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A Rare Case Report of Uterine Leiomyosarcoma in young age

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Abstract: *Leiomyosarcoma is a rare malignancy of smooth muscle origin, notorious for it's aggressive nature and poor prognosis. Uterine leiomyosarcoma accounts for 1% of all uterine malignancies. Here we are presenting a rare case of uterine leiomyosarcoma at a young age of 29 years. She is para one with one living issue, underwent lower segment caesarean section, had complaints of continuous bleeding per vaginum and lower abdominal pain for 5months. Initially, she was managed conservatively as intramural fibroid. Later, the patient presented with continuous bleeding per vaginum with severe breathlessness diagnosed as Uterine Leiomyosarcoma stage IVB with lung metastasis.*

***Keywords*:** *Uterine leiomyosarcoma, metastasis,postpartum period.*

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I. Introduction

Leiomyosarcoma is a rare malignancy of smooth muscle origin, a highly aggressive tumour, can arise from gut, uterus and blood vessels. Uterine Leiomyosarcoma accounts for 1% of all uterine malignancies and 25-36% of all sarcomas of the uterus. The incidence is 0.3-0.4/1,00,000 population[1]. The median age is usually in perimenopausal age group(43 to 53years). Disease in young patients is very rare[2]. Risk factors are prior pelvic radiation therapy, nulliparity, obesity, increasing age, African-American women have higher incidence than asian women. Presenting symptoms may not be specific to the disease which include abnormal uterine bleeding which is the most common, pelvic pain/pressure and rapidly growing abdominal mass. Mean duration of symptoms is 6 months[2-8].

The tumors spread to the myometrium, pelvic blood vessels and lymphatics, contiguous pelvic structures, abdomen and distantly most often to the lungs. Histological markers like tumours with more than 10mitotic figures/10HPF, severe cytological atypia and coagulative cell necrosis are considered as frankly malignant with poor prognosis. Tumours with 5 -10 mitotic figures/10HPF are termed as smooth muscle tumours of uncertain malignant potential (STUMP)[2]. The five-year survival rate for Leiomyosarcoma stage I is 40-70% and stage II-IV is 8-12%. The overall 5-year survival rates extend from 15-25%[1-2].

II. Case Report

A young woman, 29 year old came to our Gynaecological OPD, with complaints of continuous bleeding per vaginum and lower abdominal pain since 5 months. The present complaint started from one month after the delivery. She had history of loss of weight and loss of appetite for 4months and she also had complaints of shortness of breath for 1 week (NYHA class –IV), not associated with orthopnoea and paroxsymal nocturnal dyspnoea.

Her previous menstrual cycles were regular and associated with normal flow. Her obstetric history is para one with one living issue conceived spontaneously. Her antenatal and intrapartum period was uneventful. She underwent Lower segment caesarean section(LSCS) in view of oligohydramnios and delivered a healthy baby at a private institution 6 months back. Her post partum period was uneventful except for occasional bleeding per vaginum for one month. After that she developed continuous bleeding per vaginum with lower abdominal pain for which she was admitted in an institution, investigated and diagnosed with intramural fibroid, managed conservatively with 2 units of blood transfusions, NSAIDS, tranexamic acid 500mg thrice daily and discharged. After 2 months, she presented to our OPD with severe breathlessness.

On examination her general condition is poor, she was pale and emaciated, afebrile, dyspnoeic with tachycardia and tachypnoea. SPo2 was 99% with O2 inhalation, bilateral crepitations present over infrascapular and infraaxillary area. Per abdomen uterus was 16 weeks size, no organomegaly noted. On per vaginal examination uterus was 16weeks size, mobile, tenderness present.

Differential diagnosis:

1. Fibroid uterus.
2. Myomatous polyp.
3. Retained products of conception.
4. Placental polyp.
5. Secondary postpartum hemorrhage.
6. Uterine arterio-venous malformations.
7. Choriocarcinoma.
8. Uterine sarcoma.
9. Endometrial cancer.
10. Ovarian tumour.

