

Giant uterine STUMP in pregnancy: A rare case report with preservation of fertility

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Abstract

Background: Smooth muscle tumors of uncertain malignant potential (STUMPs) are infrequent and represent an extremely rare cause of oligohydramnios in pregnant women.

Description of the case: A 34-year-old woman at 25 weeks of gestation was referred with clinical features indicative of anemia. The transabdominal ultrasound revealed a fetus with appropriate growth for the gestational age, with oligohydramnios and a giant uterine mass. At 30 weeks of gestation, due to anhydramnios and repeated non-reassuring non-stress tests, a cesarean section was performed, followed by a myomectomy of a 9 kg mass. A neonate weighing 1,350 g was delivered (Apgar score 1': 4, 5': 7). The woman preserved her fertility and had an uneventful recovery during the puerperium, while the final histology revealed multiple STUMPs. The postoperative management included follow-up every six months for five years.

Conclusion: This is the only reported case of a pregnant woman with STUMPs of this size that underwent myomectomy during the cesarean section and avoided hysterectomy. HIPPOKRATIA 2021, 25 (4):169-171

Keywords: Giant mass, pregnancy, fibroid, myomectomy, STUMP, Smooth muscle tumors of uncertain malignant potential

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Introduction

Fibroids are found in about 3-12 % of pregnant women and may increase in size, mainly in the first trimester¹. Smooth muscle tumors of uncertain malignant potential (STUMPs) are rare tumors that cannot be classified as benign or malignant². Moreover, STUMPs have been previously described interchangeably as leiomyomas with bizarre nuclei or atypical leiomyomas³. Most cases of fibroids in pregnancy are effectively managed with analgesics. We report a unique case of a giant uterine mass during pregnancy that caused severe oligohydramnios, was removed during cesarean section, the uterus was preserved, and the histology revealed multiple STUMPs.

Case report

A 34-year-old Caucasian, primiparous woman, at 25 gestational weeks, attended the emergency obstetric department, reporting dyspnea and fatigue. The patient's vital signs were stable, but blood testing showed a hemoglobin level of 6.8 g/dL; therefore, the patient was transfused with two bags of red blood cells. The estimated fetal weight was on the 40th centile, but the amniotic fluid was reduced (deepest pool: 1.8 cm). Moreover, a

large mass was observed at the uterine fundus. There was no clinical evidence of ruptured membranes, and an AmniSure® test (AmniSure® International LLC, Boston, MA, USA) was negative. Further evaluation with magnetic resonance imaging (MRI) revealed a large uterus with multiple masses suggestive of fibroids (the diameter of the largest was 40 cm) with partial cystic degeneration and necrosis (Figure 1A).

The fetus was assessed with ultrasound and a non-stress test twice weekly. At 30 gestational weeks, anhydramnios was detected, and the non-stress test became non-reassuring. Following a course of corticosteroids, the woman underwent elective cesarean section with a classical hysterotomy for adequate exposure. The neonate (1,350 g, Apgar score 1': 4, 5': 7) was admitted to the neonatal intensive care unit. During the cesarean section, the uterine masses (total weight: 9 kg, diameter of the largest mass: 40 cm) were removed (Figure 1B, C). Due to significant intra-operative blood loss (about 1,500 ml), the woman was transfused; however, a hysterectomy was avoided. The patient was discharged on day four, and the neonate six weeks later, in good health.

The postoperative histology identified the masses as STUMPs (Figure 1D). The tumors had diffuse moderate

