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Case Report

Rare case of AUB in a postmenopausal woman endometrial stromal sarcoma: A case report

Indra Bhati and Nishu Bhandari*

Department of Obstetrics & Gynecology, Dr. S. N. Medical College, Jodhpur. India

***Correspondence Info:**
Dr. Nishu Bhandari
Third year resident
Dept of Obstetrics & Gynecology
Dr S.N. Medical College
Jodhpur, India
E-mail: nishu.4587@gmail.com

Abstract

Endometrial Stromal Sarcomas (ESSs) are very rare malignant tumors that constitute approximately 10% of all uterine sarcomas but only around 0.2% of all uterine malignancies. The annual incidence of ESS is 1–2 per million women. It is also known as endometrial stromal myosis. The median age was 44.35 ± 6 years. Women with ESS are younger than women with other uterine sarcomas, with a median age between 45 and 57 years and, generally do not have the usual risk factors for endometrial cancer. Reported cases of endometrial stromal sarcoma in a postmenopausal woman are a rare entity.

These tumors are said to be hormone dependent and therefore seems to be more common in reproductive and perimenopausal age group. Patients mainly presents with complaints of heavy vaginal bleeding and pain abdomen. ESS can be mistaken for leiomyoma. Clinical and radiology findings are suggestive of a leiomyoma. HPE remains the mainstay for diagnosis of such tumors .As these tumors are known for their recurrences close follow up of these patients is very important.

Keywords: endometrial stromal sarcoma, postmenopausal

1. Case Report

A postmenopausal, 70 year old woman (P3) visited our O.P.D with complaints of heavy bleeding per vaginum since three months, which was not associated with any pelvic or abdominal pain.
O/E- Abdomen was soft non tender and no mass could be felt.

1.1 Per vaginum examination

OS admitted one finger; a smooth large polyp like structure felt arising from lateral wall of uterus filling the endometrial cavity and uterus was regularly enlarged to 12 weeks with b/l adnexa free.

Her routine investigations (Hb, total leucocyte counts, total platelet counts, coagulation profile, renal function and liver function tests) were within normal limts.

1.2 Transvaginal USG

Findings revealed presence of a solitary solid intrauterine mass of about 7x8 cms with cystic changes? **Leiomyoma.** Endometrial thickness ET was 3mm.

Her *endometrial biopsy report* was suggestive of Endometrial Stromal Sarcoma (ESS)

CT SCAN findings negated any metastasis or lymphovascular invasion and showed the presence of a solid smooth mass in endometrial cavity of about 7.4x6x4cms.

Subsequently after counselling & oncosurgical consultation the patient with high risk consent underwent TAH & BSO. Uterus was uniformly enlarged to about 12 wks size, external surface was smooth with no growth visible on the external surface and b/l adenexa normal for age. Cut section of the specimen revealed a smooth solitary ovoid mass in the endometrial cavity measuring appx 7x5 cms in dimensions and was free from all sides from myometrium except on the left where it was attached through its broad base. Cut section of growth showed fatty degeneration but absence of any whorled appearance.

Histopathological report showed the presence of endometrial stromal sarcoma with less than 50% invasion of myometrium FIGO Stage IB There were evidences of micro invasions in the lymphovascular channels.

Patient had uneventful recovery & was discharged on D6 postoperatively with advice to remain in follow up and consult medical oncologist for further treatment.

Fig 1: Gross specimen

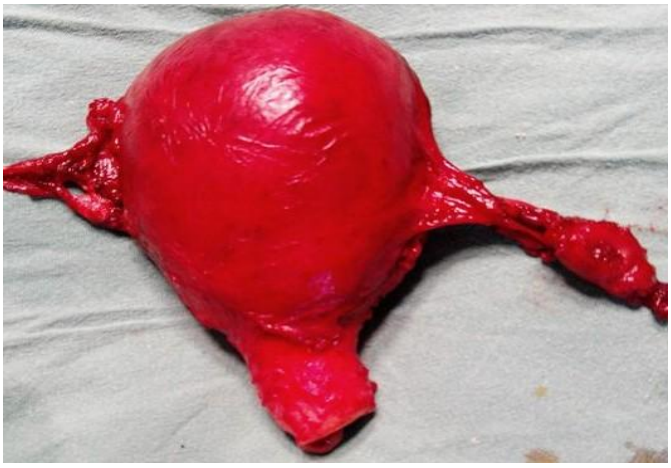


Fig 2: Cut section of the above gross specimen revealing a single large nodule in the endometrial cavity which is free from all sides of myometrium except at one side where it is attached through its broad base.

