AN INTERESTING CASE OF ENDOMETRIAL STROMAL SARCOMA CLINICALLY MIMICKING FIBROID UTERUS

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ABSTRACT

The diagnosis of uterine sarcoma in women undergoing hysterectomy with a clinical diagnosis of fibroid uterus is very low. Uterine sarcomas are clinically indistinguishable from leiomyomas since both typically present with abnormal uterine bleeding, pelvic pain and a pelvic mass. A high degree of suspicion is essential to diagnose these tumours. Endometrial Stromal Sarcomas(ESS) are rare, constitute 10% of all uterine sarcomas and 0.2% of all uterine malignancies. The mean age of presentation is 42 to 58 years. Uterine sarcomas are most commonly diagnosed following myomectomy or hysterectomy. In a premenopausal woman with bleeding disproportionate to size of uterus and significant pain, sarcoma is suspected. We report a case of ESS in a 42 year old woman with fibroid uterus following hysterectomy.

Key words: *Endometrial Stromal Sarcomas, Hysterectomy, Leiomyomas.*

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INTRODUCTION

The diagnosis of uterine sarcoma in women undergoing hysterectomy with a clinical diagnosis of fibroid uterus is 0.2 to 0.5 percent.[1] Uterine neoplasms with mesenchymal differentiation includes leiomyoma, leiomyosaromas and endometrial stromal tuomors. Endometrial Stromal tumors are rare and can be benign or malignant. Endometrial Stromal Sarcomas(ESS) constitute 10% of all uterine sarcomas and 0.2% of all uterine malignancies.[2] The International Society of Gynecologic Pathologists and the World Health Organization, classifies uterine sarcomas as purely nonepithelial or mixed epithelial-nonepithelial type. The usual clinical presentations are vaginal bleeding, pelvic pressure symptoms (eg,pressure, urinary frequency, constipation), enlarged uterus, or abdominal distension. Distinguishing leiomyoma and sarcoma in the preoperative period is difficult. Only rare cellular variants of leiomyoma progress to sarcoma. Diagnostic modalities like Ultrasound examination, magnetic resonance imaging, computed tomography, or positron emission tomography cannot reliably distinguish between a sarcoma and leiomyoma, endometrial cancer, lymphoma, intravenous leiomyomatosis, or adenomyosis. Endometrial sampling will yield the correct diagnosis in some, but not all patients.[3] We report this case to highlight the fact that even clinically innocent looking lesions may turn out to be malignant on histopathology.

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CASE REPORT

A 42 year old lady presented to gynaecology department with complaints of dysmenorrhoea of increasing intensity for the past 6 months. She essentially did not have menstrual complaints except dysmenorrhoea. Per abdomen examination showed a uterine mass of 16 weeks size. Speculum examination showed a healthy cervix and pap smear was taken. On pelvic examination she had a 16 weeks size uterus. Baseline investigations were done and were within normal limits. Transvaginal ultrasonogram showed a 9.7 x 6.9 x 8.0 cm intramural fibroid in anterior right lateral wall and endometrial thickness was 8.5 mm (Fig.1). Pap smear was negative for intraepithelial lesion or malignancy. Endometrial

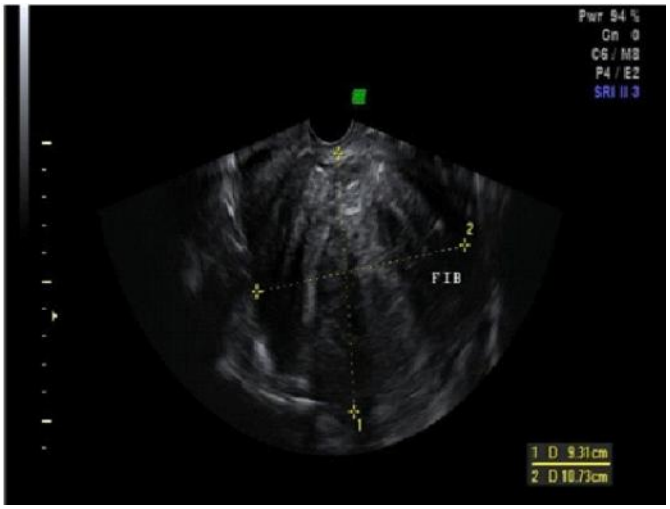

Fig-1 TVS showing anterior wall intramural fibroid 9.3x10.7cm.

aspiration showed disordered proliferative pattern. With a provisional diagnosis of fibroid uterus total abdominal hysterectomy (TAH) with ovarian conservation was planned.

