International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: 6.614
Available Online at www.journalijcar.org
Volume 9; Issue 01 (C); January 2020; Page No.21040-21042
DOI: http://dx.doi.org/10.24327/ijcar.2020.21042.4124

Review Article

LOW GRADE ENDOMETRIAL STROMAL SARCOMA: A CASE REPORT

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A R T I C L E I N F O

Article History:
Received 4th October, 2019
Received in revised form 25th November, 2019
Accepted 18th December, 2019
Published online 28th January, 2020

Key words:
Endometrial stromal sarcoma, Low grade, Tumor

A B S T R A C T

Low-grade endometrial stromal sarcoma (LGESS) is a rare malignant tumor affecting younger women presenting as abnormal uterine bleeding in peri menopausal women. Based on the mitotic state, ESS is histologically divided into two groups: high grade (HG) and low grade (LG). LGESS is relatively more common and tends to occur before menopause. LGESS exhibits a more indolent course, but has high relapse potential. For LGESS, hysterectomy is the cornerstone of treatment, however, the role of bilateral salpingo-oophorectomy (BSO), as well as lymphadenectomy including hormonal treatment, chemotherapy and radiotherapy is debated. Here we are reporting a case of low grade stromal sarcoma. By reporting our case, we wish to stress the necessity for a high grade suspicion to diagnose this tumor even in younger women. A prompt diagnosis and timely intervention are keys to improve patient survival.

INTRODUCTION

Low-grade endometrial stromal sarcoma (LGESS), also known as endolymphatic stromal myolysis, is a rare tumor, accounting for just 0.2% of malignant uterine Tumors and approximately 20% of uterine sarcomas.[1-3] The annual incidence of endometrial stromal sarcoma (ESS) is 1-2 per million women. Compared to other uterine malignancies, ESS affects younger women and the mean age is 42-58 years. Based on the mitotic state, ESS is histologically divided into two groups: high grade (HG) and low grade (LG).[5] HGESS is currently defined as an undifferentiated endometrial sarcoma (UES) characterized by more than 10 mitosis per 10 high power fields (HPFs). Additionally, this sarcoma is more aggressive and has a poor prognosis. In contrast, LGESS has fewer than 10 mitosis per 10 HPFs and the cell nuclei are not atypical or pleomorphic.[5] LGESS is relatively more common and tends to occur before menopause. LGESS exhibits a more indolent course, but has high relapse potential.[6] For LGESS, hysterectomy is the cornerstone of treatment, however, the role of bilateral salpingo- oophorectomy (BSO), as well as lymphadenectomy for complete surgical staging, is debated. Adjuvant treatment including hormonal treatment, chemotherapy and radiotherapy.[7] we are reporting a case of low grade stromal sarcoma

Case Summary

A 42 years old female (para4 living4) was referred to our hospital as cervical carcinoma with complaints of menorrhagia for the last 1 years, discharge per vaginal and mild lower abdominal pain for last 8 month. Her menstrual cycles had been normal 1 year back when she developed menorrhagia. She had periods at an interval of twenty two days and bleeding lasting for eight to ten days. Flow was excessive with history of passage of clots. She was not using any contraception.

Physical examination showed the patient was moderately built and nourished. The rest of the general vitals were stable and systemic examination was unremarkable. On per abdominal examination, abdomen was soft non tender, no organomegaly present. Per vaginal speculum examination, a large hanging type shaggy mass about 4 inches in diameter was seen coming out through cervical os, bleeding and fowl smelling discharge present. On vaginal examination, a firm proliferative growth, tubular type of about 4x4 cm in size present over the anterior and posterior cervical lip with growth filling vagina in its upper part. All fornices were free. Finger stained with blood. Per rectal examination shows rectal mucosa and bilateral parametrium free. As per routine biopsy was taken from the suspicious growth and sent for histiopathological examination which reported to be suspicious for low grade endometrial stromal sarcoma. For confirmation a rebiopsy was done under anaesthesia in operation theatre. The re-biopsy report confirmed the diagnosis of low grade endometrial stromal sarcoma.

CECT shows gross ballooning of the cervix with extremely thin wall by a heterogeneous area, measuring 7.4cms (Transverse) x 7.1cms(craniocaudal) x 5.2cms (Anterior-posterior) with approximately volume of 137cc. Post contrast images showed cranially it is extending into endometrial

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cavity, caudally into upper part of vagina, posteriorly causing mild compression over rectum and anteriorly indenting the urinary bladder. Biopsy from growth shows low grade endometrial stromal sarcoma. Radical abdominal hysterectomy with bilateral salpingo-oophorectomy with pelvic lymph node dissection was performed. Cut section showed a sessile polypoidal growth of 2.5x2cm arising from fundus and protruding into endometrial cavity. Cut surface of rest of uterus, cervix, bilateral ovaries and tubes was unremarkable. All dissected pelvic lymph nodes are free from tumour. Histopathological report confirmed the diagnosis of Low grade endometrial stromal sarcoma. According to the new 2018 FIGO Staging, it was stage IA disease (pT1aN0). Immuno-histochemical stains were not done in our case because of economic limitations.

