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Leiomyomatosis peritonealis disseminata: A case report and meticulous review of the literature

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ABSTRACT

INTRODUCTION: Leiomyomatosis peritonealis disseminata (LPD) is a peculiar benign clinical disorder characterized by proliferation of peritoneal and subperitoneal nodules. LPD is a difficultly diagnosed benign disease that rarely degenerates into malignancy.
PRESENTATION OF CASE: A 40-year-old Caucasian female with vaginal bleeding proceeded to our institution for elective excision of abdominal and pelvic masses which were firstly considered as leiomyosarcomas. The histologic diagnosis of the mass lesions revealed smooth muscle benign cells. This is the first case of LPD reported in Greece. A meticulous review of the literature was conducted as well.
DISCUSSION: The differential diagnosis of LPD is difficult due to its clinical resemblance with peritoneal carcinomatosis or metastatic lesions and with benign metastasizing leiomyoma (BML) as well. Etiological factors, pathophysiology and clinical manifestations which lead to a safe diagnosis of LPD are adequately described.
CONCLUSION: Surgeons' thorough knowledge concerning this rare clinical condition is fundamental and crucial in order to establish a correct diagnosis and assert the appropriate treatment and the minimization of the probability of malignant transformation of LPD.

1. Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a peculiar clinical disorder characterized by proliferation of peritoneal and subperitoneal nodules, primarily consisted by smooth muscle cells [1,2]. The differential diagnosis of LPD is rough due to its clinical resemblance with peritoneal carcinomatosis or metastatic lesions and its histologic resemblance with benign metastasizing leiomyoma [1,2]. Prompt diagnosis of LPD is crucial since, although benign in nature, LPD may degenerate into malignancy [3,4]. To our knowledge, this is the first case of LPD reported in Greece. The present manuscript has been reported in line with the SCARE criteria [5].

2. Case report

A 40-year-old female proceeded to our institution for elective excision of abdominal and pelvic masses. Two months ago, the patient had manifested severe vaginal bleeding and abdominal discomfort and she had presented to her obstetrician for medical advice. The ultrasound of pelvis that had been initially performed by the obstetrician had revealed a tumescent pelvic tumor that had been considered as a leiomyosarcoma. The patient had past history of one birth 7 years ago and of laparoscopic myomectomy without the use of power morcellator, 4 years ago due to the presence of a large leiomyoma of the uterus. She had no previous history of prolonged use of contraceptive pills. Furthermore, the patient had family history of nor leiomyomas, neither leiomyosarcomas of the uterus. Providing the patient's findings and clinical history she was referred to our institution for further evaluation.

Subsequent abdominal CT and MRI revealed multiple tumors with evidence of vascularity in the pelvis, varying in size from 1.2 to 6.1 cm. In addition, three more similar tumors were identified into the peritoneal cavity. The ascending colon was deviated away from the larger one of these tumors, which had a diameter of 11.2 cm (Figs. 1–3). In order to exclude the probability of malignancy, FNA biopsy of the identified tumors was suggested. In fact, image guided FNA was performed during the abdominal CT as well, and the histologic analysis of the tissue sample revealed smooth muscle tumors that were benign in nature.

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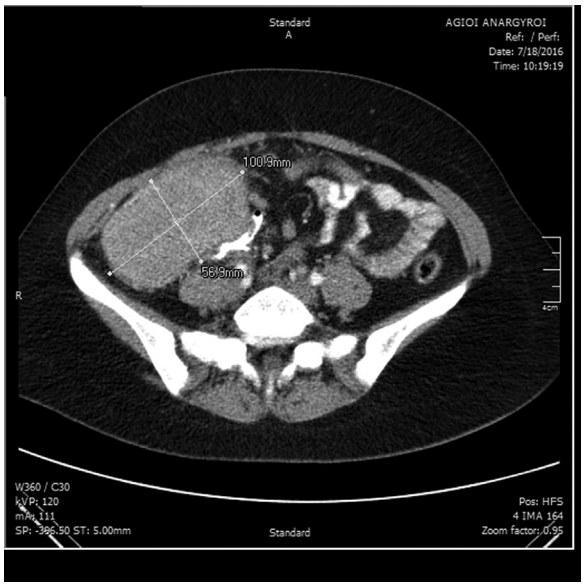

Fig. 1. Tumescent abdominal mass.

