Endometrial Stromal Sarcoma: A Rare Entity
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ABSTRACT
Endometrial Stromal Sarcoma (ESS) is a hormone sensitive tumor. It is a rare gynecological tumor and is considered to occur more often in pre-menopausal women. A proper pre-operative diagnosis is difficult and confirmed in most cases after hysterectomy for a presumed benign disease. Endometrial sampling, ultrasound, and magnetic resonance imaging can provide diagnostic clues. For early disease complete surgical cure is possible, however, adjuvant therapy is available for recurrence. This case of Low Grade Endometrial Stromal Sarcoma (LGESS) in a 21 years old woman was presented as irregular vaginal bleeding. Clinical diagnosis of fibroid was made but analysis of endometrium showed ESS confirmed on hysterectomy specimen. One should consider it in any case with rapid fibroid enlargement.

Key Words: Gynecological tumors. Endometrial stromal sarcoma. Uterine malignancy. Hysterectomy. Low grade.

INTRODUCTION
Sarcomas of the uterus are uncommon, and may arise from connective tissue, smooth muscle or the endometrial stroma. Uterine sarcoma is a rare form of malignancy, occurring in 2 - 5% of all patients presenting with uterine malignancy with an incidence of 1 - 2 /100000 women in general population. Endometrial Stromal Sarcomas (ESSs) are very rare malignant tumors that make up approximately 10% of all uterine sarcoma but only 0.2% of all uterine malignancies.1 It has good prognosis despite late recurrence.

CASE REPORT
A young 21 years old female presented with irregular vaginal bleeding for 3 months. She was para 1; vaginally delivered 3 years back. She had menarche at the age of 13 years with regular menstrual cycles up till 3 months back, without inter-menstrual or post-coital bleeding. She never practiced contraception. Now for 2 - 3 months menstrual pattern had become irregular. She took treatment from local doctor but was not relieved and was referred to a Teaching hospital.

Clinical examination was unremarkable. Her pelvic ultrasound showed an abnormal mass of 3 - 4 cm in anterior uterine wall and a diagnosis of fibroid uterus was made. Doppler showed increased vascularity. Her hematological picture revealed Hb of 9 gm% and O-positive blood group; biochemistry regarding liver and renal function were normal. Diagnostic dilatation and curettage was done and results of endometrial sampling showed an unusual entity Low Grade Endometrial Stromal Sarcoma (LGESS). The patient and her family were informed of diagnosis, and counseled about the need of surgery and different adjuvant treatment modalities. Her total abdominal hysterectomy with bilateral salpingo-oophorectomy was done and peritoneal washings taken. Pelvic and para-aortic lymph nodes were not palpable. Specimen was sent to Shaukat Khanum Laboratory for histopathology. On macroscopic examination, uterus was 8 cm, with normal looking tubes and ovaries; no infiltration or metastatic deposits were seen in them. Microscopic analysis showed no malignant cells in the peritoneal washing. Characteristically uniform and spindle shaped cells suggestive of low grade ESS were seen (Figure 1), infiltrating upto 5 cm of myometrium. Sections from fallopian tubes, ovaries and cervix were normal. According to surgico-pathological staging, it was labeled as stage Ic.

She was referred to Bahawalpur Institute of Nuclear Oncology (BINO) for adjuvant therapy. Her radiotherapy was done in 5 sessions. She was fine 6 months post-operatively.

DISCUSSION
The traditional classification of ESS into low-grade and high-grade categories has fallen out of favor. Based on tumor margin status and cytological features, in 2003, the WHO classified endometrial stromal tumors into (a) Endometrial Stromal Nodule (ESN), (b) Low-Grade Endometrial Stromal Sarcoma (LGESS) and (c) undifferentiated endometrial or Uterine Sarcoma (UES).1 ESN does not infiltrate myometrium while ESS infiltrates myometrium and is characterized by proliferate of uniform small cells closely resembling those of endometrial stroma in proliferative stage. Women with ESS are younger, no definitive risk factor known, but exposure to tamoxifen, unopposed estrogens, and...
conditions such as polycystic disease of ovary are thought to have some role in its causation. There is a relation between chromosomal aberrations and endometrial sarcomas. Chromosomal deletion on 7p was the most common finding (55.6%) in ESS and may play a role in tumor development and progression. These tumors are diploid with a low S-phase fraction. The usual clinical presentation of ESS is abnormal uterine bleeding that occurs in about 90% of women and 70% cases show uterine enlargement. They can present with pelvic pain and dysmenorrhea. An asymptomatic ESS occurs in 25% individuals. About 30 - 50% of the ESS has extra uterine spread at the time of the diagnosis. In majority of the cases, the pre-operative diagnosis is either fibroid of uterus or adenomyosis as ultrasound and MRI are inconclusive. Trans vaginal color Doppler shows low impedance flow compared to other benign tumors. As ESS involves the endometrium, endometrial sampling by curettage may be helpful in pre-operative diagnosis. ESS is almost always positive for both estrogen and progesterone receptors and CD10 that may work to distinguish it from some other pathology. Its differential diagnosis include ESN, cellular leiomyoma cellular endometrial polyp, low-grade Mullerian adenosarcoma, and adenomyosis. Surgery is the final resort for diagnosis and primary treatment of ESS as in other sarcomas. It includes total abdominal hysterectomy, bilateral salpingo-oophorectomy; and pelvic and periaortic lymph node sampling along with peritoneal washings for cytology. The FIGO staging for sarcomas has applied to ESS. Prognosis depends upon the stage of disease. Prognostic factors are still controversial. Clinical factors such as age, race, parity, menopausal status, and pathological factors including tumor size, tumor stage, nuclear atypia, mitotic activity, tumor necrosis, lympho-vascular space invasion, DNA ploidy proliferative activity, and expression of hormone receptors have been explored with varying outcomes. As recurrence occur in about half to one-third cases, many clinicians advocate adjuvant therapy in the form of chemotherapy, radiotherapy and hormone therapy (progestrone GnRH analogues, Letrozole) to suppress tumor. It is also given in stage-II - IV and un-resectable tumors.

At FIGO stage I, the 5-year survival rate for ESS is 54% to nearly 100% and in stage-II it is 30%. For advanced disease (stage-III and IV) the survival is only 11%. As these tumors have a tendency for late recurrence, long-term follow-up is essential. It should be done once in 3 months for the first year and half-yearly for next 4 years. Thereafter, annual follow-up is recommended. The relapse-free survival depends on the tumor stage, myometrial invasion, adjuvant treatment and bilateral salpingo-oophorectomy.

REFERENCES