When we operate on a uterine fibroid, and we come across a smooth muscle cell tumor of uncertain malignant potential (STUMP): A case report

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ABSTRACT Background: Smooth muscle cell tumor of uncertain malignant potential is a rare tumor. By reporting a case report, our study aims to define this tumor, its therapeutic management, and its prognosis. Case report: Our patient is a multiparous 46 years-old patient with chronic pelvic pain and no pathological medical history. Her pelvic ultrasonography showed an aspect of a 64 mm uterine fibroid. She underwent a myomectomy. The anatomopathological study showed a smooth muscle cell tumor of uncertain malignant potential. A total hysterectomy with bilateral adnexectomy was performed. No signs of recurrence were noted during surveillance. Conclusion: Smooth muscle cell tumor of uncertain malignant potential are heterogeneous with no clinical nor radiological specific signs. The treatment is mainly surgical with hysterectomy or myomectomy if fertility preservation is desired. The risk of recurrence is always present and can occur after several years hence the importance of prolonged monitoring.

KEYWORDS Smooth muscle cell tumor of uncertain malignant potential, uterine STUMP, myoma, recurrence

Introduction

Uterine smooth muscle cell tumors are the most common tumors of the uterus. Leiomyomas are the benign and leiomyosarcomas the malignant ones. Uterine smooth muscle cell tumors of uncertain malignant potential (STUMP) is a subcategory of uterine smooth muscle tumors defined by exclusion: tumors that do not fit the definition of any of the other categories of uterine smooth muscle tumors are classified as STUMP [1]. Being highly heterogeneous, the diagnostic guidelines had not been standardized, nor therapeutic one. The management and the follow-up of uterine STUMP continue to be challenging. By reporting a case of uterine STUMP, we will review the current state of knowledge about STUMP and its possible treatments.

Case report

We report the case of a 46 years-old woman, G3P2, with no particular medical history, who suffers from chronic pelvic pain with asthenia. The physical examination was normal. Her Pap smear didn’t show any abnormality. A pelvic ultrasound showed a 64 mm lateral fibroma (Figure 1). She was anemic with 9.2 d/dl of hemoglobin. The patient refused hysterectomy in the first place. She performed magnetic resonance imaging (MRI) (Figure 2) that showed the same aspect. She underwent a myomectomy.

The anatomopathological study showed a mesenchymal tumor characterized by a proliferation of smooth muscle fibers arranged in regularly tangled bundles. These cells show moderate cytonuclear atypia visible at low magnification (100). Mitotic activity is low: less than one mitosis per 10 fields with high magnification. No ischemic necrosis was seen.

A total hysterectomy with bilateral adnexectomy was performed after discussion with the patient. The same anatomopathological funding was found. The patient did not undergo any further treatment. Three years of regular follow-up didn’t show any sign of recurrence.
Discussion

STUMP is a subcategory of smooth muscle cell tumors of the uterus that can be difficult for pathologists to diagnose. It's defined histologically by the presence of any unusual combinations of the three features that do not satisfy the current criteria for leiomyosarcomas such as diffuse moderate-to-severe atypia, a mitotic count of at least 10 mitotic figures per 10 high power fields (according to Stanford criteria) and no tumor cell necrosis [1, 2].

The mitotic count is important for the differential diagnosis between STUMP and Leiomyomas in particular for leiomyomas with bizarre nuclei, although degenerating nuclei called "pseudotypical mitosis" is sometimes difficult to distinguish from true mitotic figures. So, although counting mitosis would appear to be an entirely objective procedure, assessment of nuclear atypia is subjective and prone to variations in interpretation from pathologist to pathologist [3].

It has been shown that MIB-1, p53, and progesterone receptors are useful in differentiating leiomyosarcoma from the STUMP group. MIB-1 and/or p53 expression of more than 15% is seen in all leiomyosarcomas but none of the STUMP category. Progesterone receptor is absent in most leiomyosarcomas but seen in all STUMP [4].

Regarding clinical features associates with STUMP, they are not specific. No differences are seen between STUMP and leiomyoma. The symptoms are fundamentally abnormal uterine bleeding, pelvic mass, pelvic pain, symptoms of anemia such as fatigue, pale skin, and tachycardia, or a combination thereof [5].

Like in leiomyomas and leiomyosarcomas, the mean age of diagnosis seems to be around 45 years [6]. Most of the patients are premenopausal [7]. No specific risk factors are found, neither race nor ethnic differences.

Radiologically, neither pelvic sonography nor magnetic resonance imaging (MRI) seems to provide a pre-operative orientation. We are more likely to find a single tumor, no acoustic shadowing, and free fluid in STUMP's imaging [8]. Contrast-enhanced MRI showed better accuracy than diffusion-weighted MRI, more pieces of information are provided in the diagnosis of malignancy and in identifying pelvic recurrence. Before surgery, imaging technics, such as computed tomographical scan and MRI are useful to provide additional information regarding the density and relationship with the surrounding structures if the pelvis [9].

Most STUMP's are diagnosed with endometrial biopsy or myomectomy. The strategy to be followed for the management of this disease remains controversial. Hysterectomy is considered the gold standard and is especially recommended for women who have completed their childbearing. In young women who choose to preserve their fertility, myomectomy is an alternative to a hysterectomy after discussion with the patient and exposure of the risk-benefits. Recent studies highlight the feasibility of fertility-sparing approaches despite the not insignificant risk of recurrence. Vilos et al. found 6.6% of patients who underwent myomectomy experienced relapse (1 of 76 patients). The death rate was 1.3% [5].

Heterogeneity of the STUMP's and difficulty of determination of the true malignant potential of these tumours lead to therapeutics dilemma. Their behaviour is unpredictable—the median survival is 61.5 months [2]. Five years survival is 92-100% [7]. Recurrence can be as uterine STUMP, uterine leiomyosarcoma, or distant metastasis including lymph node metastasis. Its rate ranges between 8.7% and 11% [1].

Relapse appears to occur generally after a long disease-free interval for up to several years, just as happens with borderline ovarian tumours. The mean value of time to recurrence is 54 months. That is the reason why long-term close follow-up is necessary [10]. Ip et al. suggested a follow-up interval of 6 months for the first five years, followed by annual surveillance for at least five additional years [2]. Careful chest monitoring should be done since the lung is found to be the most frequent metastatic site.

Treatment of recurrence remains surgical. Adjuvant treatment, such as chemotherapy and pelvic irradiation, has been reported without a standardized protocol, affected by the physician’s preferences. Doxorubicin and cisplatin were found to be the most used chemotherapeutic agents as adjuvant and palliative treatment. Some studies reported the use of endocrine therapy based on progesterone, but further investigations should be done [11]. Most authors agree that the individualization of treatment after relapse is necessary.

Conclusion

Uterine STUMP's remain a therapeutic dilemma. Its clinical and radiological presentations have no specificity. The diagnosis is histological, sometimes difficult. A multidisciplinary approach is mandatory. Surgery is the standard gold treatment and consists of hysterectomy or myomectomy when fertility needs to
be preserved. Follow-up is essential and should be prolonged for many years since the risk of recurrence exists. Therapeutic protocols are needed for relapse treatment. More understanding of STUMP's carcinogenesis may provide further information about the disease, its accurate diagnosis, and even therapeutic possibilities.

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Conflict of interest
There are no conflicts of interest to declare by any of the authors of this study.

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