Case Report

Dilemma in Diagnosis Surprise at Histopathology

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A 65-year post-menopausal woman presented in the Gynaecology OPD of RD Gardi Medical College, Ujjain, with complaints of pain, a lump in her lower abdomen along with difficulty of micturition, defecation and vaginal bleeding for 4 years. She has had a known case of hypertension for 10 years and is taking treatment. Clinically diagnosed as 20 weeks size uterine fibroid, ultrasonography reported 11 to 14.5 cm fibroid, and MRI showed degenerating fibroid. The patient was managed by abdominal hysterectomy with bilateral salpingo-oophrectomy. The post-operative period was uneventful. Histopathology reported leiomyosarcoma. The patient was discharged and followed by chemotherapy.

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Introduction

Uterine fibroids are benign, monoclonal tumors of smooth muscle layers of myometrium and contain large aggregations of extracellular matrix composed of collagen, elastin, fibronectin, and proteoglycan, which develop in 20 to 50% of reproductive-age women[1]. Leiomyosarcoma is a rare malignant tumor that arises from smooth muscle cells in the uterus. There is no whorled appearance nor any capsule. The incidence of uterine leiomyosarcoma is in the range of 0.5 to 0.7 per 1000,00 per w omen[2]. Uterine leiomyosarcoma is are rare tumor mesodermal origine 2 to 6% of uterine malignancy. The commonest being leiomyosarcoma (40%), carcinosarcoma (40%), endometrial stromal sarcoma (10–15%), and endometrium adenosarcoma (0–5%). Uterine leiomyoma has the ability to mimic benign uterine fibroids, and there is no preoperative diagnostic tool that has sufficient sensibility (Figure 1) and specificity to exclude malignancy in women with uterine fibroid. The incidence of sarcoma in patients operated on for leiomyoma is 0.23%.[3] This article shows its clinical presentation, diagnostic journey of ten years as uterine fibroid, treatment approach, and outcomes as a malignant entity. The rarity of leiomyosarcoma underscores the importance of a comprehensive understanding for accurate diagnosis and management. This contributes to the existing medical knowledge by shedding light on the intricacies of leiomyosarcoma and its multidisciplinary management.

Case Report

A post-menopausal, moderately built, 65-year-old woman with Para IV, menopause attained 10 years back presented with persistent abdominal pain, difficulty in micturition, defecation and vaginal bleeding for four years. She was hypertensive for 10 years, taking treatment. She underwent a laparotomy three years back with closure without any result. But, she did not have any documentation of surgery. On examination, patient was clinically stable. Abdominal examination showed mass corresponding to 20 weeks of gravid uterus, hard, non-mobile, non-tender having a smooth surface, upper and lateral margins well defined, and lower margins not reached. Per speculum examination,

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foul smelling discharge, Bim annual examination cervix hard, mass extended up to umbilicus. Pap smear report showed inflammatory cells. Endometrial biopsy reported predominantly blood clots with neutrophilic aggregates and fibrinous material. Total abdominal hysterectomy with bilateral salpingo-oophrectomy done no lymph nodes palpable, specimen sent for histopathology. The post-operative period was uneventful and the patient was discharged. Followed by chemotherapy.

Diagnosis

Ultrasonography showed a well-defined oval heterogeneous, hypoechoic lesion with well-defined internal cystic areas measuring approximately 11.5×14.5×10.8 cm, almost completely occupying the uterus with no significant internal vascularity. Ovaries were not separately visualized. Endometrial thickness 3 mm. Magnetic resonance imaging showed a uterus bulky and large well, circumscribed intramural fibroid most completely occupying the fundus and body of the uterus measuring11.4×15.6×14.7 cm and appears hypointense on T1W /T2W images with central hyperintense signal in T2W images suggestive of degeneration (Figure 2). Lesion is causing mass pressure on the surrounding structure displacing the bowel loops superior and pushing bladder antero-inferior.

Pathological Aspect

Leiomyosarcoma (LMS) a rare and aggressive malignant tumor that originates from smooth muscle cells. In most cases, the diagnosis of leiomyosarcoma is made by histopathological examination only after hysterectomy or myomectomy specimen. Leiomyosarcoma presents diagnostic and therapeutic challenges due to its diverse clinical manifestations and complex molecular profile, but confirmed by histopathological examination as leiomyosarcoma (Figure 3).

