

CASE REPORT

Rare Case of Smooth Muscle Tumor of Uncertain Malignant Potential – Clinical Case

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ABSTRACT

Introduction: Smooth muscle tumor of uncertain malignant potential (STUMP) still represents a medical and surgical challenge, being rare and hard to diagnose. Normally STUMP consists in a multitude of uterine tumors that do not meet the current histologic criteria for a specific malignant or benign tumor. Clinical, imagistic and laboratory investigations seem to only raise the presumptive diagnosis in these cases, the diagnosis being confirmed during histological examination of the post hysterectomy/myomectomy specimen.

Case report: We present the case of a 50-year-old female patient who was examined for the first time for pelvic pain and sensation of increased pressure in the lower abdomen in June 2020. A gynecology consultation was performed, during which a large uterine mass was felt on vaginal digital examination combined with pelvic palpation. Further pelvic ultrasound examination revealed a voluminous tumor of the left ovary. Then, a magnetic resonance imaging was performed, which showed a large pelvi-abdominal tumor of probable ovarian origin with a mixt tissue type and cystic structure, the presence of pathological abdominal and pelvic lymph nodes, ascites with a suspicion of peritoneal carcinomatosis. The patient was referred to a tertiary center and the multidisciplinary team recommended surgical treatment. A total hysterectomy with bilateral oophorectomy and stadial lymphadenectomy was performed. After surgery, the patient had a favorable evolution and was discharged five days after the intervention in a good health condition. Macroscopic and histological examinations of the biopsy pieces showed morphological aspects of uterine smooth muscles tumors with uncertain malignancy potential/STUMP and multiple uterine leiomyomas.

Discussion: Due to the rarity of these tumors, the scientific literature needs to be constantly updated in order to help physicians to correctly identify and treat this pathology. It is highly recommended to identify tumors with a high malignancy potential, so that the follow up will be sufficient to discover and treat recurrences before they become life-threatening.

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Conclusion: Smooth muscle tumor of uncertain malignant potential remains a difficult diagnosis due to uncertainty between malignancy and benign features. Treatment approaches are difficult in these cases and must always be in accordance with the patients' desire to maintain their reproductive functions and fertility. Recurrence of STUMP has been reported, therefore a close follow up of these patients must always be performed.

Keywords: uterine smooth muscle tumor, menopause, leiomyosarcoma.

INTRODUCTION

Smooth muscle tumors of the uterus (STUMP) represent a rarely encountered pathology throughout medical and surgical practice nowadays. About 0.01% of patients who undergo myomectomy or hysterectomy are diagnosed with STUMP (1). Diagnosis is usually confirmed after the histological examination of the biopsy pieces. Medical theories consider STUMP as a transition between leiomyoma to leiomyosarcoma or possibly an undiagnosed low-grade leiomyosarcoma (2). The World Health Organization classifies STUMP as smooth tumors with an intermediate behavior between benign and malignant. In order to facilitate the diagnosis of STUMP, Stanford's criteria for leiomyosarcoma are being taken into consideration: the presence of tumor cell necrosis, diffuse moderate to severe cytological atypia and a mitotic count of at least 10 mitotic figures. The diagnosis will be established when a tumor shows an atypical combination of these three criteria but does not totally satisfies the Stanford criteria (3).

Treatment options are not as variable in comparison to other pathologies of the female reproductive system and must always take in consideration the patient's fertility desire (4). Recurrence of STUMP ranges between 8,7%-11% but, due to underdiagnosing, this data is not always reliable (5). □

