

# Uterine Sarcomas: 15-year Experience from a Tertiary Care Cancer Hospital in India

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## ABSTRACT

**Introduction:** Uterine sarcomas are rare neoplasms with poor prognosis comprising around 2–6% of uterine malignancies. They include leiomyosarcomas, endometrial stromal sarcomas, undifferentiated sarcomas, carcinosarcomas (previously existed, but it does not exist anymore) and few other rare variants. Leiomyosarcomas are the most common uterine sarcoma. The most common clinical presentation is postmenopausal vaginal bleeding while others include lower abdominal pain and white discharge per vaginum. This study analyzes the general patient characteristics and survival data of uterine sarcoma patients treated in our institute.

**Materials and methods:** The patients treated for uterine sarcomas in our institute from 2004 to 2019 were identified and their case records analyzed. The patient's baseline characteristics, stage distribution, recurrence patterns, survival, outcomes of laparoscopic, and open approaches in treating uterine sarcomas are all analyzed.

**Results:** Totally, 31 patients were treated during the time period out of which 6 patients were lost the follow-up. The mean age of patients at diagnosis was 46.5 years. The most common type in our center was endometrial stromal sarcoma. Eleven (35%) patients had recurrence. Those with recurrences were offered surgery when it was limited to pelvis. The adjuvant chemotherapy and radiation were given as indicated. Four patients had a history of tamoxifen intake. Eight patients (25%) had died during the follow-up which makes up to 72% of the recurred patients. The median disease-free survival was 45 months after a median follow-up of 90 months.

**Conclusion:** Although uterine sarcomas have poor prognosis, our patients had better prognosis on comparison with literature. The stage of the disease and the pathological type are the important factors determining prognosis. Laparoscopy offers better postoperative outcomes and larger studies are needed to demonstrate their oncological safety.

**Keywords:** Endometrial stromal sarcoma, Laparoscopic staging, Leiomyosarcoma, Uterine sarcoma.

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## INTRODUCTION

Uterine sarcomas are rare neoplasms with poor prognosis comprising around 2–6% of uterine malignancies.<sup>1</sup> Uterine sarcomas are both histologically and pathologically different entity from endometrial cancers. Although they are considered different, the clinical presentation is the same with postmenopausal vaginal bleeding being the most common presentation in sarcomas. The various risk factors associated with endometrial adenocarcinoma such as diabetes; also, hypertension has a role in uterine sarcomas. The history of pelvic radiation therapy and tamoxifen therapy in postmenopausal women were also considered as significant risk factors in the development of uterine sarcomas.<sup>2</sup> As with any other malignancy, the patients who are diagnosed at an early stage and have a low-grade disease fair well and the most common cause of mortality is distant metastasis to lung. This analytical study aims at the analysis of the patient characteristics, recurrence patterns, and survival data of the patients with uterine sarcomas.

## MATERIALS AND METHODS

An analysis of the case records of the uterine sarcoma patients treated in our hospital from June 2004 to June 2019 was done and the results were tabulated. All patients shortlisted were histopathologically confirmed uterine sarcomas. All patients underwent a thorough clinical examination followed by imaging and a biopsy procedure. The imaging done was a magnetic resonance imaging (MRI) of abdomen and pelvis and patients underwent Pipelle biopsy. To those whom the pathology was

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inconclusive on Pipelle biopsy, fractional curettage was done. We have also included carcinosarcomas in this analysis although they are recently not considered to be uterine sarcoma at all but high-grade epithelial endometrial cancer. All these patients underwent a comprehensive surgical staging in the form of hysterectomy with salpingo-oophorectomy and pelvic lymph node sampling. The para-aortic lymphadenectomy was performed only in patients with enlarged para-aortic nodes as per the imaging or by the intraoperative finding. The staging was done either through laparoscopic or open approach. The patients were analyzed for their baseline characteristics, staging distribution, complications among laparoscopic and open procedures, recurrence patterns, and survival. International Federation of Gynaecology and Obstetrics

