

A Young Patient with Low Grade Endometrial Stromal Sarcoma: Case Report

Düşük Dereceli Endometriyal Stromal Sarkomlu Genç Bir Hasta

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ABSTRACT Endometrial stromal tumors are rare uterine neoplasms, accounting for approximately 0.2% of all genital tract malignancies. The tumor is classified into a low-grade endometrial stromal sarcoma (LGESS) and high-grade endometrial stromal sarcoma (HGESS) based on mitotic rate. The prognosis of LGESS is much better than for other uterine sarcomas, including HGESS. Endometrial stromal sarcoma is often diagnosed postoperatively on hysterectomy specimens taken from patients initially thought to have a benign condition such as leiomyoma. The recommended treatment is total hysterectomy with bilateral salpingo-oophorectomy, and lymphadenectomy is rarely performed. We described a rare uterine neoplasm, low-grade endometrial stromal sarcoma, in a 21 year-old woman diagnosed after myomectomy operation.

Key Words: Sarcoma, endometrial stromal; receptors, estrogen; receptors, progesterone

ÖZET Endometriyal stromal sarkomlar, tüm genital trakt malignitelerinin yaklaşık %0.2 kadarını oluşturan nadir uterin neoplazmlardır. Tümör, mitoz hızlarına dayanılarak düşük-dereceli endometriyal stromal sarkom (LGESS) ve yüksek-dereceli endometriyal stromal sarkom (HGESS) olarak sınıflanır. LGESS'nin prognozu HGESS içinde bulunduğu diğer sarkomlardan daha iyidir. Endometriyal stromal sarkom tanısı genellikle başlangıçta leiomyom gibi benign durumlar olarak düşünülen hastaların histerektomi materyallerinde postoperatif olarak konulur. Önerilen tedavi total histerektomi ve bilateral salpingooferektomidir ve nadiren lenfadenektomi uygulanır. Biz; 21 yaşındaki bir kadında, miyomektomi operasyonu sonrası tanı alan nadir bir uterin neoplazm, düşük-dereceli endometriyal stromal sarkom vakasını tanımladık.

Anahtar Kelimeler: Sarkom, endometriyal stromal; reseptörler, östrojen; reseptörler, progesteron

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Endometrial stromal tumors are rare uterine neoplasms, accounting for approximately 0.2% of all genital tract malignancies.¹ Although their incidence is approximately 2 per million women, the incidence of endometrial cancer is 700 per million women.² It is composed of cells closely resembling normal proliferative endometrial stromal cells. The tumor is classified into a low-grade endometrial stromal sarcoma (LGESS) and high-grade endometrial stromal sarcoma (HGESS) based on mitotic rate. Nuclear atypia or pleomorphism is not demonstrated in LGESS which has less than 10 mitoses per 10 high-power fields (HPF). HGESS is characterized by more than 10 mitoses per 10 HPF and shows a more aggressive be-

havior with frequent metastasis and poor prognosis.³

LGEES shows a strong expression for estrogen receptors (ER+) and/or progesterone receptors (PgR+). The prognosis of LGEES is good and much better than other uterine sarcomas, including HGEES.⁴

Endometrial stromal sarcoma is often diagnosed postoperatively on hysterectomy specimens taken from patients initially thought to have a benign condition such as leiomyoma.⁵ The recommended treatment is total hysterectomy with bilateral salpingo-oophorectomy, and lymphadenectomy is rarely performed.⁶ Up to present no prospective studies have investigated the merits of adjuvant radiation and/or chemotherapy or hormonal treatment following resection of ESS.⁴

CASE REPORT

A 21-year-old female who was gravida 2, para 1, abortion 1 woman presented with menometrorrhagia and abdominal distention that had lasted for 6 months. A transvaginal-ultrasound examination was performed to the patient and it showed a sharply demarcated uterine mass of 7.4 x 7.7 cm diameters that originated from the posterior wall of uterus. It was thought to be a leiomyoma (Figure 1). The complete blood cell count data were as follows: hemoglobin, 10.6 g/dL; white blood cell count, 7.600/mm³; platelet count, 222.000/mm³. Other examinations, including urinalysis, liver



FIGURE 1: Ultrasonographic view of a sharply demarcated uterine mass which is 7.4 x 7.7 cm diameters that originated from the posterior wall of uterus.

function tests and renal function tests, revealed no abnormalities. Chest radiogram showed no abnormality. The data from the tumor marker studies are as follows: cancer antigen (CA)-125, 32.4 IU/mL; CA 19-9, 29.7 IU/mL, CA 15.3 12.7 IU/mL; carcinoembryonic antigen (CEA) 1.7 ng/mL and alpha-fetoprotein (AFP), 9.8 IU/mL.

