Case Report

Endometrial Stromal Sarcoma, a Rare Gynecological Neoplasm

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Abstract

Endometrial stromal sarcoma is an uncommon gynecological malignancy. It is difficult to diagnose preoperatively and mostly diagnosis is confirmed after surgery on histopathology. This is the case report of endometrial stromal sarcoma infiltrating both ovaries that was diagnosed during stage II b. A 44year old female presented with complaint of irregular heavy vaginal bleeding and history of loss of weight, appetite and altered bowel habits for last 6 months in the outpatient department of University of Lahore Teaching Hospital in January, 2020. Clinical diagnosis of Leiomyosarcoma was suspected but on histopathology endometrial stromal sarcoma infiltrating both ovaries was confirmed.

Key words: endometrial stromal sarcoma, uterine malignancy, low grade and high grade.

Introduction

Uterine sarcomas are exceptional neoplasms that can arise from connective tissue of the endometrium or smooth muscles of the myometrium. Worldwide incidence is 1-2/100,000 in general population. It comprises only 2% in patients presenting with uterine neoplasm. Endometrial stromal sarcoma (ESSs) are very uncommon malignant tumors.[1,2,3] Overall incidence is 0.2% in all uterine malignancies and 10% in uterine sarcoma.[4,5,6]

Patient Information and Clinical Presentation

A 44-year-old mother of 4 children with no history of abortion (P4A0) and all normal vaginal deliveries presented with complaint of heavy vaginal bleeding since 3 days in the outpatient department of University of Lahore Teaching Hospital in January, 2020. Detailed history taking revealed that the bleeding had been irregular accompanied by weight loss and decrease in appetite for last 6 months.

Diagnostic Assessment and Treatment

On general examination she was weak, emaciated and extremely pale. Per abdominal examination revealed a mass in lower abdomen, firm to hard in consistency with restricted mobility, consistent with 18-20 weeks of gestation. On vaginal examination, fullness in pouch of Douglas was felt. On rectal examination, the mass was compressing rectum but rectal mucosa was mobile. Investigations showed hemoglobin of 7 mg/dl, anti HCV positive but her liver function tests and renal function tests were normal. Ultrasound pelvis showed 18 cm. mass in uterine cavity most probably fibroid uterus, with bilateral ovarian cysts 5 cm on left side and 2 cm on right side. The tumor marker CA 125 level was normal in serum. CT scan report showed 18x20 cm sized uterine mass with poor demarcation between endometrium and myometrium, left sided 5-6 cm cysts with solid component. Liver, spleen and kidney were unremarkable without any signs of enlargement or lymphadenopathy. Anemia was corrected with blood transfusion and staging laparotomy planned. Intraoperatively gut was adherent with left side of uterus. The surgeon performed total abdominal hysterectomy with unilateral (right sided) oophorectomy. Left sided cyst was posteriorly placed and removed in piece meal. Peritoneal washing and omental biopsy was also taken. Specimen was sent for histopathology which showed endometrial stromal sarcoma infiltrating both ovaries with normal cytology and normal omental biopsy.

Outcome

After 10 days of post-operative stay in the hospital, oncologist was consulted who referred her to Inmol hospital, Lahore for pelvic radiation. Unfortunately she never returned for follow up.

Figure 1: Cut sections of endometrial stromal cell sarcoma

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Discussion

World health organization (WHO) recently classified endometrial stromal cell sarcoma (ESS) into 4 categories[3,4] which are as follows:

A. Endometrial stromal nodule (ESN),

B. Low grade endometrial stromal sarcoma (LGESS),

C. High grade endometrial stromal sarcoma (HGESS)

D. Undifferentiated endometrial stromal sarcoma (UESS)

Endometrial stromal nodule (ESN) is mostly localized, only involving endometrium and not infiltrating myometrium. On the other hand endometrial stromal sarcoma (ESS) infiltrate myometrium and histologically it is composed of uniform small cells similar to endometrial stroma in proliferative stage. The mean age group is 45-50 year. Etiology is still unclear, but tamoxifen therapy, poly cystic ovarian syndrome and unopposed estrogen can be the risks factors for endometrial stromal sarcoma.[3,7] Deletion of chromosome 7p was found in 55.6% cases of ESS and supposed to be a risk factor for development and progression of tumour.[3,8]

Patients with endometrial stromal cell sarcoma mostly present with abnormal uterine bleeding, pelvic mass, pelvic pain and dysmenorrhea. 25% of cases of ESS are asymptomatic. 30-50 % of patients have extra uterine metastasis at the time of diagnosis.[3,9] Preoperative diagnosis is difficult, even USG and MRI are inconclusive and may be misdiagnosed as Fibroid or Adenomyosis. Endometrial sampling may be helpful in those cases in which endometrium is involved.[3,10,11] Its differential diagnosis includes adenomyosis, leiomyoma, endometrial polyp and low grade Mullerian adenosarcoma. Estrogen and progesterone receptors are frequently present in ESS and the tumor marker CD10 may be helpful to differentiate it from other pathological conditions.[8] Primary treatment is surgical that includes staging laparotomy followed by total abdominal hysterectomy, bilateral salphingoooophorectomy, peritoneal washings, omental biopsy and para aortic lymph node sampling. In some cases, surgery is the main source of diagnosis as well.[3,12]