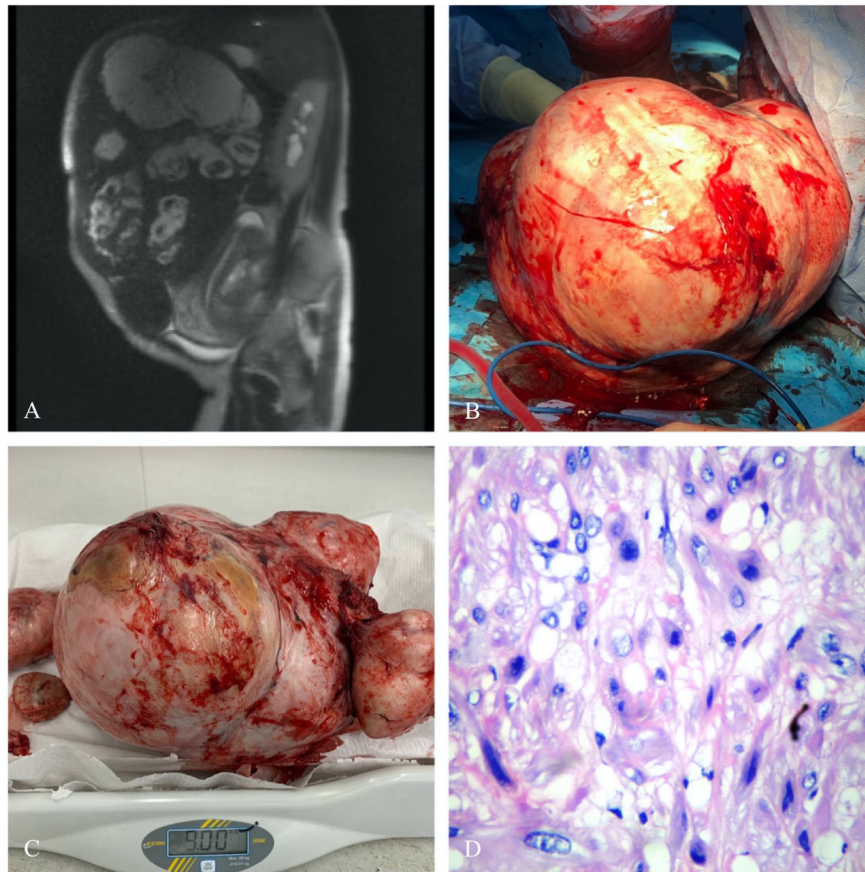


Figure 1: A) Sagittal image of pelvic magnetic resonance imaging showing the fetus and multiple uterine masses. B) perioperative and C) postoperative imaging showing uterine masses during and after the myomectomy (total weight: 9 kg). D) Histopathologic image showing smooth muscle tumors of uncertain malignant potential (STUMPs) with diffuse moderate atypia, 2-7 mitotic figures per 10/HPF, and no tumor cell necrosis (hematoxylin & eosin stain, magnification, x 400).

atypia, a mitotic index of 2-7 per 10/HPF (high-power field), and no tumor cell necrosis. A close follow-up, i.e., every six months for five years, was recommended by a gynecologic oncologist, including clinical, ultrasonographic evaluation, and MRI.

Discussion

STUMPs may cause pain, pelvic pressure, vaginal bleeding, and/or anemia⁴. The presence of benign uterine tumors, particularly those that distort the uterine cavity, has been associated with an increased risk of miscarriage, placenta previa, malpresentation, cesarean section, prematurity, and peripartum hemorrhage¹. In our case, the immense size of the uterine masses caused anemia, dyspnea, and digestive problems. In the absence of signs of ischemic placental disease, we assumed that the oligohydramnios was associated with the masses.

The finding of STUMP and/or leiomyosarcoma in pregnancy is rare. Regarding the diagnosis of uterine STUMPs, the initial evaluation is based on the patient's symptoms, while ultrasonography is the recommended initial imaging modality for benign uterine masses⁵. MRI may distinguish benign from malignant tumors; however,

there is limited evidence in differentiating the subtypes of benign uterine tumors. Moreover, a study on diffusion-weighted imaging and the apparent diffusion coefficient values to discriminate between low-risk and high-risk groups yielded 100 % sensitivity, 94 % specificity, and 94.6 % accuracy⁶. At least two of the Stanford criteria are included in the histological diagnosis of leiomyosarcoma: diffuse moderate-to-severe atypia, mitotic count of ≥ 10 mitotic figures/10 HPF, and tumor cell necrosis³, whereas STUMPs present with a combination of the features mentioned above, but without fulfilling the diagnosis of leiomyosarcoma². Furthermore, according to the WHO classification, a uterine smooth muscle tumor not unequivocally diagnosed as malignant or benign should be defined as STUMP⁷. In a study of biomarkers, it was found that the expression of the progesterone receptor, pHH3, and p16 were found to be significantly different between atypical leiomyomas and leiomyosarcomas but not between usual leiomyomas and atypical leiomyomas⁸.

