International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Kirubanandan S et al. Int J Reprod Contracept Obstet Gynecol. 2025 Mar;14(3):926-930
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20250532

Case Report

A case report of uterine leiomyosarcoma

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Received: 26 December 2024
Accepted: 29 January 2025

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ABSTRACT

The most prevalent benign tumour in women is uterine leiomyoma. Smooth muscle cells give rise to the uncommon malignant tumour known as uterine leiomyosarcoma. It makes up 1% of all uterine cancers. One out of every 800 women who are thought to have leiomyoma actually has a sarcoma. Uterine leiomyosarcoma is usually diagnosed by histopathological examination and surgery is the only treatment. We describe a 48-year-old woman who had heavy menstrual flow and was initially diagnosed with uterine leiomyoma. Histological analysis revealed that she actually had uterine leiomyosarcoma.

Keywords: Uterine leiomyosarcoma, Menstrual disorder, Uterine sarcoma

INTRODUCTION

Uterine sarcomas make up 3–7% of all uterine cancers and about 1% of all gynecological neoplasms in females. Carcinosarcomas, leiomyosarcomas, endometrial stromal sarcomas, and undifferentiated sarcomas were the categories into which it was divided.

The most prevalent uterine sarcoma is leiomyosarcoma which typically affects women over 40 years of age and usually present with palpable pelvic masses (54%), abnormal vaginal bleeding (56%) or pelvic pain (22%). In fact, one out of every 800 women who are thought to have leiomyoma has a sarcoma.[1,2] Uterine sarcoma is extremely uncommon, although it typically has a poor prognosis.[3]

An uncommon malignant tumor that develops from embryonic mesenchymal cells is leiomyosarcoma of the uterus (ULMS). The most frequent place for leiomyosarcoma is the uterus, where it accounts for 2% to 5% of all uterine cancers. It spreads quickly via the hematogenous and intraperitoneal routes. It is frequently identified after surgery for a supposed benign condition after myomectomy, hysterectomy, or supracervical hysterectomy. It usually presents in perimenopausal women with median age of 50 years. Because leiomyosarcoma resembles benign uterine leiomyomas, it might be difficult to diagnose before surgery. This condition's rarity restricts our understanding of prognostic factors and optimal adjuvant treatments.

CASE REPORT

48 years old Mrs. X with obstetric score P6L5, sterilized presented to OPD with complaints of heavy menstrual bleeding for 5 months having 10-15/30 days cycle, changes 3-4 clothes/day associated with passage of clots and dysmenorrhea. Patient had similar complaints in the past and previous records of ultrasonography not available and patient had been on hormonal pills for the same.

Previous menstrual history was unremarkable. She has no comorbidities. On general examination she appeared moderately built and nourished with BMI of 24.1 kg/m2 with stable vitals. Other systemic examination along with breast and thyroid examination were normal.

Per abdominal examination revealed soft to firm mass of size 16 to 18 weeks as compared to pregnant uterus and

has smooth surface. The mass was mobile from side to side, with no tenderness or any changes in the overlying skin. Sterilization scar present. On per speculum examination, cervix and vagina appears healthy and grade 1 cystocele noted. No growth/polyp/erosion seen. On Bimanual examination uterus irregularly enlarged to 20 weeks and mass felt through pouch of douglas, left lateral and anterior forniceal fullness present. There was no forniceal tenderness. Transmitted mobility of mass through cervix is felt.

Investigations

Blood examination with complete hemogram and other blood examinations were found to be normal. Chest X-ray found to be normal. On ultrasound abdomen and pelvis, uterus measures 8.3×6.3×5.3 cm, endometrial thickness measures 3.9 cm. A well-defined heterogeneously hypoechoic lesion of 12.4×10×8.8 cm showing peripheral vascularity noted arising from the anterior myometrium (as shown in Figure 1 and 2). Another small heterogeneously hypoechoic lesion measuring 1.8× 1.2 cm noted in right lateral wall suggestive of uterine fibroid.