SURGICAL APPROACH AND PATHOLOGICAL FINDINGS

The patient underwent myomectomy operation for leiomyoma. Intraoperative frozen-section was performed because the appearance of the myoma was unusual. Histopathological findings of the frozen-section was suspicious because of densely cellular proliferation punctuated by frequent vascular spaces. Operation was limited with myomectomy because of the patient's young age and until detailed histological results were completed.

In paraffine section of the specimen, macroscopically, a fleshy, solid mass was seen which was 11 x 6 x 5 cm and 9 x 6 x 3 cm in diameters and histologically, the neoplasm infiltrated to the myometrium as sheets and irregular nodules (Figure 2). The neoplastic cells were polygonal and elongated with mildly pleomorphic, hyperchromatic nuclei and moderate amount of eosinophilic cytoplasm (Figure 3). Per ten high power fields, 3-4 mitotic figures and foci of necrosis were seen in the mitotically active areas of the tumour. The immunohistochemical study showed that CD 10 and vimentin were positive, desmin was focally positive, keratin and actin were negative, and estrogen and progesterone receptors were strongly positive (Figure 4).

The result of the histopathological examining concluded with low grade endometrial stromal sarcoma. On the basis of these findings, we performed an exploratory laparotomy. The surgical procedure consisted of total abdominal hysterectomy and bilateral salpingo-oophorectomy, pelvic lymph-node dissection, an appendectomy, an omentum biopsy and aspiration of abdominal fluid for cytologic evaluation.

After the second operation, pathological examination was performed again. Uterus was measured as 9 x 4 x 3 cms. Serosal irregular area in the

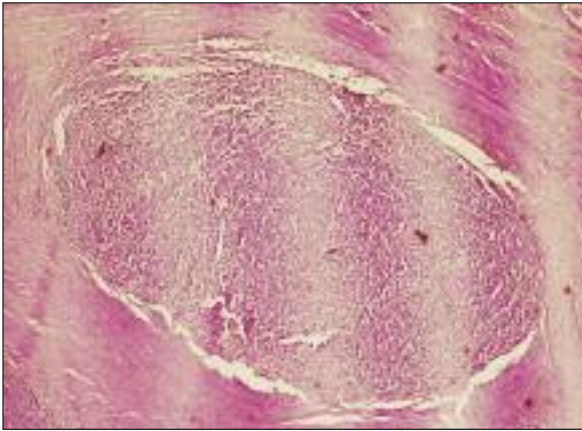


FIGURE 2: Tumoral infiltration of myometrium with low grade endometrial stromal sarcomal cells as sheets and nodules (H&E x10 HPF).

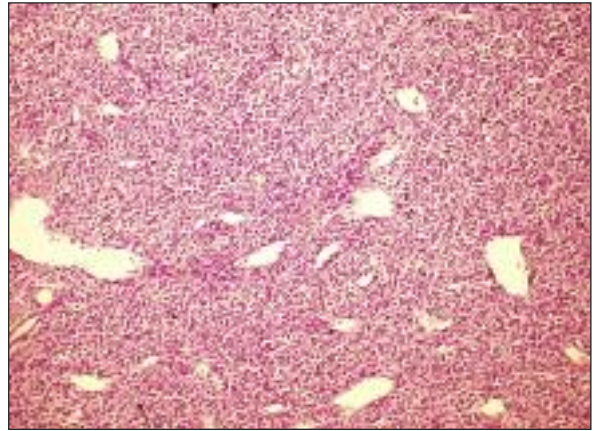


FIGURE 3: The view of low grade endometrial stromal sarcomal cells (H&E x100 HPF).

fundal region was seen and multiple biopsies were taken from that region. Only in one foci in the myometrium which measured 0.8 mm, LGESS was detected and ovaries and twenty lymph nodes which taken from external, internal iliac and obturator region were free of tumour infiltration.

Postoperative recovery was uncomplicated and she had been disease-free for the last three years in the follow-up period. Expectant management was most suitable follow-up option for this patient. In the follow-up period the patient visited the hospital for tumor marker studies, complete blood cell count, urinalysis, liver function tests and renal function tests and chest X-ray every three

months and for computerized tomography every six months.