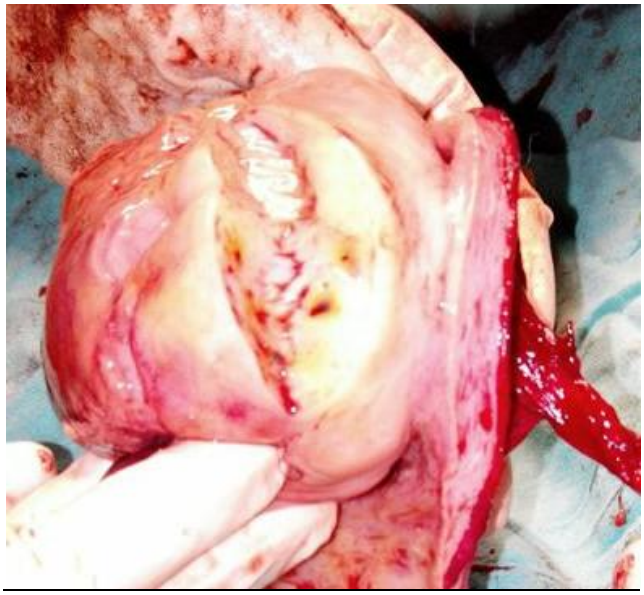


Fig 3: Histopathological findings of the above gross specimen showing perivascular invasions by malignant cells.

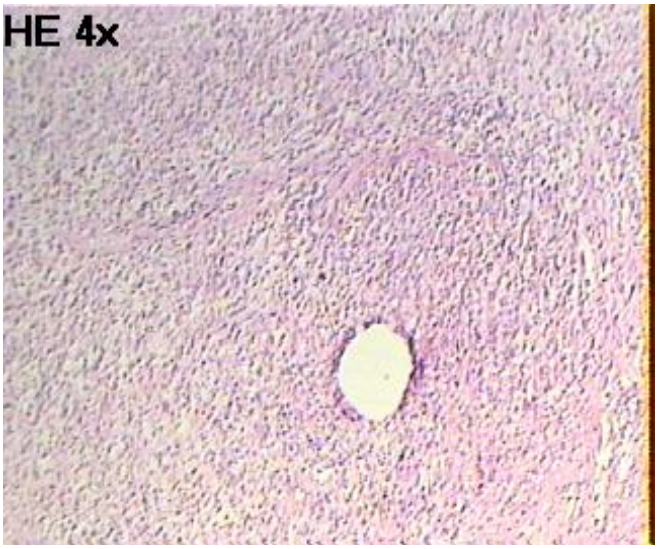


Fig 4: Histopathological finding showing invasion of myometrium by malignant stromal cells.

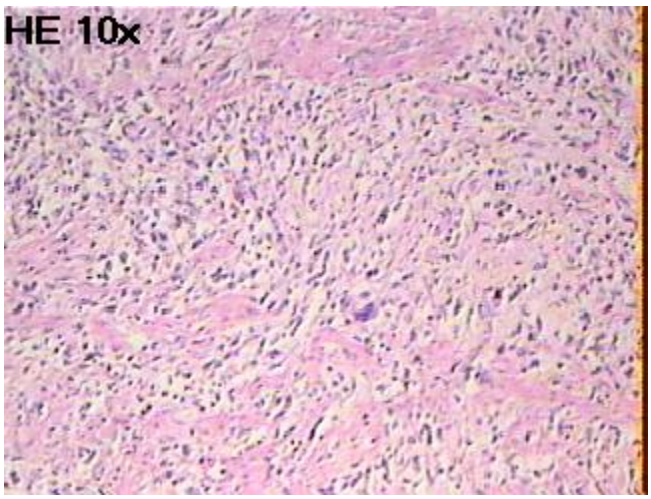
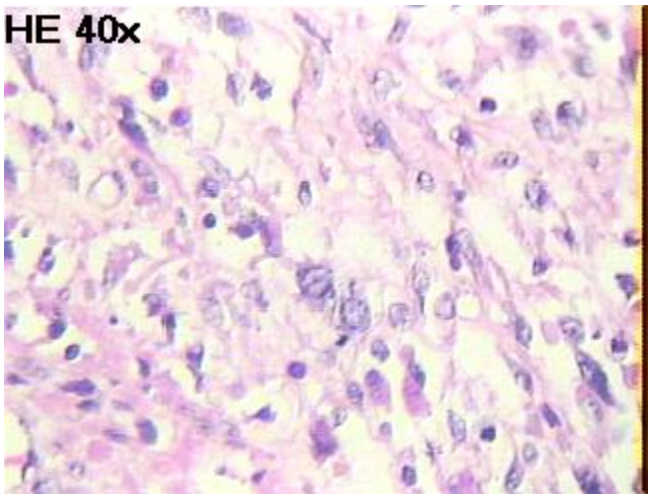