Right ovary measures-2.6×1.6 cm, normal, left ovary measures 2.7×1.4 cm, normal. Pap smear showed unsatisfactory for evaluation and endometrial biopsy taken revealed Proliferative phase endometrium. Patient was planned for total abdominal hysterectomy with bilateral salpingo oophorectomy.

Intraoperative findings

Uterus enlarged to 20-22 weeks size. Left broad ligament fibroid of size 5×5 cm noted and found to be degenerated. Adhesions noted between Posterior surface of broad ligament fibroid and serosa of bowel and the same was released. Bilateral tubes and ovaries normal. Anterior myometrial wall fibroid of size 9×8 cm noted (as shown in Figure 3). Specimen sent for HPE examination.

Histopathological examination

On gross examination of uterus along with cervix with bilateral fallopian tubes and ovaries, cut surface of anterior myometrial fibroid measures 13×10×8 cm. External surface appears nodular and shows variegated appearance with areas of haemorrhage and necrosis and surrounding myometrium is compressed.

On microscopical examination sections made from anterior wall fibroid shows tumor cells arranged in long interlacing fascicles. Individual cells are spindle showing moderate atypia, vesicular to hyperchromatic nuclei with coarse chromatin and eosinophilic cytoplasm (Figure 4).

Increased mitosis including atypical ones are seen. Mitotic rate 20-60/10 hpf (Figure 5). Also seen are geographical necrosis and hyalinized areas with no evidence of Myometrial invasion. As per CAP protocol of surgical cancer staging it was diagnosed as pT1b (Tumor limited to uterus with greatest dimension >5 cm).

Histopathologically categorized as spindle (conventional) leiomyosarcoma. After HPE examination report, patient was called and explained regarding the condition and referred to oncologist and advised for regular follow up.

Postoperatively patient did not follow up and came after 7 months. CECT pelvis was taken and revealed large peritoneal masses/peritoneal deposits, lower abdominal wall metastatic deposit extending to subcutaneous plane with pelvic lymphadenopathy involving bilateral external iliac and obturator region largest measuring 11×10 mm in left external iliac region, moderate right pleural effusion with suspicious parietal pleural thickening/ nodularity. Patient was explained regarding the condition and was sent for chemotherapy.

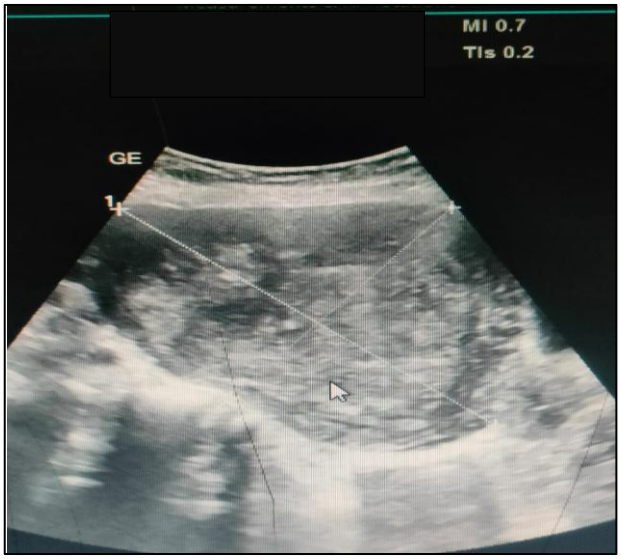

Figure 1: Large anterior myometrial fibroid.

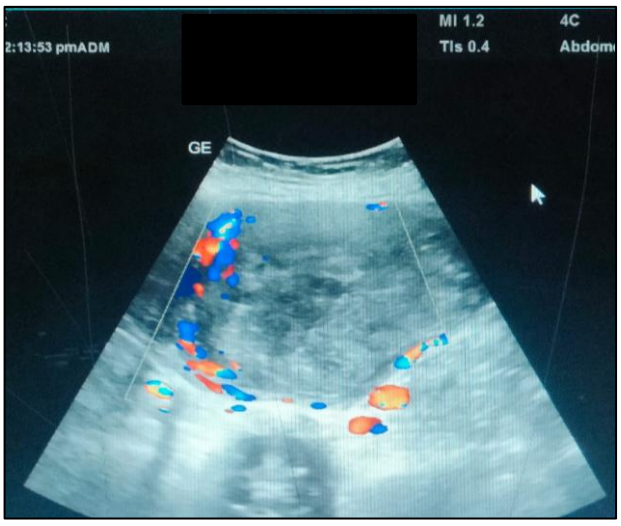

Figure 2: Image showing anterior myometrial wall fibroid with peripheral vascularity.

