Endometrial Stromal Sarcoma: Unexpected Guest!

Abstract

Endometrial Stromal Sarcoma (ESS) are very rarely encountered malignancies of uterus that account for only 1% of all uterine malignancies. ESS is commonly seen in perimenopausal women, typically presents with abnormal uterine bleeding and most commonly, pre-operative diagnosis will be leiomyomas. We report an interesting case of 42 year old female presenting with irregular cycles and excessive bleeding per vaginum. With provisional clinical diagnosis of fibroid uterus, total abdominal hysterectomy was done. Histopathological examination and immunohistochemistry staining with CD 10 confirmed the diagnosis of Low 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, which was mimicking leiomyoma both clinically and radiologically. The histopathological examination again proved as gold standard to confirm the diagnosis of this rare entity.

Keywords: *CD10, endometrial stromal sarcoma, uterus*

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Introduction

Endometrial stromal sarcoma (ESS) accounts for <1% of all uterine tumors. Sarcomas are rarely encountered malignancy of the uterus, with an incidence of 1–2 cases/100,000 women. The World Health Organization (WHO) classifies endometrial stromal tumors as a benign endometrial stromal nodule (ESN) and ESS. ESNs are termed benign, as they do not infiltrate myometrium. In contrast, ESSs are characterized by the infiltration of the myometrium. We came across this rare malignant tumor, typically presenting with abnormal uterine bleeding in perimenopausal women. The case report highlights the classical gross appearance of this rare entity and reviews the literature available on ESS.

Case Report

A 42-year-old female patient presented with complaints of irregular menstrual cycles for 2 years with excessive bleeding per vagina along with passage of clots for the past 10 days. No history of abdominal pain and abnormal discharge was reported.

On abdominal examination

Uterus was 18 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 fornices free with a healthy cervix and vagina.

Radiological findings

Ultrasound abdomen showed enlarged uterus with probable fibroid that measured 9.5 cm × 7.5 cm in the posterior wall of the uterus. Bilateral ovaries were normal. With the preoperative diagnosis as fibroid uterus, the patient underwent total abdominal hysterectomy with bilateral salphingo-oophorectomy.

Gross specimen of the uterus showed multiple gray-yellowish ropy nodular masses ranging in size from 0.1 to 1.5 cm seen over the entire endomyometrium [Figure 1].

On histopathological examination

Endometrial stromal cells were seen without any evidence of nuclear atypia, showing extensive myometrial permeation (tongue-like growth) by sharply defined tumor islands with pointed edges [Figure 2]. The endometrial stromal cells displayed round to oval bland nucleus, inconspicuous nucleoli and scanty cytoplasm. These cells were arranged concentrically around spiral arterioles [Figure 3]. Extensive lymphatic

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emboli were appreciated [Figure 5] suggesting the appropriate previous name "endolymphatic stromal myosis." These tumor cells were seen infiltrating >2/3rd thickness of myometrium, however, confined to the uterus. There was no evidence of nuclear atypia. The mitotic activity was very minimal with <10/10 high-power field (HPF). There was no evidence of hemorrhage or necrosis. On immunohistochemistry (IHC) staining for CD10 [Figure 4], the diagnosis of low-grade ESS (LGESS) was confirmed. Six months postoperative period follow-up of the patient is uneventful, and she has been advised for long-term follow-up and referred to an oncologist for further management.

Discussion

ESS account for 1% of all uterine malignancies and 15% of malignant mesenchymal neoplasms of the uterus.[1] In 2014, the WHO categorizes ESS into LGESS, high-grade, and undifferentiated sarcoma types.[2] At the time of presentation, the symptoms are nonspecific, and most of the patients will present with abnormal uterine bleeding. An early diagnosis is essential because the patient survival is directly related to tumor stage.[3] 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.[4] 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.[5] Pelvic ultrasound examinations may also go in vain to diagnose the disease accurately as happened in our case. Grossly, LGESS may be submucosal or intramural, usually with ill-defined borders and "wormlike" permeation within the myometrium and parametrial tissue, though some tumors might appear relatively circumscribed, and the cut surface is fleshy tan-to-yellow. These classical gross findings are very much evident in our case report.[6] Histologically, it characteristically demonstrates extensive permeation of the myometrium as irregular islands with a frequent lymphovascular invasion. The majority of LGESSs show bland nuclear features with monotonous oval-to-spindle nuclei that resemble proliferative phase endometrial stroma, the mitotic activity is generally low (5/10 HPF), and the necrosis is usually absent.[7] In

Figure 1: Gross specimen of the uterus showing multiple gray-yellowish ropy nodular masses ranging in size from 0.1 to 1.5 cm seen over the entire endomyometrium

