Uterine Smooth Muscle Tumors of Unknown Malignant Potential (STUMP): A Dilemma for Gynecologists and Pathologists

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Uterine smooth muscle tumors of uncertain malignant potential (STUMP) belong to a subcategory of uterine smooth muscle tumors. The clinical behavior of these tumors is also not clarified. It is hard to state which histological features of STUMPs might represent high recurrence rates. The follow up period also has not been mentioned. For clinicians, STUMPs are difficult tumors to manage since they clinically present as ordinary fibroids. For pathologists, diagnosis of these tumors could be difficult because these tumors are rare and a certain level of expertise in gynecological pathology is required to make a correct diagnosis. Here we report a patient with STUMP that subsequently recurred as a leiomyosarcoma, aiming to remind the disease and its recurrence risk even if followed strictly.

Key Words: Uterus, Smooth muscle tumors, STUMP, Recurrence, Leiomyosarcoma

Gynecol Obstet Reprod Med 2013;19:55-57

Introduction

Uterine smooth muscle tumors of uncertain malignant potential (STUMP) belong to a subcategory of uterine smooth muscle tumors.[1] These tumors are a heterogeneous group of neoplasms, from both the histological and clinical point of view. Although some investigators proposed classification systems to better categorize these tumors, controversy still exists.[2,3] The clinical behavior of these tumors is also not clarified. While majority of cases follow a benign course, some of the patients suffer from recurrent disease and even succumb to disease. The histologic distinction between benign and malignant uterine smooth muscle tumors is usually based on the assessment of combination of features including atypia, mitotic rate, and presence or absence of tumor cell necrosis.[4]

For clinicians, STUMPs are difficult tumors to manage since they clinically present as ordinary fibroids. For pathologists, diagnosis of these tumors could be difficult because these tumors are rare and a certain level of expertise in gynecological pathology is required to make a correct diagnosis.[5] In case of recurrence, histology of the recurrent tumor could be STUMP or leiomyosarcoma.[2] Here we report a patient with STUMP that subsequently recurred as a leiomyosarcoma, aiming to remind the disease and its recurrence risk even if followed strictly.

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Submitted for Publication: *28. 02. 2013*
Accepted for Publication: *15. 04. 2013*

Case Report

A 50-year-old woman was admitted to our hospital with menorrhagia and pelvic pain. Physical examination was unremarkable except for globally enlarged and myomatous uterus. Transvaginal ultrasonography showed multiple intramural myomas in various sizes ranging 0.5 cm to 10 cm. An office endometrial biopsy revealed disorganized proliferative endometrium. Her past medical history and family history was uneventful.

The patient underwent total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO). At the time of surgery, her uterus was enlarged to the size of 16-week pregnancy. Bilateral adnexal structures were normal. Patient did well after surgery and discharged on third postoperative day.

Before permanent paraffin procedure, macroscopic examination of the specimen brought out multiple intramural smooth muscle tumors in the uterus. Slices from these smooth muscle tumors showed moderate pleomorphism and hyalinization necrosis. The mitotic rate averaged 3-4 per 10 high power fields (HPF) with a maximum of 7 per 10 HPF. Permanent pathology was reported as STUMP. (Figure 1)

Patient was put on a close clinical follow-up schedule, with pelvic examination and abdominopelvic ultrasound every 6 months and chest X-ray and pelvic computed tomography

(CT) scans every year. In the second year of her surgery she presented with abdominal distention. On physical examination, a palpable pelvic mass to the level of umbilicus was noted. A CT scan of abdomen and pelvis showed a giant, complex, fixed pelvic mass; 14x12 cm in diameter which is nearby the sigmoid colon and posterolateral part of the bladder (Figure 2). There was bilateral hydroureteronephrosis which was most prominent on the left side since the mass was deviated to left side and compressing the urinary tract (Figure 3). Renal function tests and other laboratory parameters including tumor markers, liver enzyme levels, electrolytes and hemogram were all normal. The patient underwent an exploratory laparotomy with a midline vertical incision. A 25 cm, fixed pelvic mass was noted. There were dens adhesions between the mass and surrounding pelvic viscera, vascular and bowel compositions. No abnormalities were detected on the peritoneal surfaces, liver, spleen and diaphragm. Avascular retroperitoneal spaces were developed and both ureters were unroofed before excision of the mass. Despite these precautions and superb surgical skills, the amount of blood loss was approximately 2.000 mL and the patient required transfusion. Postoperative recovery was uncomplicated.

