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Abdominal leiomyosarcomatosis after surgery with external morcellation for occult smooth muscle tumors of uncertain malignant potential: A case report



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ABSTRACT

INTRODUCTION: Although rare, cases of abdominal sarcomatosis (AS) after laparotomy/laparoscopic interventions for uterine smooth muscle tumors of uncertain malignant potential (STUMP) have been reported. **PRESENTATION OF CASE:** We describe a rare case of diffuse abdominal sarcomatosis in a patient that some year earlier had undergone myomectomy for a suspected uterine myoma, which was histologically proven to be a STUMP. Once the patient was admitted at our Department, she underwent a diagnostic laparoscopy that confirmed a condition of peritoneal sarcomatosis disseminated through the entire abdomen, and then a laparotomic total hysterectomy, bilateral salpingo-oophorectomy, and total omentectomy, achieving a complete cytoreduction. Histological examination showed high-grade uterine leiomyosarcoma (LMS). Since there is no evidence of any clinical benefit of adjuvant treatment, given the risk of disease recurrence, we decided, with the patient's agreement, to conduct close follow-up with a Positron Emission Tomography (PET)/Computed Tomography (CT) scan every 3 months and diagnostic laparoscopy every 6 months, even in the absence of PET/CT positivity. After 2 years PET/CT showed a relapse of LMS in the perigastric region and, therefore the patient underwent a diagnostic/operative laparoscopy with complete removal of the neoplastic recurrence. To date, the patient is disease-free. **DISCUSSION AND CONCLUSION:** STUMPs should be submitted to a frequent surveillance for their risk of recurrence, dissemination and transformation into LMS, even many years after the first diagnosis. A follow-up including surgical re-exploration with laparoscopy and PET/CT imaging may allow early detection and timely treatment of the relapse with good long-term outcome, as demonstrated by our case.

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1. Introduction

Abdominal sarcomatosis (AS) is a rare condition that most often arises from primitive uterine leiomyosarcoma (LMS) [1]. AS is typically described as soft tissue sarcoma dissemination throughout the abdomen. There are also a growing number of cases involving peritoneal sarcomatosis from occult uterine LMS following the use of internal morcellation, which is performed during laparoscopic hysterectomies or myomectomies for supposed uterine fibroids [2–5]. Although they are rare, cases of AS after laparotomy or laparoscopic interventions for uterine smooth muscle tumors of uncertain malignant potential (STUMP) have been also described [6,7]. The World Health Organization classifies STUMPs as a very large set

of neoplasms, including morphological variants of uterine smooth muscle tumors that cannot be clearly defined or classified as benign or malignant; these tumors also have undefined behavior and clinical prognosis [8].

According to a previous study, the incidence of STUMP among patients who underwent hysterectomy or myomectomy for presumed benign fibroids was 1/1004 [9]. The evidence about the natural history of STUMP is debatable. Despite their indefinite nature, uterine STUMPs can metastasize, relapse as STUMPs, or develop into LMS [10,11]. Here, we describe a clinical case of diffuse AS resulting from dissemination following laparotomic myomectomy for an occult STUMP that developed into LMS.

2. Presentation of case

The research work has been reported in line with the SCARE criteria [12]. Written informed consent was obtained from the patient for the publication of the case report and the accom-

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Fig. 1. The profile of the enlarged abdomen of the patient lying on the operating room bed before surgery.

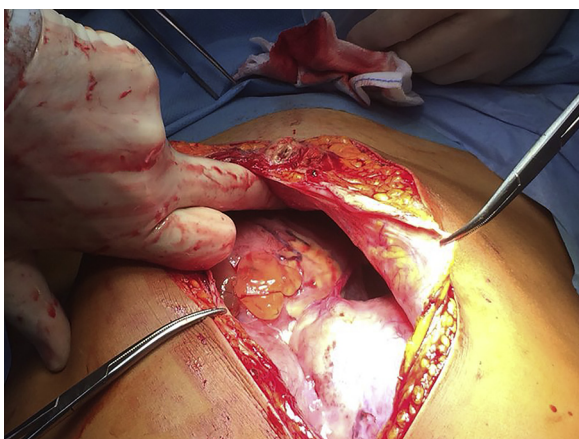


Fig. 2. View at the laparotomic opening of the abdominal cavity.

