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Case Report

A CASE OF MALIGNANT LOW GRADE ENDOMETRIAL STROMAL SARCOMA AND REVIEW OF THE LITERATURE

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ABSTRACT

Low grade endometrial stromal sarcoma is a rare pelvic malignancy that arises from the endometrium. This article describes the morphological features of one such tumour discovered as finding in a hysterectomy specimen of a 32 year lady with a clinical diagnosis of dysfunctional uterine bleeding with multiple fibroids. Morphological and immunohistochemical evaluations were performed and a final diagnosis of low grade endometrial stromal sarcoma was given. This report is aimed to present a case of endometrial stromal tumor because of its rare existence and difficulties in establishing histological diagnosis.

Keywords: uterine sarcoma, low grade endometrial stromal sarcoma

INTRODUCTION

Uterine mesenchymal tumors pose many problems to the surgical pathologist in prediction of their biological behavior, i.e. whether benign, low malignant potential or frankly malignant. Differentiation of endometrial stromal and smooth muscle tumors can be done in most instances by routine light microscopic examination, has not been a subject of discussion in the literature. However, highly cellular leiomyomas can be misinterpreted as endometrial stromal tumors and vice versa. The morphological features of one such lesion, a low grade endometrial stromal sarcoma is presented.

Endometrial stromal sarcomas (ESS) are rare neoplasms, comprising approximately 0.2% of all uterine malignancies. [1] The tumors are 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 in prognosis. The endometrial stromal nodule is a lesion confined to the uterus, with pushing margins, less than three mitosis per ten high power fields and absence of lymphatic or vascular spread. The disease usually has good prognosis with no reported recurrences or deaths following surgical removal of the tumor. Low grade ESS is defined as infiltrative stromal tumor show less than ten mitosis per ten high power fields, frequently extending into and growing within large vascular spaces. It has a five year survival rate of 100%. [2] High grades ESS is characterized by more than ten mitosis per ten high power fields. 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]

CASE REPORT

A 32 year old woman presented with menorrhagia and dysmenorrhoea of four months duration. She underwent hysterectomy for bulky uterus with suspicion of multiple fibroids from clinical examination.

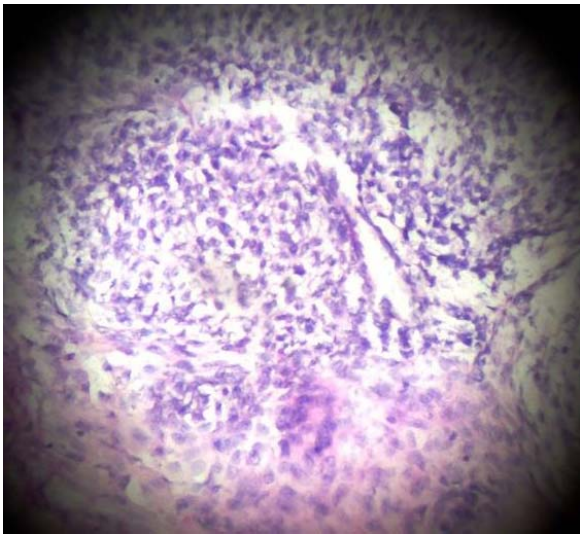

Fig 1: H & E stain. (400x)

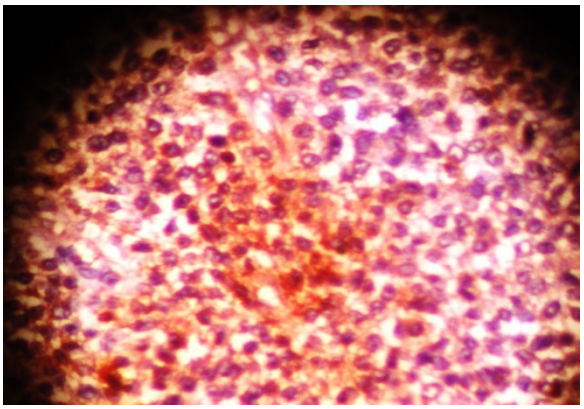

Fig.2 CD 10 positive. (DAB chromogen, 400x)

