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Case Report

Myxoid leiomyosarcoma-decoding the puzzle in the Pandora's box-pre to post operative

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ABSTRACT

Myxoid leiomyosarcomas (MLMS) are a rare form of uterine cancer developed from the smooth muscles of the uterus. It is a very aggressive tumour and usually presents as abdomino pelvic mass. This is a case report of a 47-year-old parous, perimenopausal lady with huge abdominopelvic mass mimicking ovarian cancer preoperatively, but postoperatively turned out to be MLMS. It was a tumor of size 40×30×9 cm, weighing 12 kg. Histopathological examination revealed myometrial neoplasm composed of atypical spindle to stellate cells disposed as loose sheets within an abundant myxoid stroma. Immunohistochemically, SMA and Ki-67 were positive, which clinched the diagnosis. Postoperatively, chemotherapy was decided for the patient, and she is presently doing well after a year of surgery.

Keywords: Myxoid leiyomyosarcoma, Immunohistochemistry, Abdominopelvic mass, Aggressive tumor, Uterine tumour

INTRODUCTION

Uterine sarcomas are uncommon malignant mesenchymal tumors, accounting for approximately 3% of all uterine cancers, and include high- or low-grade endometrial stromal sarcoma (ESS), undifferentiated uterine sarcoma (USS), uterine leiomyosarcoma (uLMS), and other sarcomas.[1] According to a 2012 systematic review of data from 1970 to 2011, uLMS was the most common subtype (63%), followed by ESS (21%), and less common subtypes such as UUS.[2]

uLMS are usually of the spindle cell (conventional) type, but less common variants are of myxoid or epithelioid morphology. Less than 90 cases of uterine MLMS have been reported in the English literature up to the present.[3-9] MLMS is a tumor with an annual incidence of 0.64/100 000 women.[10]

Its diagnosis is extremely challenging owing to the clinical and histological disparity.[3,11]

CASE REPORT

A 47-year-old parous, perimenopausal lady presented to OPD with complaints of abdominal distension for 8 years with a rapid increase in size for the past 3 months. She had history of easy fatigability. Her past history showed no menstrual irregularity or menorrhagia. In general appearance, she looked cachectic with a distended abdomen. Vitals were within normal limits. On examination, pallor was present; no inguinal or supraclavicular lymph nodes were palpable. The abdomen was fully distended with an abdominopelvic mass of 40×30 cm, lobulated, occupying all quadrants of the abdomen, extending up to the xiphisternum.

On per vaginal examination, the cervix was healthy and felt high up, the uterus could not be palpated separately,

and there were no deposits in the pouch of mass was felt through pouch of douglas. Rectal examination done was normal.

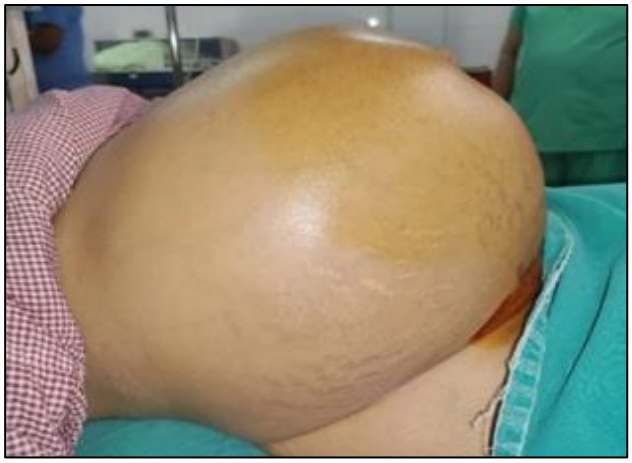

Figure 1: Preoperative picture showing a huge mass.

Laboratory tests showed anemia (Hb-9 gm/dl). Tumor markers, including CA125, CEA, and CA-19-9, were within normal limits.

MRI revealed an enlarged uterus measuring 15.9×19.2×10.8 cm with multiple fibroids, the largest measuring 5.8×5 cm. Few of the fibroids with calcific degeneration. Ovaries not visualized. Thickened and heterogeneous endometrium 4.6 cm. A large multiseptated abdominopelvic cystic lesion m/s 30×23×21 cm occupying the right-sided abdominal cavity and the pelvis, with the epicenter in the right adnexa-probably an ovarian mucinous cystadenoma.

