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Uterine Sarcoma an Elusive Enigma- Case Series

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Introduction

Uterine sarcomas previously called as mixed mullerian tumours of uterus, are rare highly malignant tumours arising from smooth muscles and connective tissue elements and represent about 1% of all malignant gynaecologic tumours and about 3–7% of all uterine malignancies. [1] The etiology remains unclear, however, chromosomal translocations result in different histological subtypes, so, as a result, each tumour is differently malignant and has a different response to chemotherapy. According to new WHO 2020 classification uterine sarcomas are classified as uterine leiomyosarcoma (LMS)[25%–60%], endometrial stromal sarcoma (ESS)[10%–20%]subdivided into high grade(HG-ESS) and low grade(LG-ESS), high-grade undifferentiated sarcoma (HG-US)[3%–5%] and uterine adenosarcoma (5%–9%) [2,3] .Uterine sarcomas have few identified risk factors which includes chronic estrogen exposure ,tamoxifen use ,prior pelvic irradiation and genetic cause as seen in women with retinoblastoma. It occurs most commonly in postmenopausal women in sixth decade, but can be found in younger women in second decade of life. Abnormal uterine bleeding is the most frequent symptom followed by palpable pelvic mass and /or pelvic pain . The symptoms are nonspecific and overlap with that of myoma. Neither preoperative imaging i.e., ultrasound, CT scan, MRI can differentiate between benign and malignant smooth muscle tumours .They are usually mistaken as Leiomyoma, Adenomyosis or intrauterine polyps and majority of time they are diagnosed postoperatively on histopathology. The prognosis of uterine sarcoma remains poor, five-year survival for stage I disease is 50–70% and for higher stages 0–20%. The present series evaluated different types of uterine sarcoma seen in Indian scenario.

Case Report 1

A 29-year-old P2L2A1 presented to the Gynaecology outpatient department with complains of prolonged heavy menstrual bleeding associated with dysmenorrhea for 2 years. The patient denied any weight loss, urinary or bowel symptoms. On examination, pallor present and abdominal examination revealed a 22-week size mass. Pelvic examination demonstrated a uterine mass of 22 weeks size, firm in consistency, smooth surface, with restricted mobility. and a fleshy mass about 6x4 cm, irregular surface, causing cervical effacement and occupying upper half of vagina. Haemoglobin was 6g/dl for which she received 2 units of blood transfusion. On Ultrasound examination the uterus was 14.8x8 cm with diffused increased thickening of the junctional zone with complete loss of myometrial-endometrial interface. A non-homogenous collection was seen in the endometrial cavity with a lobulated isoechoic mass in the lower uterine segment and cervix measuring 7.3x5.4 cm with increased vascularity, suggestive of diffuse adenomyosis with an endocervical mass and haematometra. MRI pelvis also showed bulky ,globular uterus 14cmx 10cmx7.6 with diffuse thickening of junctional zone on the right side with a polypoidal soft tissue in the endocervical canal causing its distention and same bulging into the upper part of vagina, measuring 12.2 cm x5cmx3.4cm.Both ovaries were normal in size and appearance with no evidence of lymphadenopathy .MRI was suggestive of uterine adenomyosis predominantly the right lateral wall with a haemorrhagic polyp in the endocervical canal with a mildly dilated ureter due to extrinsic compression. Histopathological examination of biopsy from polyp revealed features of polypoidal tissue with ulcerated surface covered with fibrinous exudate, Mitosis <1/10HPF with no necrosis

or significant atypia giving an impression of? stromal tumour, endometrial tumour? epitheloid leiomyoma.