DISCUSSION

Uterine sarcomas comprise a rare group of cancers that mostly affect postmenopausal women. Common presenting symptoms are abnormal bleeding and abdominal pain.⁷ Because of the rarity of this tumor, neither preoperative diagnostic procedures nor standard therapy have yet been established.⁸

This patient was only 21-years of age at the time of diagnosis. Although there are a few reported cases within this age group, this case represents an unusual presentation of a LGESS.

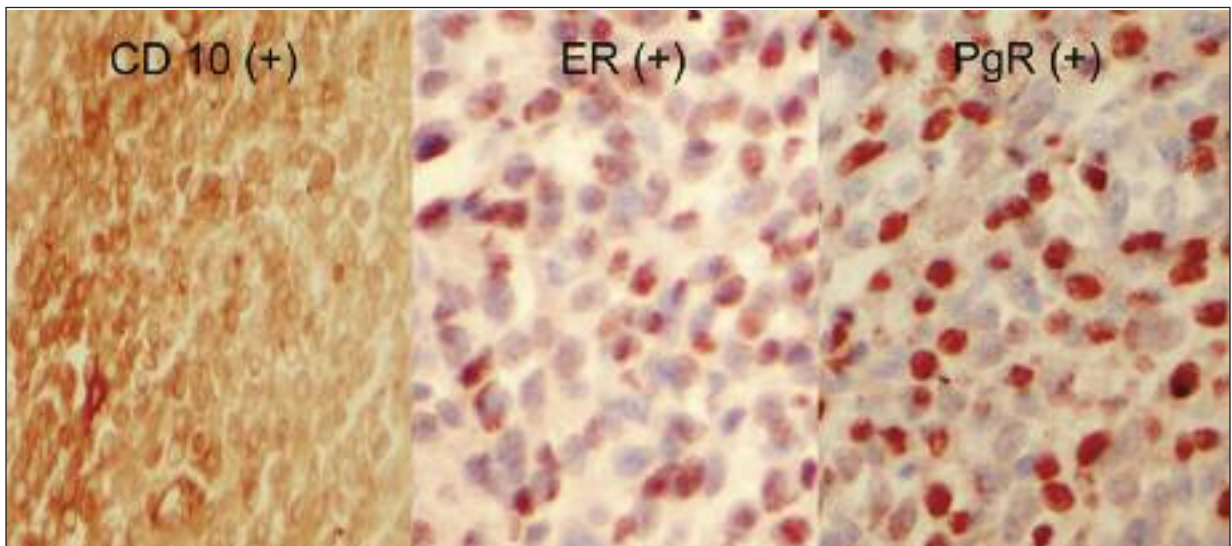


FIGURE 4: The immunohistochemical study showed that CD 10 were positive, estrogen and progesterone receptors were strongly positive.

Surgery has always been described as the most effective treatment in uterine sarcomas.^{9,10} Taking away of all tumor mass seems to optimize the chance of survival for patients with ESS. Removal of the ovaries is recommended for these low-grade tumors because they are characterized by high levels of estrogen and progesterone receptors and often respond to hormonal therapy. In one series, patients with LGESS and retained ovaries had a recurrence rate of 100% (6/6), but the recurrence rate was 43% for the patients who had oophorectomies at the initial surgery.¹¹ We performed hysterectomy and bilateral salpingoopherectomy after treatment modalities were discussed with patient who had no desire to preserve her fertility and high recurrence rates of this tumour.

Adjuvant treatment of ESS is controversial. One study reported, no difference in recurrence rates in patients who had surgery with adjuvant therapy versus surgery alone.¹² Huang et al. found that chemotherapy did not decrease the progression of ESS.¹³ Postoperative radiotherapy has been shown to reduce central tumour but not distal recurrence and has not improved overall survival.^{12,14}

After extensive surgical procedures had been performed to the patient the expectant management was the most suitable option for this patients' follow up.

There is no firm evidence from a prospective study that adjuvant chemotherapy or radiation therapy is substantial benefit for patients with uterine sarcoma. Postoperative pelvic radiotherapy reduces local recurrence but has not been consistently shown to prolong survival.¹⁵

Several clinical observations link sex steroids as growth factors in LGESS. It is postulated that progestins may be tumourostic by interfering with the stability, availability or turnover of the estrogen receptor complex or by turning off specific estrogen responsive genes. Additional research and clinical experience, however, are needed to define the exact role of hormonal therapy for ESS.¹⁶

As a result; LGESS could be detected coincidentally after myomectomy operation in younger patients and the surgical treatment should be individualized according to the grade and invasion of myometrium by tumour.

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