panying images. The retrospective observational nature of the study did not necessitate the local institutional ethics committee approval. A 43-year-old female patient presented at our attention at the Department of Gynecologic Oncology, in June 2014 with a notable increase in abdominal circumference and symptoms caused by abdominal pressure and constipation. Three years earlier, at another Institution, she had undergone laparotomic myomectomy for a suspected uterine myoma, which was histologically proven to be a STUMP. The patient did not receive a specific follow-up. Ultrasound examination performed at our first observation revealed a greatly enlarged uterus deformed by the presence of multiple ovoidal nodules. The abdomen was entirely occupied by a large, highly vascularized neof ormation with several areas of necrosis. **Fig. 1** shows the profile of the enlarged abdomen of the patient lying on the operating room bed before surgery. Diagnostic laparoscopy confirmed a mass with macroscopic characteristics indicative of peritoneal sarcomatosis. Laparotomy was then performed. Upon opening the abdominal cavity (**Fig. 2**), the pelvis was found to be completely occupied by a huge bilobate mass that was fed from the right adnexa by a thick vascular pedicle with large, newly formed vessels (**Fig. 3**). The mass had adhered to the omentum, several intestinal loops, and the small intestinal mesentery, although there was no evidence of infiltration. We performed cautious dissection of the large neof ormation, which proved to be relatively straightforward. **Fig. 4** shows the entire large mass extracted from the abdominal cavity. Other ovoidal masses were present in the abdominal wall, most notably in the anterior abdominal wall of the previous laparotomic wound, abdominal and



Fig. 3. Picture of the large mass, showing its feeding from the right adnexa by a thick vascular pedicle and its large newly formed vessels.

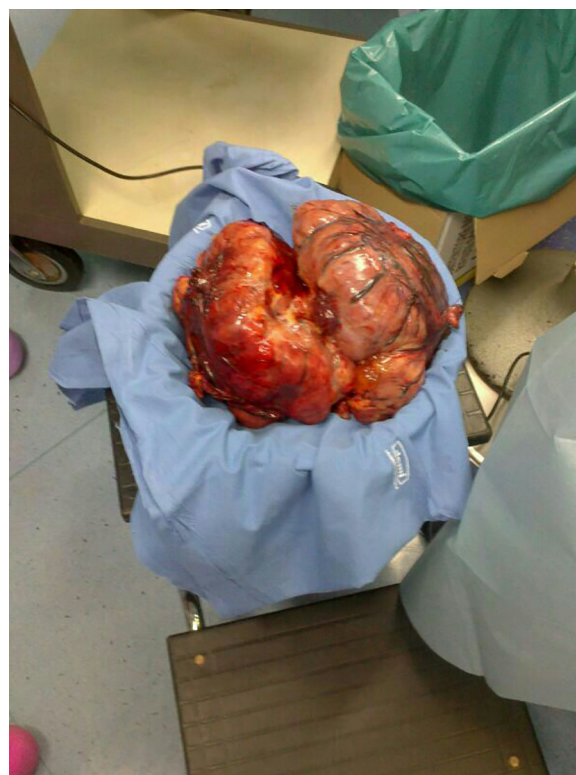


Fig. 4. Picture of the large bilobate large mass extracted from the abdominal cavity.

pelvic peritoneum, and peri-sigma region. Small nodules (≤ 1 cm) were present over the entire peritoneal surface. None of these nodules had infiltrated the adjacent organs, and the dissection was smooth without blood loss. Total hysterectomy, bilateral salpingo-oophorectomy, and total omentectomy were performed, and complete cytoreduction was apparently achieved. Histological examination showed high-grade uterine LMS with extensive areas of necrosis and 25 mitoses/10 High Power Field. The patient was discharged in good condition 5 days after surgery. There was a lack of evidence of any clinical benefit of adjuvant treatment and a high probability of disease recurrence; therefore, we decided, with the patient's agreement, to conduct a close follow-up with a Positron Emission Tomography (PET)/Computed Tomography (CT) scan every 3 months and diagnostic laparoscopy every 6 months, even in the absence of PET/CT positivity. After 2 years (May 2016), PET/CT showed a relapse in the perigastric region. The patient

underwent diagnostic and operative laparoscopy with complete removal of the neoplastic recurrence. To date, after 1 year from the last recurrence, the patient is free of disease after the most recent PET/CT examination.