CASE REPORT

We present the case of a 50-year-old female who came to a gynecology consultation in June 2020 because she had pelvic and abdominal pain not responding to pain killers for three weeks as well as an enlarged abdominal and pelvic region with constant sensation of abdominal pressure. The patient entered menopause two years before and had two vaginal births during

her reproductive life. No particularities were noticed on speculum examination, except for the presence of one Naboth cyst. A large uterine mass was felt on vaginal digital examination combined with pelvic palpation. Ultrasound showed a pelvic tumor difficult to measure due to technical limitations, which was limited in the anterior part by the umbilical region and laterally left and right reaching the iliac crests. Tumor had a mixt structure consisting of both tissue type and cystic componence. The cystic area presented multiple septa with hyperechoic signal and no Doppler signal and the tissue type area with heterogeneous aspect and presence of Doppler signal. Ultrasound examination has also revealed a bilateral renal micro lithiasis but could not help identify the organ origin of the tumor; therefore, magnetic resonance imaging (MRI) was indicated.

In July 2020, the MRI examination of the abdominal and pelvic region showed the presence of a voluminous left ovarian tumor with a mixt structure of parenchymatous tissue and septate cysts with an effect of lateral uterine mass that modified the uterine position by deviating it towards the right area, with compression of the intestines, urinary bladder and the ureters, and without direct signs of invasion. The tumor had the following dimensions: 190 mm (transverse)/128 mm (antero-posterior)/195 mm (lateral). The solid part of this mass, measuring 150/118 mm, was predominantly towards the right abdominal area, whereas the cystic component 131/140 mm-T was predominantly towards the left abdominal area and applying pressure to the transvers area of the colon. The uterus was deviated to the right and posterior, measuring 73 mm/32 mm, with a homogenous endometrium of 7 mm and a junctional area and myometrium tissue without particularities. Left and right adnexa were difficult to assess. Intraperitoneal ascites liquid was identified in moderate quantity (maximum of 20 mm). Lymphatic nodes

were present on MRI as well in the lombo-aortic, left external iliac and the left obturator areas. The imagistic investigations lead to the following conclusion: voluminous abdominal pelvic tumor probably of ovarian origin, with a mixt tissue type and cystic structure, presence of pathological abdominal and pelvic lymphatic nodes, ascites liquid in moderate quantity and peritoneal carcinomatosis, without hepatic lesions. Considering the large extension of this tumor and its imagistic aspect, the patient was further referred to a surgical evaluation.

In August 2020, the patient was submitted to the Regional Institute of Oncology Iasi, Romania, for re-examination. After full examination, she was given a presumptive diagnosis of malignant tumor of the ovary. Her case was presented in the multidisciplinary meeting, and a recommendation for surgical resection was made. Thoracic radiography and cardiologic consultations were performed, but no abnormalities were detected. Laboratory exams identified a mild anemia (Hb 8.8 g/dL). All therapeutic and surgical options were explained to our patient and she agreed for a radical surgical intervention. On 26 August, she underwent a total hysterectomy with bilateral oophorectomy and stadial lymphadenectomy was performed. Postoperative the evolution was simple and she was discharged on the fifth postoperative day, with the presence of the physiological digestive transit, physiological urinary function and no vaginal bleeding or other complaints.

The macroscopic evaluation of the surgical resection specimen showed a deformed uterus measuring 19/18/16 cm with a uterine cavity of 5 cm length. Throughout the uterus section there were multiple white colored nodules with fasciculate aspect and a maximum size of 15 cm (the biggest nodule identified in our uterine section). A cystic degeneration, an edematous aspect of the solid part and unregulated serous has been also described. Adhesions of the uterus to an epiploic segment on an area of 6.5/2.5 cm and another area of 8.5 cm were noticed, hemorrhagic deposits being described at the uterine serous level. Right adnexa and salpinx measured 10 cm in length and the right ovary 2.7/2/0.8 cm with integrity of the ovarian capsule maintained and the presence of numerous corpus luteum. Left adnexa and salpinx measured approximately 7 cm in length and an ovary of 1/1.2/0.7 cm