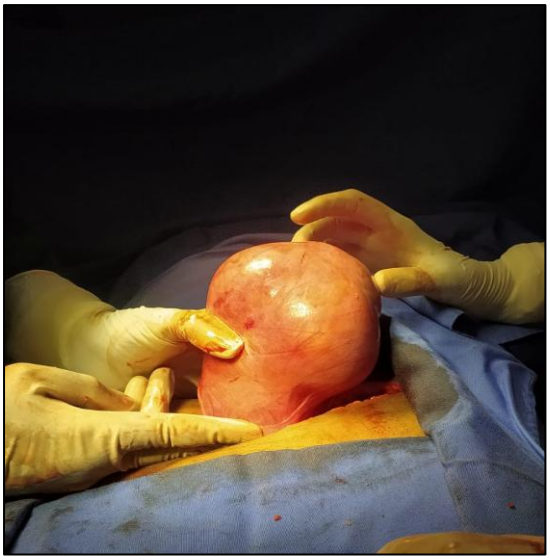

Figure 3: Enlarged uterus with anterior myometrial fibroid.

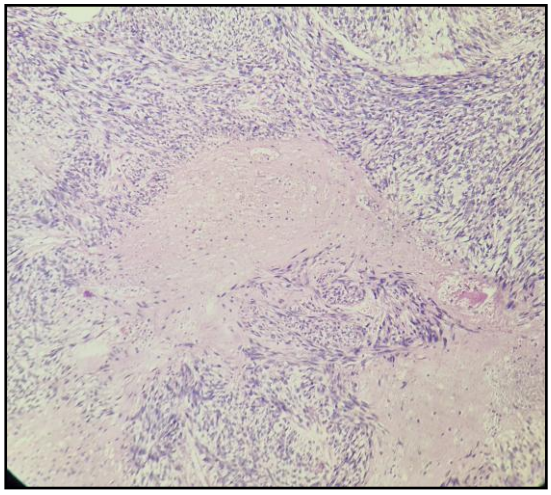

Figure 4: Microscopy showing fascicles of spindle cells with pleomorphic hyperchromatic nuclei.

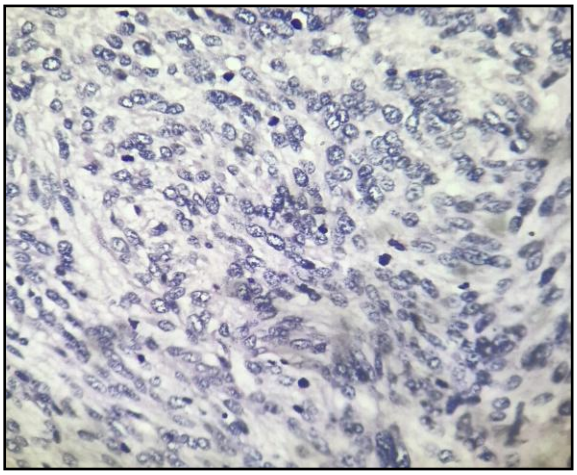

Figure 5: Microscopy showing increased mitosis. Mitotic rate of 20-60/hpf.