Intraoperatively uterus was 22 weeks size, b/l tubes were oedematous and b/l ovaries were healthy. Dense fibrous tissue present on the right side of the uterus invading Right ureter and right uterine artery and bowel on the posterior side of the uterus. Uterus was bisected after b/l uterine A ligation as cervix was about 6cm broad, so mass was pulled up from upper vagina, in order to protect the ureter while taking vaginal angles. A 14x 8 cm leiomyoma like, soft and doughy with yellowish hue mass, was present in form of bunch of grapes adherent to right uterine wall anteriorly and extending into the vagina assuming an hourglass appearance, impacted in vaginal canal (Figure 1). Intraoperative frozen section revealed Leiomyomatous polyp and as she was 29 years old hence decision of not doing oophorectomy was taken and total abdominal hysterectomy with b/l salpingectomy was performed.

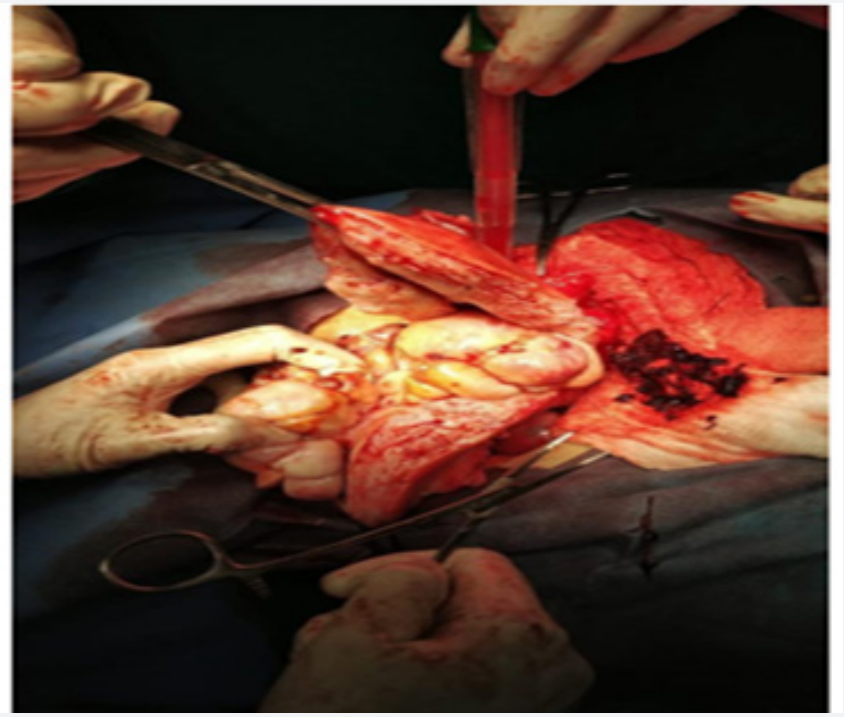

Figure 1

Histological examination of the sample revealed a cut specimen of uterus and cervix measuring 16x 11x 10 cm. A multilobulated polypoidal structure was seen filling the endometrial cavity measuring 13x8x4,5 cm. Microscopy showed tumour cells in spindle shape with scant cytoplasm. Immunostaining was positive for Vimention, Bcl2, CD10 and negative for SMA, Ki 67 proliferative index was 2%. Histology and immunostaining favoured low grade endometrial stromal sarcoma. So, in consultation with the multidisciplinary tumour board meeting, bilateral oophorectomy was performed later.

Case Report 2

A 62-year-old postmenopausal female presented with a pelvic mass without abnormal vaginal bleeding. On examination patient was moderately built and Vitals were stable. On per abdominal examination a 22–24-week size firm mass was palpable with smooth surface, mobile from side to side with no tenderness. On per vaginal examination, the cervix mid position, a firm mass measuring 18x20 cm with restricted mobility. On CT scan there was a large solid cystic mass with internal septations and few calcifications measuring 115x140x115mm. The mass was abutting the fundus of the uterus, compressing bilateral ureters causing mild bilateral hydroureteronephrosis with no retroperitoneal lymphadenopathy probable diagnosis of Leiomyoma was made. Intraoperatively uterus was enlarged about 22 weeks size with multiple fibroids and increased vascularity on surface with bilateral atrophic ovaries. Total abdominal hysterectomy with bilateral salpingo oophorectomy was planned (Figure 2).