**Table 1:** Patient baseline characteristics

Number of patients	31
Mean age at diagnosis (in years)	46.5 (range 34–71)
Menopausal status	
Premenopausal	2 (6.4%)
Postmenopausal	29 (93.5%)
Diabetes	18 (58%)
Hypertension	16 (51.6%)
H/O tamoxifen intake	4 (12.9%)
H/O pelvic RT	2 (6.4%)
H/O, history of; RT, radiation	

**Table 2:** Staging distribution by histology

Histology/Stage	I	II	III	IV	Total
Endometrial stromal sarcoma	13	2	2	–	17 (54.8%)
Leiomyosarcoma	–	2	1	–	3 (9.6%)
Carcinosarcoma	4	4	3	–	11 (35.4%)

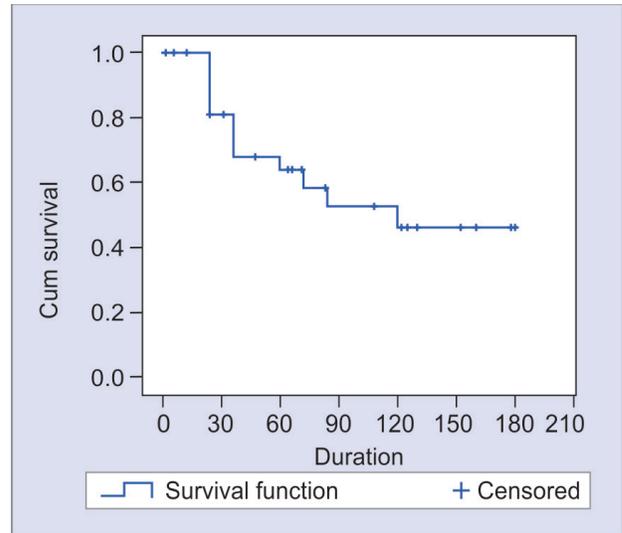
(FIGO) staging was used. Follow-up of the patients included clinical examination and annual imaging in the form of computed tomography (CT) abdomen and pelvis and further investigations done in case of specific symptoms. The survival analysis curves were made using Kaplan–Meir curves. The defaulted patients were contacted by telephonic conversation to assess outcomes.

**RESULTS**

The baseline patient characteristics were analyzed. There was a total of 31 patients diagnosed to have uterine sarcoma. The mean age at diagnosis was 46.5 years (Table 1). The histologies included endometrial stromal sarcoma, carcinosarcoma, and leiomyosarcoma. There were 17 patients (54.8%) with endometrial stromal sarcoma, 3 patients (9.6%) with leiomyosarcoma, and 11 patients (35.4%) with carcinosarcomas. A total of 93.5% of patients were of postmenopausal. Four patients (12.9%) had a history of tamoxifen intake as a part of their breast cancer treatment. Two of the patients (6.4%) had a history of pelvic radiation as a part of their cervical cancer treatment. The stage distribution (Table 2) of the patients varied based on histology. Overall, there were 17 patients (54.8%) in stage I, 6 patients (19.3%) each in stages II and III, and 2 patients (6.4%) in stage IV.

All patients underwent surgery after the metastatic workup. Seven patients underwent laparoscopic staging whereas others underwent open staging. Two patients were referred following hysterectomy for a benign diagnosis preoperatively, one with vault recurrence and one with lung metastasis—4 months after surgery. No adjuvant treatment was offered to patients who had their disease confined to uterus. An external beam radiation was given to five patients and palliative chemotherapy was given for the patients who had metastasis.

Mean follow-up period was 90 months (0–180 months). A monthly follow-up visit during the first year; once-in-2-months follow-up visits during the second year; and 3 monthly follow-up visits from the third year, and thereafter, were done. A thorough clinical examination and chest X-ray were done periodically. The CT scans were done as warranted by the symptoms the patients presented with. The median disease-free survival was 45 months



**Fig. 1:** Overall survival analysis curve

**Table 3:** Recurrence pattern

Histology/Stage	I	II	III	IV	Total
Endometrial stromal sarcoma	5	1	1	–	7 (22.5%)
Leiomyosarcoma	–	–	1	–	1 (3.2%)
Carcinosarcoma	–	1	2	–	3 (9.6%)

