

# Two cases of leiomyomatosis peritonealis disseminata and Review of the Literature

## Abstract

Leiomyomatosis peritonealis disseminata (LPD) is an extremely rare condition. Here we present two cases of LPD in our hospital. We discuss the aetiology and treatment of this disease.

## Review Article

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## Introduction

Leiomyomatosis peritonealis disseminata (LPD) is an extremely rare condition. It is also referred to as disseminated peritoneal leiomyomatosis which is characterized by the presence of multiple peritoneal and subperitoneal nodules composed of bland smooth muscle cells. This disease was first described by Willson & Peale [1]. It was designated as LPD by Taubert et al. [2]. It was diagnosed mainly in the reproductive-aged females. But a few cases occur in postmenopausal women and in men. Since then, less than 200 cases have been reported in the literature. Here we present two cases of LPD in our hospital.

## Case Series

### Case 1

A 38 years old patient (granda1, para1, aborta0) admit to our hospital because of the gynecological ultrasound reveal a 5.0 cm mass in the pelvis. The patient present with no specific symptom. No abnormality was identified in the liver, spleen ultrasound. The heart and lung assessment no abnormality. In addition, cancer antigen 125 and epididymis protein 4 and alpha-fetoprotein in normal range. She underwent laparoscopic myomectomy for uterine fibroid in local hospital five years ago. During surgery a myoma with 5cm diameter was identified in the posterior uterine wall. It was removed by morcellator. The post-operative pathology reported that rich in leiomyoma cells. 2 years later she had laparoscopic hysterectomy due to the relapse of the uterine fibroid in the other hospital. During surgery two myoma was identified in the uterine. The post-operative pathology reveal that leiomyoma and positive for alpha-smooth muscle antibody and desmin. The exploratory laparotomy was performed. During surgery myoma tubercle were identified on the surface of rectum. The size was about 6cm. There were no abnormalities in the omentum, mesentery, sigmoid colon, intestine and liver. The lesion excision was performed. The frozen section indicated mesenchymal tumor. The post-operation pathology determined a diagnosis of leiomyomatosis peritonealis disseminate with no mitosis and necrosis. The result of immunohistochemistry was mostly desmin+, SMA+, ER+, PR+. The patient was uneventful and monitor by fellow up. There is no recurrence in 60months.

### Case 2

A 34 years old patient (granda1, para1, aborta1) admit to our hospital because of the gynecological ultrasound reveal pelvic mass. The patient present with no specific symptom. The CA125 and epididymis protein4 was normal. She had laparotomy hysterectomy and salpino-oophorectomy in other hospital because of uterine myoma 10 years ago. The final pathology revealed uterine leiomyoma. There were no abnormality in the heart and lung. The exploratory laparotomy was carried out. In numerous leiomyoma-like implants were present in the abdomen and pelvis. There were also prominent at the surface of multiple organs including the omentum, mesentery, sigmoid colon, and intestine. The size varied from a diameter of a few millimeters (miliary seeds) to that of 6 cm. Partial omentectomy and palliative debulking of the large nodules and appendicectomy were performed. On histopathology all implanting tumors consisted predominantly of bland smooth muscle cells. On immunohistochemistry the tumor cells were mostly desmin+, SMA+, ER+, PR+. The final diagnosis was LPD. The patients was well and no recurrence in fellow up (48 months).

## Discussion

LPD predominantly occurs in females of reproductive age. But the pathogenesis of LPD is poorly understood. The condition may be caused by estrogen and progesterone, peritoneal metaplasia, growth factors, iatrogenic factors or genetics [3-9]. A possible origin of the smooth muscle cells in LPD is thought to be the submesothelial multipotential cells. These cells have also been referred to as the secondary müllerian system. It seems to maintain its ability to differentiate into specialized epithelia or stroma of the müllerian system in the adult life. But it lacks organization of the fallopian tubes, uterus, and upper vagina. The stimuli to induce smooth muscle differentiation is hormonal, genetic, or

both. This hypothesis is persuasive because LPD occurs mainly in the reproductive-aged females. Moreover, the results of positive immunoreactivity for estrogen and progesterone receptors in the smooth muscle cells of LPD. Furthermore the cases of spontaneous regression of LPD after discontinuation of hormonal agents. So that it suggested that an increased sensitivity to estrogen in susceptible patients predisposes them to the development of LPD.

Although the prognosis of LPD is good. It is rare and difficult to be diagnosed [10]. It generally presents with no clinical symptoms. The symptom of gastralgia or abdominal distension do occasionally occur. It exhibits recurrent and malignant tendencies. The minority of this disease become malignant [11,12]. The histological atypia nuclear polymorphism, hyperchromasia, tumor cell malignant and increased mitotic figure are sign of malignant transformation. Until now there is a lack of pre operative diagnostic methodology for LPD. The diagnosis is predominantly relied on observations during surgery intraoperative or post operative pathology results. LPD lesions always appear at the uterine bladder peritoneal reflection, the omentum majus, mesentery and surface of the small intestine, or at the colon, uterus, ovary, oviduct or pouch of Douglas. The LPD follow laparoscopic myomectomy as our case one. It has several different characters from the classical LPD. First the patient had history of laparoscopy with tissue morcellation and original pathology was benign leiomyoma. Second the nodule appear to be large and less numerous.

There is no standard treatment currently exists for LPD. The majority of studies propose that females of reproductive age should undergo lesion excision followed by discontinuation of hormonal stimulation. such as the termination of oral contraceptive drugs, or the administration of gonadotropin releasing hormone agonists (GnRh a), aromatase inhibitors. The treatment of LPD should be individualized. It is determined by the age, the severity of disease, the symptom. The total abdominal hysterectomy, salpingo oophorectomy, omentectomy and debulking may be the most appropriate for the patients who do not desire to bear children [13,14].

Some cases of LPD have a medical history of laparoscopic myomectomy for uterine fibroid. The use of laparoscopic power morcellation may lead to the development of LPD.

Therefore the FDA published the following advice in April 2014:

- i) Laparoscopic power morcellation during hysterectomy or myomectomy for uterine fibroids should be discouraged;
- ii) all the possible treatment strategies should be considered for the treatment of female patients with symptomatic uterine fibroids, and the benefits and risks of each should be discussed with the patient; and;
- iii) for those patients in whom laparoscopic power morcellation is considered to be the most appropriate therapeutic procedure, a specimen bag should be used during morcellation, with the aim of containing the uterine tissue and minimizing the risk of dissemination throughout the abdomen and pelvis.

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