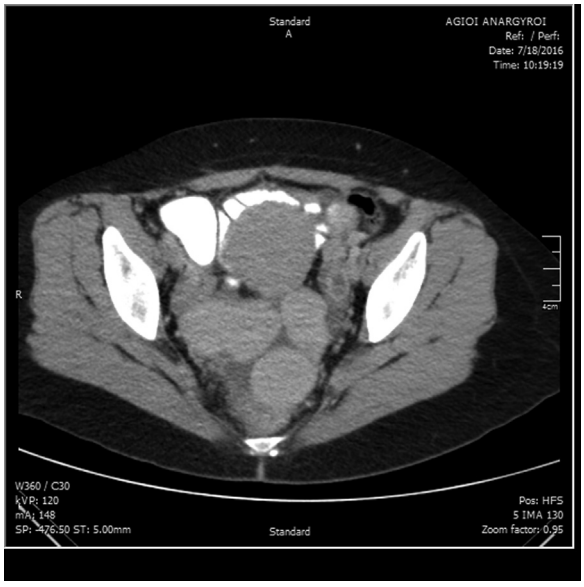

Fig. 2. Multiple tumescent masses.

A CT scan of the chest was also performed and it had no any pathological findings. The physical examination of the patient was unremarkable. The vital signs and blood test results were all into the normal spectrum. Following this, laparotomy was scheduled.

During the operation, the oncology-surgeons identified six-seven round, white-grey nodules into the peritoneal cavity and the lesser pelvis in close relation to the large intestine and the mesocolon, and they dissected them off (Fig. 4). A tumescent nodule was detected on the surface of the greater omentum and was carefully excised by the surgeons as well (Fig. 5). Furthermore, the large tumor that applied pressure to the ascending colon was meticulously dissected. Finally, a mass lesion was detected on the surface of the rectum and the surgeons attained to dissect the masses off, but the excision was not feasible due to the major potentiality of perforation and subsequent hemorrhage. Careful hemostasis was performed and two drainages were placed in the abdomen. They were finally removed the 3^{d} postoperative day when the patient was discharged with instructions, without any complication.

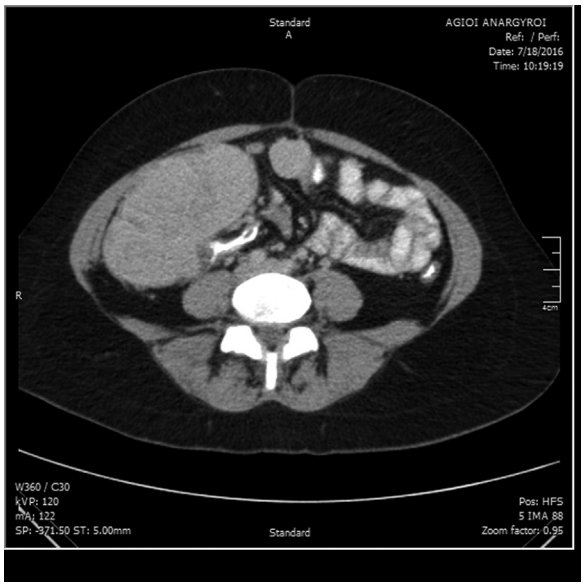

Fig. 3. Multiple benign nodules into the peritoneal cavity.

Intraoperative frozen section of the nodules revealed mesenchymal tumors, without mitotic figures or any other evidence of malignancy. The histopathologic analysis of the tissue sample from the excisional biopsy revealed tumors consisted by benign smooth muscle cells, without mitotic figures or cell atypia, or tumor cell necrosis, with negative PRs and ERS, similar to leiomyomas. The histopathologic analysis in combination with the position of the detected benign tumors and the patient's history confirmed the diagnosis of LPD.

Both surgeons and oncologists recommended close observation of the patient by abdominal CT and MRI for the first postoperative year, and no further treatment was administrated.

3. Discussion

Leiomyomatosis peritonealis disseminata (LPD) is an extremely rare clinical condition [6] that was first described in 1952 by Wilson and Peale [7]. There are approximately 100 cases of LPD in the literature, but the true prevalence of the disorder remains vague, since the majority of patients remain asymptomatic and subsequently, LPD is underdiagnosed [2].

Although rare, LPD mostly affects premenopausal women [6] but there are also reported cases of LPD in postmenopausal women [8] and in male patients as well [2,9].

The etiology and pathophysiology of LPD is yet not elucidated [1,6]. Nevertheless, as it is considered, LPD originates from metaplasia of submesothelial, multi-potential mesenchymal cells [10]. More specifically, LPD is associated with exposure to high levels of endogenous or exogenous estrogens, such as in cases of prolonged administration of contraceptive pills and pregnancy [11]. In fact, in LPD tumor cells both ERs and PRs have been identified [12].