**Table 4:** Postoperative complications compared between laparoscopy and open procedures

Complications	Laparoscopy (n = 7)	Open (n = 24)
Wound infection	–	7 (29%)
Bladder morbidity	2 (28%)	2 (8.3%)
Intestinal obstruction	–	1 (4.1%)
Small bowel prolapse	–	1 (4.1%)
Burst abdomen	–	1 (4.1%)

(0–90 months) (Fig. 1). Eleven patients (35%) had recurrence (Table 3). Seven patients had local recurrence whereas four had lung metastasis. Eight patients had died during the follow-up which made up to 25% of all patients and 72% of recurred patients. The postoperative complications were also studied between the laparoscopic and open procedures (Table 4). The patients operated by laparoscopic approach had bladder morbidity same as those who got operated by open method. Otherwise, wound morbidity, intestinal obstruction, and bowel prolapse were observed in patients who underwent open surgery.

**DISCUSSION**

Uterine sarcomas include endometrial stromal sarcomas, leiomyosarcomas, undifferentiated sarcomas, and much rarer variants such as adenosarcoma, rhabdomyosarcoma, and PEComa.<sup>1</sup> Uterine sarcomas account for around 2–6% of uterine cancers and the worldwide incidence is about 1.7 in 100,000 women.<sup>2</sup> Carcinosarcomas were considered as uterine sarcomas previously but it is not existing anymore. They are considered and treated now as high-grade endometrial cancers. They are also referred to as malignant mixed Mullerian tumor. Uterine sarcomas are relatively

rare entities and have limited literature reviews, although there are quite a few case series and reports.

The mean age of our patients was 46.5 years which was also consistent with various literature reports.<sup>3</sup> Among the uterine sarcomas, leiomyosarcomas are the commonest with an incidence of 55% while endometrial stromal sarcomas the least common.<sup>4</sup> In our study, it is exactly the opposite with endometrial stromal sarcomas being most common (54.8%) and leiomyosarcomas being the least common. The most common presenting symptom was abnormal vaginal bleeding in postmenopausal women. This is the one which brings them early for treatment. However, most leiomyosarcomas are diagnosed postoperatively with hysterectomy being done for benign diagnosis.

Surgical management is the cornerstone of the treatment and is considered as one of the most important prognostic factors for uterine sarcoma.<sup>5</sup> The standard treatment for any uterine sarcoma would be total hysterectomy with bilateral salpingo-oophorectomy. Considering the adverse effects of early salpingo-oophorectomy, ovarian function may be preserved in stage I disease.<sup>6</sup> Lymphadenectomy is also controversial.

The recurrence rate is high in uterine leiomyosarcomas with rates up to 50–70% as per literature.<sup>7</sup> However, in our study, although the numbers are less to be considered significant, recurrences are more in endometrial stromal sarcomas (22.5%) than uterine leiomyosarcomas (3.2%).

Laparoscopic surgery for uterine sarcomas has not been studied extensively. Our study shows that the early postoperative outcomes are better with laparoscopic staging than open staging in the treatment of uterine sarcomas. There are several reports of uterine sarcomas being diagnosed in the postoperative specimen of hysterectomy done for benign lesions. In such cases the use of laparoscopic power morcellator has to be strictly avoided as it would result in intraperitoneal dissemination.<sup>8</sup> For comparison between laparoscopy and open staging, with regard to oncologic safety, larger multicenter trials are needed.

The survival patterns differ according to the stage of the disease. As expected, the survival is better in the early-stage disease than in the advanced-stage disease. The overall median survival of uterine sarcomas is around 45 months. The literature reports an overall 5-year survival of close to 70% in early-stage disease.<sup>7</sup> The survival of our patients matches with that of various literature reports in spite of difference in pathological patterns.

## CONCLUSION

Although uterine sarcomas have poor prognosis, our patients had better survival on comparison with literature. The survival is far better in our group of patients mainly because most of them are low-grade endometrial stromal sarcoma. The stage of the disease at diagnosis and pathological variety dictates the prognosis. However, the treatment does not change much with adjuvant chemotherapy, radiation, and hormonal therapy as indicated. The evaluation of the role of laparoscopy and its oncological safety needs much larger studies.

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