present hospital, applying morcellation. Some studies suggest that residual fragments in the abdominal cavity can be parasitized in the abdominal wall, intestine, large and small omentum, and other parts, by drawing from the adjacent tissue nutrition to continue to grow. Although the second surgery removed the myoma seen by the naked eye, using specimen bags, and by thoroughly washing the abdominal cavity, this did not prevent recurrence and progression of LPD. Thus, considering the

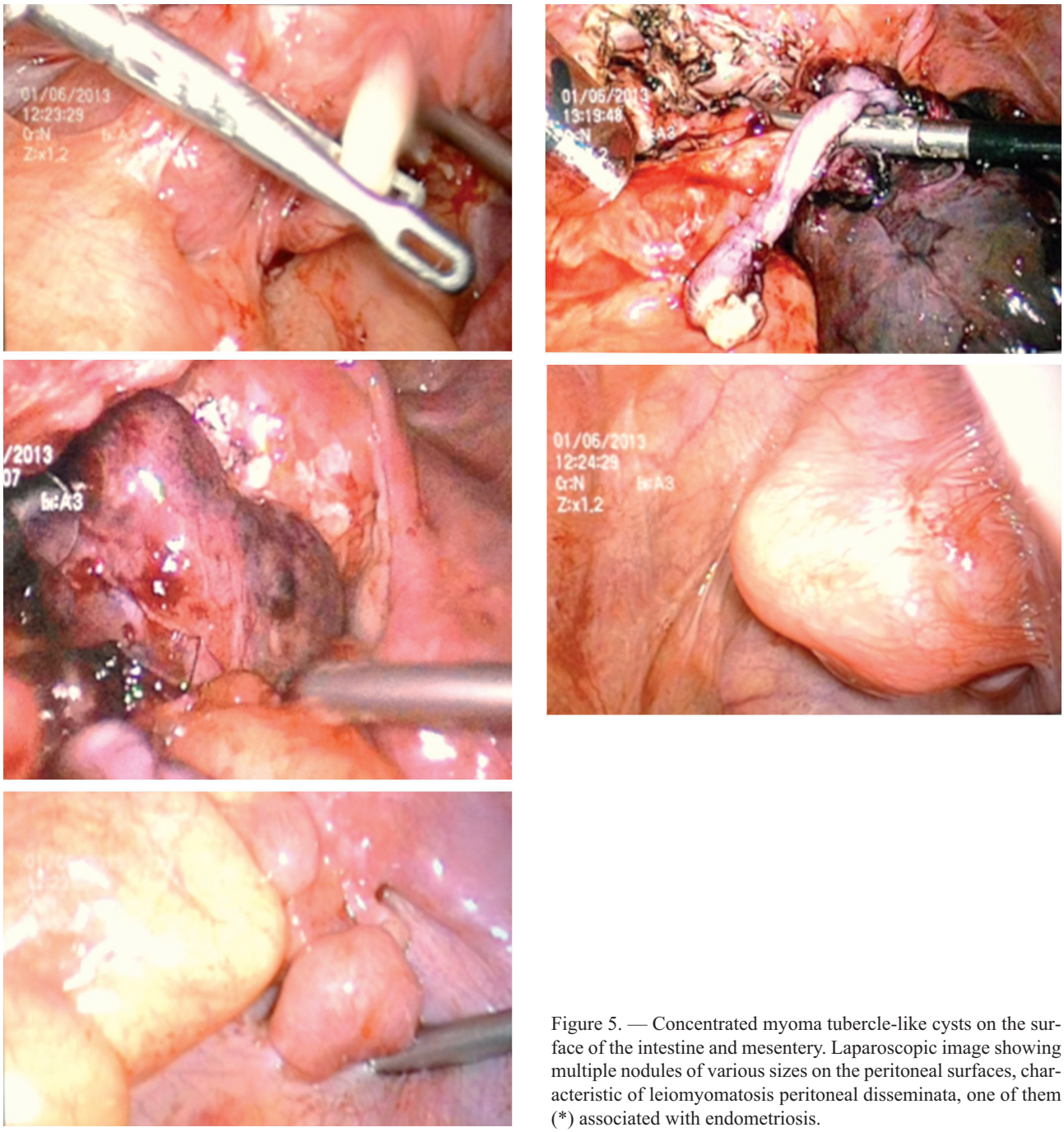

Figure 5. — Concentrated myoma tubercle-like cysts on the surface of the intestine and mesentery. Laparoscopic image showing multiple nodules of various sizes on the peritoneal surfaces, characteristic of leiomyomatosis peritoneal disseminata, one of them (*) associated with endometriosis.

invisible debris of myoma, the incidence of LPD in 2013 and the recurrence of LPD in 2015 may be related to iatrogenic factors, that is, the application of the intraoperative fibroid morcellator.

