Wide spread Low Grade Endometrial Stromal Sarcoma

Abstract: Endometrial stromal sarcoma is an unusual malignant neoplasm of the uterus. It mainly occurs in the age of 40-50 years. It accounts 2 to 6% of uterine malignancies. Usually, the diagnosis is confirmed postoperatively. Common symptoms are abnormal vaginal bleeding, abdominal mass and mild abdominal pain. We herein report a case of endometrial stromal sarcoma in a 40 years old woman. Biopsy specimens were received at the pathology lab of ANMCH. She presented with abnormal vaginal bleeding since 4 months. Histopathological findings confirmed the diagnosis of endometrial stromal sarcoma which was then confirmed on immunohistochemistry. We aimed to present a case of endometrial stromal sarcoma because of its rare occurrence and problems in establishing histopathological diagnosis.

Key words: Endometrial stromal sarcoma, Uterine malignancy

Introduction
Uterine sarcomas are very rare and may arise from endometrial stroma, smooth muscle and connective tissue. Sarcoma of the uterus is an unusual form of malignancy affecting 2 - 5% patients. It has incidence of 1 - 2 /100000 females in overall population.1 Uterine sarcomas as a group have tendency for rapid progression along with bad prognosis. Histopathologically, uterine sarcomas are classified as endometrial stromal sarcomas, leiomyosarcomas and mixed mesodermal tumors.2 The second most common mesenchymal tumor of the uterus is endometrial stromal tumor.3 It constitutes to about 0.2% of all uterine malignancies and less than 10% of all uterine sarcomas.3 Endometrial stromal sarcoma occurs mainly in the age group of perimenopausal women along with one third being in post menopausal women.4 The World Health Organization classifies endometrial stromal tumors in four categories i-e endometrial stromal nodule, low grade endometrial stromal sarcoma, high grade endometrial stromal sarcoma and undifferentiated uterine sarcoma.5 The endometrial stromal nodule lies at the benign end of the spectrum of stromal tumors. It is a solitary sharply circumscribed mass that is confined to uterus. It does not invade veins, lymphatics or the myometrium. The prognosis is excellent. Recurrences do not occur after complete removal of the tumor.6 Low grade endometrial stromal sarcoma has less than ten mitosis per ten high power fields. It has propensity to infiltrate the large vascular spaces. Five year survival rate of low grade endometrial stromal sarcoma is 100%.7

Case Report
A forty year old woman presented with abnormal vaginal bleeding since 4 months. Ultrasound and Doppler imaging revealed a heterogeneous mass in the wall of uterus with internal vascularity. Total abdominal hysterectomy with bilateral salpingooophorectomy was done with clinical suspicion of fibroid. On gross examination, specimen consisted of uterus, cervix and both tubes and ovaries measuring 11x10x8cm. The outer surface was lobulated. Cut surface of uterus showed slit like lumen and a necrotic whitish firm growth involving whole thickness of wall in a whorling pattern. The upper cervix also appeared to be involved by the growth. One ovary and tube were enlarged in size, ovary measured 5x5cms and tube had a diameter of 3cm. Cut surface of ovary was whitish. Other ovary and follician tube appeared normal. There was also a separate nodule in container, measuring 5x4 cms with a white cut surface. Microscopically, multiple sections from nodule revealed a tumor composed of uniform small cells having round to oval nuclei and scanty cytoplasm. These cells tend to encircle small vessels in a concentric manner typical of endometrial stromal tumors. There were 5 mitosis per 10 high power fields. Vascular invasion was seen. Despite its low
mitotic count the tumor was remarkable in its multifocal spread. These multiple foci of tumor sheets had a pushing expansile pattern of growth (Figure 1). The tumor was present in the myometrium and upper cervix and metastasizing to one ovary and foppian tube (Figure 2,3&4). Other ovary and foppian tube were free of tumor. Immunohistochemical staining revealed that the neoplastic cells were positive for Vimentin, CD 10 and estrogen and progesterone receptors. The diagnosis of low grade endometrial stromal sarcoma with multifocal spread was established.

**Discussion**

Endometrial Stromal Sarcoma is a very unusual malignant neoplasm with incidence of about 1-2%. Endometrial stromal sarcomas are divided into different types on the basis of mitotic activity, vascular invasion, presence of hemorrhage and necrosis. The tumor has a malignant potential and can spread to fallopian tubes, ovaries, ureters, bladder and vagina. Jain R, in his study, had reported metastasis of endometrial stromal sarcoma to distant sites i-e lung, heart and other sites. Involvement of adnexal structures and cervix is also reported as in a case documented by Geetha P wherein endometrial stromal sarcoma infiltration was seen in the myometrium and the right foppian tube. However such extensive involvement with low grade tumor like in our case has not been reported. The common preoperative differential diagnosis of endometrial stromal sarcoma include leiomyoma of uterus and uterine leiomyosarcoma. One study conducted by Jin Y in 2010 revealed that diagnosis of endometrial stromal sarcoma preoperatively was difficult and most cases were diagnosed as benign leiomyoma preoperatively in his study. So early accurate diagnosis is very essential and also beneficial for the patient survival.

Immunohistochemistry helps in distinguishing the above mentioned differential diagnosis of endometrial stromal sarcoma. Diffuse CD10 is a useful positive marker for endometrial stromal sarcoma whereas leiomyomas are negative for it. Jassal C, in his article, had same results of positive immunohistochemical staining for endometrial stromal sarcoma. As compared to endometrial adenocarcinoma the uterine sarcomas as a group have a worse prognosis. Mukhopadhyay P reported two year survival rate of 50% in uterine sarcomas even at an early stage. However the low grade stromal sarcomas are remarkable in having a better survival rate as compared to other uterine sarcomas as documented by Xue W, therefore early and accurate diagnosis is important for proper patient management.

**Conclusion**

Endometrial stromal sarcoma is a rare malignant neoplasm. Usually leiomyoma uterus is the preoperative diagnosis and confirmed only after histopathology of uterus. By reporting our case, we wish to stress the necessity for a high degree of suspicion to diagnose this tumor even in any age group of women. A quick diagnosis and timely intervention are necessary strategies to improve patient’s health and survival.

Figure 1 Sheets of bland oval tumor cells arranged concentrically around spiral arterioles (H&E 40x)

Figure 2 Section from myometrium showing tumor infiltrating the myometrium (H&E 40x)
Figure 3 Tumor infiltrates in the cervix stroma. (H&E 10x)

Figure 4 Section from left ovary showing tumor deposits. (H&E 4x)

References


### HISTORY

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### CONTRIBUTION OF AUTHORS

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### KEY FOR CONTRIBUTION OF AUTHORS:

A. Conception/Study Designing/Planning
B. Experimentation/Study Conduction
C. Analysis/Interpretation/Discussion
D. Manuscript Writing
E. Critical Review
F. Facilitated for Reagents/Material/Analysis