Smooth muscle tumor of uncertain malignant potential (STUMP)

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ABSTRACT

Uterine smooth muscle tumor of uncertain malignant potential (STUMP) is a rare tumor belonging to a group of smooth muscle tumor that possess both benign and malignant features, complicating the diagnosis. STUMP is a rare uterine tumor with a paucity of literature available regarding management and subsequent malignant potential.

A 22-year-old unmarried lady presented as uterine fibroids with 24 weeks size uterus. There was one intramural and another subserosal mass both of which revealed smooth muscle tumor of uncertain malignant potential in histopathology.

Keywords: abdominal lump, fibroid uterus, STUMP

INTRODUCTION

The World Health Organization defines uterine smooth muscle tumor of uncertain malignant potential (STUMP) as a borderline tumor between benign leiomyoma (LM) and malignant leiomyosarcoma (LMS)[1]. It is a rare slow growing tumor, with an incidence of 0.1% in histological specimens of women undergoing myomectomy or hysterectomy for a preoperative diagnosis of leiomyoma[2]. The diagnosis of STUMP is based on presence of three histopathologic features which are mitotic count activity or mitotic index (number of mitotic figures per 10 high power fields (HPF), presence of coagulative tumor cell necrosis (CTCN) and degree of cytological atypia. Of these, the presence of CTCN is pathognomonic of diagnosis of STUMP. The correct diagnosis of STUMP tumors however, can be challenging as many histologic characteristics overlap with rare subtypes of leiomyoma variants.[3] Accurately diagnosing uterine smooth muscle neoplasms is clinically significant for optimal patient management.

CASE

We present a case of 22- year- old unmarried nulliparous woman who presented to gynecology OPD of our hospital with the chief complaints of secondary amenorrhea for one year. Her urine pregnancy test was negative. Her menstrual history since menarche had

reduced flow in the form of spotting for 1 day with a regular cycle length of 28-30 days associated with dysmenorrhea. There was no history of tuberculosis or any other chronic illness. On general examination her vitals were stable and body mass index was 20 kg/m^2. Abdominal examination revealed a firm, regular mass arising from pelvis that extended up till the umbilicus corresponding to 24 weeks size gravid uterus. The mass was mobile side to side with well-defined borders. On bimanual examination same findings were confirmed. Her baseline investigations revealed a hemoglobin of 13 g/dl with normal liver and renal functions and normal hormone profile i.e., Day 2 LH, FSH, prolactin and thyroid function test. Her CA 125 was 21 IU/ml. Transabdominal ultrasound revealed a large intramural fibroid at the anterior wall of the uterus measuring 10×15 cm and another sub-serosal fibroid at the posterior wall of the uterus of 3 × 4 cm. Both the adnexa were normal looking. Magnetic resonance imaging (MRI) pelvis was done for mapping of leiomyoma before surgery as patient was nulliparous and the finding were found similar to ultrasonography with no features suggestive of malignancy. After informed and written consents, patient was taken up for myomectomy. Intraoperative findings were of an enlarged uterus with two fibroids in the anterior (intramural) and posterior wall (sub-serosal). Grossly, the uterine mass measured 20x20 cm irregular and an another subserosal fibroid of 3x4 cm. Both tubes appeared beaded and blocked at their fimbrial ends. Left ovary was not visible separately due to tubo-ovarian complex of 4 × 3 cm. Right ovary was normal looking. Peritoneal biopsy was taken to rule out tuberculosis in view of beaded Fallopian tube with tubo-ovarian complex, however there was no evidence of tuberculosis on HPE. There was no free fluid in the pelvis and no visible findings suggestive of malignancy. Myomectomy was done. Specimen retrieved was sent for histopathology. Postoperative recovery was uneventful and the patient was discharged on day 3. Histopathological examination revealed STUMP. The tumor had a low mitotic index with five mitotic figures per 10 high-power fields (HPF) and no coagulative tumor necrosis however, diffuse significant cytological atypia was seen. [Figure-1]

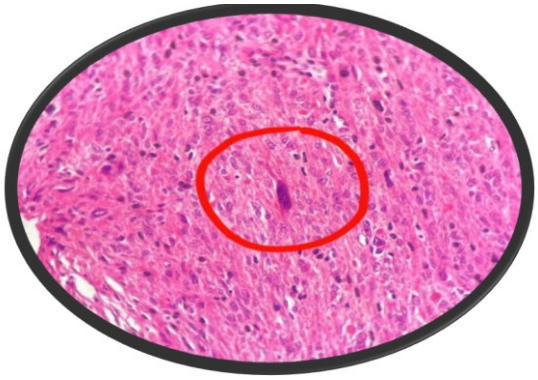

Figure-1: STUMP showed spindle-cell proliferation marked by red outline, moderate atypia, and absence of CTCN.