DISCUSSION

Uterine leiomyosarcoma is an uncommon malignancy accounting for approximately 1% of uterine cancer with an estimated annual incidence of 0.64 per 100,000 women. Although leiomyosarcoma can occur elsewhere in the pelvis, including the cervix and urinary bladder, it is more commonly found in the uterus, as seen in our case.[4] Most occur in women over 40 years of age who usually present with abnormal vaginal bleeding (56%), palpable pelvic mass (54%) and pelvic pain (22%). Signs and symptoms resemble those of the far more common leiomyoma and preoperative distinction between the two tumors may be difficult.[5]

Risk factors

Uterine sarcoma was more likely to be diagnosed in patients with uterine mass, age ≥40 years, postmenopausal status and postmenopausal bleeding, AUB, palpable mass and identification of fast-growing mass, and ultrasonography detected solitary uterine mass.[6] In women with breast cancer, long-term adjuvant tamoxifen also increases the risk of sarcomas.[7,8]

Prognosis

A favorable prognosis was independently linked to early tumor stage, age <50 years, and lack of vascular space involvement. Lymph node metastasis and histological type have prognostic implications.[9] The prognosis is poor for tumors more than 5 cm and ULMS that extending beyond the uterus and cervix.[10] In early tumor stages, mitotic count was found to be a powerful predictive factor; however, in patients with tumor stage II–IV disease, it did not act as an independent prognostic factor.[11]

Metastasis

The lung, liver, brain, kidney, and bones are the primary sites of metastasis. However, only 3.5% of uterine LMSs are secondary to the ovary.

Diagnostic criteria

Regardless of the stage at which it manifests, ULMS is an aggressive tumor that is different from other forms of endometrial cancer and carries a significant risk of recurrence and death. According to the Stanford criteria, ULMS is diagnosed when at least two of the following traits are present. 1) Areas of coagulative tumor cell necrosis, 2) moderate to severe cellular atypia, and 3) a high mitotic rate >10 figures per 10 high-power fields.[12]

Macroscopic appearance

The uterine wall is invaded by these tumors, which often grow as solitary, irregular, bulky masses with a grey-white, lobulated cut surface with hemorrhagic foci.

Histopathology

ULMS are malignancies of smooth muscle differentiation. Broad fascicles of plump spindle cells intersecting at right angles with varied degrees of hyalinization, containing copious amounts of highly eosinophilic fibrillary cytoplasm, prominent cell boundaries, and cigar-shaped nuclei with characteristic architecture of smooth muscle found in well-differentiated tumors. Microscopic analysis shows hypercellularity and extensive mitosis (>10 MF/10 HPF) together with coagulative necrosis of tumor cells.

Immunohistochemistry

Ki 67 proliferation index: 8–10%; smooth muscle markers, such as smooth muscle actin and h-caldesmon are commonly expressed by ULMS.[13] SMA (Smooth muscle actin), Desmin and vimentin shows positive.

Management

Hysterectomy and BSO (bilateral salpingo-oophorectomy) with the removal of any obvious metastatic disease should be part of the surgical staging. At initial diagnosis, around 60% of women with ULMS had disease limited only to uterus. Neoadjuvant chemotherapy is required in case of advanced staging of ULMS.

When used as first-line therapy for patients with locally advanced or metastatic ULMS, the doxorubicin plus trabectedin combination has demonstrated remarkable effectiveness in terms of survival.[14] There is no evidence to support the use of further chemotherapy or radiation therapy following total surgical removal.

The disease stage at diagnosis affects survival rates of the patients. For stage I, the five-year survival rate is 50–55%, while for stages II–IV, it is 8–12%. For all stages, the overall five-year survival rate falls between around 30% and 50%. Surgery can be used to save local recurrences. Isolated pulmonary metastases can be resectable with overall survival rates at five and ten years are 45% and 35%, respectively.

CONCLUSION

A 48 years old female who presented with abnormal uterine bleeding with leiomyoma after surgical intervention and histopathological examination found to have uterine leiomyosarcoma. It contributes to 1% of uterine cancer. So high index of suspicion is needed to diagnose and treat such patients timely. Because of their rarity, there is no screening method applicable in such patients and there is also non-availabilities of sensitive and specific tumour markers. Surgery is the primary treatment for leiomyosarcoma. Currently there is no proven benefit of using chemotherapy or radiotherapy after complete surgical removal.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kirubanandan S, Jaget N, Sekar S, Saranya S, Rajamanickam P. A case report of uterine leiomyosarcoma. Int J Reprod Contracept Obstet Gynecol 2025;14:926-30.