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Case Report

Endometrial Stromal Sarcoma of the Uterus: A Rare Entity

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ABSTRACT

Endometrial stromal Sarcoma (ESS) is a rare disease with probably less than 700 new cases per year. We report a rare case of low grade ESS, presenting with the symptom of abnormal uterine bleeding and diagnosed as fibroid uterus at ultrasonography. Preoperative endometrial aspiration showed proliferation of stromal cells, giving a presumptive diagnosis of a stromal tumor. Gross examination showed a fibroid uterus but it showed low-grade ESS on histopathology examination of the total hysterectomy specimen. Despite the rarity of the tumor, one has to consider the possibility of ESS in cases with presentation of abnormal uterine bleeding with fibroid uterus.

Key words: Endometrial stromal sarcoma, abnormal uterine bleeding

INTRODUCTION

Endometrial Stromal Sarcoma (ESS) is a very rare malignancy that constitutes approximately 10% of all uterine sarcomas but only 0.2% of uterine malignancy. [(1)] The annual incidence of ESS is 1-2 per million population accounting for 400-700 new cases each year. [(2)]

CASE REPORT

A 35yearold woman presented with increased bleeding per vagina during her menstrual periods since six months. Her last child was born 10 years back and there was no history of contraceptive use. An ultrasound scan showed enlargement of the uterus with diffuse coarse heterogeneous myometrial echoes and an intramural fibroid 2 ×2 cms in posterior wall. A dilatation and curettage was performed and microscopy showed endometrium with cystically dilated glands. A focus of endometrium showed a stromal growth pattern with areas of evenly distributed arterioles. A presumptive diagnosis of endometrial stromal tumor was suggested. (Fig 2) Based on histopathology report of D & C, a total abdominal hysterectomy with bilateral salpingooopherectomy hysterectomy was performed. The hysterectomy specimen was sent to the Department of Pathology for histopathological examination.

Grossly, the specimen consisted of uterus, cervix and bilateral adnexa measuring 12 cm

× 8 cm × 5 cm) The cut surface showed an intramural fibroid measuring 2 cms x 2 cms with a yellowish tan.[Figure 1] Microscopically, the H & E sections from the uterus showed proliferating uniform oval and spindle-shaped shaped endometrial cells resembling the proliferative phase of the endometrium with an infiltration of the myometrium showing a distinctive growth pattern as worm like cords or islands of tumor cells placed between smooth muscles. Mitosis was not seen in the sections examined. [Figure 2]. The cervix and ovaries were normal. The patient was referred to a regional cancer centre with the report of low-grade endometrial stromal sarcoma.

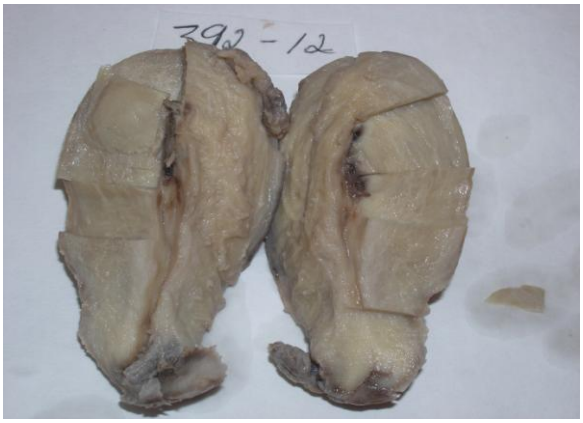

Fig 2: Gross: shows a well circumscribed nodule in the myometrium

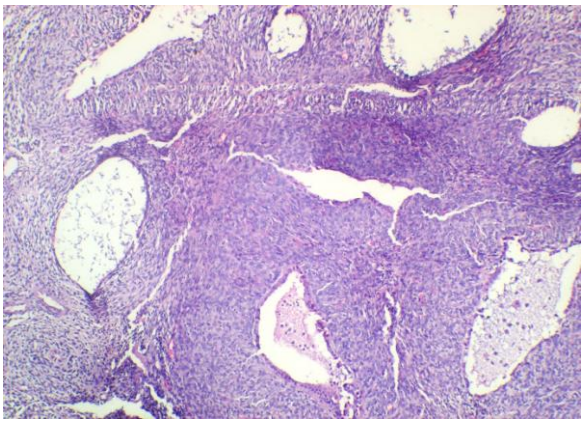
Fig 1:H & E x 400 Proliferation of the stromal cells on endometrial biopsy

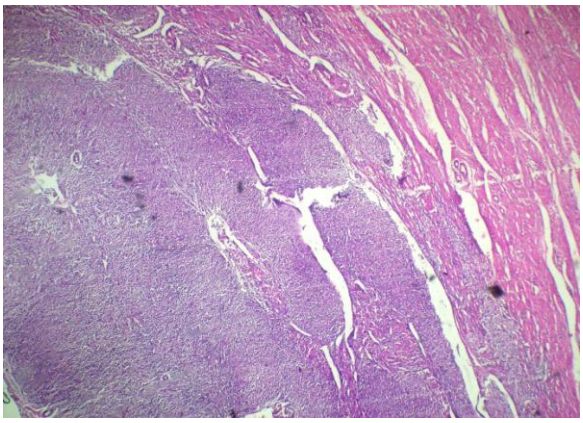
Fig 3a: H & E x 100 Endometrial stromal tumor with infiltrating margin

