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Leiomyomatosis peritonealis disseminata: A case report of recurrent presentation and literature review

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ABSTRACT

INTRODUCTION: Leiomyomatosis peritonealis disseminata (LPD) is an uncommon disease featured by the presence of multiple nodules of smooth muscle cells scattered in the abdominal cavity. To date only about 150 cases have been reported in literature. We report a case of recurrent LPD after laparotomy.
CASE PRESENTATION: In March 2016 a 36-year-old female, with a history of multiple previous laparoscopic myomectomies, consulted her gynaecologist complaining abdominal pain; a MRI was performed and reported multiple pelvic masses, subsequently excised during laparotomy. The patient refused a total hysterectomy with bilateral salpingo-oopherectomy so a close follow-up was recommended. In November 2017 when a new MRI revealed recurrency of the disease, a second laparotomy is performed and all visible nodules are excised. The histological exam confirms LPD diagnosis. On follow-up after three months the patients is completely asymptomatic.
DISCUSSION: Differential diagnosis of LPD is challenging due to its similarity to carcinomatosis and to other benign abdominal disorders. Malignant transformation is rare, but it may occur, so a close follow-up is necessary. Even if there is no consensus regarding the treatment, hormonal therapy is probably the best first line approach, while surgery should be the second choice.
CONCLUSIONS: LPD is an uncommon but potentially severe disease. In our opinion larger studies are necessary to improve our diagnostic effectiveness and to define the best therapeutic strategy.

1. Introduction

Leiomyomatosis peritonealis disseminata (LPD) is an uncommon disease characterized by the presence of multiple nodules composed of smooth muscle cells, located in the abdominal cavity, both in peritoneal and subperitoneal spaces. This disease was first described by Wilson and Peale in 1952 [1], and later named LPD by Taubert et al. [2].

The disease is usually observed in premenopausal women and rarely seen in postmenopausal women and men. Up to now only about 150 cases have been reported in literature.

The aetiology remains unclear, but different hypotheses have been proposed: a hormonal theory with subperitoneal mesenchymal stem cells metaplasia, a genetic theory and an iatrogenic origin subsequent to laparoscopic surgery [3].

We report a case of recurrent LPD after laparotomy in woman with a history of multiple laparoscopic myomectomies, occurred in our academical institution.

2. Case report

The work has been reported in line with SCARE criteria [4].

A 36-year-old female was admitted to our Institution for elective laparotomy after a follow-up MRI reported disease recurrence, with appearance of two oval nodules (6 cm and 5 cm respectively), located subcutaneously in the left-anterior abdominal wall (Fig. 1); three nodules (2 cm) behind them, one nodule (2 cm) strictly adherent to the left ovary, two nodules (1 cm) behind the cervix uteri, one nodule (1 cm) below the cecum and several small nodules in the greater omentum. There was neither ascites nor lymphadenopathies and all the lesions showed benign appearance with non-invasive behaviour.

The patient had previous history of one Caesarean section in 2011, an abortion in August 2017 and multiple laparoscopic

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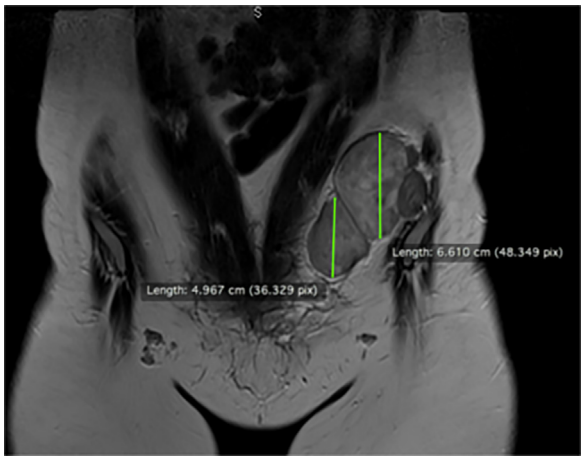

Fig. 1. Nodules of the left-anterior abdominal wall.

