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Case Report

Metastatic endometrial stromal sarcoma: a case report

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ABSTRACT

Endometrial Stromal Sarcoma (ESS) is a rare slow growing tumour of mesodermal origin arising from the stroma of the endometrium and accounting for less than 1% of all uterine cancers. It is characterized by late recurrences and distant metastases. This report presents a case of ESS in a 40 year old nulliparous woman who had a myomectomy for a clinically suspected Leiomyoma uterus in a local hospital. The histopathological examination of the specimen revealed ESS and the patient was referred to our tertiary institute. Here after investigations including a CT scan which also revealed pulmonary metastases, patient underwent Modified Radical Hysterectomy with Bilateral Salpingo-oophorectomy with pelvic lymph node sampling. Histopathological Examination of the uterine specimen confirmed the diagnosis. The patient was given the option of referral to a thoracic surgeon for resection of the isolated lung metastasis, but she refused this and opted instead for hormone therapy which she is presently undergoing. ESS is a very rare tumour often presenting with clinical and examination findings suggestive of leiomyoma of the uterus and hence misdiagnosed. In cases of rapidly growing tumours and suspicious radiological features, suspect sarcoma and initiate timely diagnosis and proper treatment. Recommended long-term follow up in view of late recurrences.

Keywords: Hormone therapy, Endometrial stromal sarcoma, Myometrial invasion, Pulmonary metastasis, Uterine sarcoma

INTRODUCTION

Sarcomas are tumours that arise from the mesodermal structures like muscles and connective tissue. ESS arises from the supporting connective tissue (stroma) of the endometrium and is composed of cells that resemble the proliferative phase endometrial stromal cells.[1]

ESS is a relatively rare malignant tumour, accounting for 0.2% of all uterine cancers and <10% of all uterine mesenchymal tumours.[1] Only limited reports with small number of patients are available in literature due to the rarity of the tumour. The annual incidence of ESS is 1-2 per million women accounting for 400-700 new cases in Europe. These are most common in the age group of 40-50 years with about 25% of the patients being premenopausal. Most of the patients present with complaints of irregular vaginal bleeding, pelvic pain and dysmenorrhea, but about 25% of patients are asymptomatic.[2] Cases of ESS have been reported in patients with polycystic ovarian disease and with exposure to unopposed estrogens and tamoxifen.2 Chromosomal aberrations like deletion on 7p have been noted in >55% of patients with ESS. In ESS, the risk of recurrence is thought to be as high as 50% and could occur even 20 years after the initial diagnosis.[3]

CASE REPORT

A 40-year-old nulliparous woman presented with complaints of abdominal pain since 10 months and irregular heavy vaginal bleeding since 6-7 months. The patient had been married since 11 years and had never conceived. She was investigated and took treatment for infertility from several local hospitals. She was unable to produce complete details of these previous treatments but

there was history of usage of clomiphene, letrozole and, estradiol valerate. She also underwent a pelvic scan in April 2013, which revealed a 10 × 7 cm mass in the posterior wall of the body of the uterus with necrotic changes. Preoperative diagnosis of Leiomyoma of uterus with degeneration was made and the patient underwent myomectomy. The histopathological examination of the myoma specimen showed islands of endometrial stromal tumour cells infiltrating the myometrium. The cells were round or oval with pleomorphic nuclei and scanty cytoplasm with 1-2 mitoses/10 HPF. Spiral arterioles seen within the tumour with areas of hemorrhage, necrosis and hyalinization and the diagnosis was low grade endometrial stromal sarcoma. The patient was hence referred to our medical college for further management but reported to our institute only 5 months after her myomectomy.

On admission, her general and systemic examinations were normal. Abdominal and pelvic examination showed the uterus to be enlarged to 14 weeks size, firm, mobile with no palpable adnexal masses. Ultrasound of the abdomen and the pelvis was done which showed a bulky uterus 9.7 × 5.4 × 8.2 cm with a mass 6.7 × 5 cm with coarse echotexture and poor endo-myometrial differentiation. An axial contrast enhanced CT scan of the abdomen and pelvis showed an enlarged uterus with a moderately enhancing mass lesion in the posterior myometrium about 6.8 × 6 cm in size. A diagnosis of sarcomatous changes in fibroid was made. No infiltration was noted into the parametrium, both fallopian tubes and ovaries were normal. Retroperitoneum showed normal anatomy, aorta and paraaortic areas were normal. There were no ascites, no bowel masses, or bowel wall thickening. Two nodular lesions measuring 10 × 9 mm and 14 × 10 mm were detected in the basal segments of the lower lobe of right lung suggestive of metastases.

The patient underwent a laparotomy. Intraoperative findings revealed no free fluid. Peritoneal washings were taken for cytology. Uterus was enlarged to approximately 12 × 12 × 10 cm in size, omental adhesions present to the uterine fundus and, the sigmoid colon was adherent to the uterus posteriorly. Both tubes and ovaries looked grossly normal. Pelvic lymph nodes were enlarged with largest being 2 × 1 cm. Omentum was grossly normal with no other visible or palpable metastatic deposits. A modified radical hysterectomy taking parametrium on both sides, bilateral salpingo-oophorectomy, bilateral pelvic lymphadenectomy and, infracolic omentectomy were done.

