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Low grade endometrial Stromal sarcoma with microscopic limited Infiltration: A rare case report.

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Abstract: EST (endometrial stromal tumors) are the second most common pure mesenchymal tumors of the uterus even though they account for less than 10 % of all such tumors. Endometrial stromal nodule (ESN) and Low grade endometrial stromal sarcoma (ESS) falls in the lower end of spectrum and undifferentiated endometrial stromal sarcoma (UES) falls in other malignant end of spectrum. ESN's are very rare and low grade ESS accounts for only 0.2% of all uterine malignancies. Here we describe a case of Low grade ESS with limited infiltration which has been recently described in literature; purpose of which is to propose to practicing surgeons and gynaecologists that although these tumors behave in benign fashion however, these patients should be carefully followed than the usual low grade ESS as for possible chances of late metastasis.

Keywords: Endometrial stromal tumors, limited infiltration, rare

INTRODUCTION

EST (endometrial stromal tumors) are rare cancers arising predominantly in the endometrium. These are the second most common pure mesenchymal tumors of the uterus. According to 2003 WHO classification EST includes Endometrial stromal nodule (ESN), Low grade endometrial stromal sarcoma (ESS) and undifferentiated endometrial stromal sarcoma (UES) [1]. The common differential diagnosis of EST is from other mesenchymal tumors showing smooth muscle differentiation [2,3] so, a panel of CD 10 and any two smooth muscle markers are usually used for diagnosing EST [4].

The exact diagnosis of EST carries importance because in addition to its rarity some of these tumors have a very different and sinister prognosis. Here we report and discuss a case of Low grade ESS with limited infiltration describing its morphological and immunohistochemical features in context to other mesenchymal tumors of endometrial.

CASE REPORT

A 40 years- old multigravida presented with irregular bleeding per vaginum, for 4 to 5 months along with dysmenorrhoea. Her general physical condition was satisfactory vaginal examination showed anteverted bulky uterus. Ultrasonography showed a submucosal

growth with? Fibroid/polyp measuring 4.0 cm x 3.5 cm in size. Routine lab investigations were within normal range except for mild anemia. Special investigations like CT / MRI were not done in the patient. Patient underwent panhysterectomy and specimen sent for histopathological examination. Grossly, the specimen measured 12.0 cm x 7.5 cm x 5.5 cm in size. Cut surface of cervix, tubes and ovaries were unremarkable. The cut section of uterus showed a well circumscribed greyish yellow soft submucosal polypoidal growth arising from endometrium and partially filling the endometrial cavity. No myometrial invasion was seen.

Microscopically, tumor was cellular and composed of sheets of uniform looking small to medium sized blue cells resembling the proliferative endometrial stroma (fig 1). Mild cellular atypia and occasional mitotic figures were seen. Focal irregularities of tumor cells in form of finger like projections varying 3 to 6 in number into adjacent myometrium was seen. No vascular invasion was seen. Immunohistochemistry for CD 10 was positive and for smooth muscle cell desmin, vimentin were negative which ruled out the possibility of smooth muscle tumor. Patient was followed up for eighteen months after which contact with the patient was lost. Patient was asymptomatic during the follow up period.

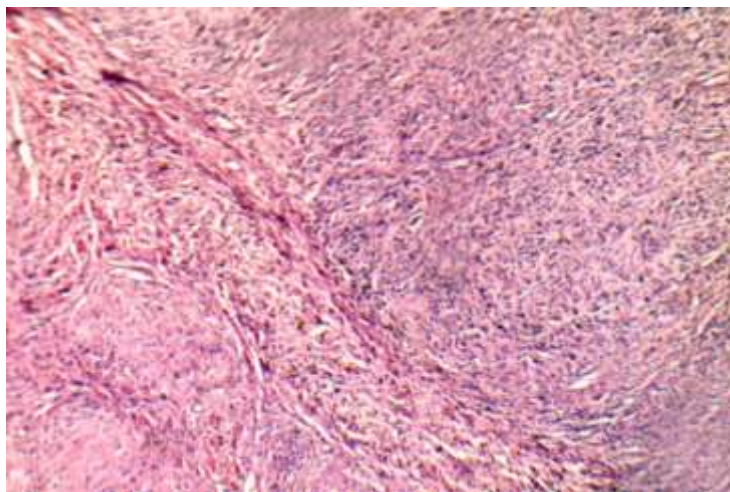


Fig-1: Section showing tumor composed of sheets and nodules of uniform looking small to medium sized blue cells resembling the proliferative endometrial stroma. (400X, Hematoxylin & Eosin)

DISCUSSION

ESTs (endometrial stromal tumors) of the uterus accounts for less than 10% of all uterine mesenchymal tumors. According to 2003 WHO classification they are divided into Endometrial stromal nodule [ESN], Low grade ESS [Endometrial stromal sarcoma] and [UES] undifferentiated ESS [1]. ESN are those tumors which are well circumscribed tumoral proliferation of uniform cells having non – infiltrative, expansile border [2]. Low grade ESS with limited infiltration has rather been described recently in the literature. Low grade ESS with limited infiltration are those lesions which are grossly indistinguishable from ESN but microscopically that do not fulfill the criteria for an ESN and does not have the overt permeative growth of low grade ESS or associated vascular invasion [2]. In our case 3 to 6 irregular tongues and detached nodules from the main mass ranging from 3 to 5 mm in size were seen with no deep myometrial or vasular invasion. The UES are those tumors which are pure sarcomas arising within endometrium but shows no evidence of endometrial stromal differentiation. Vaginal bleeding is the common clinical presentation in most of the patients [1,2]. The common differential diagnosis includes cellular leiomyoma. ESTs are positive for CD 10 and negative for smooth muscle cells [3]. So, the most common panel used for diagnosing EST includes CD 10 and two smooth muscle markers [4]. Grossly, tumor circumscription with well defined non infiltrative expansile border is the main differentiating feature between ESN and UES / ESS [4]. Extrauterine spread at the time of diagnosis is found in upto one third of the patients with low grade ESS and rarely this tumor may initially be present at an extrauterine site, most commonly the ovary [5]. Microscopically, ESN and ESS are typically composed of diffuse growth of small blue cells with scant cytoplasm and oval to spindle nuclei that resemble the stromal cells of proliferative phase of endometrium [1,2]. Focal irregularities in form of lobulated or finger

like projections into the adjacent myometrium that are not more 3 mm in size and are not more than 3 in number may be seen [2,4,5]; Myometrial and vascular invasion are the two most important features seen in UES and low grade ESS [2,6]. Total abdominal hysterectomy with bilateral salpingo-oophorectomy is the treatment of choice in these cases however, some patients may be benefited with adjunct progestational agents [4]. Owing to limited experience and unpredictable behaviour of these tumors, as studies with long follow –up are scant; for practical purpose it is currently best to report these tumors as low grade ESS with explanatory note describing that this tumor is not as overtly invasive as a typical low grade ESS and for this reason it may behave in a more benign fashion however the patient should be followed than the usual low grade ESS as for possible chances of late metastasis.. By emphasizing this point to all practising surgeons and gynaecologists, we aim to report this case.

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