Furthermore, etiological correlation of LPD and previous myomectomy, hysterectomy and endometriosis has been reported as well [13,14]. Indeed, it is documented that laparoscopic hysterectomy with tumor morcellation may augment the potentiality of tumor implantation and dissemination [15]. More specifically, LPD may present even years after myomectomy or hysterectomy [16], as in the presented case. In addition, recently, autosomal-dominant model with varying level of penetrance has been reported in the literature for LPD, occurring to the identification of a case of familial clustering of LPD [2].

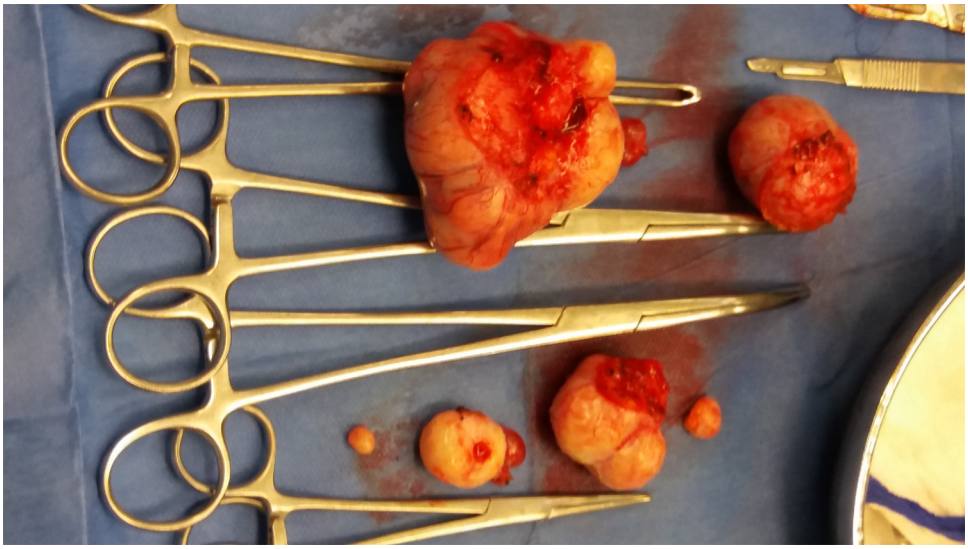

Fig. 4. Another tumescent benign nodule excised from the lesser pelvis.

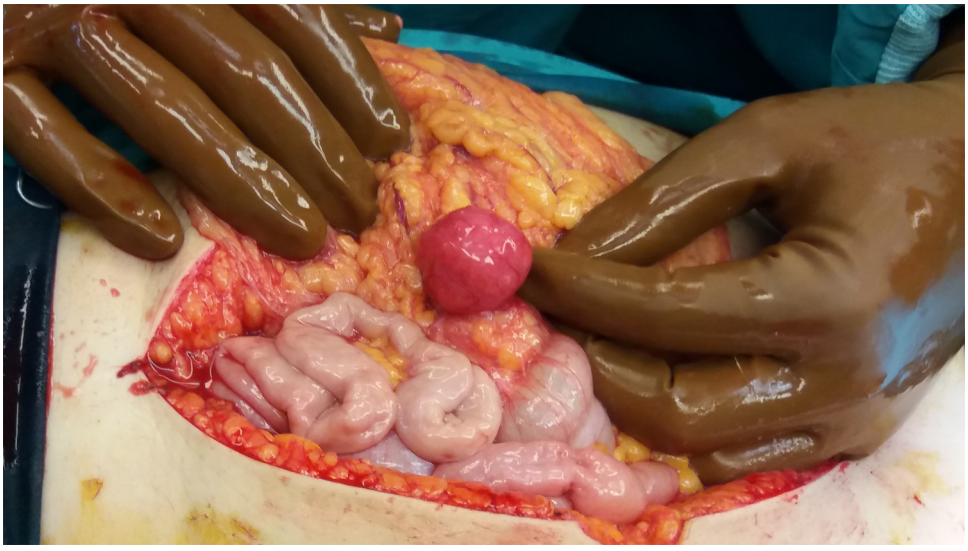

Fig. 5. Large benign nodule detected on the surface of greater omentum.