The final pathology result revealed leiomyosarcoma with atypia and limited necrosis. The mitotic rate averaged 18 per 10 high power fields (HPF). (Figure 4)

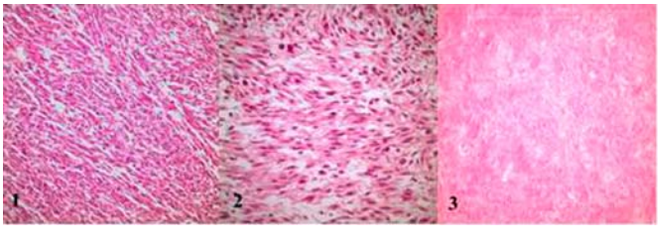

Figure 1: Microscopic features of STUMP
1: Tumor displays moderately high cellularity, 2: Mild cytologic atypia and sparse mitotic figures, 3: An area of hyalinization like necrosis.

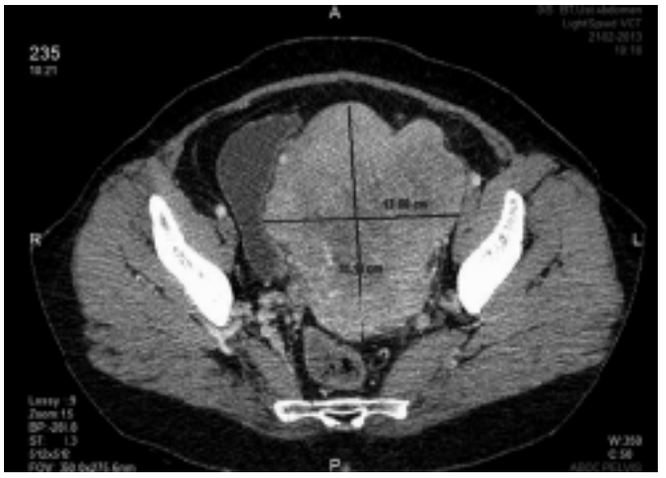

Figure 2: A giant, solid-cystic mass overflowing the pelvis

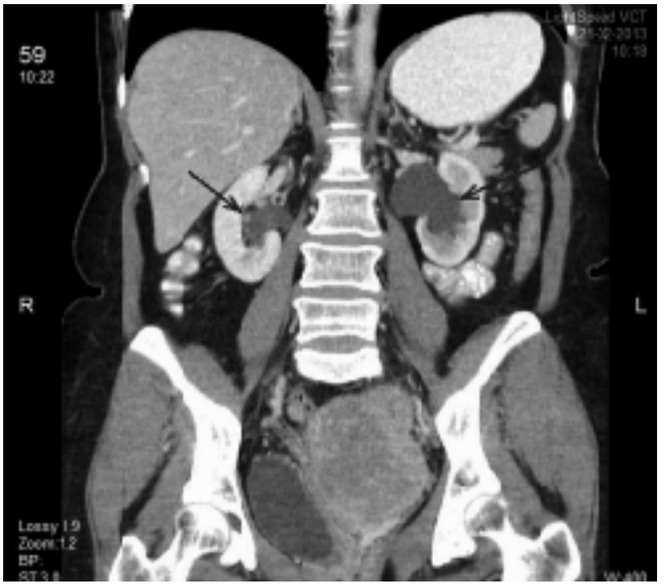

Figure 3: Bilateral hydronephrosis

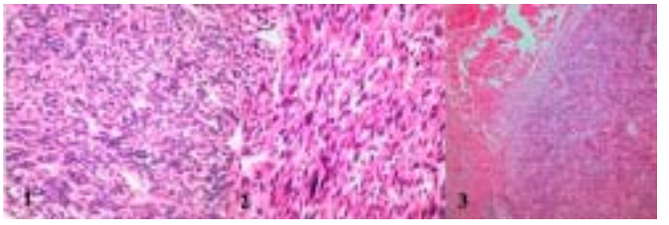

Figure 4: Microscopic features of leiomyosarcoma
1: Tumor displays marked cellularity, 2: Moderate cytologic atypia and numerous mitosis, 3: A small focus of necrosis.

