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Case Report

Rare but Relentless: The Leiomyosarcoma Enigma

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Abstract:

Uterine sarcomas comprise less than 1% of gynaecological malignancies and 2-5% of all uterine malignancies. Leiomyosarcoma accounts for less than 1% of all uterine malignant mesenchymal tumour. As there is no specific pattern of symptoms to differentiate leiomyosarcoma from leiomyoma,the diagnosis of leiomyosarcoma is usually made postoperatively. MRI is a useful mode of investigation for detection of LMS and helps in determination of appropriate management. LMS is commonly seen in perimenopausal women, hence preoperative imaging with MRI and in bag morcellation may be considered in such cases.

Keywords: LMS-Leiomyosarcoma; Myoma; hysterectomy

Introduction

Leiomyosarcoma is a rare mesenchymal uterine malignancy. It is a aggressive tumour with high mortality and morbidity. Preoperative diagnosis of leiomyosarcoma is still a real challenge.

63 yearold Multipara, has been married for 47 years and had 4 normal child birth. Last child birth was 39 years back. Postpartum sterilisation was done. She had attained menopause at 45 years of age. She came for consultation with complaints of abdominal pain and dribbling of urine for 4 weeks duration. She was on homeopathic medication for 2 weeks. She was admitted in view of urinary retention, relieved by bladder catheterisation. MRI showed a large lobulated T2 hypo-intense mass in the pelvis measuring 9.5 x 8.8 x 10.9 cm, as can be seen in Fig1. The mass shows T2 hyper-intense areas of cystic degeneration within. Anteriorly, it is causing significant compression of the urinary bladder. Posterolateral, it is reaching up to the lateral pelvic wall on the left side abutting the obturator internus and the pyriformis muscle, suggestive of broad ligament fibroid. She was taken up for total abdominal hysterectomy with bilateral salpingo oophorectomy with unilateral DJ stenting.

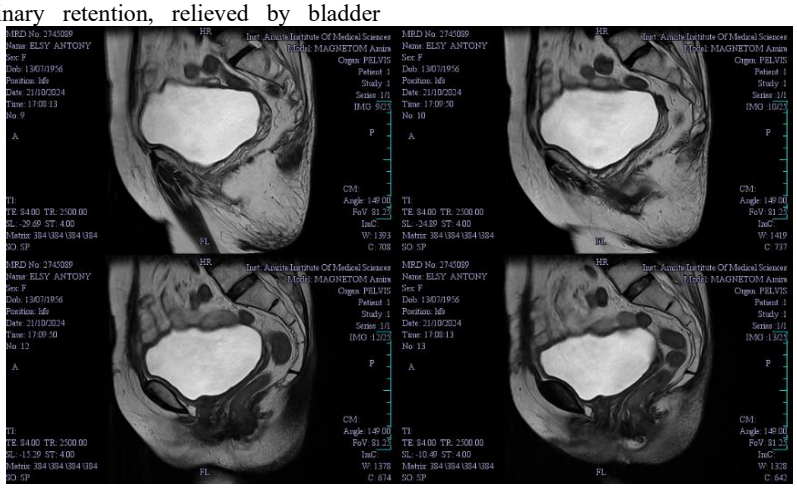


Figure 1: MRI Findings: Large lobulated T2 hypointense mass in the pelvis measuring 9.5 x 8.8 x 10.9 cm with T2 hyperintense areas of cystic degeneration within,causing significant compression of the urinary bladder anteriorly

Intra operative Findings

A large lobulated mass of 10 x 9cm seen anterior to the uterus and bladder stretched over the myoma, extending through the anterior vaginal wall upto the level of external cervical os and has extension into left broad ligament. The mass was friable with high vascularity. The mass was found to have extension into vagina, which was dissected with finger and pushed the mass pervaginally above and the mass bulk of tumor removed. Capsule of left broad ligament opened and mass enucleated.

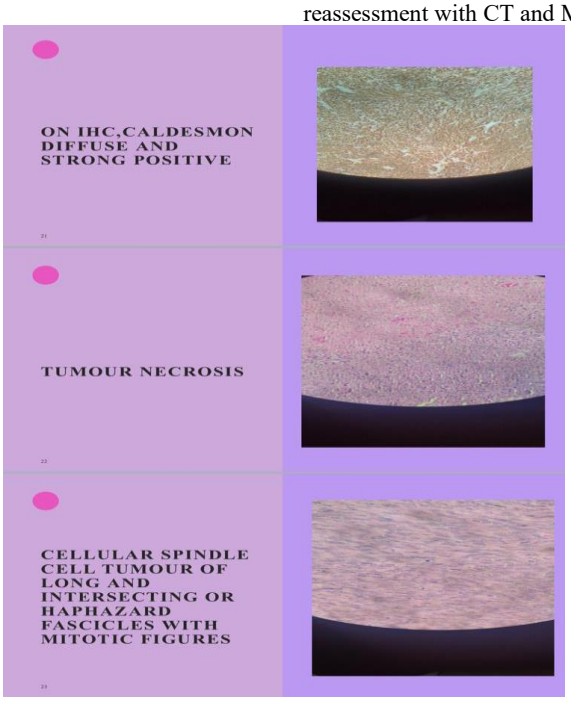
Histopathology revealed Leiomyosarcoma

Immunohistochemistry confirmed Leiomyosarcoma. Cells are positive for Caldesmon, SMA,negative for Cyclin D1,CD10,p53(wild)

Post operative CT done showed Residual lesion

Per vaginal and per rectal examination showed fixed mass in left side of vault Tumour board discussion done, planned for chemotherapy followed by reassessment for radiation therapy

She was started on Inj Gemcitabine 675 mg/m2 Day 1 and 8 with Docetaxel 75 mg/m2 day 8 only Q 3 weekly for 4 cycles followed by reassessment with CT and MRI for further treatment.



Discussion

Uterine sarcomas comprise less than 1% of gynaecological malignancies and 2-5% of all uterine malignancies. Leiomyosarcoma accounts for less than 1% of all uterine malignant mesenchymal tumour[1], which spreads through intraperitoneal and hematogenous pathways. Malignant pure mesenchymal uterine tumour/uterine sarcomas encompass endometrial stromal sarcoma(ESS), Leiomyosarcoma (LMS) and undifferentiated sarcoma[1].

It is more common in perimenopausal women with median age of 50 years(5). Clinical picture is fairly similar to leiomyoma which includes Abnormal uterine bleeding, pelvic pain or pelvic mass[2]. Regional lymphnode positivity and distant metastasis are rare [3].

It is often diagnosed post operatively following hysterectomy or myomectomy following a benign disease by histopathology [4].

Investigations

Surgery remains the mainstay of treatment, hysterectomy with bilateral salpingo oophorectomy is the surgery of choice, ovarian preservation can be considered in young women.

Adjuvant chemotherapy and radiotherapy is not beneficial

Chemotherapeutic drugs like Gemcitabine, Docetaxel and Doxorubicin are the treatment of choice in advanced or recurrent disease.

Conclusion

Broad gynecological complaints such as abnormal uterine bleeding,pelvic pain or pelvic mass.High clinical suspicion of LMS should be there in fibroids diagnosed in perimenopausal women.MRI is a good assessment tool. Pre operative imaging with MRI and in bag morcellation may be considered in such cases.

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