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High-Grade Endometrial Stromal Sarcoma Presenting As a Polyp: A Rare Case Report

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ABSTRACT

Endometrial Stromal Sarcoma (ESS) is commonly seen in perimenopausal women, typically presents with abnormal uterine bleeding and most commonly, pre-operative diagnosis will be leiomyomas. We report a case of 40 year old female presenting with excessive bleeding per vaginum since 2 months with provisional clinical diagnosis of bulky uterus with leiomyomatous polyp, abdominal panhysterectomy was done. Histopathological examination is helpful in the diagnosis of High Grade ESS. The case is presented in view of its rarity and showing very much distinguishing gross and microscopy appearance. It highlights the unsuspected uterine malignant tumor. The histopathological examination again proved as gold standard to diagnose this rare entity.

Keywords: Endometrial Stromal Sarcoma, Uterus, Abnormal uterine bleeding.

INTRODUCTION

Endometrial Stromal Sarcoma (ESS) are very rarely encountered malignancies of uterus that account for only 1% of all uterine malignancies.[1] Endometrial stromal tumors are divided into endometrial stromal nodule and endometrial stromal sarcomas (ESS). ESS further divided into low-grade, high-grade, and undifferentiated stromal sarcomas. It is difficult to detect it preoperatively, and thus the diagnosis is usually, histopathological after the hysterectomy has been done for a clinically suspected benign condition. The prognosis of the disease is largely based on the stage of the disease at presentation. However, ESS has a better life expectancy than other sarcomas. The treatment is surgical, and extent depends on the type and staging of the tumor. We, thus, report this case to highlight the diagnosis of high-grade ESS by histopathology in a patient which presented clinically as per vaginal bleeding.

CASE REPORT

A 40-year-old female patient presented with complaints of excessive bleeding per vagina along with passage of clots for the past 2 months. No history of abdominal pain and abnormal discharge was reported.

On abdominal examination

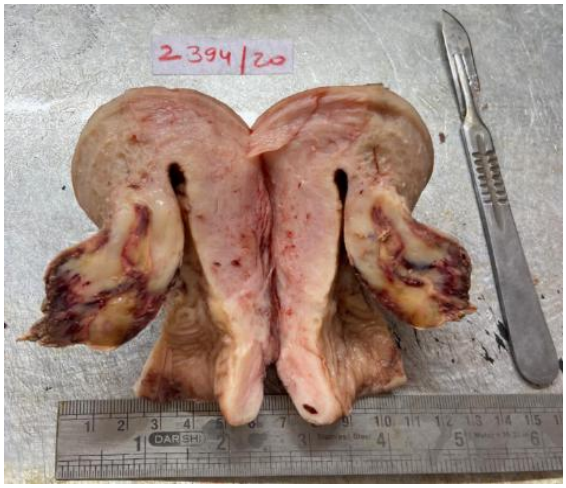
Uterus was 16 weeks size with a firm, nontender mass in the uterine fundus with side-to-side mobility and regular margins.

On per vaginal examination, the uterine movement transmitted to the cervix and clinically suspected as fibroid polyp which was fungating and bleeds on touch.

Radiological findings

Ultrasound abdomen showed enlarged uterus with elongated Endometrial Polyp. Endometrial thickness was 17mm. Bilateral ovaries were normal. The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy.

Gross examination

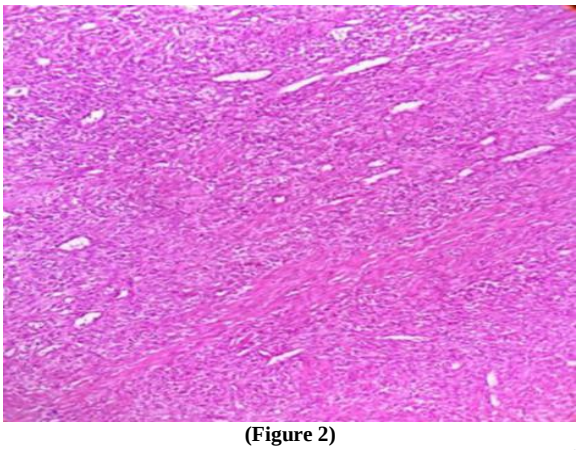


(Figure 1)

Uterus with cervix measures 10.5 x 7.5 x 5.7 cm and weighs 200gms. External surface of uterus and cervix was unremarkable. Endometrial canal measures 5.3cm in length. On cutting open, in the fundal region endometrial cavity showed a polypoidal mass measuring 5.1 x 3.9 x 3cmwhich was firm, grey white to grey yellow, fleshy with focal areas of hemorrhage and necrosis. The polypoidal mass was not invading the body, isthmus of uterine cavity as well as endocervical canal grossly. Both ovaries and fallopian tubes with attached fimbriae appear unremarkable. (Figure 1)

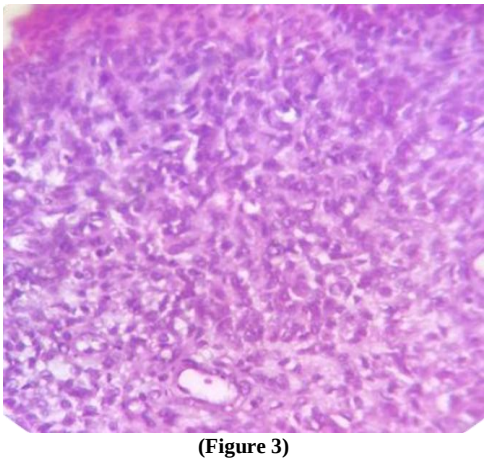
Microscopic features

H and E stained sections studied show a tumor composed of neoplastic endometrial stromal cells arranged in loose fascicles. The individual cells are moderate to severely pleomorphic round to oval to spindle shaped cells with nuclei having moderate to severe atypia and hyperchromatic nuclei with inconspicuous nucleoli and scant to moderate eosinophilic cytoplasm. The mitotic activity was > 10/ 10 Hpf. Areas of hemorrhage and necrosis were also seen. Tumor was seen invading inner half of myometrium of the fundal region. The histopathological diagnosis was given as High grade Endometrial Stromal Sarcoma.

