Abstract:
Uterine sarcomas are rare and aggressive cancers constituting 3-6 of all uterine malignancies. Uterine leiomyosarcomas are rare smooth muscle tumors accounting for 30 of all uterine sarcomas. Non-puerperal inversion of uterus is an uncommon presentation of uterine leiomyosarcomas. We report this case of uterine leiomyosarcoma with inversion in a 50 year old woman due to its rarity of clinical presentation. 50 year old perimenopausal woman, para 4 live 4, presented in May 2010 with complaints of mass descending per vaginum for 3 months associated with abnormal uterine bleeding and lower abdominal pain of 6 months duration. She was thin-built and anemic. Examination revealed no mass palpable abdominally. Local examination showed a large irregular proliferative mass at the introitus covered with necrotic slough. Metastatic work up normal with no evidence of liver metastasis on ultrasound and chest Xray was normal. A preliminary biopsy from the mass showed it to be high grade uterine leiomyosarcoma. Surgery planned after anemia correction. Under general anesthesia, uterine inversion confirmed through midline abdominal incision. Excision of the mass from the fundus done vaginally followed by correction of uterine inversion and vaginal hysterectomy with bilateral salpingo-oophorectomy done. Histology proved to be HIGH GRADE UTERINE LEIOMYOSARCOMA STAGE 1B. Post operative pelvic radiotherapy given. Recovery was good. Out of 28 cases of uterine sarcomas reported in our institute from 2006-2010, we had 32.1 (9 cases) of leiomyosarcoma out of which only this case presented with inversion. Patient on regular follow up with disease-free survival of 19 months. Current literature describes only 17 cases of inversion with uterine sarcomas reported so far. Although uterine leiomyosarcomas are tumors of high metastatic potential, our patient had a good clinical outcome with
INTRODUCTION
Uterine sarcomas are rare and aggressive forms of uterine malignancies constituting 3-6% of all uterine neoplasms. They are primary neoplasms of uterine mesenchymal tissues. The Gynecologic Oncology Group (GOG) classified uterine sarcomas into 4 histological types:

1. Uterine leiomyosarcomas
2. Malignant mixed mullerian tumors or carcinomas
3. Endometrial stromal sarcomas
4. Others. Uterine leiomyosarcomas are rare smooth muscle tumours accounting for 30% of all uterine sarcomas. They occur primarily in women of 40 to 60 years of age about a decade earlier than the other types of sarcomas uterus. Non-puerperal inversion of uterus is an uncommon presentation of uterine leiomyosarcomas.

CASE REPORT
We report this case of uterine leiomyosarcoma with inversion in a 50 year old perimenopausal woman due to its rarity of clinical presentation and to evaluate the clinical features, stage of the disease, histopathological features and response to treatment of this interesting condition. 50 year old perimenopausal, para 4 live 4 sterilised woman, presented to us in May 2010 with complaints of mass descending per vaginum for 3 months associated with dull aching lower abdominal pain of 6 months duration and foul smelling vaginal discharge of 3 months. She gave history of menorrhagia of 6 months duration followed by amenorrhoea for the past 3 months. She denied any history of voiding difficulty. Her vital signs were within normal limits. She was thin-built (BMI 20.3 kg/m²) and anaemic. Examination revealed no mass palpable abdominally. Local examination showed a large irregular proliferative mass 10 by 5 cm at the introitus covered with necrotic slough with the upper vagina everted along with the mass. Cervix could not be made out. There was no active bleeding from the mass. Bimanual examination could not be performed due to the irreducibility of the mass. Her hemoglobin was 9 gm%. Other blood investigations were normal. Metastatic work up done. Uterus was not imaged clearly and there was no evidence of liver metastasis on ultrasound and chest Xray was normal. A preliminary biopsy from the mass showed it to be high grade uterine leiomyosarcoma. Surgery planned after anaemia correction under prophylactic antibiotic coverage. Under general anaesthesia, uterine inversion confirmed through midline abdominal incision. Excision of the mass from the fundus done vaginally followed by correction of uterine inversion and vaginal hysterectomy with bilateral salpingo-oophorectomy done on 10th June 2010. Grossly, uterus measures 8*6*4 cm with the fundal areas showing irregular ragged appearance. Cervix was normal. Tumor measuring 10*8*6 cm with irregular external surface. On cut section, the mass was fleshy, grey-white in colour with focal areas of necrosis. Both ovaries and tubes were normal. Microscopically section from the tumour showed malignant spindle cells arranged in fascicles. The cells showed high grade pleomorphic nuclei with 8 to 10 mitoses per high power field. There were areas of necrosis seen. The myometrium was infiltrated with tumour cells with similar morphology. The diagnosis of HIGH GRADE UTERINE LEIOMYOSARCOMA STAGE 1B was made. Post operative period was uneventful. Patient was...
given post op pelvic radiotherapy. Recovery was good. She is on regular follow up with a disease-free survival of 19 months with no evidence of disease recurrence or metastasis.

