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A case report of postmenopausal leiomyomatosis peritonealis disseminata

H. Zhou[1*], W. Chen[2*], X. Zhang[1], L. Wang[1]

[1] Department of Gynecology, the First Hospital of Jilin University, Changchun
[2] Department of Orthopedics, China-Japan Union Hospital, Jilin University, Changchun (China)

Summary

Leiomyomatosis peritonealis disseminata (LPD) is a rare and benign disease, mainly occurred in reproductive age. The authors report a postmenopausal Chinese woman with a large, hard, and unmovable mass in lower abdomen. Computerized tomography revealed a mass of 13.4× 12.0 cm with irregular edges adhering to surrounding organs, with multiple nodules around the abdominal aorta, and in the pelvic peritoneum, mesenteric, and ileocecal regions. She had abnormal higher levels of serum carbohydrate antigen (CA) 125, but normal levels of serum CA 19-9, alpha-fetal protein, and carcinoembryonic antigen. After being treated with epirubicin, carboplatin, and interventional embolization, her abdominal mass was reduced by about 10% and she received a successful radical surgery, including pelvic tumor resection, total hysterectomy, bilateral oophorectomy, omental resection, appendectomy, rectal resection, and anastomosis. Her pelvic mass weighed 1,350 grams and histologically diagnosed as LPD. Immunohistochemistry of the mass showed positive staining of vimentin, caldesmon, smooth muscle actin (SMA), b-catenin, and Bcl-2, but negative for CD10, CD34, CD99, CD117, S-100, progesterone receptor, and DOG1 staining. She had no disease recurrence during a year of follow up.

Key words: Leiomyomatosis peritonealis disseminata; Embolization; Postmenopausal women.

Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a rare and benign disease characterized by multiple nodules of proliferative smooth muscle cells. Most LPD occur in reproductive age; there are only a few cases of LPD in postmenopausal women in the literature [1-4]. Here, the authors report a postmenopausal Chinese woman with a large LPD mass who received a radical surgery following an interventional embolization to reduce the tumor size.

Case Report

The case report was approved by the Ethics Committee of The First Hospital of Jilin University and written informed consent was provided by the patient.

A 64-year-old Chinese woman was admitted in March 2010 because of acute dysuria and urinary retention. She became menopausal at the age of 48 and once had an ovary surgery due to ovarian teratoma. She was incidentally found with a "pelvic mass" in 2007 and found to have a large and unresectable mass at the local hospital in May 2009. In December 2009, she was diagnosed with a smooth muscle tumor by biopsy examination.

Her admitting physical examination showed a palpable large mass in lower abdomen. Both pelvic and digital rectal examinations showed a large, hard, and unmovable mass. Color Doppler ultrasound showed a substantive echo of a large irregular shaped mass and a mixed echo measuring 59×48 mm in the right upper region of this mass. Abdominal CT scan revealed a soft mass of 13.4×12.0 cm with irregular edges appeared to adhere to surrounding organs, with multiple nodules around the abdominal aorta, and in the pelvic peritoneum, mesenteric, and ileocecal regions (Figure 1A). Laboratory examinations revealed 127.8 U/ml of serum carbohydrate antigen 125 (CA125, normal range: < 35 U/ml) and normal levels of CA 19-9, alpha-fetal protein, and carcinoembryonic antigen. The primary diagnosis was pelvic tumor with potential malignancy.

Because the tumor was large and adhered to other organs, it was very difficult to resect completely. The patient was treated with 60 mg epirubicin and 200 mg carboplatin through uterine arterial perfusion twice and treated with an interventional embolization. The treatments alleviated her dysuria and reduced the pelvic mass by about 10%. In following exploratory laparotomy, a large tumor (14×13×11 cm) was found in the pelvic cavity which was irregular, hard, and adhering to the surrounding bowel, uterus, and left ovary, as well as adnexa, with multiple hard nodules on the surface of the sigmoid colon mesentery, peritoneum, and appendix (Figure 1B).

The patient underwent pelvic tumor resection, total hysterectomy, bilateral oophorectomy, omental resection, appendectomy, rectal resection, and anastomosis. The tumor weighed at 1,350 grams and postoperative pathological diagnosis was LPD (Figure 1B). Microscopic examination revealed high mitotic activity, spotty haemorrhagia, and anemic infarcts. Immunohistochemistry showed positive staining of vimentin, caldesmon, smooth muscle actin (SMA), b-catenin, and Bcl-2 (Figure 1C), but negative for CD10, CD34, CD99, CD117, S-100, progesterone receptor, and DOG1 staining, consistent with previous findings in post-menopausal LPD [5-8]. The patient recovered from the surgery and was discharged on day 13 post operation. One year later, her abdominal ultrasound showed no obvious recurrence of the tumor.

[*] These authors ontributed equally to this work.

