CASE REPORT

Smooth Muscle Tumor of Uncertain Malignant Potential: An Unexpected Diagnosis in a Young Female

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Abstract

A 25-year-old married nulliparous female with suspected fibroid uterus was admitted to the emergency ward with complaints of heaviness in the pelvic region, increased uterine bleeding for the past year, and torrential bleeding episodes. She was clinically very pale and presented with shock symptoms in an emergency. Conservative management with a blood transfusion was performed. Radiological imaging revealed a fibroid uterus with gross cystic degeneration. Her nulliparous state necessitated an abdominal myomectomy. However, the planes were not well-defined, and the cystic fluid was profuse; therefore, the final diagnosis was inconclusive. Her histopathology report revealed a uterine smooth muscle tumor of uncertain malignant potential (STUMP).

Keywords: Case report, Fibroid, Malignancy, Myomectomy, Recurrence, Smooth muscle tumor of uncertain malignant potential.

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Introduction

Uterine smooth muscle tumors are classified as leiomyoma and leiomyosarcoma, being benign and malignant, respectively, depending upon the presence of tumor cell necrosis, cytological atypia, and mitotic activity. Uterine smooth muscle tumor of uncertain malignant potential (STUMP) is a rare tumor (reported incidence 0.01%).[1,2] It is "a smooth muscle tumor with features that preclude an unequivocal diagnosis of leiomyosarcoma, but it does not fulfill the criteria for leiomyoma; it may behave in a malignant fashion".[2] A case series of six patients with over 10 years of study is reported from Turkey.[1] Additionally, two more cases were reported from India, one as a metastatic pulmonary nodule with a history of hysterectomy three years ago and another as a huge fibroid with myomectomy followed by hysterectomy in the same setting due to a frozen section positive for malignancy.[3,4]

Case Description

A 25-year-old married nulliparous female presented to the emergency room with complaints of abdominal lumps and pelvic heaviness for the last year and excessive bleeding per vaginum for 7 days. She had a history of gradually increasing heavy periods over the last year, with a sense of heaviness and pressure in the pelvic and lower abdomen. Her bladder and bowel habits were normal. Upon general examination, she appeared clinically pale, and exhibited tachycardia, hypotension, and dehydration, prompting immediate resuscitation. Upon examination of the abdomen, we observed a firm, mobile, and tender 26-week-old abdominoplevic lump. Upon speculum examination, we found massive clots in the vagina, pale vaginal mucosa, and a nulliparous cervix with active bleeding. Upon bimanual examination, we identified the same abdomino pelvic mass with restricted mobility, obliterated the anterior fornix, and found both lateral fornices free.

Her urgent complete blood count was conducted, which showed Hb: 4.5 gm/dL, TLC: 13, 450/mm^3, DLC–N65 L28 E1 B1. While, liver function tests, renal function tests, electrolytes, PT-INR, urine routine, and culture were within normal limits.

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Her transabdominal ultrasound showed a huge abdomino pelvic lump of size 16 × 12 × 12 most likely a huge fibroid uterus with gross cystic degeneration. In view of such an atypical presentation of fibroid MRI was performed further. MRI showed large uterine masses with multiple moderate sized, lobulated, well-defined and smooth lesions present, the largest measuring 120 × 93 × 127 mm, suggesting a bulky uterus with multiple fibroids and cystic degeneration with mild hydroureteronephrosis (Fig. 1).

Management

She received four units of packed red blood cells (pRBC) for very severe anemia. Her abdominal myomectomy was performed after