Discussion

The term ‘‘smooth muscle tumor of uncertain malignant potential’’ was first used in the literature by Kempson in 1973.[4] Since these tumors are rare, the literature on this topic is limited. STUMP could be diagnosed following myomectomy or hysterectomy. The diagnosis of these tumors is challenging for pathologists as interpretation of cellular atypia, mitotic index and coagulative tumor cell necrosis is subjective. There are no available guidelines regarding appropriate management of these tumors. The majority of cases follow a benign clinical course, however a few can recur as either tumor of low malignant potential or leiomyosarcomas.[5]

In 1994, Peters et al.[6] reported a recurrence rate of 27% and an overall survival rate of 92% at 5 years. However, their report did not specify the histology of recurrent tumors (STUMP vs. leiomyosarcoma). Barretta et al.[7] reviewed outcomes of three patients with STUMP. During follow-up, one of these three patients developed distant metastasis. Histologic examination of metastasectomy specimens revealed STUMP. The study with largest series of patients was published by Guntupalli et al.[2] of M.D. Anderson Cancer Center in 2009. 41

patients with STUMP were included in the study. Three patients (7.3%) had a recurrence and one of the patients was found to have a leiomyosarcoma at the time of recurrence. Another important finding of this study was the similar recurrence rates between patients who underwent myomectomy and who underwent hysterectomy.

Since the lack of patient series diagnosed with STUMP, it is hard to state which histological features of STUMPs might represent with high recurrence rates. However Bell et al.[1] reported tumor cell necrosis and atypia as the most correlative factor with malignant potential. Our patient's pathology result revealed a moderate atypia and necrosis after the first operation.

The majority of these tumors are seen in the reproductive age, especially between 30-39 years and abdominal distention with rapidly growing dimensions of uterus and vaginal bleeding are the most seen symptoms.[8] Both of these symptoms were present in our patient and the recurrent giant mass caused hydroureteronephrosis.

The literature mentions most of these tumors with a benign clinical behaviour, so that myomectomy or hysterectomy could be enough for the treatment.[9] Adjuvant therapy is not required after the initial diagnosis of STUMP.[2]

In conclusion, both diagnosis and clinical behavior of these neoplasms are poorly understood. There is a notable risk of recurrence and the follow-up interval for women who have STUMP should probably be a minimum of every 6 months until the 5th year and, thereafter, annual surveillance should be practiced for a further 5 years. The choice of treatment for recurrence is surgical excision and it would be advisable to consult these patients to a gynecologic oncologist or a generalist with advanced surgical skills.

Malignite Potansiyeli Bilinmeyen Uterin Düz Kas Tümörleri (STUMP): Jinekolog ve Patologların Çıkmazı

Malignite Potansiyeli Bilinmeyen Uterin Düz Kas Tümörleri (STUMP) uterus düz kas tümörlerinin bir alt grubudur. Bu tümörlerin klinik seyri net kesinlik kazanmamıştır. Hangi histolojik STUMP tiplerinin daha yüksek rekürans oranı göstereceğini söylemek güçtür. Belli bir takip konsensusu oluşmamıştır. Klinisyenler için STUMP'un yönetimi sıradan miyomlar gibi prezente oldukları için güçtür. Patologlar için ise bu tümörlerin tanısı nadir görüldükleri için güç olmakla beraber doğru tanı için jinekolojik patoloji üzerinde belli bir süre deneyim gerekmektedir. Biz bu vakada histerektomi spesimeninde STUMP saptanan bir olgunun leiomyosarkom reküransı şeklinde prezente olmasını tartıştık.

Anahtar Kelimeler: Uterus, Düz kas tümörleri, STUMP, Rekürans, Leiomyosarkom

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