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Case Report

Endometrial stromal tumor in a young woman – A rare presentation

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Abstract

Endometrial stromal tumors (EST) arise from stromal component of endometrium and are rare and common in peri or postmenopausal age group. EST are partially estrogen dependent tumors and their histopathology resemble the stromal cells of normal proliferative endometrium. They present in varied forms pathologically ranging from most benign to most malignant variants. Diagnosis essentially is by histopathology and prognosis is based on the mitotic activity of the tumor. We have presented here a case of EST presenting in a young nulliparous woman.

Key words

Endometrial stromal tumor, Mitotic activity, Sarcoma.

Introduction

Endometrial stromal tumors arise from stromal component of endometrium and the cells forming the stromal tumors resemble the cells of normal proliferative phase endometrium. These include the benign endometrial stromal nodule and endometrial stromal sarcoma. Endometrial stromal sarcoma can again be low and high grade. Still controversy exists in the criteria dividing these three variants. Variants of endometrial stromal tumors are as per **Table – 1**.

Generally EST show no relationship to parity, oncogenic precursors or association with metabolic diseases. These tumors essentially show vascular and lymphatic permeation even macroscopically.

At surgery uterus is typically uniformly enlarged. These tumors are unencapsulated and invade the veins and lymphatics of broad ligament. These extensions can be pulled out as worm like threads from the broad ligament during hysterectomy.

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Treatment is only surgical removal of uterus along with both fallopian tubes and ovaries. Removal of ovaries is essential because of possible recurrence of the tumors under estrogenic influence if ovaries are left behind. High grade tumors may require postoperative chemo or radiotherapy as adjuvant treatment.

Recurrence is a common feature. Local recurrences more with low grade tumors and distant and early recurrence is encountered with high grade tumors.

Prognosis depends on stage at presentation, mitotic activity of tumor, metastasis and receptor status.

Case report

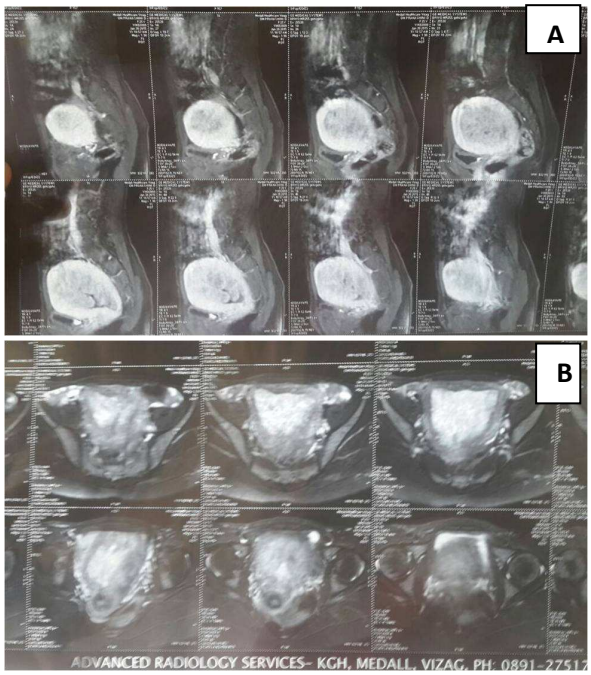
A 25 year old woman hailing from tribal belt of Visakhapatnam district in north coastal Andhra Pradesh, of low socio economic background was admitted at our medical college hospital. Her chief complaints were on and off bleeding per vaginum and pain lower abdomen since past 5 years. The woman had irregular on and off bleeding per vaginum with no rhythm or cyclical pattern, occasionally heavy changing 10 – 12 sanitary pads a day and severely painful. She also has chronic and persistent dull aching suprapubic pain with no relief on consuming any type of analgesics or anti spasmodics. Leucorrhea was also complained on rare occasions and foul smelled. There were no constitutional clinical features.

This lady after attaining menarche had regular menstruation for 5 years, got married but did not conceive. The family members took her to different gynecologists at different times and thrice underwent dilatation and curettage procedure but histopathology was not done so far.

On general examination, she looked normal built for her age except for mild pallor. On per abdomen palpation uterus found to have enlarged to 16 weeks pregnant uterine size, and same findings were confirmed on bimanual pelvic examination.

Imaging of the pelvis sonologically revealed a 13x9x7 cm mass in the uterine cavity with calcifications here and there and peripheral functional arterial pattern with high resistance noticed on Doppler enhancement. CECT imaging showed hypo isoT2 mixed intense lesion in the uterine cavity with poor planes of cleavage between endo and myometrium. CECT imaging suggested the possibility of endometrial malignancy in stage Ib as diagnosis. **(Figure - 1A, 1B)**

Figure – 1A, 1B: Enlarged uterus with uterine cavity mass lesion and no demarcation between endo and myometrium.



Endometrial biopsy revealed only proliferative endometrium without any malignancy, hyperplasia or atypia.

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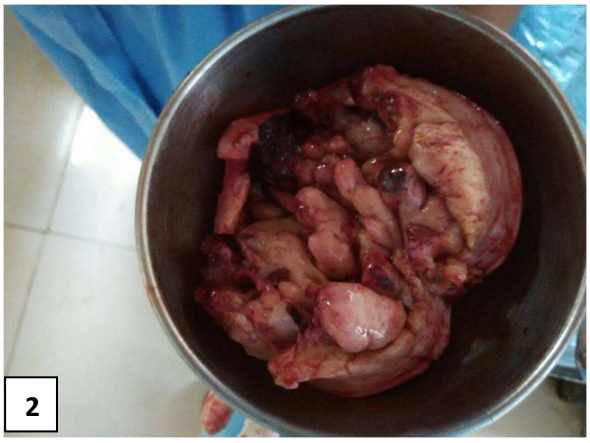
Since the woman did not respond to either antifibrinolytics or progesterones in highest possible doses, and CECT prompting the possibility of endometrial carcinoma, the woman was taken up for surgery after necessary evaluation and preoperative hematological stabilization.