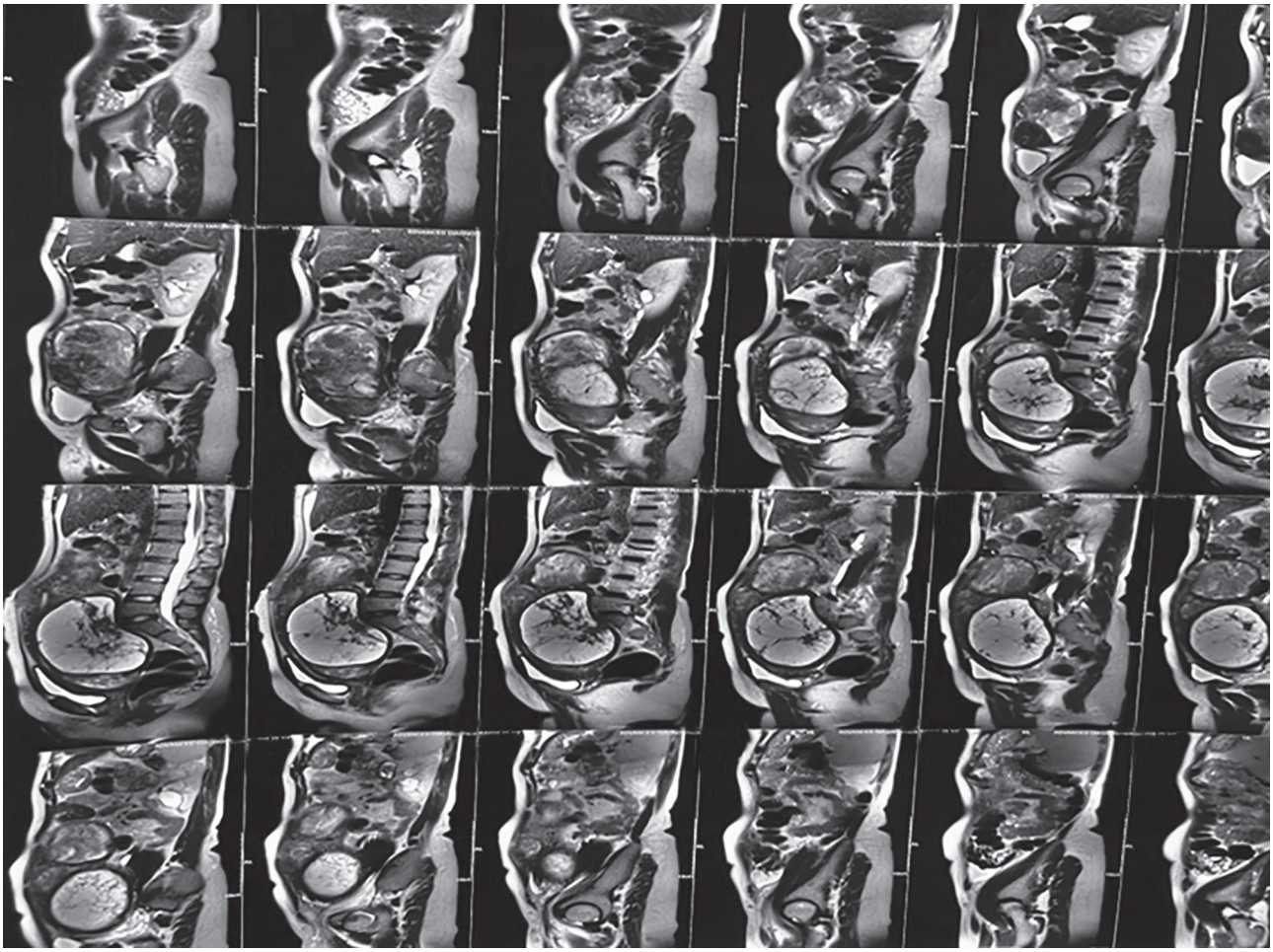


Fig. 1: MRI-multiple moderate sized, lobulated lesions, well defined and smooth lesions present, largest measuring 120 × 93 × 127 mm, suggestive of bulky uterus with cystic degeneration with mild hydroureteronephrosis

ureteric stenting (in view of hydroureteronephrosis). During the laparotomy, the uterus revealed large myomas (Fig. 2). Peroperative findings were different from usual leiomyoma: (A) Planes of differentiation between myoma and healthy myometrium were poorly formed; (B) The amount of cystic fluid that was present within the lesions was copious; (C) Four myomas were removed without opening the cavity. The patient received one unit of PRBC and two units of FFP intraoperatively. She did well in the postoperative period and was discharged on the 10th postoperative day.

Histopathology showed predominantly spindle cells with indistinct borders, eosinophilic cytoplasm, and cigar-shaped nuclei. Bizzare shaped cells with hyperchromatic, multifocal nuclei have a fair number of mitotic figures (5–6/10 HPF) with necrosis evidently suggestive of a (Fig. 3A).

Immunohistochemistry showed smooth muscle antigen (SMA) (Fig. 3B), H-caldesmon, ER, PR positive; CD-10, STAT-6, HMB-45-negative and Ki67PI-15% positive.

A literature search revealed this tumor to have a high recurrence rate. So, this case was discussed on the tumor board of the institute where it was opined that she should undergo a transvaginal scan and MRI pelvis every 6 months to see the growth of the myomas, and she was encouraged to complete her family as she might require a hysterectomy later on in life.

