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Case Report

Endometrial Stromal Sarcoma Presenting As Puberty Menorrhagia

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Abstract:

Endometrial stromal sarcomas are rare uterine tumours usually seen in perimenopausal females. We report here a case of low grade malignant endometrial stromal sarcoma in an adolescent girl, presenting as puberty menorrhagia. She underwent total hysterectomy with bilateral salpingo-oophorectomy and pelvic node sampling. She also received adjuvant chemotherapy and radiotherapy. She is disease free at completion of one year of follow-up.
Key Words: Endometrial stromal sarcoma, puberty menorrhagia, sarcoma uterus

Introduction

Endometrial stromal sarcomas (ESS) are rare neoplasms, comprising approximately 0.2% of all uterine malignancies.[1] The tumour is composed of cells resembling normal endometrial stroma. Endometrial stromal tumours are divided into three types on the basis of mitotic activity, vascular invasion and observed differences prognosis. The endometrial stromal nodule is a lesion confined to the uterus with pushing margins, less than three mitosis per ten high power field and no lymphatic or vascular spread. The prognosis of this disease is usually good and there are no reported recurrences or deaths following surgical removal of the tumour. Low grade ESS is defined as an infiltrative stromal tumour with less than ten mitosis per ten high

power field which frequently extends into and grows within large vascular spaces. It has a five year survival rate of 100%.[2] High grade ESS is characterized by more than ten mitosis per ten high power field. It is a highly lethal neoplasm with a aggressive clinical course and a five year survival of 55%.[2] ESS occur primarily in the perimenopausal age group, between 45 and 50 years with about one-third being in post menopausal age group.[3,4,5] We are presenting a case of a 17 year old girl who was investigated for puberty menorrhagia and was found to have low-grade ESS.

Case Report

A 17 year old unmarried girl presented with menorrhagia of six months duration. Her past history revealed no significant childhood diseases and no previous operations performed. Menarche had occurred at the age of twelve and her periods were regular. For the last six months she has been having heavy bleeding lasting for 10-12 days and occurring at three weeks interval and not associated with dysmenorrhoea. Physical examination revealed a pale girl with normal secondary sexual characteristics. Rectal examination showed the uterus to be bulky. Pelvic ultrasound showed the uterus to be enlarged with the endometrial cavity filled with a tumour. Hysteroscopy was performed and it showed a friable, necrotic tumour in the upper left lateral uterine wall. Biopsy was taken from the tumour and the report was endometrial stromal sarcoma. Investigations for distant metastasis were negative. She underwent hysterectomy with bilateral salpingo oopherectomy and bilateral pelvic lymphnode sampling on May 2003. The uterus was found to be enlarged to eight weeks size with normal serosa.

The pathologic report revealed the following: Uterus measured 9x5x3 cms. Cut section showed the endometrial cavity filled with a polypoid fleshy growth with areas of hemorrhage. Tumour was infiltrating more than half of myometrial thickness and extending approximately 3-4 mm from the serosa. Histologically the neoplasm was seen involving the endometrium and infiltrating the myometrium in sheets and irregular nodules. The neoplastic cells were polygonal and elongated with pleomorphic, hyperchromatic nuclei and moderate amount of eosnophilic cytoplasm. About 6-8 mitotic figures were seen per ten high power fields in the mitotically active areas of the tumour. Foci of necrosis were also seen. The immunohistochemical study showed Vimentin positive, smooth muscle actin (SMA) positive and Desmin negative. The conclusion was that it was ESS of low grade malignancy. Ovaries and lymphnodes were free of tumour infiltration.

Post operative period was uneventful. She received adjuvant pelvic radiotherapy with 45 Gy in 23 fractions followed by chemotherapy with four courses of Adriamycin 40mg/m^2 and Cyclophosphomide 70 mg/m^2. She is now disease free and has completed one year of follow up.

Discussion

ESS is most frequently encountered in 40 to 50 year age group but is extremely rare in adolescents.[6,7] According to Bellone et al[7] only 5 cases of ESS in adolescent patients has been published. According to Larsen et al[3] the mean age for low grade ESS is 47.2 years and for high grade ESS is 50.2 years.

The main symptom of ESS is menometrorrhagea and postmenopausal bleeding Diagnosis of such cases especially during adolescence is difficult because even if a mass is palpated, the physician may confuse it with a ovarian tumour or hematometra which are more common. Pelvic ultrasound and computerised tomography are helpful in diagnosis.

Treatment of ESS is surgical[8] and regardless of patients age, preservation of ovarian tissue is not recommended because of likelihood of ovarian metastasis. In addition since ESS has steroid receptors, the possibility exits that estrogen production by

retained ovaries may stimulate any residual disease, oopherectomy is recommened.[9]

Combined therapy with surgery and radiotherapy decreases chance of local recurrences and improves survival.[8] The propensity for hematogenous spread in ESS makeschemotherapy also an attractive treatment modality.[10]

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