Intra-operatively uterus was 16 weeks enlarged with healthy tubes and ovaries. Total abdominal hysterectomy with bilateral salpingoopherectomy was performed. On gross cut section of the specimen, uterine cavity showed a 10x10 cm soft friable and necrotic mass with no cleavage plane between the endo and myometrial interface. **(Figure – 2)** Histopathological examination (HPE) reported the tumor as endometrial stromal nodule. **(Figure – 3)** Postoperative period was uneventful. Since this is benign variant of EST we are not planning any adjuvant treatment.

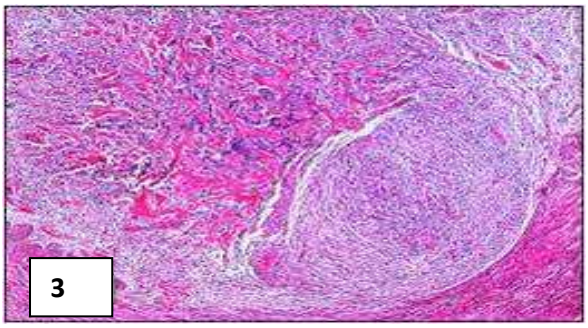
Figure – 2: Hysterectomised specimen in cut section showing friable necrotic cavitary mass.



Discussion

Uterine sarcomas are relatively rare tumors of mesodermal origin. They constitute 2-6% of uterine malignancies. There is an increase in incidence of uterine sarcomas after radiation therapy for cancer cervix or any other pelvic irradiation [1]. The relative risk is 5.38% with an interval of 10 – 20 years. Uterine sarcomas are more malignant than endometrial cancers and differ from them in diagnosis, clinical behavior pattern of spread and management. Uterine sarcomas are classified into histological variants as per **Table – 2** [1, 2]. EST are different from routine endometrial malignancies as per **Table – 3** [1, 3].

Figure – 3: Endometrial stromal tumor on HPE.



The 3 distinct variants of EST are endometrial stromal nodule, low grade and high grade stromal sarcomas are based on their mitotic activity, vascular invasion and observable differences in prognosis as per **Table – 4** [1, 4, 5, 6, 7].

Treatment is total abdominal hysterectomy with bilateral salpingo-oopherectomy. Response to hormonal treatment noted by various authors [8, 9, 10].

In our case, the EST occurred at a relatively young age, could not be diagnosed by preoperative endometrial sampling, misdiagnosed as endometrial malignancy on CECT imaging and diagnosed only by histopathology.

Conclusion

EST are rarely encountered in gynecological practice. Histopathology is only the diagnostic tool. Mitotic activity is the key criterion to classify and prognosticate these tumors.

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Vascular and lymphatic invasion is the hallmark of these tumors. Prognosis is poor in high grade ESS even with prompt treatment. Progesterones may be beneficial in low grade tumors.

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Table – 1: Variants of endometrial stromal tumors.

Stromal nodule	Low grade stromal sarcoma or endolymphatic stromal myosis	High grade stromal sarcoma or undifferentiated sarcoma
Histology similar to proliferative stromal cells with little atypia	Like proliferative stromal cells with atypia	Highly cellular and pleomorphic tumor with little resemblance to normal stromal cells
Plexiform arrangement of small vessels	Peripheral arrangement of vessels	Variable vascular pattern
Less than 3 mitosis per 10 HPF	Between 3-10 mitosis per 10 HPF	Mitotic rate more than 10 per 10 HPF
Circumscribed margin	Infiltrative margin	Extensive invasion
No vessel invasion	Vessel invasion macroscopically visible and worm like	Microscopicalls visible vessel invasion
No metastasis	Metastasis over long duration	Early and extensive metastasis
Estrogen receptors present	ER /PR present and respond to progesterones	Absence of ER /PR and nonresponse to progestogens

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Table – 2: Histological variants of uterine sarcomas.

1.Endometroid stromal sarcoma (ESS) a. Endometrial stromal nodule b. Low grade stromal sarcoma c. High grade stromal sarcoma
2. Leiomyosarcomas a. Variants like cellular leiomyoma, leiomyoblastoma b. Benign metastazing lie intravenous and disseminated peritoneal leiomyomatosis
3. Malignant mixed mullerian tumors MMMT a. Homologus b. Heterologus
4. Uterine tumor resembling ovarian sex cord tumor (UTROSCT)
5. Heterologus sarcomas like rhabdo, chondro, osteoliposarcomas

Table – 3: Difference between endometrial malignancy and endometrial stromal tumor.

Endometrial malignancy	**Endometrial stromal tumor**
Cellular origin from glands	Cellular origin from stromal cells
More in post menopause	Perimenopause
Uterine enlargement less	Significant uterine enlargement
Parametrial invasion rare	Rubbery parametrial invasion
Vascular encroachment less	Worm lie extension to pelvic vessels

Table – 4: Prognostic differences in 3 variants of EST.

Endometrial stromal nodule	**Low grade ESS**	**High grade ESS**
Less than 3 mitotic figures /10 HPF	3-10 mitotic figures /10 HPF	More than mitotic figures /10 HPF
Expansive non infiltrating solitary lesion confined to uterus with pushing margins	Extra uterine spread confined to pelvis Distant metastasis rare	Aggrieve rapidly metastasizing tumor Recurrences common
No lymphatic or vascular invasion	present	extensive
Prognosis good after surgery No recurrences	Recurrences if present take more than 5 years	Poor prognosis even after adequate treatment Need adjuvant treatment