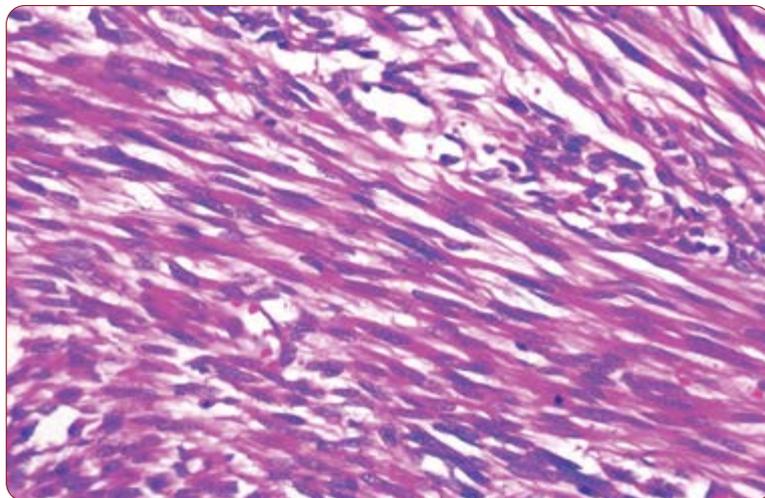


FIGURE 1. Hematoxylin eosinophilic coloration 40x. showing fusiform tumorous cells with smooth muscle phenotype

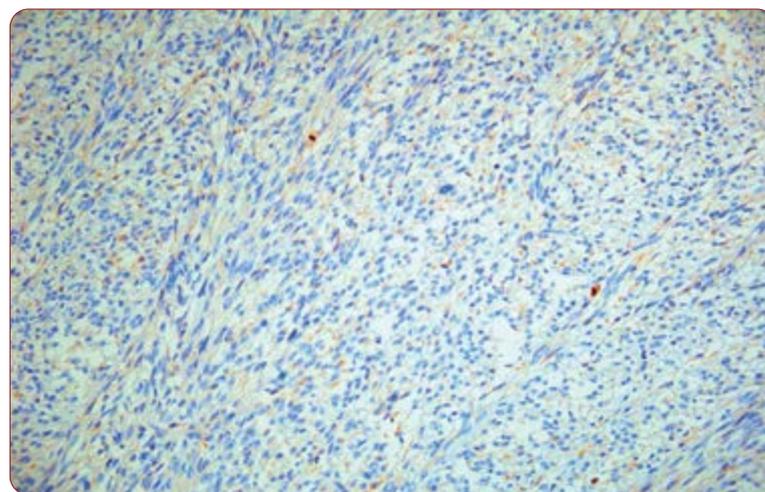


FIGURE 2. Phospho-H3 coloration 20x showing areas of stromal edema and cystic degeneration

also with the integrity of the ovarian capsule maintained and numerous hemorrhagic luteal bodies.

The microscopic examination of multiple uterine biopsies showed at the myometrium level a mesenchymal tumor proliferation with a fasciculate architecture composed by fusiform tumorous cells with smooth muscle phenotype (Figure 1). The biggest tumorous proliferation presented an irregular shape, high cellular density and reduced cytology atypia, high mitotic activity – more than 15 mitoses /10 HPF. Areas of ischemic necrosis accompanied by hyalinization process were also identifies as well as areas of stromal edema and cystic degeneration (Figure 2). No tumorous necrosis areas were identified. The

omentum adhered to the uterine serosa but had no signs of tumorous infiltration. Endometrium with signs of unbalanced estrogenic-progestative stimulation.

