Case Report

Low Grade Endometrial Stromal Sarcoma in 40 years old female - A rare case report

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Abstract

Endometrial uterine sarcoma is a very rare tumour of the uterine cavity with an incidence of 1-2 cases per 100,000 women. Low grade Endometrial stromal sarcoma (LGESS) is an occasional diagnosis in a patient presenting as leiomyoma uterus. The symptoms are nonspecific, mostly abnormal uterine bleeding in perimenopausal women. Clinically and radiologically it is difficult to diagnose this entity. Histopathological examination and immunohistochemistry confirmed the diagnosis of LGESS. In addition of surgery chemotherapy, radiotherapy and immunotherapy treatments will be quite useful in all cases of LGESS. This case report of Low grade Endometrial stromal sarcoma (LGESS) is presented here because of its rarity.

Key words

Low grade Endometrial stromal sarcoma, Uterine bleeding, Uterine cavity tumor.

Introduction

Low grade Endometrial stromal sarcoma (LGESS) is a rare type of endometrial cancer [1] that is mainly present in older women. ESS is an occasional diagnosis in a patient presenting as leiomyoma uterus. The symptoms are nonspecific, mostly abnormal uterine bleeding in perimenopausal women. In comparison with HGESS, the age group of LGESS is usually younger (45-55 years). An early diagnosis is essential because patient survival is directly related to tumor stage. The usual preoperative diagnosis of LGESS is difficult to make by radiological investigations and definitive diagnosis is achieved only after histopathology of uterus. We reported a case of low-grade ESS in a 40-year-old woman, presenting as irregular and excessive per vaginal bleeding since 4 months. In addition of surgery chemotherapy, radiotherapy and immunotherapy treatments will

be quite useful in all cases of LGESS. Here in we just want to stress rarity of LGESS and we are able to find and document the typical features of LGESS.

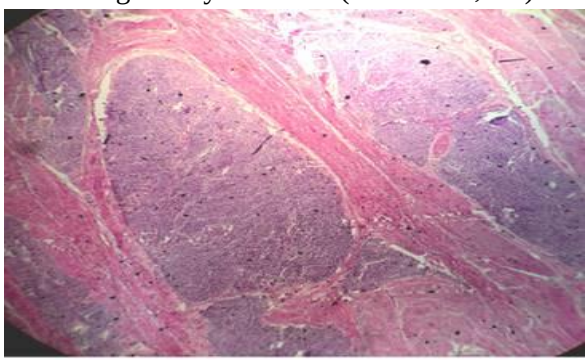
Case report

A 40-year-old female came to the Outdoor patient Department of Obstetrics and Gynaecology Department of Dhiraj General Hospital and Research Institute due to complaint of the irregular and excessive per vaginal bleeding since 4 months. All the haematological, biochemical and serological examinations were normal. Ultrasonography (USG) was done and an impression of leiomyoma was made. Hysterectomy with bilateral salpingo-oophorectomy was done and the specimen was sent for histopathological examination in the pathology department. On gross examination, hysterectomy specimen comprising of uterus with cervix and bilateral adnexa measured 13x7x4.6 cm. On cut section poorly demarcated lesion with multiple solid masses and multiple cystic areas measured 5.2 x4cm seen on endometrial wall. There was also presence of tumor nodules infiltrating the myometrium (**Photograph - 1**).

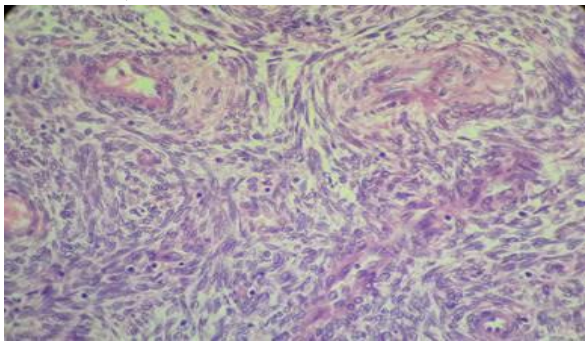
Photograph – 1: Poorly demarcated uterine mass lesion with multiple solid and cystic areas.

On microscopic examination, hematoxylin and eosin stained sections demonstrated that the irregular nests and islands of blue cells with a prominent delicate vascular network diffusely infiltrate the myometrium. In the myomertrium there was presence of tongue like infiltration of tumor cells. There was presence of abnormal mitotic figure but they were <5 / 10 hp field. The tumor cells were closely resembles endometrial stromal cells (**Photograph - 2, 3**). Immunohistochemistry demonstrated that the tumour cells were immunopositive for CD 10 and Vimentin (**Photograph - 4, 5**). Based on the histopathological and immunohistochemical features, a diagnosis of Low grade Endometrial stromal sarcoma (LGESS) was made.