RESULTS: Out of 28 cases of uterine sarcomas reported in our institute from 2006-2010, uterine leiomyosarcomas were the most common histological type (9 out of 28 cases) followed by mixed malignant mullerian tumors 28.6% (8 out of 28), endometrial stromal sarcomas were 17.9% (5 out of 28) and others 21.4% (including 2 cases of rhabdomyosarcomas, 3 undifferentiated sarcomas & 1 case of adenosarcoma). Out of all the 9 cases of leiomyosarcomas we had only one case associated with non-puerperal uterine inversion managed successfully by surgery and post-operative radiotherapy with a disease free survival of 19 months.

DISCUSSION
Uterine leiomyosarcomas are rare smooth muscle tumours accounting for 1.5% of all uterine sarcomas. They are neoplasms of high metastatic potential. The older literature indicates that leiomyosarcoma is the most common type of uterine sarcomas however the data from GOG (Gynecologic Oncology Group) shows that it is 2nd most common next to mixed mesodermal sarcomas. The mean age of patients with leiomyosarcoma is mid 50’s (40-60 years) about a decade earlier than the other types of sarcoma uterus. The age adjusted incidence of leiomyosarcoma for black and white women respectively are 1.5 and 0.9 per 100,000. Most of the uterine inversions are puerperal and non-puerperal uterine inversions are extremely uncommon events accounting for 1/6th of all cases of inversions. It usually results from a mass or tumor implanted on the fundus of the uterus. About 150 cases of non-puerperal uterine inversions were published in literature between 1882-2006. Most of them were due to benign myomas and rarely by tumors like leiomyosarcoma like in our case. All cases of non-puerperal inversion are usually chronic but about 8.6% are sudden in onset. Current literature indicates that only 17 cases of uterine inversion associated with uterine sarcomas have been reported since 1887. The most frequent presenting symptoms are abnormal uterine bleeding and pelvic or abdominal pain. Our patient had similar complaints. The amount of bleeding ranges from spotting to menorrhagia sometimes associated with foul smelling vaginal discharge. Less common symptoms
includes weight loss, weakness, fever. Rarely the tumor may prolapsed through the cervical os and into the vagina or even outside the introitus presenting as uterine inversion as in our rare case report. Due to the rarity of this tumor data regarding parity, onset of menarche, age of menopause as risk factors are inconclusive. However a history of pelvic irradiation is noted in 5 – 10% of cases. Uterine leiomyosarcomas usually arise ‘de novo’ from uterine smooth muscles or rarely from pre-existing benign leiomyomas. Uterine inversion is suspected when tumor is palpable in vagina or at the introitus but uterine fundus is not palpable abdominally. Leiomyosarcomas are rarely diagnosed before surgery as pre-op uterine curettings are diagnostic only in 10-20% of submucosal tumors. Most cases are diagnosed after hysterectomy at the time of histological review of surgical specimen. Restaging is not recommended because unlike other types of uterine sarcomas, leiomyosarcomas rarely spread to pelvic and para-aortic lymph nodes. Hence performing lymphadenectomy is not associated with improved survival rates. Extensive local growth is a hallmark of leiomyosarcomas and spreads by local, hematogenous to intra abdominal viscera, lungs and liver parenchyma and rarely lymphatic spread to involve para-aortic nodes. Grossly the tumor appears as large, soft fleshy consistency, grey-yellow to pink with foci of hemorrhage and necrosis. Microscopically, most uterine leiomyosarcomas are overtly malignant with hypercellularity, coagulative tumor cell necrosis, abundant mitoses (>10 mitoses per 10 high power fields), atypical mitoses, cytological atypia. Our patient had all the features with 8-10 mitoses/hpf indicating the high grade of the tumor stage 1B due to myometrial invasion. Imaging studies like CT or MRI do not reliably distinguish a sarcoma from a benign leiomyoma. However inversion can be seen in MRI as U-shaped uterine cavity and thickened inverted uterine fundus on sagittal image. The only treatment of proven benefit for malignant leiomyosarcomas is surgery (total abdominal hysterectomy with bilateral salpingo-oophorectomy). If associated with inversion a combined abdomino-vaginal approach is preferred as in our case. Adjuvant pelvic radiation can help to decrease the incidence of local recurrence but studies have shown to have no impact in improving survival rates. There is no change in the survival rates after chemotherapy as well. The most active drugs include doxorubicin, ifosfamide, docetaxel and gemcitabine are under phase ii and phase iii trials. Though leiomyosarcomas of uterus express oestrogen & progestogen receptors, hormonal therapy has not been extensively evaluated. A reported c-kit immuno-reactivity in 0 to 83% cases of uterine leiomyosarcomas may help future researches in molecular biology of this rare entity. Prognosis of patients with uterine leiomyosarcomas may help future researches in molecular biology of this rare entity. Prognosis of patients with uterine leiomyosarcomas is poor. The reported 5 year survival rate varies from 30% to 48%. Although associated with inversion and high grade disease, our patient had a better outcome with a disease free survival of 19 months.

CONCLUSION:

Current literature describes only 17 cases of inversion with uterine sarcomas reported so far. Although uterine leiomyosarcomas are tumours of high metastatic potential, our patient had a good clinical outcome with no distant spread or recurrence.
To conclude, non-puerperal uterine inversion may be due to uterine sarcomas which are aggressive neoplasms and hence pre-operative biopsies would aid in planning proper treatment as in our patient.

REFERENCES:


