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Case Report

Long-Term Survival in a Patient With Abdominal Sarcomatosis From Uterine Leiomyosarcoma: Role of Repeated Laparoscopic Surgery in Treatment and Follow-Up

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ABSTRACT Uterine leiomyosarcoma (LMS) in some cases may disseminate through the abdominal cavity, without extra-abdominal spreading, determining a condition of abdominal sarcomatosis, which represents a peculiar situation. Only radical surgical removal offers a chance of long-term survival in such cases of LMS. Here we describe a case of diffuse abdominal sarcomatosis from uterine LMS in a 51-year-old perimenopausal woman who underwent laparoscopic radical hysterectomy, bilateral salpingo-oophorectomy, total pelvic peritonectomy, pelvic lymphadenectomy to the mesenteric inferior artery, and omentectomy. Then, given the high probability of disease recurrence, the patient underwent a close follow-up consisting of positron emission tomography (PET)/computed tomography every 3 months and diagnostic (and if necessary operative) laparoscopy every 6 months. To date, the patient had 11 laparoscopies; 5 of them were preceded by a PET indicative of the presence of disease with high metabolic activity, which was confirmed at surgery and each time completely removed laparoscopically with no evidence of residual disease. To date, 5 years from diagnosis the patient is alive and continues her follow-up. Our report brings to light the ability of laparoscopic surgery to obtain disease control in a case of LMS with abdominal dissemination. Moreover, laparoscopic surgery, as demonstrated in our case, may have an important role in the close follow-up of the disease and allow a timely and early radical surgical approach of relapses before they become extremely large and difficult to remove radically. Journal of Minimally Invasive Gynecology (2016) 23, 1003–1008 © 2016 AAGL. All rights reserved.

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The dissemination of soft tissue sarcoma all over the abdominal cavity without extra-abdominal spread is known as abdominal sarcomatosis (AS), an uncommon disease most often arising from uterine leiomyosarcoma (LMS) [1]. Although most uterine LMS are confined to the uterus, many cases involve local spread to peritoneal surfaces and adjacent organs and distant metastasis [2]. Tumor stage is the strongest prognostic factor for all uterine sarcomas, with a 5-year survival rate of 50% to 55% for stage I patients and 8% to 12% for patients with more advanced tumors [3]. The current management of uterine LMS does not take into account individual clinical pathologic prognostic factors, such as tumor size (>5 or ≤5 cm), mitotic activity (≤10 or >10 mitosis/10 high-power fields), age, and vascular invasion [3].

In view of this evidence, the prognosis of peritoneal sarcomatosis cannot be well established. Only radical surgical removal offers a chance of long-term survival in cases of LMS [4]. In this context, AS associated with primary uterine LMS represents an unusual situation. There are also growing cases involving peritoneal sarcomatosis from occult uterine LMS after the use of internal morcellation for laparoscopic hysterectomies or myomectomy for presumed uterine fibroids [5,6]. The management of AS from uterine LMS is therefore difficult because optimal radical resection may be complicated by disease spread and frequent recurrence, and very few data are available about a laparoscopic approach.

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To date, conventional therapeutic modalities have failed to improve the outcome of patients with uterine LMS associated with extrapelvic spread. Retrospective analyses [7–10] and phase II studies [11,12] analyzed the morbidity and mortality of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy perfusion (HIPEC) in the treatment of uterine LMS with abdominal spread. The lack of effective chemotherapeutic agents coupled with the hematogenous spread of sarcomas means that the use of cytoreductive surgery and HIPEC remains controversial [1,13]. Furthermore, some authors found no difference between patients treated with or without HIPEC after complete cytoreduction, suggesting that resection status is more important for survival than HIPEC use [10,13]. These discordant results may reflect large variations in the behavior of these tumors, and their best definition is mandatory.

In this context, we describe a clinical case of diffuse AS from uterine LMS, with a long survival obtained through repeated cytoreductive laparoscopic surgeries. The description of this case may contribute to clarify the central role of surgery, and in particular of a laparoscopic approach, in the treatment of this disease and implement our knowledge of their biologic heterogeneity, which may have therapeutic implications.

Case Report

Written informed consent was obtained from the patient for publication of the case report and accompanying images. The retrospective observational nature of the study did not necessitate the local institutional ethics committee approval.

Five years ago, a 51-year-old perimenopausal pluriparous (gravida 2 and para 2) white woman who was not taking oral contraceptives presented with menometrorrhagia. Physical examination revealed an enlarged uterus believed to be a result of uterine fibromatosis with multiple myomas. Abdominal ultrasonography confirmed an enlarged uterus deformed by the presence of probable multiple myomas. Serum CA-125 levels were 75 U/mL (normal range $<$ 34 U/mL). For this reason, we performed laparoscopy to examine the uterus and help determine the most suitable surgical approach.

Because of the large volume of the uterus, a port was placed above the umbilicus and a pneumoperitoneum of 14 mm Hg was established and maintained throughout the surgery. Intra-abdominal visualization was achieved using a 10 mm, 0-degree telescope (Karl Storz, Tuttlingen, Germany), and three 5-mm trocars were introduced under laparoscopic visualization through ports in each lower quadrant and in the suprapubic region. Our initial observation showed a greatly enlarged uterus, deformed by numerous reddish-brown nodules in its anterior and posterior walls. These neoformations also extended throughout the pelvic peritoneum (Douglas' cavity, vesicouterine recess, and right pararectal lodge), the great omentum, the mesosigma, the small intestine mesenterium, and the abdominal peritoneum (Fig. 1). Extemporaneous examination revealed a mesenchymal neoplasm with signs of atypia

Fig. 1

Laparoscopic visualization of the uterus and multiple peritoneal pelvic metastatic localizations of LMS at diagnosis.