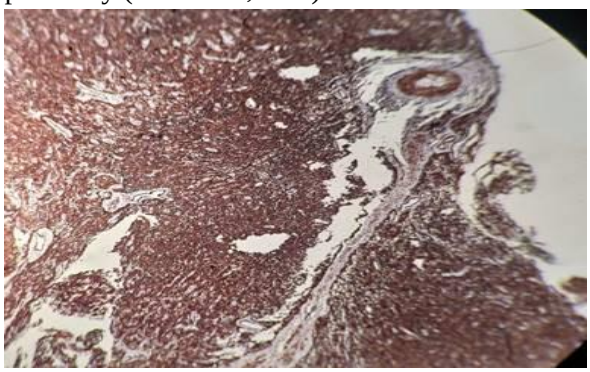
Photograph – 2: Islands of blue cells diffusely infiltrating the myometrium (H&E stain, 4X).



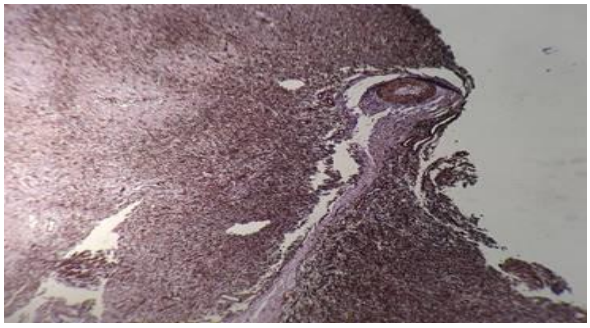
Photograph – 3: The tumor cells which were closely resemble endometrial stromal cells (H&E stain, 40X).



Photograph – 4: Tumor cells with strong CD 10 positivity (IHC stain, 20X).



Photograph – 5: Tumor cells with strong Vimentin positivity (IHC stain, 10X).



Discussion

Endometrial uterine sarcoma is a very rare tumour of the uterine cavity with an incidence of 1-2 cases per 100,000 women. The site of origin may be connective tissue, smooth muscle, or endometrial stromal. The latter (endometrial stromal sarcoma, ESS) is still rarer tumors that made up approximately 10% of all uterine sarcomas [1].

Most of the patients of the LGESS presented at the age of 45-55 years. Our patient presented at 40 years, which is a rarity in itself. The presenting features are similar to uterine leiomyoma, abnormal vaginal bleeding, abdominal pain, pressure caused by an enlarging pelvic mass and some patients may be asymptomatic [2]. Our patient was presented with irregular and excessive per vaginal bleeding since 4 months.In most of the cases radiological investigations like Ultrasound and magnetic resonance imaging are not useful for the diagnosis of LGESS.

The pathogenesis of these tumors is yet not known. Probable risk factors are past exposure to pelvic radiation therapy, long-term tamoxifen use, and unopposed estrogen use. However, Halbwed, et al. [3] study shown a strong correlation between chromosomal deletion on 7p and tumor development.

In the 2014, WHO has classified endometrial stromal tumors into Endometrial stromal nodule (ESN), Low grade endometrial stromal sarcoma, High grade endometrial stromal sarcoma, and undifferentiated endometrial sarcoma (UES). The histopathology reveals uniform small cells bearing resemblance to the proliferative stage endometrial stroma. In the myomertrium there will be presence of tongue like infiltration of tumor cells. From histopathological examination the differential diagnosis could be cellular leiomyoma, adenomyosis in post-menopausal age group and leiomyosarcoma. The immunohistochemical markers such as h-caldesmon and CD 10 may solve the diagnostic problem as CD 10 staining is positive in ESS but not in leiomyoma. We had performed CD 10 and vimentin staining to make the diagnosis. In our case also the USG and clinical diagnosis was leiomyoma but histopathological examination made diagnosis of Low grade endometrial stromal sarcoma with the help of IHC. In support to our case there was another rare presentation of LGESS was as a low grade endometrial sarcoma of endocervix presenting as a soft hemorrhagic mass in the posterior cervix looking like a degenerated leiomyoma [4].

The definitive management of LGESS is surgical, both to establish the diagnosis as well as for treatment. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and excision of all grossly detectable tumor is the commonest surgical approach for the treatment of LGESS [5]. Since the recurrence rate is very high with this type of tumor, it is essential to have a very thorough follow-up. It shall be once in 3months for the first year, and half yearly for next 4 years. Thereafter, annual follow-up is recommended. The relapse free survival depends on the tumor stage, myometrial invasion, adjuvant therapy, and bilateral salpingo-ophorectomy.

Conclusion

LGESS is a rare malignant tumor and the recurrence rate is very high. As radiological investigations are not a reliable way to diagnose endometrial sarcoma, histopathological examination with IHC stain remains gold standard method for final confirmation and by

timely diagnosing this entity we can definitely improve patients' survival.

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