On gross examination the uterus with cervix measured 13x10x6 cms. Cut section showed the endometrial cavity filled with multiple nodular fleshy growths with areas of hemorrhage, largest nodule measuring 3x3cms. Tumor was infiltrating more than half of myometrial thickness and extending approximately 3-4 mm from the serosa. Histologically, the sections from various parts of nodule show densely cellular tumor made up of round/oval/oblong uniform cells (resembling endometrial stromal cells) with scanty cytoplasm. (fig.1) Cells are arranged in diffuse sheets, at places intersecting and anastomosing cords arranged around spiral arteriole. 6-8 mitotic figures were seen per ten high power fields in the mitotically active areas of the tumor. Irregular shaped, tongue shaped, and circumscribed nests of tumor cells are seen infiltrating the bundles of myometrium. Foci of necrosis and hemorrhage were also seen. The immunohistochemical study showed CD 10 positive (Fig.2), CD34 negative, EMA negative, CK negative, ER (Fig.3) and PR (Fig.4) positive. The conclusion was that it was ESS of low grade malignancy.

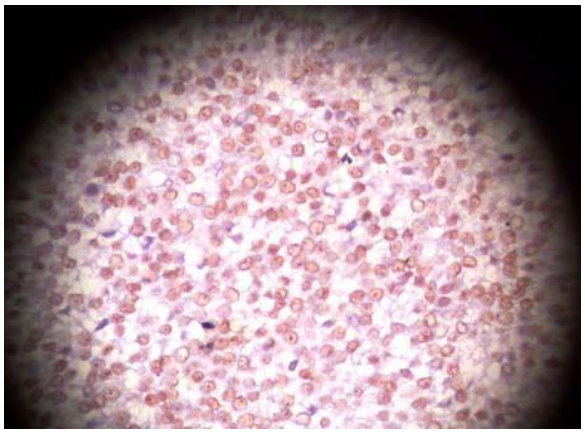

Fig.3 ER positive (DAB chromogen, 400x)

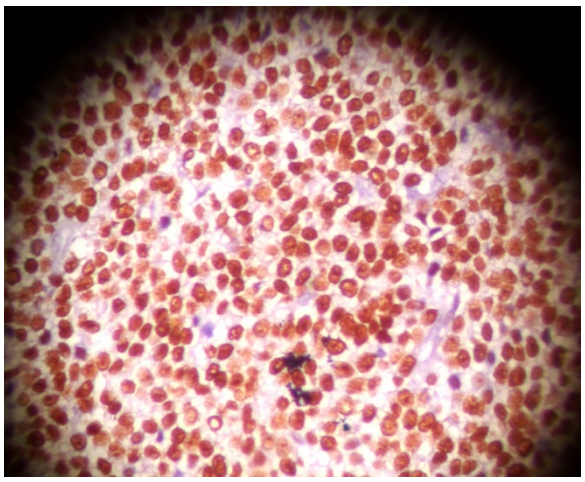

Fig. 4 PR positive (DAB chromogen, 400x)

DISCUSSION

Endometrial sarcoma constitutes 15 to 25% of uterine sarcomas. These tumors are seen most commonly in older postmenopausal women. However women in the reproductive ages may be affected. [6] LGESS have typically a mitotic count of 5 and less than 10 hpf with minimal atypia in the cells. Some tumors may be positive for estrogen and progesterone receptors which may affect treatment modalities.[7] Patients with LGESS typically present with abnormal vaginal bleeding, pelvic and abdominal pain. In some cases it might be without any complaints. [8] Most tumors grow through the intramural sections of the uterus rather than intra cavitary, hence

making it difficult for preoperative histopathology diagnosis.

Patients most commonly undergo surgery with the presumptive diagnosis of uterine fibroid or pelvic mass. Olive et al. emphasized the presence of large thick-walled muscular vessels as a feature that serves to distinguish a highly cellular leiomyomas from a stromal proliferation. [10] Immunohistochemistry is helpful to differentiate both neoplasms. The addition of new immunohistochemical markers such as h-caldesmon and CD10 may solve the diagnostic problems. CD10, expressed by lymphoid cell precursors, is a cell-surface neutral endo peptidase and it stains endometrial stroma in the uterus but not glands. Strong and diffuse CD10 staining was observed in ESN and LGESS whereas most leiomyomas were negative. Between EST and leiomyomas, correct classification is important due to the differences in clinical behavior and treatment.

LGESS usually behaves in an indolent clinical fashion; however recurrences and distant metastases can occur. Prolonged survival as well as cure is common despite the development of recurrent or metastatic disease. [5]

The mitotic count is an important independent prognostic factor for these tumors. [5] In addition, it has been suggested that early tumor stage, low myometrial invasion, and low mitotic count are associated with a lengthened overall survival in patients with endometrial stromal sarcomas. [9]

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