The majority of patients with LPD remain asymptomatic. Symptoms- if any- are nonspecific and they include abdominal pain and discomfort, bleeding from the rectum or the vagina, as in the present case, abdominal distention or abdominal masses which may lead to intestinal obstruction [1,2]. Furthermore, LPD has been associated with the presence of both ascites and endometriosis [17] or both ascites and lymph nodes enlargement [16]. Finally, the first case of LPD and ovarian leiomyoma, which is a scarce tumor, has been recently documented [18].

Preoperative detection of LPD with the utilization of imaging procedures, such as abdominal ultrasound, CT or MRI may be beneficial but they are not a great help for the differential diagnosis of malignancies. Hence, preoperative biopsy and histopathologic analysis are essential for the diagnosis of LPD, as performed in the presented case [1]. In fact, only the histological examination may permit the differential diagnosis between LPD and leiomyosarcomas of the uterus due to their exceptional clinical resemblance.

Microscopically, LPD is typically characterized by smooth muscle cells in interdigitating fascicles with or without mitotic figures [19] and they lack nuclear polymorphism, hyperchromasia, tumor cell necrosis and cellural atypia in contrast to a leiomyosarcoma [1,11]. Although histological diagnosis excludes the diagnosis of peritoneal disseminated metastasis and other malignancies, differential diagnosis of LPD from benign metastasizing leiomyoma (BML) remains difficult.

In contrast to LPD though, BML is considered as a smooth muscle mass in a solid organ [2]. In particular, BML usually presents as one or multiple pulmonary nodules [6] but the benign tumors may also metastasize in female patients who have undergone myomectomy or hysterectomy to distant positions, such as skin, bone, muscular tissue, vascular channels, lymph nodes, mediastinum and even to heart [20]. When identified in the abdomen or the pelvis, BML is typically detected close to the round ligament or the iliac veins [6].

Intraoperatively, LPD is presented as multiple, round nodules, varying in size from several millimeters to centimeters may be detected on any peritoneal or omental surface of the abdominal cavity, the small or large intestines, the mesentery and the retroperitoneum [2,13].

LPD is a scarce benign disorder with a good prognosis [13]. Nevertheless, degeneration of LPD into malignancy has been reported

in the literature [3,4]. Moreover, simultaneous identification of LPD and leiomyosarcoma has been reported, although it is not elucidated if the malignancy occurred due to transformation of the LPD tumors [6].

Therefore, it is obvious that further studies concerning molecular and cytogenetic features of the tumors are essential so that clinicians will be able to understand the rare probability of malignant transformation of LPD.

Finally, concerning the treatment of LPD there are no elucidated guidelines [1]. Recently, determination of the therapy according to the patient's age, comorbidities and severity of symptoms of LPD has been proposed [13]. In fact, for women with reproductive desire, a conservative therapeutic approach, including surgical castration or stopping the contraceptive pills, is preferred [1,11]. More aggressive surgical treatment is strongly recommended in cases of high risk of malignant degeneration. Such cases are no exposure to estrogens, no previous history of leiomyoma and negative PRs or ERS in the benign nodules [11]. Close observation of the patients by abdominal CT or MRI should be performed to high-risk patients as well.

4. Conclusion

LPD is a scarce benign disorder that may mimic clinically peritoneal carcinomatosis or metastatic leiomyosarcomas. Its differential diagnosis is difficult and requires an ample history of the patient, clinical evaluation, preoperative guided FNA and histopathologic analysis of the FNA tissue and of the tumor resection. Surgeons' knowledge concerning this rare clinical condition is fundamental in order to establish a correct diagnosis and warrant the appropriate treatment and the minimization of the probability of malignant transformation of LPD.

Conflicts of interest

All authors declare that there are not any competing interests.

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Ethical approval

This is a Case Report for which the patient provided written informed consent.

Consent

Written consent for the publication of this case report and accompanying images was obtained from the patient. The consent can be provided to the Editor if he asks so.

Author contribution

Psathas conceived of the study. Mariolis- Sapsakos was senior consultant at this case report and participated in its coordination. Kaklamanos and Birbas contributed to the acquisition of clinical data, its analysis and interpretation and to the preparation of images. Zarokosta and Zoulamoglou carried out the literature review. Psathas and Zarokosta contributed to the preparation of the manuscript. Thivaios, Chrysikos and Mariolis-Sapsakos contributed to the refinement of the case report. All authors have approved the final article.

Registration of research studies

This is a Case Report and according to the Research Registry, its registration is not essential.

Guarantor

The Guarantor who is responsible for the present case report is Theodoros Mariolis-Sapsakos. He coordinated the preparation of the case report and revised it critically for important intellectual content.

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