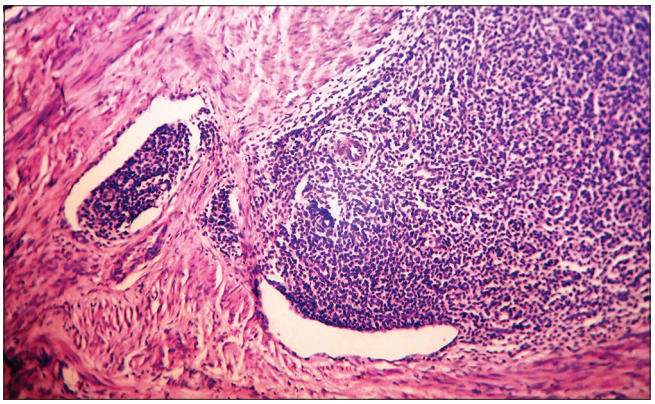

Figure 2: Endometrial stromal cells showing extensive myometrial permeation (tongue-like growth) by sharply defined tumor islands with pointed edges (H and E, ×4)

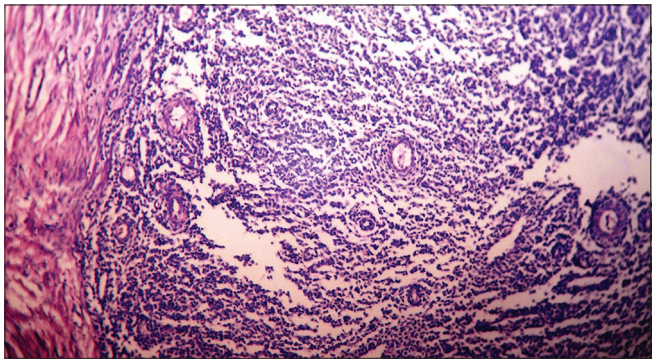

Figure 3: Endometrial stromal cells displaying round-to-oval bland nucleus, inconspicuous nucleoli, scanty cytoplasm, and arranged concentrically around spiral arterioles (H and E, ×4)

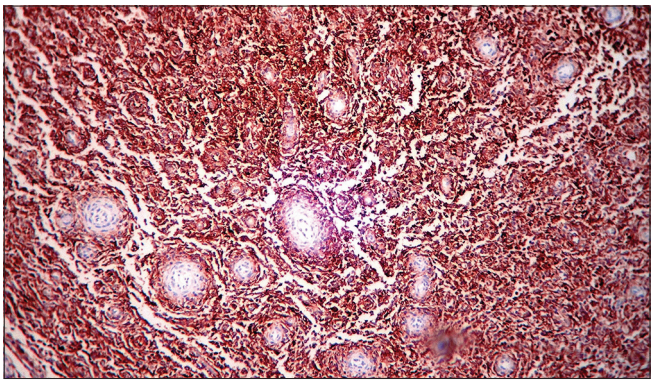

Figure 4: Endometrial stromal cells strongly positive for immunohistochemistry marker CD 10 (×10)

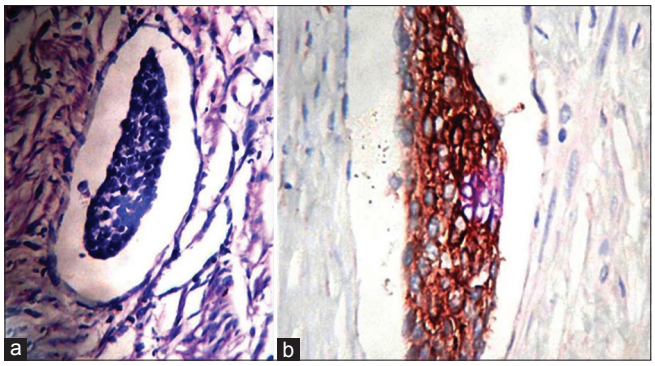

Figure 5: Extensive lymphatic emboli suggesting appropriate previous name "endolymphatic stromal myosis" ([a] is H and E, and [b] is immunohistochemistry with CD10. ×10)

our case report, we have not considered any of the other lesions, as the histopathology findings were very much convincing of LG-ESS. The most common cytogenetic abnormality of LGESSs is a recurrent translocation involving chromosomes 7 and 17 t(7;17) (p15;q21), which results in a fusion between JAZF1 and SUZ12 (formerly designated as JJAZ1).[8] We did not carry-out any other molecular diagnostic tests in our case due to financial constrains with the patient. The surgical stage appears to be the most important prognostic factor.[9] Patients with LGESS have an excellent prognosis with a 90% 5-year disease-free survival if low stage (I/II); 5-year survival drops to 50% if high stage (III/IV)[10] At times, it is very difficult to differentiate ESS from the cellular leiomyoma. In these cases, IHC is especially helpful to arrive at the final diagnosis. The IHC markers such as h-caldesmon and CD 10 may solve the diagnostic problem as CD 10 staining is positive in ESS but not in leiomyoma.[3] We performed CD 10 staining to ascertain the diagnosis. In our case, the tumor cells of ESS were strongly positive for CD10 marker and tumor emboli within lymphatic channels also showed positivity thereby confirming the diagnosis of ESS. A prompt diagnosis and timely intervention are keys to improve patient survival. Our case referred to Chemotherapy at regional cancer institute for further treatment and 6 months follow up of the patient is uneventful.

Conclusion

ESS are 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 uterine tumor mimicking leiomyomas clinically and radiologically. Histopathological examination again proved as the gold standard to confirm the diagnosis of this rare entity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their 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, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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