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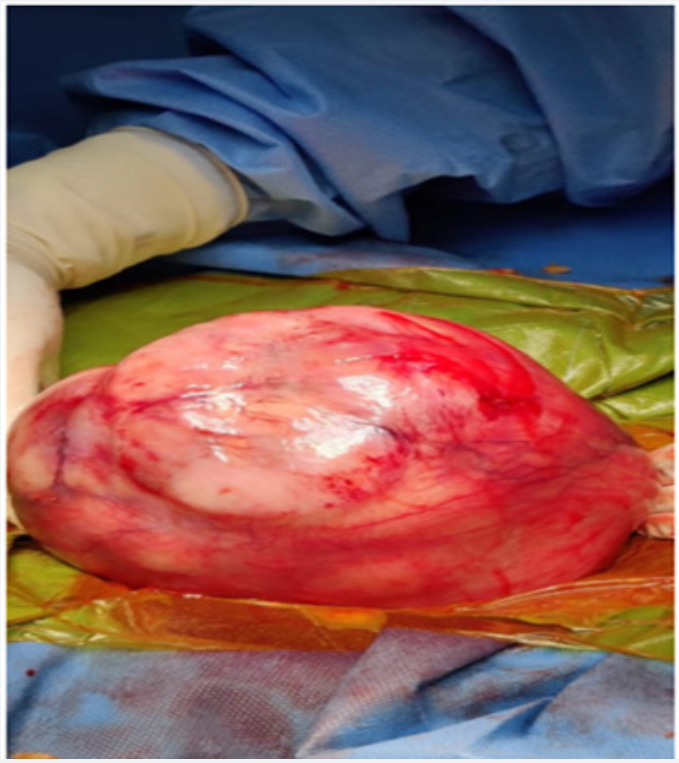

Figure 2

Uterus measuring 12x10x6 cm firm in consistency, on cut section, serosa was free of tumour, endometrial cavity was obliterated with grey, white mass. Histologically, tumour cells display pleomorphism with hyperchromatic nuclei with moderate cytoplasm, mitosis including atypical forms along with foci of necrosis also observed. Tumour cells were positive for Vimentin, Desmin and focally for SMA and negative for CD10, ER, PR, CD117, MyoD1, Ki 67proliferation index was 40 – 50 % making diagnosis of Leiomyosarcoma. Cervix, Right ovary has a simple serous cyst and b/l fallopian tubes, and left ovary were unremarkable. Patient was advised to follow up in medical oncology.

Case Report 3

A 28-year-old P2L2 reported to Gynaecology OPD with a histopathological report after Transabdominal hysterectomy with right salpingo oophorectomy for abnormal uterine bleeding –Leiomyoma at another hospital. HPE showed that dissection of the enlarged uterus revealed endometrial cavity filled with blood clot and 4 well circumscribed masses were identified ranging from 3x2x1 cm to 6x5 cm, cut surface were homogenous ,soft tan to brown in colour. Cut surface of the right ovary showed a single cyst filled brownish material . Histopathology showed nodules and sheets of epitheloid to round cells having round to oval vesicular nuclei, dispersed chromatin, few showing prominent nucleoli and scant eosinophilic cytoplasm with infiltrating pattern and >10/10/hpf mitosis making diagnosis of high grade endometrial stromal sarcoma. Section from the right ovary shows corpus luteal cyst and was free of tumour. On examination women was stable and no systemic signs. Per vaginal examination vault was healthy, no mass or growth was felt and CBC, LFT, KFT were all within normal limits and Ca125 was8.40iu/ml, sBhcg was<1.20mu/l.