As recurrence is common, therefore, many clinicians advocate adjuvant therapy in the form of chemotherapy, radiotherapy and hormone therapy (progesterone, GnRH analogue and aromatase inhibitors) to suppress tumor. Above mentioned therapies are primary treatments for stage II-IV and unresectable tumors. Prognosis factors are still controversial and includes age, parity, race, menopausal status, tumor size, stage, atypical appearance of the nucleus, abnormal mitotic activity, tumor necrosis, lymph vascular invasion, DNA ploidy, proliferative activity and hormone receptors expression.[3,13,14] The 2018 International Federation of Gynecology and Obstetrics (FIGO) uterine cervical cancer staging system has been applied to ESS.15

Five year survival rate for ESS stage I is 54-100%, stage II 30%, stage III and IV is about 11%.[3,4] As late recurrence is commonly observed, therefore, it needs long term follow up. Follow up schedule is 3 monthly for 1st year, 6 monthly for next 4 years then annually thereafter. The relapse free survival depends on staging, myometrial infilteration, adjuvant treatment and bilateral salphingo-oophorectomy.[3,16]

References

1. Eamudomkarn N, Itarat Y, KleebKaow P and Kiet Peerra Kol. A case report of high grade Endometrial sarcoma: A Rare cause of abnormal uterine bleeding in a young woman.case report obstet Gynecol 2018, Nov.28.doi 10.1155/2018/5906760

2. Sato S, Ojima Y,Kanda M,Kizki t,Ohara N. Endometrial stromal sarcoma arising from endometrial polyp. A case report, Kobel J Med Sci 2018 Sep 11;64(2):E36-E42

3. Jabeen S, Anwar S, Fatima N. Endometrial stromal sarcoma: A Rare Entity.J.Coll.physician surgical Pak.2015 March; 25(3):216-7.doi:03.2015/JCPSP.216 217.

4. Geetha P,Krishnan NM.Endometrial stromal sarcoma: A review of the literature. Indian J Med Paediatr Oncol 2012; 33:1-6.

5. Jain R,Batra S,Ahmad A,Elahi AA,Gupta M,Saith P,Low grade endometrial stromal sarcoma: A case report. Iran Med J Med Sci.2015 Jan, 40(1):81-4.

6. Puliyah G, Nair VR and Singh. Endometrial stromal sarcoma. Indian J Med Paediatr Oncol 2010 Jan-Mar; 31(1):21-23.

7. Cohen I, Endometrial pathologies associated with postmenopausal tamoxifen treatment: Gynecol Oncol 2004; 2:256-66.

8. Halbwed I, Ullmann R, Kremser ML, Man YG, Moud NI, Lax S, ET al.Chromosomal alteration in low grade endometrial stromal sarcoma and undifferentiated endometrial sarcoma as detected by comparative genomic hybridization.Gynecol Oncol 2005.2:582-7.

9. Tavassoli FA,Devila P,WHO classification of pathology and genetics of tumors of breast and female genital organs:Tavassoli FA,Devilee P,editors,lyon,France:IARC Press;2003.P.233-6.ss

10. Ganjoei TA, Behtash N, and Shariat M, Low AM.Grade endometrial stromal sarcoma of uterine corpus: A clinicopathological and survey study in 14 cases. World J surg Oncol 2006.4:50.

11. Jin y,Pan L,Wang X,Dai Z,Hnang H,Guol,et alClinical characteristics of endometrial stromal sarcoma from an academic medical hospital in china:Int J Gynecol cancer 2010;116:131-9.

12. Somoye G, Lawton H, Havenga S.Endometrial stromal sarcoma: experience from district hospital and literature review.Eur J Gynecol Oncol, 2009; 3010):664-7.

13. NCCN clinical practice guidelines in oncology. Soft tissue sarcoma, Version2 National Comprehensive CancerNetwork, Inc; 2011.

14. Weitmann HD,Knocke TM,Kucera H,Potter R.Radiation therapy in the treatment of stromal sarcoma,Int J Radiat Oncol Biol Phys 2001;41:739-48.

15. D'Angelo E, Part Uterine sarcoma: A review.Gynecol Oncol 2010, 116:131-9.

16. Bodner K,Adler BB,Obermann A,Windbichler G,Petrn E,-Mayerhofer S,et al.Prognostic parameters in endometrial stromal sarcoma: A clinic pathological study in 31 patients.Gynecol Oncol 2001;81:160-5.