The gross examination of the cut section as shown in Figure 1 delineates a well circumscribed lesion within the myometrium with worm like yellowish appearance about 5 × 4 cm seen extending from myometrium to within 2 mm from serosa. The microscopy findings of the endometrium showed corkscrew shaped glands and columnar epithelium. Section from myometrium, which is shown in Figure 2, is an infiltrating neoplasm composed of round to oval cells with scanty cytoplasm, nuclei with coarse chromatin clumping with mitotic figures 4/10 HPF. Figure 3 shows the image in high magnification (40x) of the infiltrating sarcoma cells. Network of delicate arterioles were seen between tumour cells. The fallopian tubes, both ovaries, pelvic lymph nodes, parametrium and, omentum did not show tumour cells and peritoneal washings had no malignant cells. The Histopathological diagnosis was ESS.

Figure 1: Gross examination of the cut section of the uterus. Worm like pattern of tumour growth (arrow) into the myometrium, typically seen in ESS.

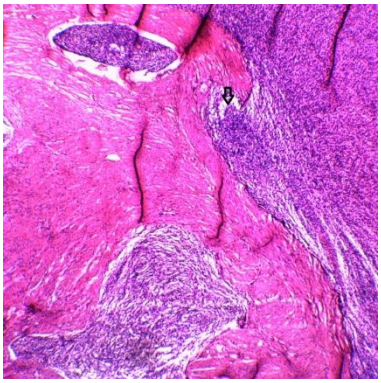

Figure 2: Sarcoma cells seen infiltrating (arrow) in between myometrium.

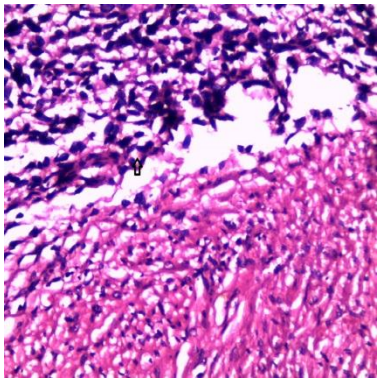

Figure 3: ESS high power view (40x) showing sarcoma cells seen infiltrating (arrow) myometrium.

The postoperative period was uneventful. The patient was advised referral to a thoracic surgeon for resection of the isolated lung metastasis, but she refused to undergo any other surgical procedure. The patient has now been referred to a medical oncologist for adjuvant hormone therapy.

DISCUSSION

ESS is a subset of uterine mesenchymal tumour and it arises from the stroma of the endometrium. Stromal tumours are classified as[4]

1. Endometrial Stromal Nodule (ESN)
2. Endometrial Stromal Sarcoma (ESS)
3. Undifferentiated Endometrial Sarcoma(UES)

This is based on vascular invasion, mitotic activity, nuclear pleomorphism and necrotic changes. ESN are well circumscribed, non-capsulated with non-infiltrating borders. The projections into myometrium, if present are < 3mm and behave like benign lesions. ESS (previously known as low grade endometrial stromal sarcoma) shows irregular nodular growth with myometrial invasion, characteristic tan- yellow color with worm like plugs of tumour that also spill and extend into myometrial and parametrial veins. The cells closely resemble the endometrial stromal cells and mitotic activity is usually less than 10/10 HPF.[4] ESS usually occur in women in their 40s, pursue a more protracted course with good 5 year survival and late recurrences while UES (previously known as High Grade Endometrial Stromal Sarcoma) is seen in older women in their 60s, follow a more aggressive course with early recurrences and distant metastasis. Highly cellular leiomyomas and intravenous leiomyomatosis often histologically mimic ESS. Use of immunohistochemistry with CD 10, h-caldesmon and desmin may be of help in differentiating them; of these, h-caldesmon is more sensitive and specific.[5]

Most patients of ESS present with an enlarged uterus, abnormal uterine bleeding, pelvic pain and dysmenorrhea. About 25% of patients are asymptomatic. Extrauterine pelvic extension is more commonly seen to the ovaries.[4] Occasionally the diagnosis is established by endometrial biopsy or a diagnostic curettage in cases with endometrial involvement, but in most cases the tumour being mainly intramyometrial, curettage will not diagnose ESS. Most often ESS is misdiagnosed as leiomyomas and the diagnosis is made only postoperatively on histopathological examination of the specimen as happened in this case.

The International Federation of Gynaecology and Obstetrics (FIGO) staging for uterine sarcomas is followed to stage ESS as well. Prognosis depends on age, race, parity, menopausal status, the stage of the disease, mitotic activity, nuclear pleomorphism, atypia, lymphatic and vascular invasion and the presence or absence of hormone receptors.[1]

The treatment recommended is total hysterectomy with bilateral salpingo-oophorectomy with selective pelvic and para aortic lymphadenectomy.[6] In Stage I disease in premenopausal women, ovarian conservation can be considered.[7] Stage I patients need only long term follow up. The efficacy of adjuvant therapy is still not proven, but in view of abundance of estrogen and progesterone receptors in the tumour, hormone therapy has been shown to be effective. Medroxy progesterone acetate, megestrol acetate, GnRH analogues like leuprolide and aromatase inhibitors - letrozole and anastrazole have been used as adjuvant therapy and to treat recurrences and advanced or unresectable metastases.[6,8] Radiotherapy can also be used as adjuvant therapy in stage III and IV and to treat local recurrences. The role of chemotherapy in the treatment re mains unclear without sufficient supporting data.

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