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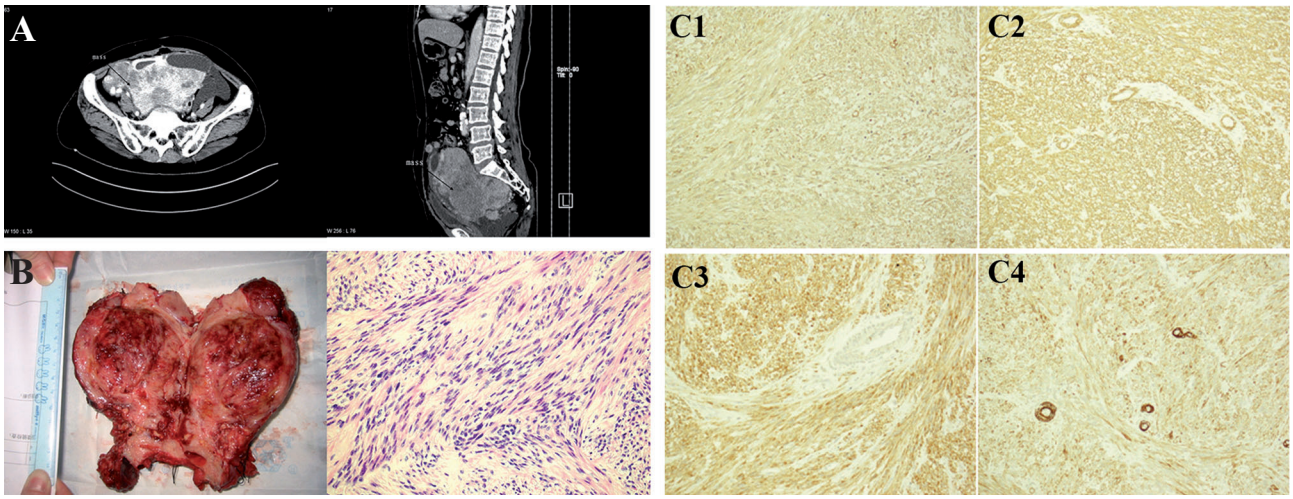

Figure 1. — (A) Abdominal CT scan before surgery. Left panel is horizontal view of the CT scan in the lower pelvic cavity. Right panel is side view of the CT scan in the abdomen. The arrows indicate the location of large masses. (B) Histological examination of tumors. The dissected tumors are measured and subjected to tissue sections, followed by H&E staining, and examined under a light microscope (magnification ×200). (C) Immunohistochemistry analysis of tumor sections. Tumor tissue sections (five mm) are prepared and stained with anti-Vim (C1), anti-SMA (C2), anti-Bcl-2 (C3), and anti-caldesmon (C4), respectively. Subsequently, the bound antibodies are detected by HRP-conjugated second antibodies and DAB, followed by imaging. Data shown are representative images from each group of the sections.

Discussion

Most patients with LPD are asymptomatic or have nonspecific symptoms and are difficult to be accurately diagnosed [9]. The present authors did not definitively diagnose the patient before surgery because of several masses in the pelvic cavity, like a metastatic cancer from CT scanning. It should be considered a LPD if a patient presents with a long history of pelvic tumor, but without ascites, other malignancy-related clinical manifestations, and laboratory findings. Pathologic examination is critical for the diagnosis of LPD in both menopausal and postmenopausal women. Histological examination resembles normal smooth muscle cells and a little mitotic activity or atypical nuclei in LPD. Immunohistological staining usually reveals typical smooth muscle tumors with high levels of vimentin, desmin, SMA, and muscle-specific actin expression in tumor sections. Conservative and surgical treatments to preserve fertility are reasonable for women in the reproductive age, particularly for those with fertility requirement, and radical surgery is recommended after giving birth or for those who are at a high-risk for malignant transformation [10].

Conclusion

Currently, there is not a standard protocol on the management of patients with LPD. Personalized management of LPD may be valuable, and selection of individual approaches depends on age, fertility requirement, hormonal status, complications, and complaints of individual patients.

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References

[1] Brumback R.A., Brown B.S., Sobie P., Shapiro M.J., Wallinga H.A.: "Leiomyomatosis peritonealis disseminata". *Surgery*, 1985, *97*, 707.
[2] Halama N., Grauling-Halama S.A., Daboul I.: "Familial clustering of Leiomyomatosis peritonealis disseminata: an unknown genetic syndrome?". *BMC Gastroenterol.*, 2005, *5*, 33.
[3] Sutherland J.A., Wilson E.A., Edger D.E., Powell D.: "Ultrastructure and steroid-binding studies in leiomyomatosis peritonealis disseminata". *Am. J. Obstet. Gynecol.*, 1980, *136*, 992.
[4] Zheng P., Xu Y.: "Leiomyomatosis peritonealis disseminata. Two case reports". *Chin. Med. J. (Engl.)*, 1996, *109*, 334.
[5] Danikas D., Goudas V.T., Rao C.V., Brief D.K.: "Luteinizing hormone receptor expression in leiomyomatosis peritonealis disseminata". *Obstet. Gynecol.*, 2000, *95*, 1009.
[6] Sharma P., Chaturvedi K.U., Gupta R., Nigam S.: "Leiomyomatosis peritonealis disseminata with malignant change in a post-menopausal woman". *Gynecol. Oncol.*, 2004, *95*, 742.
[7] Strinic T., Kuzmic-Prusac I., Eterovic D., Jakic J., Scukanec M.: "Leiomyomatosis peritonealis disseminata in a postmenopausal woman". *Arch. Gynecol. Obstet.*, 2000, *264*, 97.
[8] Takeda T., Masuhara K., Kamiura S.: "Successful management of a leiomyomatosis peritonealis disseminata with an aromatase inhibitor". *Obstet. Gynecol.*, 2008, *112*, 491.
[9] Al-Talib A., Tulandi T.: "Pathophysiology and possible iatrogenic

cause of leiomyomatosis peritonealis disseminata". *Gynecol. Obstet. Invest.*, 2010, *69*, 239.

[10] DeAngelis S.L., Harris R.R., Maiman M.A., Knochenhauer E.S.: "Live birth of a normal mature healthy baby by in vitro fertilization-embryo transfer (IVF-ET) after an exploratory laparotomy and multiple myomectomy for leiomyomatosis peritonealis disseminata". *Fertil. Steril.*, 2009, *91*, e1915.

Corresponding Author:
L. WANG, M.D.
No. 71 Xinmin Street,
Changchun 130021
Jilin Province (China)
e-mail: wanglina1973@163.com