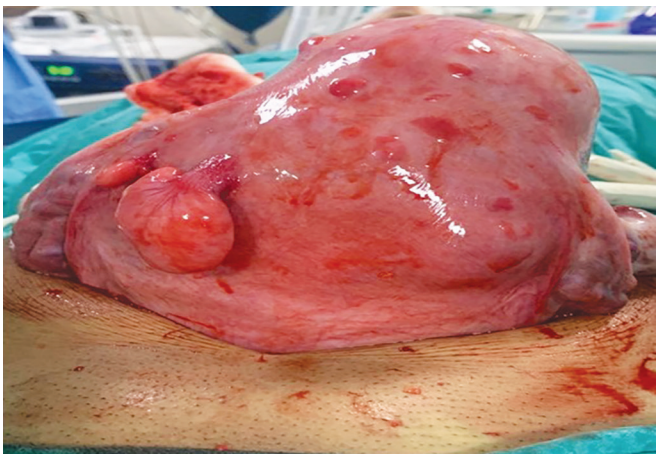
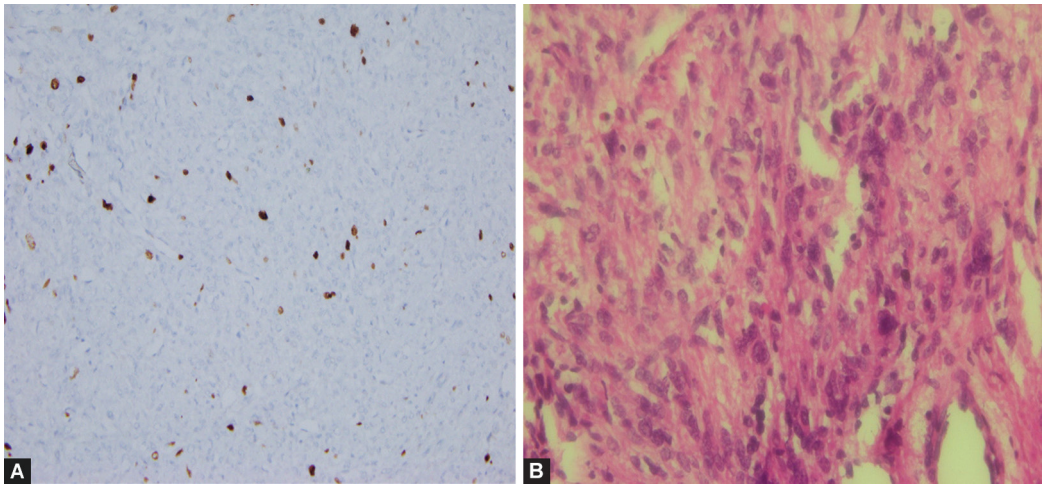


Fig. 2: Gross appearance of the tumor with multiple uterine fibroids

She received advice to attempt conception due to her nulliparous status. She conceived spontaneously after a few months. Furthermore, she was under our regular follow up during the entire antenatal period, which was uneventful. Her pregnancy was terminated at 37 completed weeks of pregnancy by a lower



Figs 3A and B: (A) Immunohistochemistry (SMA+) Ki67PI-15% positive; (B) Histopathology showing bizzare shaped cells with mitotic figures (5–6/10 HPF)

segment cesarean section, and she delivered a healthy male baby weighing 3 kg. There was no myoma in the lower segment of the uterus, so stitching was done without any difficulty. The body of the uterus was palpated, which revealed 4–5 round, smooth masses of sizes around 3–4 cm on the anterior surface of the upper part of the body of the uterus near the fundus. She did well and was discharged in satisfactory condition from the hospital on the 5th postoperative day.

Discussion

The World Health Organization (WHO) classifies STUMP as a smooth muscle tumor that falls between benign and malignant criteria. It is a subtype of uterine smooth muscle tumors, with an incidence of about 0.01%.[1] Usually, the histopathology report of post-hysterectomy and occasionally myomectomy specimens makes this diagnosis.[1] Clinical signs and symptoms are similar to those of leiomyoma and leiomyosarcoma.[1] Those are pelvic pain, abnormal uterine bleeding, pelvic mass, pelvic pressure sensation, or a combination of them. Smooth muscle tumor of uncertain malignant potential has a recurrence rate of 7–27%.[3] Stanford criteria are used for histopathological classification, which is based on the presence of abundant mitosis (≥10 per HPF), the presence of atypical cells, and areas of coagulative tumor cell necrosis. Smooth muscle tumor of uncertain malignant potential shows these features but does not fulfill the diagnostic criteria for leiomyosarcoma.[4]

This case, however, presented in an early age group, was a nulliparous female with all findings in favor of benign fibroid with cystic degeneration. It was planned for fertility-preserving surgery. The HPE report showed a STUMP. She conceived spontaneously and had an uneventful antenatal, intranatal, and postnatal course.

Bahadur BR et al.[4] reported a case where a 28-year-old patient presented to out-patient department (OPD) with an abdominal pain complaint without any menstrual disturbance. Myomectomy was done, but on the frozen section, malignancy was identified, and so hysterectomy was done. In our case, the patient had gross menorrhagia, leading to very severe anemia. A myomectomy was done for her, and she conceived spontaneously and delivered a healthy baby.

Conclusion

We reported a STUMP case that appeared in the second decade of life in a nulliparous female. The definitive treatment was surgery (hysterectomy ± bilateral salpingo-oophorectctomy, depending on age and other factors). The prognosis depends on the mitotic index and the extent of the disease. Patients with STUMP, especially those with myomectomy, should be informed of the risk of recurrence and closely monitored.[4,5] Thus we conclude that preoperatively, sonographic discrimination from leiomyoma or leiomyosarcoma is not possible. When dealing with a rapidly growing fibroid uterus, we should keep this clinical entity (STUMP) as one of the differential diagnoses and confirm the final diagnosis through histopathological and immunohistochemical analysis.

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