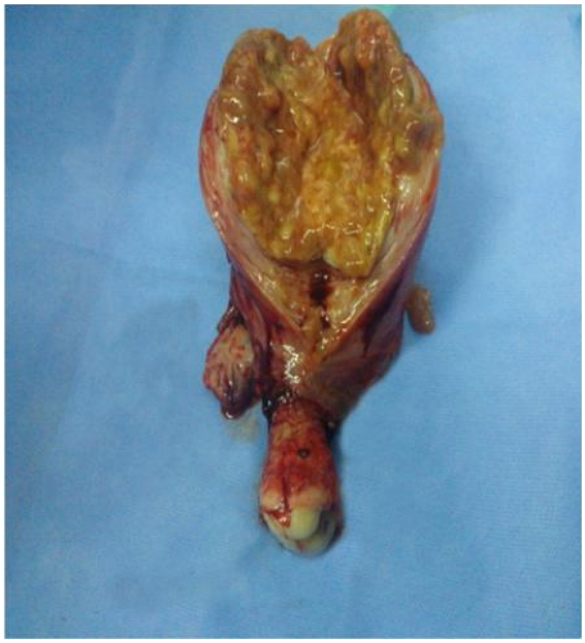

Fig. 2: Picture showing cut section of uterus with the mass and right adnexa.

Intraoperatively TAH with right salpingo oopherectomy was done as right adnexa was adherent to uterus and left ovary was conserved. (Fig.2). On cut section fibroid with degenerative changes was observed and didnot arouse a suspicion for frozen section. Histopathological examination of the specimen showed a tumor composed of small round to oval cells with uniform size and shape(Fig. 3). There were only 3 mitosis/10 HPF with no nuclear atypia or necrosis. With these features a diagnosis of Endometrial stromal sarcoma -low grade was given. Full thickness of myometrium was involved. By immunohistochemistry Estrogen Receptor and

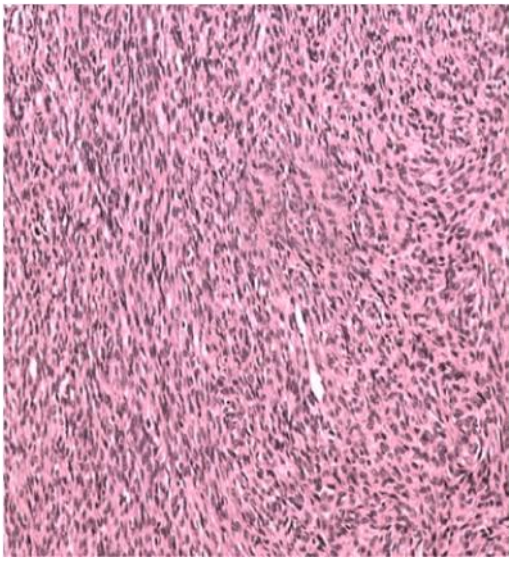

Fig.3: Endometrial stromal sarcoma -Photomicrograph showing the tumour with small round to spindle cells(H &E X 100)

Progestrone Receptor was positive. The positivity of these markers can be used as a targeted therapy. Also C-Kit (CD 117) was done to rule out a GIST (Gastro intestinal Stromal Tumor).

Medical oncologist opinion was obtained. Computerised tomography plain and contrast was done which showed a post hysterectomy status ,left ovary

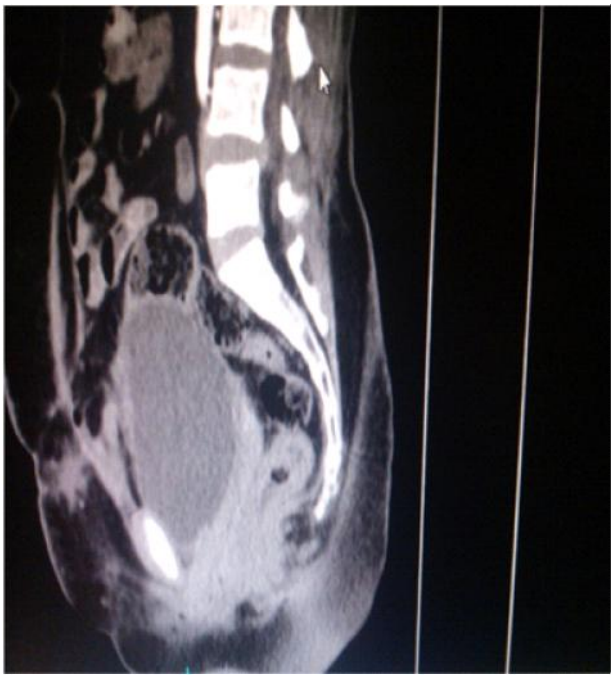

Fig. 4 : Computerised tomography of abdomen and pelvis showing no evidence of extra uterine spread in the post operative period.