After a multidisciplinary team discussion, it was decided that her management should include a bi-annual clinical review and pelvic ultrasound and that she should have a simple hysterectomy once she completes her family. Patient was explained about the same and is currently under follow up.

DISCUSSION

Studies have shown that it affects women in the perimenopausal age group with a mean age of 45 years similar to LM and LMS. The clinical signs and symptoms of STUMP mimic those of LM and LMS. These include presence of pelvic pain, abnormal uterine bleeding, pelvic mass, symptoms secondary to anemia or compression or a combination of them.

Sonographic presence of single tumor, absence of acoustic shadowing and presence of free fluid to be more commonly associated with STUMP/ malignant mesenchymal tumor if detected. MRI has been used to differentiate benign leiomyoma and LMS utilizing increased signal intensity, but evidence is still lacking to distinguish STUMP from leiomyoma. So, the diagnosis is in majority cases will be post-surgery. Histological distinction between malignant and benign smooth muscle tumors remains challenging, therefore researchers evaluated the role of immune- histochemical (IHC) markers expression to aid diagnosis.[3] The most commonly studied markers are p16, p21, p53, Ki 67, Bcl-2, progesterone and estrogen receptors and their expressions are highlighted. [Table-1]

Table-1: Expression of IHC markers

IHC markers	Leiomyoma	STUMP	Leiomyosarcoma
p16	+	-	++
Bcl 2	++	+	+
p53	-	-	+
Ki 67	-	-	+
Progesterone receptor	+++	++	+

The use of immunohistochemistry has a definite role in diagnosis and risk stratification of the tumors but its utility should be weighed against the cost of the tests.

Uncertain malignant potential, indolent behavior, and prolonged survival of the tumor, leaves the management at the crossroads for the patient as well as the treating gynaecologists and oncologists. A postoperative diagnosis of STUMP on myomectomy specimen does not warrant a reoperation and hysterectomy. Hysterectomy is currently considered the gold standard and it is especially recommended for women who have completed their childbearing. On the other hand, the choice between myomectomy and hysterectomy represents an extremely important issue in the management of STUMP in young women balancing the risk of recurrence and the preservation of fertility and the need for strict surveillance.

The recurrence rates range from 8.7% to 11% irrespective of the type of surgery performed. Time to recurrence ranges from 2 to 194 months in the published datas.[4] This wide range is due to the unpredictable behavior of STUMP. Although standard guidelines for treatment are not available, the common strategy for recurrence remains surgical treatment. Role of adjuvant therapy in the form of hormone therapy, pelvic irradiation or chemotherapy is not clear as the clinical course of the tumor has been found to be similar in absence of such treatment.

Metastasis of STUMP is rare, but a reported phenomenon. The lung has been the most common extra-uterine site for metastasis followed by the bone. Miller et al in a retrospective review identified 10 patients with benign metastasizing leiomyoma to the lungs [5]. The most common distant metastatic sites were found to be lung (15/46, 33%), bone (7/46), liver and peritoneum.[5]

CONCLUSIONS

Uterine STUMP has posed as an enigma for the last 3 decades. Concerns regarding over or under diagnosis exists due to lack of specific diagnostic criteria, indolent clinical course and possible malignant potential. A multidisciplinary approach is mandatory and future perspective studies should be undertaken to identify the molecular basis of STUMP using molecular biology techniques. The identification of key genes directly

involved in the carcinogenesis of STUMP may suggest novel opportunities in the management of the disease and provide further information in understanding the process of carcinogenesis. To conclude, it is suggested that a detailed pathological evaluation by experienced gynecological pathologists will go a long way in correctly diagnosing and managing such cases.

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