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Case Report

An Unexpected Cause of Severe Anemia: Endometrial Stromal Sarcoma; Case Report

Ciddi Aneminin Beklenmeyen Bir Nedeni: Endometrial Stromal Sarkoma; Olgu Sunumu

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Abstract

Endometrial stromal sarcoma (ESS) is a rare malignant tumour of the uterus that usually occurs in perimenopausal women. Such a case was a hospitalized 42-year-old woman with acute heavy vaginal bleeding and severe anaemia. The patient refused to have a gynaecological examination and endometrial sampling. However, during the transabdominal ultrasonographic evaluation, an intramural submucous mass of 9 cm × 6 cm was detected in the fundal area of the uterus. The diagnosis of ESS was discovered unexpectedly by examining a frozen section of the tumour that was thought to be leiomyoma preoperatively. After intraoperative consultation with the Gynaecologic Oncology Department, total hysterectomy and bilateral salpingectomy were performed. Once the final pathological report was presented, the patient was recommended to have bilateral oophorectomy in order to proceed with the treatment.

Keywords: Endometrial stromal sarcoma (ESS); malignant tumour; endometrium

Öz

Endometrial stromal sarkom (ESS), genellikle perimenopozal kadınlarda görülen uterusun nadir bir malign tümörüdür. Bu olguda akut ağır vajinal kanama ve şiddetli anemi ile başvuran 42 yaşında jinekolojik muayeneyi ve endometriyal örneklemeyi reddeden bir kadın hastayı ele aldık. Transabdominal ultrasonografik değerlendirmede uterusun fundal bölgesinde 9x6 cm boyutlarında intramural submuköz kitle tespit edildi. ESS tanısı, ameliyat öncesi leiomyom olduğu düşünülen tümörün frozen kesiti ile tesadüfen konuldu. Jinekolojik Onkoloji Bölümü ile intraoperatif konsültasyon sonrası total histerektomi ve bilateral salpenjektomi yapıldı. Nihai patolojik raporun ardından hastaya tedavinin sonlandırılması için bilateral ooferektomi önerildi.

Anahtar Kelimeler: Endometrial stromal sarkom (ESS); malign tümör; endometrium

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1. Introduction

Endometrial stromal tumours (ESTs) are rare and complex subset of mesenchymal uterine neoplasms with heterogeneous morphological, immunohistochemical and genetic features. ESTs constitute ~10% of uterine mesenchymal tumours (1). In the most recent classification by The World Health Organization (WHO) in 2020, Endometrial Stromal Tumours (EST) have been divided into four categories: Endometrial Stromal Nodule (ESN), Low-Grade Endometrial Stromal Sarcoma (LG-ESS), High-Grade Endometrial Stromal Sarcoma (HG-ESS) and Undifferentiated Uterine Sarcoma (UUS) (2). While ESN is clinically benign and LG-ESS shows a low malignant potential, HG-ESS is a highly aggressive tumour (3). The mean age for LG-ESS is 52 years, ranging between 16 and 83 years. The risk factors are pelvic radiation and prolonged use of tamoxifen or oestrogen. The most common findings are abnormal uterine bleeding and pelvic pain.

A rare case of LG-ESS in a premenopausal woman will be presented in order to draw attention to the presence of this rare malignancy that manifests itself with acute bleeding and severe anaemia.

2. Case Study

A 42-year-old woman had an emergency admission to the "Reproductive Endocrinology Department" of the Health Science University of Zübeyde Hanım Women's Disease Training and Research Hospital, Ankara, Turkey in March 2021. The patient had severe persistent vaginal bleeding that had been present for the past 20 days. The subject was a virgin, non-smoker with no history of contraceptive or hormonal method use. The patient was asymptomatic and had regular menstrual cycles 20 days prior to her hospital admission. The subject's past medical history (PMH) was rather clean - there were no medical disorders, no previous surgeries nor any gynaecological examinations. In admission, the patient was cooperative, with a pale skin, a blood pressure of 90/60 mmHg, and a pulse rate of 84 beats/min, accompanied with moderate vaginal bleeding. She refused to have a speculum and/or bimanual gynaecological or rectal examination. Transabdominal ultrasonography revealed a submucous-intramural mass of 9cmx6cm located in the fundal area of the uterus while the remaining regions of the myometrium were heterogeneous. The endometrial thickness was not evaluated clearly due to the compression of the presumed leiomyoma on the uterine cavity. Ultrasonography of the abdomen was otherwise normal.