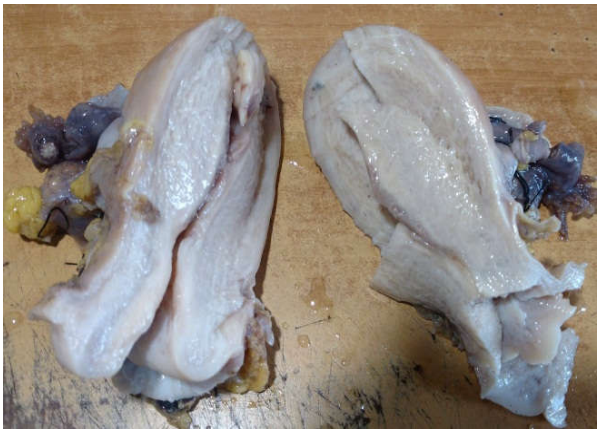


Figure 1-2 showing gross and histopathological view of the resected tissue

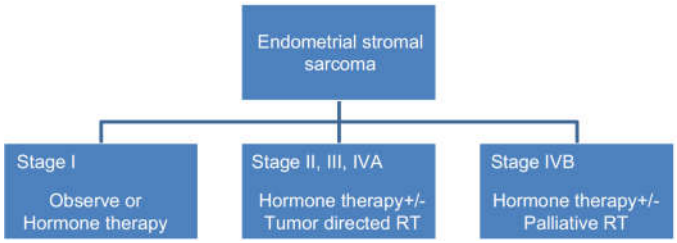
DISCUSSION

Uterine sarcomas are rare tumours of mesodermal origin. They constitute 2 to 6% of uterine malignancies. Of these, endometrial stromal sarcomas are very rare. They are divided into three types depending upon mitotic activity, vascular invasion and observed differences in prognosis.[8]

1. Endometrial stromal nodule(ESN)
2. Lowgrade endometrial stromal sarcoma(ESS) and
3. High grade or undifferentiated endometrial stromal sarcoma(UES)(8)

Boardman *et al.* differentiated low grade ESS by cellular uniformity, less frequent mitosis (<3/10 HPF versus >10/HPF), and lack of haemorrhage and necrosis.[9] Uterine sarcomas usually affect post-menopausal females. Women with LGESS are younger with a median age being 45 and 57 years. Symptoms at presentation include abnormal vaginal bleeding, menorrhagia and abdominal pain.[10] The tumours have an indolent growth with a tendency for late recurrence.[11] Metastasis are rarely detected before the diagnosis of the primary lesion.[12] Grossly, low grade ESSs involves the endometrium, occasionally extensively. By definition tumours are infiltrative. Rarely, the tumours manifest as polyps, usually with haemorrhage and infarction. Because of its prominent intravascular growth a soft tan to yellow cut surface and appear as cords and nodules infiltrating through the uterine smooth muscle. The histologic features recapitulate the gross appearance with cords of tumour cells infiltrating between smooth muscle and within lymphatic spaces. The neoplastic stromal cells resemble those of the proliferating endometrium, are monotonous in appearance, and have relatively uniform size and shape.[12] Tumour cell nuclei are round to ovoid and have fine chromatin, and small, inconspicuous nucleoli may be seen. A small amount of cytoplasm is present, and cell borders are indistinct. Mitotic activity is usually low (<10/10 HPF). It should be noted that rare causes of low-grade ESS will have a greater number of mitotic figures, although this is not associated with an adverse prognosis.[13] Proliferating small vessels resembling the endometrial spiral arterioles are characteristic, and tumours can have bands of hyaline connective tissue separating islands and clusters of bland neoplastic stromal cells.[14] The differential diagnosis of LGESS includes ESN, cellular leiomyoma, cellular intravenous leiomyomatosis, cellular endometrial polyp and various soft tissue neoplasms.[15]

Surgery is the final resort for primary treatment of LGESS consisting of total abdominal hysterectomy with bilateral salpingo-oophorectomy. Regardless of patient's age, preservation of ovarian tissue because of likely-hood of ovarian metastasis. In addition, since ESS has steroid receptors the possibility exists that estrogen production by retained ovaries may stimulate any residual disease, oophorectomy is recommended. Due to high recurrence risk even with localized tumours, many clinicians advocate use of adjuvant chemotherapy, radiation therapy and/or hormonal therapy to suppress tumour growth.[16]



Hormone therapy with medroxy-progesterone, tamoxifen, gonadotropin releasing hormone analogues(GnRH) and aromatase inhibitors are suggested for LGESS with recurrent disease.[17-18] Uterine sarcomas have a poorer prognosis and survival is much worse than that reported for endometrial adenocarcinoma, with an overall survival of less than 50% at 2 years, even when presenting at an early stage.[18] A higher survival is reported with LGESS as compared to other uterine sarcoma.[10,19]

CONCLUSION

LGESS is a rare malignant tumor, presenting as abnormal uterine bleeding in peri menopausal women. The usual preoperative diagnosis is uterine leiomyoma and definitive diagnosis is achieved only after histopathology of uterus. By

reporting our case, we wish to stress the necessity for a high grade suspicion to diagnose this tumor even in younger women. A prompt diagnosis and timely intervention are keys to improve patient survival.

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How to cite this article:

Dr Mranalini verma and Dr Laxman Pandey (2020) 'Low Grade Endometrial Stromal Sarcoma: A Case Report', *International Journal of Current Advanced Research,* 09(01), pp. 21040-21042. DOI: http://dx.doi.org/10.24327/ijcar.2020. 21042.4124

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