4.2x2.4x4.3 cms and no significant lymph node enlargement and no evidence of extrauterine spread (Fig. 4) Oncologist advised revision surgery to remove left ovary and for pelvic lymphadenectomy. In the event of no surgery, to decide for chemoradiation. The patient was administered 6 cycles of chemotherapy with Paclitaxel, Adriamycin and Cisplatin. External Radiation Therapy was given to deliver a dose of 50.4 Gy in 28 fractions. Patient had a good response and is on a regular follow up.

DISCUSSION

The American College of Obstetricians and Gynecologists advises that there is a risk of uterine sarcoma in women taking tamoxifen.[4] Post menopausal status , pelvic irradiation and a childhood history of Retinoblastoma are other risk factors.[5] The overall survival rate at 10 years is 65 to 76 percent. Uterine sarcomas are referred to as homologous or heterologous. The majority are homologous and they differentiate in ways similar to normal uterine tissues.It includes those arising from endometrium (ESS-endometrial stromal sarcomas), muscle (leiomyosarcoma), or sarcomas of nonspecific supporting tissue (eg, connective tissue, blood vessels, lymphatics). Heterologous tumors contain elements with non-native differentiation like skeletal muscle, cartilage, bone. The World Health Organization classifies endometrial stromal tumors into three categories: endometrial stromal nodule (ESN), endometrial stromal

sarcoma (ESS), and undifferentiated endometrial sarcoma (UES). ESN is a benign entity that can be cured with simple hysterectomy. Endometrial stromal sarcoma - ESS is a low-grade sarcoma with metastatic potential. They exhibit myometrial and/or vascular invasion[6] ESS is characterised by distinctive finger-like projections that invade the myometrium, veins, and lymphatics. Histologically, they are characterized by densely uniform stromal cells with minimal cellular pleomorphism, mild nuclear atypia, and rare mitotic figures. Low-grade ESSs have less-frequent mitosis (<3 per 10 high-power fields) and they do not show hemorrhage and necrosis. They are immunoreactive for the estrogen and progesterone receptors (ER and PR).They are typically immunohistochemically positive for CD10 and negative for desmin and h-caldesmon.[7] Endometrial stromal sarcoma (ESS) has a non-specific appearance on ultrasound, typically characterized as a heterogeneous hypoechoic endometrial mass, which can show extensive myometrial involvement. On magnetic resonance imaging (MRI), these tumors appear as large masses with or without evidence of myometrial invasion. The characteristic pattern of ESS consists of worm-like tumor projections along the vessels or ligaments, which are best visualized on MRI with diffuse weighted imaging.[8]

ESS are staged according to the 2010 International Federation of Gynecologic Oncology (FIGO) staging system. If sarcomas are diagnosed following hysterectomy for a fibroid uterus it is essential to do imaging to look for metastasis. Further surgery is not needed if the imaging is negative. If extrauterine disease is detected, surgical staging and cytoreduction are performed only if there is no extraabdominal disease and the intraabdominal metastases are resectable. Lymphadenectomy is performed only in patients with preoperative evidence of enlarged lymph nodes (based on imaging), intraoperative findings of lymphadenopathy, and those with extrauterine disease.[9]

Observation is needed for patients with surgical stage I ESS. For women with surgical stage II to IV ESS, adjuvant endocrine therapy is needed. Radiotherapy may be administered (in addition to endocrine therapy) to reduce the risk of a locoregional recurrence rather than chemotherapy. For patients with recurrent or metastatic ESS who progress despite endocrine therapy chemotherapy is given. Hormone therapy with medroxy progesterone, tamoxifen, gonadotropin releasing hormone (GnRH) analogues and aromatase inhibitors are suggested for low-grade ESS stage 3-4 and for recurrent disease.[10] Available treatment combinations include gemcitabine plus docetaxel and doxorubicin-based regimens. As these tumors have a tendency for late recurrence, long-term follow up is essential. It shall be once in 3 months for the first year and half-yearly for next 4 years. Thereafter annual follow up is recommended. Our case had no evidence of metastasis on computerised tomography and was treated with radiotherapy and chemotherapy to prevent metastasis. She is advised to come for a regular follow up in future.

This case is highlighted to show the significance of histopathological examination in all clinically benign and innocent looking lesions, as a correct diagnosis will help in the proper management of the patient.

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