Fig 5: Histopathological finding showing bizarre malignant cells with high nuclear pleomorphism



2. Discussion

Endometrial stromal sarcomas (ESS) are rare malignant tumors of the uterus and their incidence in postmenopausal age group is even rarer entity.

Endometrial stromal sarcoma has some genetic predilections. Study reveals a specific translocation t{7;17} {p15;q21} with involvement of two zinc fingers genes juxtaposed with another zinc protein 1.[2] CD 10 is expressed in ESS along with inhibin which helps it to differentiate with cellular leiomyoma that does not express CD 10.[3] Estrogen and Progesterone receptors are found to be positive in ESS which further helps in evaluating the prognosis of tumor and thereby there rare occurrence in postmenopausal age group.

2.1 Differential Diagnosis

This includes several soft-tissue neoplasms demonstrating arborizing vasculature, highly cellular leiomyoma, cellular endometrial polyp, low-grade mullarianadenosarcoma, and adenomyosis[4]. Extra genital ESS may be confused with gastrointestinal stromal tumors, hemangiopericytoma, lymphangiomyomatosis, or mesenchymal cystic hamartoma of the lung[5,6]. ESS of the ovary is difficult to distinguish from sex-cord stromal tumors.

Surgery is the final resort for diagnosis and primary treatment of ESS. Preoperative diagnosis is mostly a uterine fibroid. Ultrasound and magnetic resonance imaging are inconclusive. If the diagnosis is known, the extent of surgery is planned according to the stage of the tumor. The FIGO staging for carcinoma of the corpus uteri has been applied to ESS.

The treatment is total abdominal hysterectomy, bilateral salpingooophorectomy and pelvic and periaortic selective lymphadenectomy. Cytologic washings are obtained from the pelvis and abdomen. Hormone therapy with medroxy progesterone, tamoxifen, gonadotropin releasing hormone (GnRH) analogues and aromatase inhibitors are suggested for low-grade ESS stage 3–4 and for recurrent disease[7,8].

3. Conclusion

The traditional classification of ESS into low-grade and high-grade categories has fallen out of favor, and high-grade tumors without recognizable evidence of a definite endometrial stromal phenotype are now termed undifferentiated endometrial sarcomas (UES) instead of high-grade ESS. UES represents a high-grade sarcoma that lacks specific differentiation and bears no histological resemblance to endometrial stroma and therefore the term ESS is now considered best restricted to neoplasm that are formally referred to as low-grade ESS. Therefore, ESS is used for previous LGESS and UES is used for previous high-grade endometrial stromal sarcoma (HGESS). Since myometrial and vascular invasion

are the two features that help us to differentiate ESS from ESN and the UES resembles the sarcomatous component of carcino sarcoma, extensive sampling of the tissues is required for confirmation of diagnosis[9].

At FIGO stage I, the 5-year survival rate for ESS is 54% to nearly 100% and at 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 shall be once in 3 months for the first year and half-yearly for next 4 years. Thereafter annual follow up is recommended. Because of concern about radiation exposure, frequent routine asymptomatic surveillance imaging is not recommended after primary treatment[10]. The relapse free survival depends on the tumor stage, myometrial invasion, adjuvant treatment, and bilateral salpingoopherectomy[11,12].

Hormone therapy is a new promising adjuvant treatment modality. Multianalysis from a large group of patients is necessary for predicting prognosis and to define proper treatment of endometrial stromal sarcoma.

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