MRI demonstrated thickened vault, with no specific deposits and left ovary was seen. Slides were reviewed showing tumour cells arranged in sheets and large islands surrounded by fibrovascular stroma. Individual tumour cells have a small monomorphic nucleus with dense scant to moderate amount of cytoplasm. Immunohistochemistry studies showed tumour cells positive for Dog 1, ER, PR, focally CD10 and negative for CK, Cyclin D1, SMA, Ki67, Proliferation index was very high (approx60%) confirming the diagnosis of high grade endometrial stromal sarcoma. Liquid based cytology of the vaginal vault was negative for malignancy. Following the advice of the multidisciplinary tumour board, decision for left salpingoophorectmy was taken. Intraoperatively, no growth seen at the vault No metastatic deposits and enlarged lymph nodes seen. Histopathology of left ovary and tube showed normal ovarian tissue and corpus luteum.

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Discussion

Uterine sarcoma is an enigma as neither curettage nor preoperative imaging are unable to clinch its diagnosis. MRI however can be used as preoperative diagnostic tool with some accuracy. The definitive diagnostic modality is histopathology coupled with Immunohistochemistry of hysterectomy specimen. Imaging may mislead to Adenomyosis and uterine Leiomyoma. In the above cases initial diagnosis was Leiomyoma and Adenomyosis and none of the imaging techniques were able to diagnose it preoperatively. Risk of occurrence of unexpected sarcoma during hysterectomy or myomectomy for presumed benign fibroid, according to the FDA, is 1 in 1100 women [4]. Around 90 % present with abnormal uterine bleeding,70 % with enlarged uterus and pelvic pain,30-40% with metastases outside the uterus and in 25% it can be an incidental finding [5]. Tumour often affects postmenopausal women but, in the literature, there is increasing evidence of tumour in young age group as seen in present series [6]. Diagnosing different types of sarcoma is also difficult. LMS is most common and appears as single large masses with soft, fish-like cut surfaces, LG-ESS is less malignant, mostly associated with endometriosis and tumour can be located in the endometrium, myometrium and HG-ESS often present as intrauterine polyp-like bulges or myometrial nodules [7]. MRI features can overlap with that of degeneration in myoma, however recently there have been reports of differentiation by DWI and apparent diffusion coefficient value. Newer calculations like ADC and PRESS scoring can be valuable in raising the early suspicion [8] ADC values<1.23x103 mm2/s are more likely to have malignant diagnosis [8,9]. Primary treatment is surgery including hysterectomy with bilateral salpingo oophorectomy and in advance stage complete cytoreduction. For endometrial stromal sarcoma, nodal metastases, negative CD10, Lack of ER, PR receptors are independent prognostic factor. For early-stage hormonal treatment in the form of medroxyprogesterone acetate, GnRh analogues and aromatase inhibitors is suggested. For advanced disease hormonal treatment and/or radiotherapy can be used while for recurrent disease surgery with radiotherapy, hormone therapy, or chemotherapy can be used. Leiomyosarcoma have a propensity for hematogenous spread and Lymph nodes spread is rare. Lung metastases are particularly common, and more than half of patients will have distant spread when diagnosed with recurrent disease. In contrast, ESS and adenosarcoma have an indolent growth pattern with long disease-free intervals. Surgical staging is the only way by which we can determined prognosis regarding recurrence and survival in LG-ESS. All these tumours invade, to some degree, by direct extension and due to a high risk of recurrence even LG- ESS is managed with adjuvant chemotherapy, radiotherapy, or hormonal therapy to reduce the local recurrence.

Conclusion

Uterine sarcoma are rare to encounter and difficult to diagnose fibroids. Surgery diagnosis is not clinched based on clinical features, imaging, frozen section and confirmation made mostly on histopathology, A high index of suspicion should be there for pre-operative diagnosis of ESS particularly in those women who presents with rapidly enlarging uterus > 20-week size and irregular bleeding leading to severe anemia and ultrasound findings are suggestive of uterine fibroids. Surgery i.e., Total abdominal hysterectomy with bilateral salpingo oophorectomy is the main line of management. Key for increased survival is high index of suspicion and the use of diagnostic modalities in women with uterine size> 20weeks and atypical presentation and multidisciplinary approach involving medical oncologist, radiologist, Gyneoncologist.

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