Cervix with chronic cervicitis and multiple Naboth cysts. Parameters without tumorous modifications, right ovary with recent hemorrhagic luteal body and left ovary with multiple luteal bodies being in process of formation. Bilateral salpinx with congestive aspect, and vaginal margins without tumorous modifications. Based on macroscopic and microscopic examination of the biopsies the following diagnostic was confirmed: tumor of the smooth muscular cells with uncertain malign behavior/ STUMP and multiple uterine leiomyomas. □

DISCUSSIONS

Simple classifications of smooth muscle tumors were used before 1994 and divided these tumors into three categories, STUMPs, leiomyomas and leiomyosarcomas, but a more complex classification, that helped physicians to imply a certain diagnostic, was proposed by Bell *et al* during the same year. Bell *et al* classified these tumors in accordance with their histopathological prognostic: presence or absence of tumor necrosis, nuclear atypia and mitosis index. These criteria have been adopted by Stanford as well. This last classification was accepted and proposed for medical use by world health organization in 2003. Further on, Bell *et al* classified STUMP into three other groups: 1) atypical leiomyoma with low chance of recurrence that has a diffuse moderate to severe atypia, less than 10 mitosis/HPF and no tumor cell necrosis; 2) atypical leiomyoma but with limited experience that has severe atypia, less than 20 mitosis/10 HPF and no tumor cell necrosis; and 3) smooth muscle tumor of low malignant potential that has tumor cell necrosis, mitosis less than 10/10 HPF and absent to mild atypia (6, 7). Therefore, taking into consideration the histological description of our biopsy piece, we could certainly say that it was a STUMP tumor, most probably being placed in the second group.

Even though multiple classifications and histological subdivisions of these tumors were made, certainty diagnosis and prognosis remain difficult to impose and represent a crucial issue in correctly treating patients. Patients with

STUMP have symptoms that are commonly found in patients with benign or malignant conditions such as abdominal and pelvic pain, abnormal uterine bleeding, pelvic pressure and the sensation of a pelvic pressure. The mean age of onset for this pathology is 45 years old, and it mostly appear during the premenopausal stage of reproductive life (8, 9).

As observed in our case report, imagistic investigations face difficulties in diagnosing and differentiation of STUMP from other uterine tumors. The first study that took into consideration the difficulties encountered in diagnosing STUMP during imagistic investigations was published by Bonneau *et al*. It compared the images retrieved from MRI and sonography investigations from 105 patients, out of which 85 had leiomyomas and 23 malignant mesenchymal tumors (MMT) and STUMP. The authors concluded that for the majority of patients with STUMP and MMT, the imagistic investigation showed the presence of a single tumor and free abdominal fluid as well as absence of acoustic shadowing (10).

The recurrence type of tumor is either leiomyosarcoma or STUMP and it could be located in the uterine, pelvic or retroperitoneal area, or even neighbor sites such as the liver, lung or bone tissue (11, 12).

When deciding to use a certain therapeutic scheme, the patient's age, fertility status and desire to maintain it or not, pathological features, recurrences and location of the tumor should be carefully taken into consideration. Studies have shown a 52% rate of pregnancy achievements after myomectomies in such cases. When having to deal with recurrences of these tumors, physicians should consider adjuvant therapies such as radiotherapy, chemotherapy or progestins gonadotroph releasing hormones analogs (11, 13, 14).

As mentioned above, the follow-up of these patients is essential when considering the recurrence rate. Therefore, even if we do not have a well-established protocol yet, studies suggest that these patients should have a follow-up every six months for the first five years and then annual surveillance for the next five years. During their consultations, patients should benefit from full body examination, laboratory investigations, imagistic investigations such as thoracic X-rays, pelvic ultrasound or MRI whenever the case, and PET-CT in order to identify recurrences (11). □

CONCLUSIONS

The rarity of these tumors, difficulties encountered in posing a pre-interventional diagnosis and establishment of a well conducted follow-up represent a real challenge for all surgeons and physicians nowadays. Smooth muscle tumor of uncertain malignant potential diagnosis can only be confirmed after a myomectomy or hysterectomy by histopathological examination. Therefore, it is difficult for us to inform our patients that a successful surgical intervention does not

necessarily mean a 100% certainty that the tumor was malignant or benign. Patients must also face the burden of biannual and annual follow-up for at least 10 years as well as recurrence possibility.

In conclusion, STUMP is a challenge for both the physicians and patients and more literature data is welcomed in order to finally establish a complex therapeutic protocol. □

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