Due to the lack of specific clinical manifestations and signs, LPD is difficult to diagnose preoperatively, with a misdiagnosis rate of almost 100%. LPD could accidentally be found in a cesarean section or during gynecological surgery because of the pregnancy or other gynecological diseases, physical examination or uterine or pelvic tumor treatment [4]. Diagnosis should be combined with intraoperative findings including tumor morphology, pathology, immunohistochemistry, etc. Laparoscopic exploration is the preferred surgical diagnostic of the disease. Diffuse nodules of varying sizes are scattered in uterine myoma and ovarian and uterine rectal fossa, peritoneum, omentum, even involving lung, spleen, and other distant organs. They have smooth surface, hard texture, and clear boundaries, exactly like malignant tumors planted. Microscopic spindle-like ripe smooth muscle cells could be seen by patho-

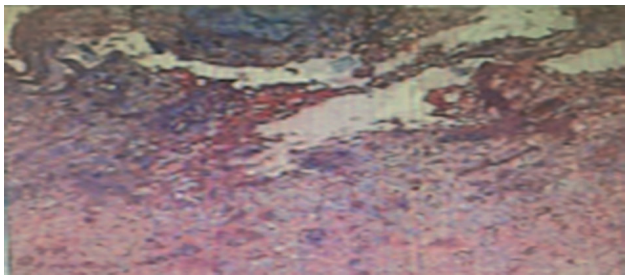

Figure 6. — Postoperative pathological analysis.

[6]. Combined with intraoperative findings and potential malignancy of leiomyoma suggesting in pathology, the final diagnosis of LPD was confirmed. In addition, no standard of treatment currently exists for LPD. Certain studies have considered individualized treatments. An animal experiment demonstrated that long-term high-level progesterone administration may cause mesenchymal stem cells to develop into leiomyomatosis peritoneal lesions. However, a clinical study determined that estrogen stimulates subcelomic mesenchymal cells to proliferate and differentiate into myoblasts, myofibroblasts, fibroblasts, and decidua-like cells.

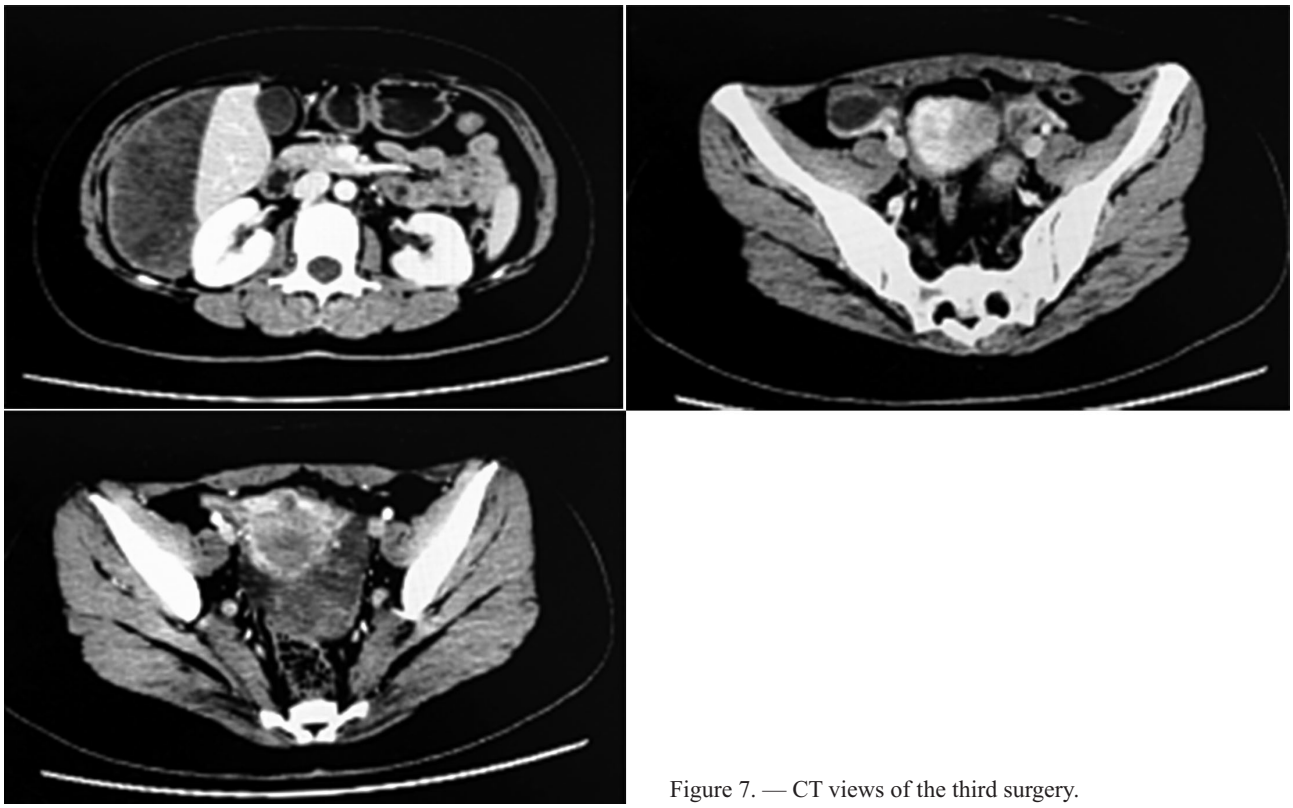

Figure 7. — CT views of the third surgery.

logical examination, and most of them have no cell-free atypia and mitosis [5]. There is no significant difference between its structure and benign leiomyoma.