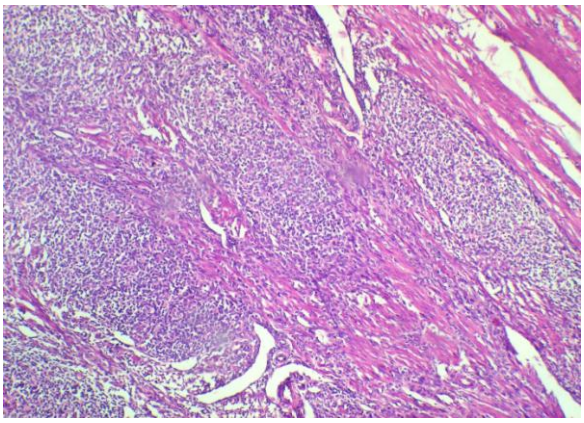
Fig 3b :H & E x 400 LG ESS showing minimal atypia with tongue like protrusions of the stromal tumor into the myometrium

DISCUSSION

Low grade ESS occurs frequently in women in age group of 45 – 55 years with common complaint of abnormal uterine bleeding and abdominal pain. [3] The patient in our case was a young female of 35 years of age who also presented with excessive uterine bleeding. [3] The differential diagnosis considered in this lesion included adenomyosis, Endometrial Stromal Nodule (ESN) and cellular leiomyoma. In our case, the gross appearance was that of solitary, well circumscribed mass indistinguishable from leiomyoma. Histologically, Cellular

leiomyoma are composed of cells having spindle shaped nuclei arranged in fascicular growth pattern with thick muscular walled vessels and cleft like spaces where as in this case, the lesion showed predominately stromal cells of uniform size and shape resembling those of endometrial stroma in the proliferative phase. [4] The nuclei of the stromal cells were round to oval with minimal atypia. The mitotic activity was < 1/10HPF. [3] However, the tumor was seen infiltrating the smooth muscle bundles of the myometrium in a tongue like fashion. ESN are well circumscribed lesion, with a pushing margin and does not infiltrate the myometrium. When there is a difficulty in diagnosing between ESS and cellular leiomyoma, immunoreactivity with antibodies to CD10 and smooth muscle actin and desmin are used. [5] In adenomyosis, beingn endometrial glands with surrounding stroma are seen within the myometrium without any evidence of infiltration.

As per the review of the literature, although the tumor is always intramyometrial, most of the endometrial stromal sarcomas involve the endometrium and so uterine curettage usually helps in the diagnosis. [6] Endometrial biopsy of Dilatation and curettage in our case showed fragments of proliferative endometrium with areas showing highly cellular stromal cells, on which, a possibility of a stromal tumor was suggested.

These tumors are indolent but late reoccurrence and distant metastasis may occur. Recurrence and distant metastasis of low-grade ESS are considered to be related to direct extrauterine development or vessel permeation. [7] Our patient didn't show no any lymphatic vessel permeation and metastasis and is disease free even 6 months later.

Management usually consists of total abdominal hysterectomy and bilateral oophorectomy as adnexal nvolvement is not always evident macroscopically. LGESS is associated with a good prognosis in most early stages (5-year survival rate, 100%), and most require no further therapy after complete surgical resection. [8] Progestin therapy has been reported to reduce the risk of reoccurrence when used in the adjuvant settings. [9]

CONCLUSIONS

Because of the rarity of the tumor, ESS may not be familiar to the gynaecologists. In young patients presenting with abnormal uterine bleeding, it can be mistaken for a fibroid. As diagnosis of ESS clinically as well as radiographically are inconclusive, the diagnosis of low grade endometrial stromal tumour may be established on morphology alone on a hysterectomy specimen. However the authors like to emphasize that a possibility of a stromal tumor can be suggested on endometrial biopsy as in this case and thus helping the gynaecologist towards further management.

REFERENCES

1. Boardman CH, Webb MJ, Jefferies JA. Low grade endometrial stromal sarcoma of the ectocervix after therapy for breast cancer. Gynaecologic Oncology 2000, 79: 120-123.
2. Pink D et al. Harm or treatment of hormonal treatment in metastatic low grade endometrial stromal sarcoma: single centre experience with 10 cases and review of literature. Gynaecol Oncol 2006, 101: 464-469.
3. Christopher D.M. Fletcher. Diagnostic Histopathology of Tumors, 3rd edition, Churchill Livingstone Elsevier, 2007; pages 667-668.

4. Koyama T, Togashi K, Konishi I, Kobayashi H, Ueda H, Kataoka ML, et al. MR imaging of endometrial stromal sarcoma: Correlation with pathologic findings. AJR Am J Roentgenol. 1999;173:767–72.
5. Stadvold JL et al. Case report, Conservative treatment of myxoid endometrial stromal sarcoma in a 16 year old nulliparous woman. Gynaec Oncol 2005, 99: 243-245.
6. Chang KL, Crabtree GS, Lim-tan SK, et al: Primary extrauterine endometrial stromal neoplasms. a clinicopathologic study of 20 cases and a review of the literature. Int Gynecol Pathol 1993; 12:282-296
7. Norris HJ, Tayler HB: Mesenchymal tumor of the uterus. a clinical and pathological study of 53 endometrial stromal sarcomas. Cancer 1966; 19:755-766
8. Feeley KM, Burton JL, Wells M. A diagnostic approach to endometrial biopsies: selected topics. Current Diagnostic Pathlogy 2000;6: 13-20.
9. Haseeiny GE, Bareedy NA, Mourad WA et al. Prognostic factors and treatment modalities in uterine sarcomas. Am J Clin Oncol 2002, 25: 256-260.

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