The patient had a haemoglobin level of 3.8 g/dL and haematocrit of 14.7%. The serum levels of ferritin 2.37 µg/l, iron 3.0 µg/dl and iron-binding capacity 370 µg/dl. The β-hCG was negative. Liver, kidney and thyroid function tests were normal. The patient received 6 units of erythrocyte suspencion transfusion (ESP) and 2 units of fresh frozen plasma (FFP). The serum hormone values were as follows: FSH 6.48 U/L, E2 211 µg/l, LH 16.1 U/L.

Since the patient refused to have an endometrial sampling, she was counselled about myomectomy and simultaneous endometrial sampling with frozen section analysis under general anaesthesia. Further, laparotomy was performed. During the intraoperative exploration bilateral fallopian tubes, ovaries and peritoneal surfaces showed to be normal. A 10cm bulky uterine mass in the fundal area which was quite distinct and red-brown soft tissue was sent to frozen section analysis followed by myomectomy (Figure 1a, 1b). After receiving the histopathological result of the tumour that was reported as a mesenchymal tumour with malignant properties, total hysterectomy, bilateral salpingectomy and bilateral pelvic

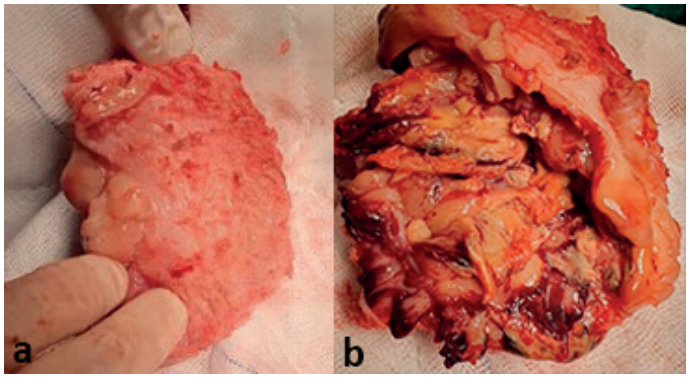

Figure 1a, 1b. Intraoperative appearance of uterine mass (a) from the posterior side (b) from the inside

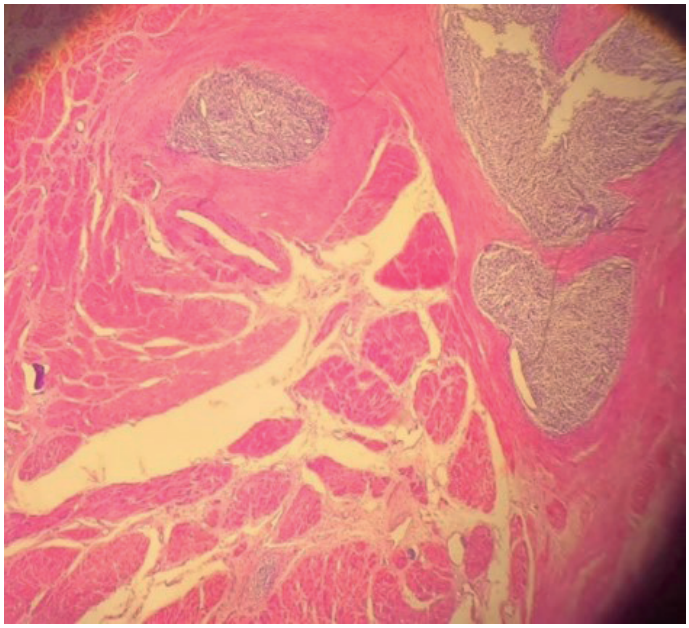

Figure 2. Histopathological appearance of the uterine mass [x40, (H&E)]

lymphadenectomy were performed after intra-operative consultation with the Gynaecologic Oncology Department. Bilateral oophorectomy was not performed because it was planned to discuss the final pathology and treatment plan with the patient. The final histopathology result was reported as low-grade endometrial stromal sarcoma, stage 1B and the endometrial sampling was also reported as a LG-ESS (Figure 2). The immunohistochemical examination reported oestrogen and progesterone receptors positivity, while cell membrane metallopeptidase CD 10 was positive and desmin Cyclin-D1 was negative. After the final pathological report, the patient received a gynaecologic oncology consultation and a bilateral oophorectomy was recommended in order to proceed with the treatment.