Before the first clear diagnosis of LPD, the present case was misdiagnosed as ovarian tumor torsion. Not only the myoma necrosis hemorrhage was found in laparoscopic exploration, but the growth of some myoma nodules near the abdominal wall. Pathological diagnosis confirmed it as cellular leiomyoma, combined with intraoperative findings of tumor morphology, distribution, and pathological microscopic findings. The second diagnosis was made due to the giant mass of the pelvis and the abdominal cavity were found in the post-natal physical examination at short-term, combined with a history of diagnosis of LPD recurrence

The pathology results of the three surgeries undertaken by the patient were all benign. However, the disease relapsed and exhibited malignant tendencies. The pathological results of the first surgery reported abundant uterine leiomyoma cells. In the third surgery, the lesion exhibited intestinal and mesentery involvement; however, all of the leiomyoma was removed, therefore, close follow-up examinations are still required.

In conclusion, LPD is rare and difficult to diagnose, however, the majority of prognosis is good, as the minority become malignant. The majority of LPD patients have a medical history of laparoscopic myomectomy for uterine fibroid. The use of laparoscopic power morcellation may contribute to the development of LPD, therefore, the spe-

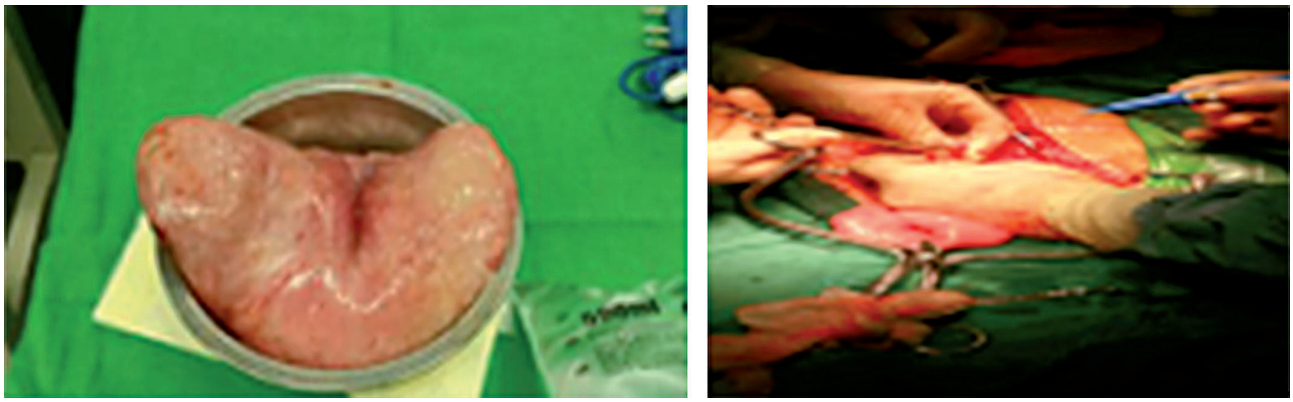

Figure 8. — Abdominal giant fibroids and the small intestine in myoma.

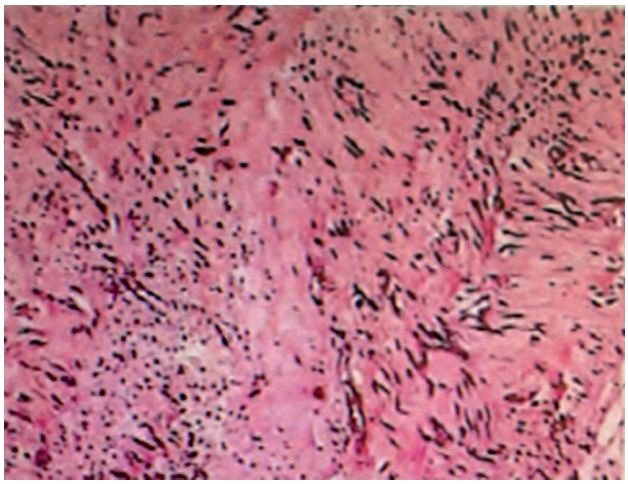

Figure 9. — Post-operative pathological analysis.

cific surgical approach used in laparoscopic myomectomy should be carefully considered, and protective measures should be taken to prevent myoma fragment spreading if laparoscopic power morcellation is used.

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