There are no guidelines for managing women with suspected STUMPs, and few data are available on the management of uterine masses during pregnancy. Women with risk factors or non-responsive to medical

therapy may be offered myomectomy during pregnancy, especially in cases of intractable pain. However, due to the potential risks, surgery is usually avoided. Other techniques, such as artery embolization or MRI-guided focused ultrasound surgery, may be useful. A meta-analysis of 2,301 cesarean myomectomies found that concomitant myomectomy had a higher blood transfusion risk compared to cesarean alone⁹.

Regarding the prevention of recurrence, neither progesterone nor GnRH analogs or chemotherapeutic agents have proven effective¹⁰. Furthermore, the postoperative follow-up of patients with STUMPs is unclear; six-month reviews (clinical, ultrasound, and MRI evaluation) for five years postoperatively may be a reasonable approach for early detection of recurrence¹¹. Once the patient with myomectomy has no desire to further preserve her fertility, counseling regarding hysterectomy may be reasonable due to the possibility of recurrence.

Conclusion

We presented a case of giant STUMPs in a pregnant woman that underwent cesarean delivery and concomitant excision of the masses with uterus preservation. To date, this is the only reported case of a pregnant woman with STUMPs of such size that underwent myomectomy during cesarean section and avoided hysterectomy. In terms of the prognosis of such cases, increasing knowledge of molecular genetics or immunohistochemistry and data regarding long-term follow-up will aid in future surveillance guidelines and management. Future research should focus on finding biomarkers and histological or immune-histochemical markers to facilitate the diagnosis of tumors with malignant transformation potential.

Conflict of interest

The authors have no conflict of interest to declare.

References

1. Parazzini F, Tozzi L, Bianchi S. Pregnancy outcome and uterine fibroids. *Best Pract Res Clin Obstet Gynaecol*. 2016; 34: 74-84.
2. Gadducci A, Zannoni GF. Uterine smooth muscle tumors of unknown malignant potential: A challenging question. *Gynecol Oncol*. 2019; 154: 631-637.
3. Bell SW, Kempson RL, Hendrickson MR. Problematic uterine smooth muscle neoplasms. A clinicopathologic study of 213 cases. *Am J Surg Pathol*. 1994; 18: 535-558.
4. Tsakiridis I, Evaggelinos D, Mamopoulos A. Multiple STUMPs in a 19-year-old woman: a rare case report. *Hippokratia*. 2019; 23: 96.
5. Vilos GA, Allaire C, Laberge PY, Leyland N. The management of uterine leiomyomas. *J Obstet Gynaecol Can*. 2015; 37: 157-178.
6. Sato K, Yuasa N, Fujita M, Fukushima Y. Clinical application of diffusion-weighted imaging for preoperative differentiation between uterine leiomyoma and leiomyosarcoma. *Am J Obstet Gynecol*. 2014; 210: 368.e1- 368.e8.
7. World Health Organization. International Agency for Research on Cancer. WHO Classification of Tumours. Female Genital Tumours. 2020. Available at: <https://tumourclassification.iarc.who.int/>, date accessed: 20/8/2021.
8. Liang Y, Zhang X, Chen X, Lü W. Diagnostic value of progesterone receptor, p16, p53 and pHH3 expression in uterine atypical leiomyoma. *Int J Clin Exp Pathol*. 2015; 8: 7196-7202.
9. Pergialiotis V, Sinanidis I, Louloudis IE, Vichos T, Perrea DN, Doumouchtsis SK. Perioperative Complications of Cesarean Delivery Myomectomy: A Meta-analysis. *Obstet Gynecol*. 2017; 130: 1295-1303.
10. Ip PP, Tse KY, Tam KF. Uterine smooth muscle tumors other than the ordinary leiomyomas and leiomyosarcomas: a review of selected variants with emphasis on recent advances and unusual morphology that may cause concern for malignancy. *Adv Anat Pathol*. 2010; 17: 91-112.
11. Basaran D, Usubutun A, Salman MC, Narin MA, Boyraz G, Turkmen O, et al. The Clinicopathological Study of 21 Cases With Uterine Smooth Muscle Tumors of Uncertain Malignant Potential: Centralized Review Can Purify the Diagnosis. *Int J Gynecol Cancer*. 2018; 28: 233-240.