3. Discussion

Based on statistics, a patient with an enlarged uterus caused by fibromatous corporeal tumors is unlikely to have a final diagnosis of malignant uterine smooth muscle tumors. In fact, LMS is rare and the majority of cases arise *de novo* rather than from the malignant transformation of benign myomas [13]. Therefore, there are no standardized guidelines on the clinical management of a patient with a rapidly enlarging uterus. In 2014, the FDA warned against using laparoscopic power morcellators to treat uterine fibroids, which resulted in considerable debate on the issues inherent to the late diagnosis of uterine occult malignant smooth muscle tumors and their potential dissemination in the abdominal cavity. Which is the most correct follow-up or the clinical behavior of occasionally diagnosed uterine malignant smooth muscle tumors after laparoscopic hysterectomies or myomectomy performed both by laparoscopy and laparotomy with morcellation, is still undefined. For a retrospective diagnosis of occult LMS after myomectomy, well-defined surgical procedures are performed. In contrast, in the case of its potential abdominal dissemination after morcellation, currently there are no well-defined guidelines for its evaluation, prevention, and follow-up. This is even less clear for a diagnosis of occult uterine STUMP when the pathology is of uncertain biological significance. Because of the rarity of these tumors, existing literature on the topic remains scarce and, therefore, a consensus regarding the diagnosis, malignant potential, treatment of choice, and follow-up has not yet been reached [14]. Clinical presentation of STUMPs is similar to that of uterine leiomyomas and typically includes abnormal vaginal bleeding, anemia, an enlarging pelvic mass, pressure symptoms, and pelvic pain [15]. The median age at presentation is similar to that of patients diagnosed with benign leiomyomas, as well as of those with LMS. In a retrospective analysis of 41 women affected by occult STUMP, Guntupalli et al. [11] reported similar long-term outcomes between patients who had undergone myomectomy and those who had undergone radical hysterectomy. These results are in accordance with other small case series where the surgical approach and the type of surgery did not influence the long-term outcomes, which were largely positive [16]. Interestingly, patients affected by STUMP complicated by subsequent disease recurrence were younger than those with an uneventful follow-up [17], as in our case.

The novelty of our work lies in the rarity of the described clinical case in terms of diagnosis of STUMP after a laparotomic myomectomy with external morcellation for supposed uterine leiomyoma. Other relevant findings were the diffusion in the abdominal cavity, the exceptionally large main metastatic nodule, and the development of LMS. This highlights the importance of obtaining precise information about the biology of STUMPs before conservative surgery, and of standardizing the follow-up and any relevant useful examinations. Hysterectomy must be performed unless a woman has future reproductive wishes. If uterus preservation is required for younger patients after myomectomy for occult STUMP, we believe that laparoscopy is essential in the follow-up because of its ability to diagnose early dissemination of these tumors, which also have a high risk of relapse and malignant transformation. Successful pregnancies following fertility-sparing surgery have been reported; however, patients who do not undergo hysterectomy should be adequately informed of the risk of recurrence and a strict follow-up program using clinical, imaging techniques and diagnostic laparoscopy is mandatory. Furthermore, PET/CT may be

used to assess the malignant degeneration of STUMP. Then, STUMP requires closer surveillance than a yearly examination, and should involve a consultation with a gynecologic oncologist [18]. Ip et al. [17] suggested an intense follow-up program with an evaluation performed every 6 months in the first 5 years followed by annual surveillance for the next 5 years. At our institute, patients treated using myomectomy for STUMP usually undergo a clinical evaluation every 6 months accompanied by PET imaging and eventually laparoscopy; we have previously reported this protocol in patients with abdominal sarcomatosis who underwent complete cytoreductive surgery [19]. Thus, a new approach along these lines could offer a broader role for laparoscopy in the close follow-up of STUMP that can evolve over time into LMS [6]. Surgical re-exploration procedures after morcellation of uterine STUMP have a high likelihood of detecting peritoneal implants, which can be benign or malignant [7], and in the same time performing the appropriate surgical treatment, as reported in our case. In this view, investigating the role of PET is warranted.

4. Conclusions

Patients with STUMP should be counseled regarding the potential for recurrence, dissemination, and transformation into LMS. A multidisciplinary management team composed of a gynecologist, dedicated pathologist, and oncologist is essential to diagnose the disease and to establish the treatment of choice and a suitable follow-up program, in particular in patients who undergo conservative surgery. As demonstrated by our case, STUMPs, although demonstrate a low-grade malignancy, prolonged survival rate, and delayed recurrence, must require more frequent surveillance than a yearly examination because of the risk of LMS transformation and metastases, even many years after the initial diagnosis. This must be implemented within a system that allows early detection of recurrence and timely treatment of the same.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Author contribution

AM contributed to the conception and design of the study, acquisition of data, analysis and interpretation of data, writing the manuscript; GC contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; PK contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; FL contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; MS contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; RD contributed to analysis and interpretation of data, revising the manuscript critically for important intellectual content; CM contributed to analysis and interpretation of data, writing the manuscript. All authors have approved the final article.

Conflict of interest

None.

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Ethical approval

The retrospective observational nature of the study did not necessitate the local institutional ethics committee approval.

Consent

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Author contribution

AM contributed to the conception and design of the study, acquisition of data, analysis and interpretation of data, writing the manuscript; GC contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; PK contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; FL contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; MS contributed to acquisition of data, analysis and interpretation of data, drafting the manuscript; RD contributed to analysis and interpretation of data, revising the manuscript critically for important intellectual content; CM contributed to analysis and interpretation of data, writing the manuscript. All authors have approved the final article.

Guarantor

Antonio Macciò.

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