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Case Report

Endometrial Stromal Sarcoma- Case Report

Dr. Ujwala Maheshwari[1], Dr. Mithila K.B[2], Dr. Priyanka Lad[2], Dr. Kalyani M[2]

[1]Professor, [2]Senior Resident,
Department of Pathology, MGM Medical College, Navi Mumbai, India

Corresponding Author: Dr. Mithila K.B

ABSTRACT

Endometrial stromal sarcoma [ESS] is a rare malignant tumour of the endometrium. Usually occurs in perimenopausal women between the age group of 40-50 years. Patients come with symptoms similar to uterine leiomyoma, abnormal vaginal bleeding or abdominal pain, some patients may be asymptomatic. There is a high chance of it to be mistaken for leiomyoma clinically. Although rare, ESS should be kept in mind in women presenting with bulky uterus. A definitive diagnosis can only be made on histopathology. Surgery and postoperative radiotherapy is the most effective treatment.

Key words: Endometrial stromal sarcoma [ESS], malignant tumour, endometrium.

INTRODUCTION

Sarcoma is a very rare malignancy of uterus, with an incidence of 1-2 cases per 100,000 women. [1] Origin of it may be connective tissue, smooth muscle or endometrial stroma. The latter (endometrial stromal sarcoma, ESS) is still rarer tumor that makes up approximately 10% of all uterine sarcomas. [2,4] Depending on mitotic activity, vascular invasion or prognosis, there are three categories of endometrial stromal tumors: endometrial stromal nodule, low-grade endometrial stromal sarcoma, high-grade endometrial stromal sarcoma. [3]

CASE

A 49 year old female came to gynaecology out patient department complaining of abnormal vaginal bleeding and pain in abdomen. Ultrasonography report showed a cervical polyp measuring 2x1cm and endometrial thickness of 3.5 cm. Endometrial curettage was send for histopathology.

Grossly- multiple, red-brown soft tissue bits measuring 3.5cm was received along with a polypoidal mass measuring 2 x 0.8cm.

Microscopy

Cervical polyp- section studied showed a polypoidal structure lined by stratified squamous epithelium on three sides. Underlying stroma showed foamy cells and myxomatous cells with mild inflammatory infiltrate.

Endometrium- section studied showed endometrial glands and stroma. Glands were round to oval, distorted at places and lined by single layer of columnar epithelium. The stroma showed atypical monomorphic oval small cells encircling small blood vessels resembling spiral arterioles. Foci of hyalinization and scattered foamy cells were also appreciated. Further immunohisto-chemical markers were also done to confirm the diagnosis of Low grade endometrial stromal sarcoma.

CD10 - Positive
Reticulin - Positive
Vimentin- Positive

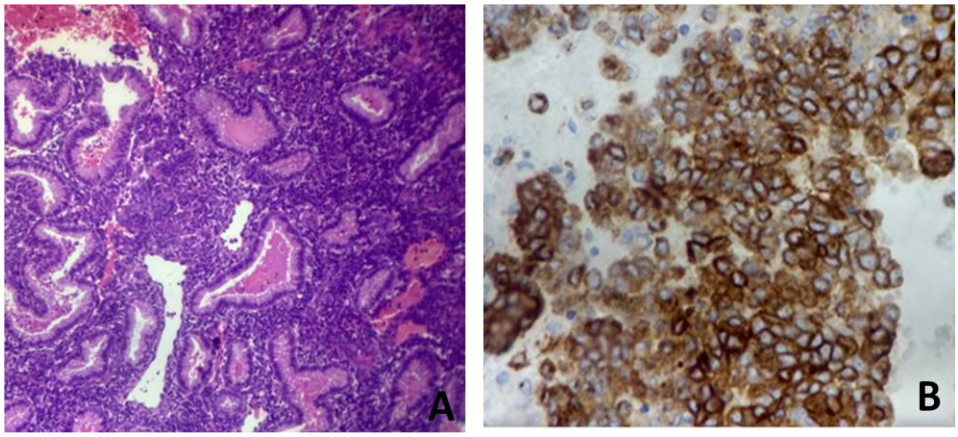

Fig A- endometrial glands are round to convulated lined by single layered columnar epithelium.
Fig B- tumour cells showing CD 10 positivity

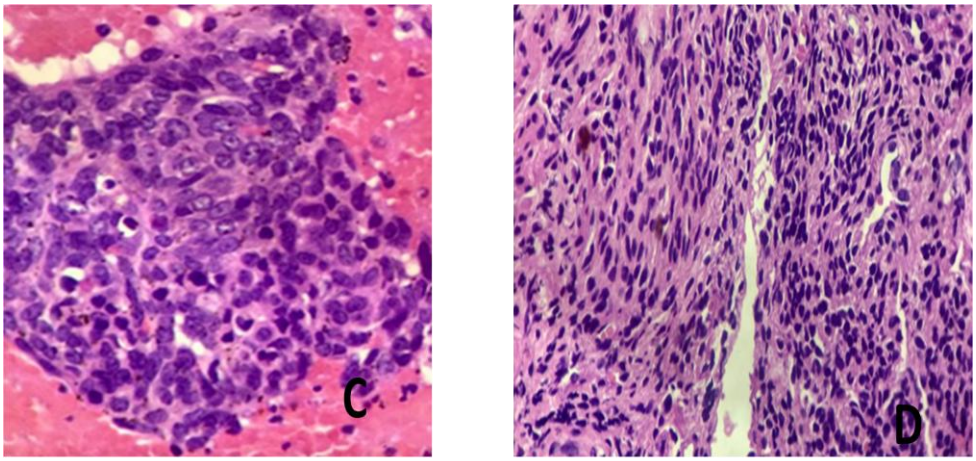

Fig C - showing marked nuclear atypia, nuclear enlargement and nuclear pleomorphism (H&E,40X)
Fig D - showing stroma atypical and monomorphic oval small cells (H&E,40X)

DISCUSSION

Endometrial stromal sarcoma usually affects women of postmenopausal age groups (45-57 years). [1] Usually patients present with abnormal vaginal bleeding, progressive menorrhagia, and abdominal pain. It can be mistaken for leiomyoma clinically making it difficult to diagnose, hence diagnosis is usually made on postoperative histopathological examination.

Grossly it is usually seen as a single nodule or multiple solid-cystic masses, and a poorly demarcated lesion with occasional cystic degeneration or rarely cystic multilocular lesion is seen. The pathogenesis of ESS remains unknown, but exposure to tamoxifen and unopposed estrogens has been implicated in some cases. [2] An early diagnosis is essential because patient survival is directly related to tumor stage. Although indolent in nature, ESS is malignant and can spread to the vagina, fallopian tubes, ovaries, bladder and ureters. Metastasis to lung, heart and to other sites can also occur. [3]

Histologically it is very difficult to differentiate ESS from leiomyoma. In these cases, immunohistochemistry helps in final diagnosis. Markers such as h-caldesmon and CD 10 are used. CD 10 shows strong positivity for ESS and not leiomyoma.

The primary line of management is surgery - total abdominal hysterectomy, bilateral salpingo-oophorectomy and excision of all grossly detectable tumour.

Surgery has always been described as the most effective treatment. Postoperative radiotherapy or progesterone is effective in control of reoccurrence of uterine sarcoma. Reoccurrence is usually seen 50% of cases.

CONCLUSION

Although rare, a clinical diagnosis of endometrial stromal tumours should be kept in mind, especially when a patient presents with a bulky uterus and symptoms similar to leiomyoma. [3] Definitive diagnosis is only achieved on histopathology. A prompt diagnosis and timely intervention are keys to improve patient survival. [1]

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