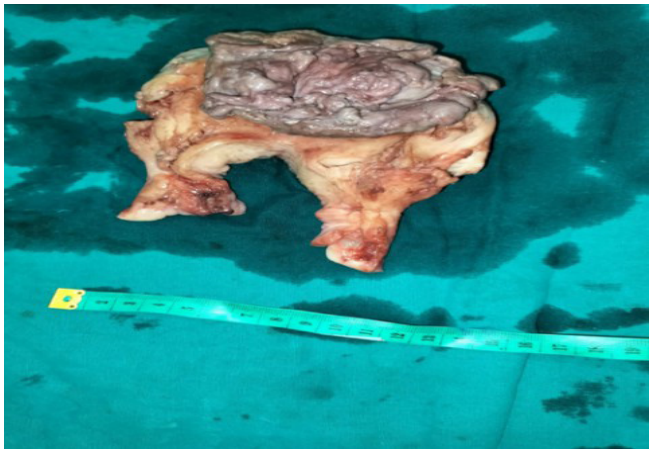

Figure 2: Degenerative fleshy mass arising from posterior wall of uterus

Histopathology

Grossly specimen had a pushing border and the capsule was present.

Myometrium with large intramural fibroid non-invasive border with an extensive area of coagulative necrosis along with an area of ischemic necrosis. The adjacent area shows foci of foamy histiocytic collection at a place with an abrupt transition of fascicle of visible spindle cells. Only a small focal area shows mild atypia of

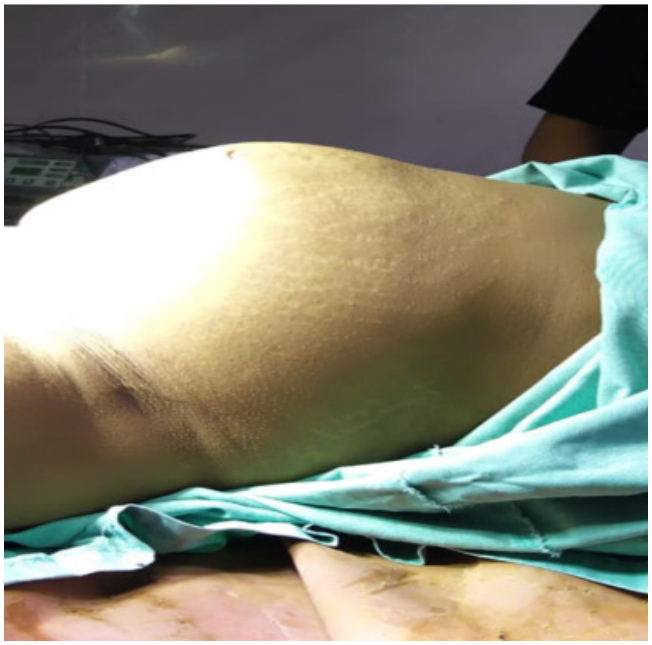

Figure 1: Size of uterus preoperatively

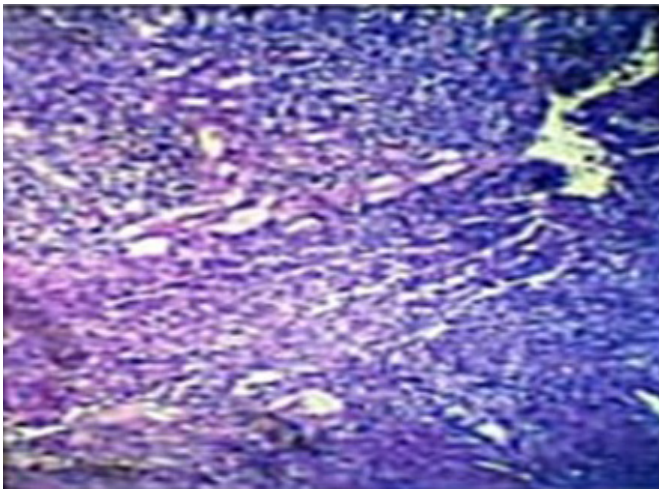

Figure 3: Post-operative pathological immunohistochemistry analysis of the specimen revealed uterine leiomyosarcoma (K i67 ~25 %).