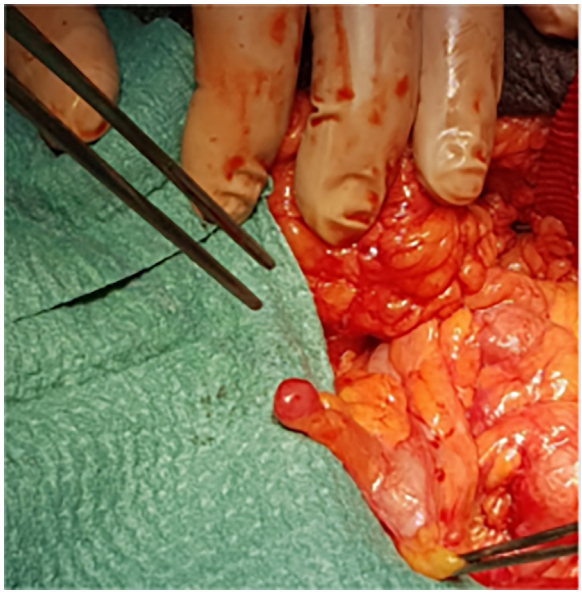

Fig. 2. Nodule of the sigmoid peritoneal surface.

myomectomies. She had no previous history of prolonged use of contraceptive pills. In 2005 the patient went to her gynaecologist after the appearance of vaginal bleeding and a uterine myoma was found, so she underwent a laparoscopic myomectomy. In the next ten years two other myomectomies were performed. In 2016 the patient consulted again her gynaecologist complaining of abdominal pain and abdominal swelling: an abdominal and pelvic ultrasound were performed detecting multiple pelvic masses, subsequently confirmed by MRI. The patient was referred to our medical centre on March 2016, and underwent an exploratory laparotomy that revealed multiple pelvic and omental neoformations, which were excised. Intraoperative frozen section revealed spindle cells without atypia, while at histopathology the tumors were composed by benign smooth muscle cells, without cell atypia or necrosis, similar to leiomyomas. The histology in combination with patient's history and intraoperative findings, confirmed the diagnosis of LPD. The patient was discharged after a ten days regular postoperative course. After the histological diagnosis a total hysterectomy with bilateral salpingo-oopherectomy was proposed to the patient but she refused because of her reproductive plans. A close radiological follow-up of the patient was then recommended and no further treatment was administered.

On November 2017, after MRI found new nodules, the patient was referred again to our department. At the moment of hospitalisation, she was completely asymptomatic, her physical examination was unremarkable and blood test, ECG and chest X-ray showed no anomalies, so a new exploratory laparotomy was scheduled. The patient preoperatively refused again the possibility of total hysterectomy with bilateral salpingo-oopherectomy.

At laparotomy, multiple grey nodules (Figs. 2 and 4), varying in size between 2 mm and 2 cm, were found on the surface of right and left ovary, uterus, cecum, sigmoid colon and greater omentum. The surgeons carefully excised all the visible lesions and partial omentectomy was made.

Another two bigger nodules, about 6 cm and 5 cm of diameter respectively, were founded in the left-anterior abdominal wall, below a previous laparoscopic incision, and carefully excised (Fig. 3). Accurate haemostasis was performed and no drainage was placed to prevent further spread of neoplastic cells in the peritoneal cavity and to the abdominal wall. No intraoperative or postoperative complications were reported and patient was discharged 4 days after surgery. On follow-up after three months the patient was asymptomatic, with no evidence of recurrence clinically and on ultrasound; treatment with ulipristal acetate was about to be initiated, but in consequence of the review of ulipristal acetate started by EMA [5], due to cases of serious liver injury in women taking ulipristal acetate for uterine fibroids, we decided to wait for the results.

Histological exam showed multiple nodules with features of typical leiomyoma: tumors were formed by whorled bundles of smooth muscle cells separated by vascularized connective tissue, with scattered lymphocytes. Neither necrosis nor atypia were present. Tumor cells expressed almost 100% positivity for ER and

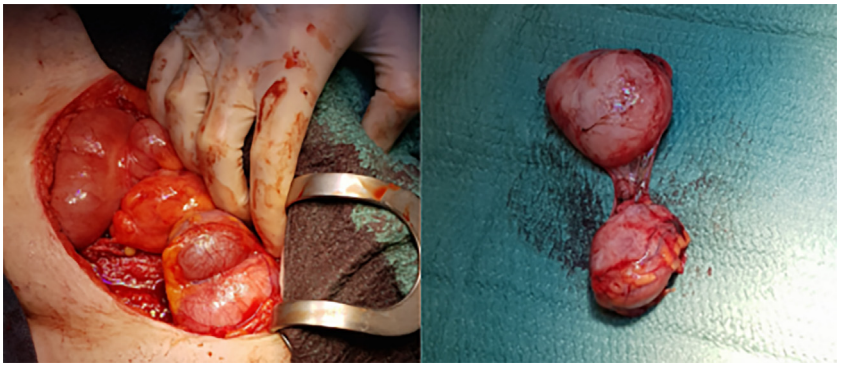

Fig. 3. Two large nodules of the anterior abdominal wall and the two nodules once removed.