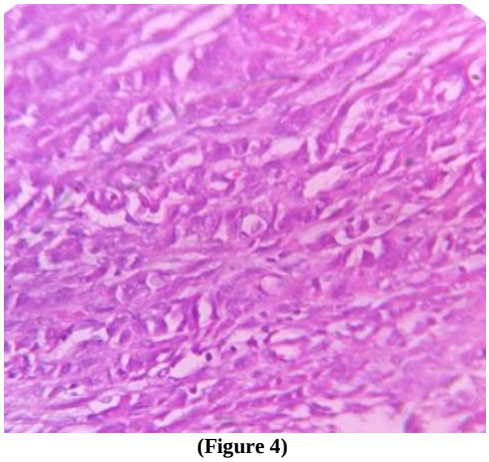


(Figure 2)

Figure 2: Scanner view showing tumor composed of endometrial stromal cells.

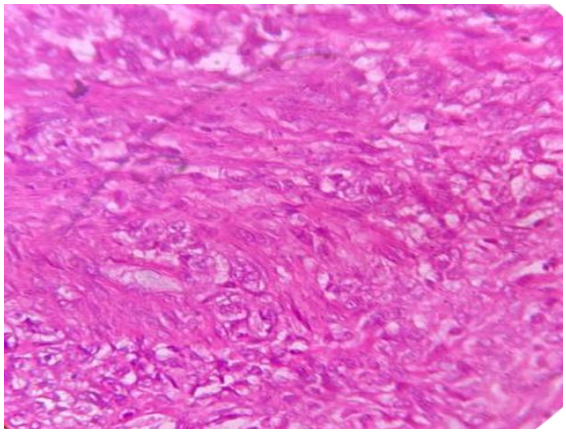


(Figure 3)



(Figure 4)

Figure- 3 and 4, High power view. The endometrial stromal cells displayed round to oval moderately pleomorphic nuclei with inconspicuous nucleoli, hyperchromasia and scanteosinophilic cytoplasm. (Fig 3,4)



(Figure 5)
Figure 5: Myometrial invasion by tumor

She had been referred to an oncologist for further management by gynaecologist.

DISCUSSION

ESS account for 1% of all uterine malignancies and 15% of malignant mesenchymal neoplasms of the uterus usually seen in the age group of 42-58years.[2] In 2014, the WHO categorizes ESS into LGESS, high-grade, and undifferentiated sarcoma types.[3] At the time of presentation, the symptoms are nonspecific, and most of the patients presents with abnormal uterine bleeding like our case. An early diagnosis is essential because the patient survival is directly related to tumor stage.[4] The uterine corpus is the most frequent location though it can also primarily arise in a variety of extrauterine locations such as the ovary, pelvis, abdominal cavity, vulva, and vagina.[5] Up to 30% of women with LGESS have an extrauterine disease at the time of presentation. Preoperative diagnosis is often difficult and around 75% are diagnosed and operated as fibroid uterus. Preoperative endometrial dilatation and curettage sampling usually do not help to arrive the diagnosis, due to similarity with normal endometrium.[6] Pelvic ultrasound examinations may also go in vain to diagnose the disease accurately as happened in our case. Low-grade ESS has a metastatic potential. The mitosis in low-grade ESS is<3/10 high-power field. The treatment is panhysterectomy with removal of bilateral adnexa. This can be followed by hormonal therapy. High-grade ESS is rapidly growing neoplasms. These do retain evidence of endometrial stromal derivation but have high-grade round cell morphology. The treatment is panhysterectomy with chemotherapy and radiotherapy. At immunohistochemistry, they typically present with high Cyclin D1 and low CD10 expression.[7] The 5-year survival rate is <57%.[8] Undifferentiated ESS represents a high-grade sarcoma that lacks specific differentiation and has no histological resemblance to endometrial stroma. These are aggressive in nature with poor prognosis. Hence, it is the need of the hour to report cases with ESS so that a proper management plan can be deciphered. About 90% of patients present with AUB and 70% present with uterine mass. In 40% cases, they present as an incidental finding. In our case the clinically diagnosis was fibroid polyp.[9] There is a high rate of lymph node involvement in ESS. Almost 10% of those who underwent lymph node dissection had nodal metastasis, so lymphadenectomy is for both prognostic and therapeutic purpose.[10] Radiotherapy, chemotherapy, or tyrosine kinase inhibitors can be used as adjuvant therapy but shows limited relapse-free survival rate.[11] In reviewing the literature, very few cases of high-grade ESS have been reported. Our case is unique in itself in many aspects

CONCLUSION

ESS is rare tumors, presents in perimenopausal women with abnormal uterine bleeding, most of the time, the preoperative diagnosis will be uterine leiomyoma. This case report highlights the unsuspected malignant endometrial tumor mimicking leiomyomatous polyp clinically. Histopathological examination again proved as the gold standard to confirm the diagnosis of this rare entity. This study recommended Endometrial Stromal Sarcoma should be included in differential diagnosis of endometrial polyps.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her gross and histopathological images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity.

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Conflicts of interest: There are no conflicts of interest.

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