3. Discussion

Uterine sarcomas are rare tumours originating from mesenchymal cells and represent about 3% of all uterine cancers (4). In the early stages, malignant uterine tumours are frequently asymptomatic and mimic the clinical nature of benign uterine tumours, such as uterine leiomyoma. In this case, the patient reported to have no complaints until the last 20 days with the ultrasonographic appearance of the mass resembling a leiomyoma.

In neoplasms with focal smooth-muscle differentiation, the neoplasm is categorized as LG-ESS if the smooth-muscle component involves < 30% of the total volume. However, they have myometrial and/or vascular invasion as distinguishing characteristics (5).

The majority of the ESS are immunoreactive for the oestrogen receptors (ER) and progesterone receptors (PR). Typically, they are strongly positive for CD10 positive for smooth muscle actin and negative for h-caldesmon and histone deacetylase 8 (HDAC8) (3). In the patient's case, the histopathological examination reported an infiltrating tumour between the myometrial muscle fibres that showed an invasion into the vascular spaces with haematoxylin and eosin stain on x40 magnification (Figure 2). Furthermore, the immunohistochemical examination reported positive oestrogen and progesterone receptors, positive CD 10, and positive desmin Cyclin-D1.

A treatment of uterine sarcomas is determined by a histopathological diagnosis and the stage of the disease. The surgical management of LG-ESS includes hysterectomy with or without bilateral salpingo-oophorectomy (6). Even if the uterus is removed by minimally invasive surgery and the diagnosis of ESS was accidental, complete and en bloc resection of the tumour without morcellation may be of significant importance for an early stage (uteus-limited) ESS diseases (7). However, the prognostic value of lymphadenectomy and adjuvant therapy is not clear yet in LG-ESS tumours (8). As reported in the literature, LG-ESS are hormone-dependent tumours and bilateral salpingo oophorectomy prevents occult metastases and endogenous hormone production of the ovary (4). The European Society of Medical Oncology guidelines concluded that the value of bilateral salpingo-oophorectomy has not yet been specifically determined for premenopausal women (9). A multi-institutional, retrospective study was conducted in a total of 124 patients, who received a curative-intent surgery in Turkey. Factors within the Stage I and Stage ≥II subgroups were analyzed and no significant prognostic factor was found for stage I; however, lymphadenectomy and adjuvant chemotherapy were significantly associated with disease outcomes for stage ≥II. Lymphadenectomy was associated with improved DFS, while chemotherapy was associated with poor DFS and OS (10).

In this patient's case, the diagnosis of ESS was discovered incidentally by frozen section of the tumour that was thought to be leiomyoma pre-operatively. This case report aims to emphasize the importance of performing a frozen section after excision of the mass that changed the approach to the case.

4. Conclusion

It can be concluded that endometrial sampling is very important for the evaluation of the patient with abnormal uterine bleeding. However, it may be rejected by the patient due to the patient's personal reasons. So in this case, laparoscopy with morcellation was not an option. But methodically participating line of actions such as macroscopic evaluation of the mass that was removed with a presumed-diagnosis of leiomyoma, presence of frozen section evaluation and immediate consultation of the Gynaecological Oncology Department enabled a reach to the diagnose and a treatment with LG-ESS for the patient.

Author contribution

Study conception and design: AA, BGÖ; data collection: AA, BGÖ; analysis and interpretation of results: BD; draft manuscript preparation: AA, BGÖ. All authors reviewed the results and approved the final version of the manuscript.

Confirmation

The written consent was received from the patient who was presented in this study.

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Conflict of interest

The authors declare that there is no conflict of interest.

Yazar katkısı

Araştırma fikri ve tasarımı: AA, BGÖ; veri toplama: AA, BGÖ; sonuçların analizi ve yorumlanması: BD; araştırma metnini hazırlama: AA, BGÖ. Tüm yazarlar araştırma sonuçlarını gözden geçirdi ve araştırmanın son halini onayladı.

Onay

Kayıt sırasında katılım için bilgilendirilmiş onam formu imzalanmıştır.

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