Table 1: Leiomyosarcoma of deep soft tissue Stanford medical pathology criteria

Leiomyoma *(requires all below)*	Smooth Muscle Tumor of Uncertain Malignant Potential *(used for any of below)*	Leiomyosarcoma *(requires any one of below)*
Cytologically bland	Bland but 1-4 mitotic figures/50 High-Power Field	Cytologic pleomorphism or atypia
<1 mitotic figure/50 High-Power Field	Multiple recurrences but lacking other atypical features	>4 mitotic figures/50 High-Power Field
No tumor cell necrosis		Coagulative tumor cell necrosis

these spindle cells with six mitotic figures/10 hp. Smooth muscle tumor of uncertain malignant potential in the body of the uterus with cervical leiomyoma.

The Stanford criteria for the histologic diagnosis of malignant SMT (leiomyosarcoma) reported by Bell *et al*. include at least two of the following criteria: diffuse moderate-to-severe atypia, a mitotic count of at least 10 mitotic figures (MF)/10 high power fields (HPFs) and tumor cell necrosis (Table 1).

According to Barek and Novak's Gynecology South Asian Edition

- 5 MF/10 HPF- Benign tumor
- 5–10 MF/10HPF- Smooth muscle tumors of uncertain malignant potential.
- 10 MF/10HPF- Frankly malignant.

Discussion

Leiomyosarcoma, a malignant soft tissue tumor arising from smooth muscle cells, is a rare and challenging clinical entity. The rarity of leiomyosarcoma underscores the importance of a comprehensive understanding for accurate diagnosis and management. According to the FIGO classification, this leiomyosarcoma comes under IB. While leiomyosarcoma represents a small portion of all soft tissue sarcomas, its clinical complexity, varied presentation, and challenging management make it a topic of significant medical interest. These tumors are most commonly seen in women above 40 years of age, with a median age of 60 years. The clinical features are similar to that of leiomyoma and include abnormal vaginal bleeding (56%), palpable pelvic mass (54%) and pelvic pain (22%). Less frequently, they can present with hemoperitoneum due to tumor rupture or other symptoms from extrauterine metastases.[4] Uterine sarcoma is not diagnosed by Preoperative endometrial sampling because sarcomas originate in the deep myometrial layer of the uterus.[5]

According to Barek and Novak's Gynecology South Asian Edition, endometrial biopsy, although not as useful, may establish the diagnosis in as many as one- third of cases if the lesion is submucosal. The preoperative distinction between the two tumors may be difficult. Immunohistochemical expression of Ki67, p53, and p16 is significantly higher in leiomyosarcomas and undifferentiated endometrial sarcomas than in endometrial stromal sarcomas. Understanding leiomyosarcoma is paramount for healthcare professionals and researchers alike, as its rarity demands a heightened level of vigilance for early detection and a multidisciplinary approach to the incidence of recurrence, but they do not aid in improving overall survival.[6] Metastasis most commonly occurs in the peritoneal cavity, lungs, liver, kidney, and bones. The 5-year survival rate is 50 to 55% for patients with early uterine sarcoma and 8 to 12% for advanced cases.[7] The surgical approach depends on the pathological type and tumor stage and mainly consists of a total hysterectomy with bilateral salpingo-oophrectomy.[s] The National Comprehensive Cancer Network (NCCN) clinical practice guidelines for uterine sarcomas recommend hysterectomy as the initial treatment of choice for all medically operable patients with disease limited to the uterus.[9] However, in pts with suspected extrauterine spread, additional surgical resection of metastatic disease or chemotherapy is recommended after radiological staging.

The above case was diagnosed as a big intramural uterine fibroid ten years back. Surgery with positive results in tertiary centers is a specialty of this case. The reason is that she underwent laparotomy without success three years back in a remote area. Preoperative examination, either EB ultrasonography MRI-PET is not capable of differentiating benign from malignant smooth muscle mass. Concluding that all huge fibroids should be justice for pre-operative marking for malignancy.

Conclusion

Long 4 years, which is not usual in any malignancy- due to her rural background and negligence patient took 4 years to come to hospital, due to which tumors size increased.

Because it was on mostly involved myometrium only, and must not involved endometrial cavity, so that report of endometrial biopsy was not in favour of leiomyosarcoma.

Yes, this is true irregular vascularity with low impedance flow on USG was more common but in our case due to some USG machine Technical issue color doppler does not show internal vascularity.

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