She was investigated and her complete blood picture, renal function tests, liver function tests were found to be within normal limits. Her CA125 levels are elevated (113IU/ml), serum alphafetoprotein, serum CEA, β-HCG levels were within normal limits. Serum Lactate dehydrogenase levels were raised. ECG and 2D ECHO were done, no abnormality detected. Chest X-ray showed multiple cannon ball like lesions. CT chest showed multiple well-defined soft tissue nodules of various sizes seen in both lungs involving all the lobes and a well-defined soft tissue mass lesion -7.5x6x4.2cm along the greater curvature of stomach. CT abdomen showed heterogeneously enhancing lesion measuring 14.7x8.5cm in uterus, extending superiorly up to umbilical region and also a metastatic deposit noted in left gluteal region. Endometrial biopsy confirmed the diagnosis of pleomorphic leiomyosarcoma and then we diagnosed her with Uterine Leiomyosarcoma Stage IVB with lung metastasis.

Figure no 1: Chest X-ray showing cannon ball metastasis.

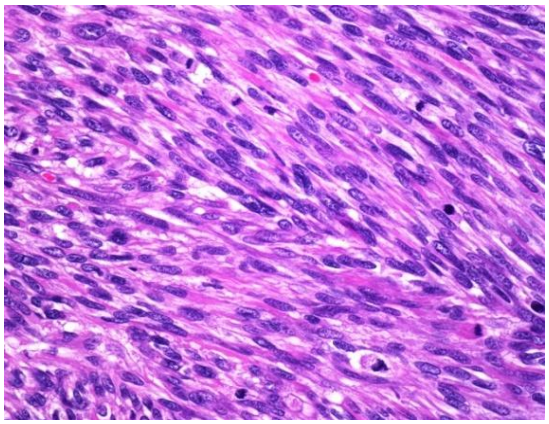

Figure no2: Leiomyosarcoma-fascicles of spindle cells exhibiting smooth muscle differentiation with nuclear atypia,moderate to severe pleomorphism.

She was started immediately on palliative chemotherapy with single agent adriamycin in the Department of oncology, but she ultimately faced the doom of death within a week**.** This is a rare presentation of uterine leiomyosarcoma in young age.

III. Discussion

Uterine Leiomyosarcoma is very rare in young patients. Although the median age group is perimenopausal women, 15% of patients are younger than 40years of age. Younger patients with leiomyosarcoma seem to have more favourable outcome than postmenopausal women[3]. High index of suspicion is mandatory for the accurate diagnosis. The present case was managed conservatively interpreting the uterine mass as leiomyoma. Leiomyosarcomas frequently coexist within a fibroid uterus and approximately 0.5% of women who have hysterectomies for uterine fibroids are found to have leiomyosarcomas[4]. Endometrial biopsy should be done for all patients in postpartum period complaining of irregular vaginal bleeding, keeping in mind the possibility of malignancy[5]. MRI imaging of pelvis may be helpful to delineate the extent of uterine involvement.

The prognosis depends on the gross presentation of tumor at the time of diagnosis and the mitotic index[6]. Vascular invasion is seen in up to 25% of leiomyosarcomas.Women with tumor size more than 5 cm in diameter have a poor prognosis[7]. Early stages can be treated by total abdominal hysterectomy with bilateral salpingo-oophorectomy and treatment of lymphatics by radiation or surgery. Late stages can be treated with combination chemotherapy. Doxorubicin (Adriamycin) is the single most active agent in the treatment of leiomyosarcoma producing 25% response rate. Gemcitabine and docetaxel combination is effective for metastatic leiomyosarcoma with response rate of 53%[9], although recurrence rate is very high.

IV. Conclusion

Uterine leiomyosarcomas being rare, are not suitable for screening. Histopathological examination plays a vital role in making the diagnosis of leiomyosarcoma. Timely diagnosis and management increase the chances of survival in these patients.

Abbrevations:
HPF- High Power Field
NHYA- New York Heart Association
CEA- Carcino Embryogenic Antigen
HCG- Human Chorionic Gonadotrpin
CT-Computed Tomography

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