Further assessment with PET CT was done, which showed a large predominantly cystic abdominopelvic mass of 33×24×23 cm with numerous thick septations, contrast enhancement, and hypermetabolism arising from right adnexa-likely malignant right ovarian neoplasm. No evidence of hypermetabolic lymph nodes or distant metastasis. Other organs were normal.

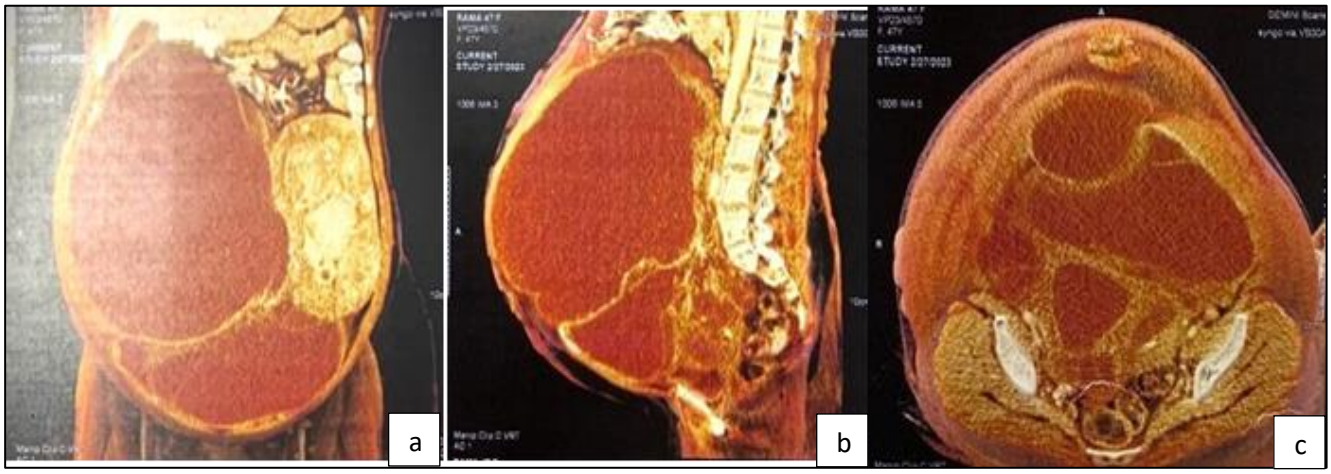

Figure 2 (a-c): PET CT pictures.

Staging laparotomy was done under general anesthesia through a vertical incision. It revealed a huge lobulated mass encasing the right ureter and a uterine subserosal fibroid 15×20 cm. Enlarged multiple pelvic lymph nodes-largest 4×3 cm on right side. Omentum, liver, and peritoneum were normal. Minimal serosal fluid was in the pelvis. The whole solid multicystic mass measuring 40×30×9 cm and weighing 12 kg with ovaries was removed. Pelvic lymph node dissection and infracolic omentectomy with peritoneal fluid cytology done.

Postoperative period was uneventful. Histopathology report showed uterine corpus-myxoid leiomyosarcoma with tumor dimensions of 40×30×9 cm. Myometrial invasion was present. Adnexal involvement-identified, one side fallopian tube wall involved. One side ovary adherent to tumor, other side not involved, lymphovascular invasion not identified, peritoneal fluid-malignant cells not identified, tumor extends upto the serosal surface, pelvic nodes free of tumor, infracolic omentum not involved.

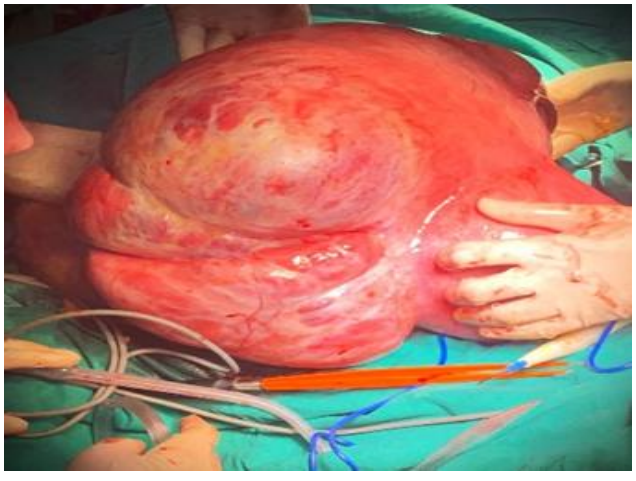

Figure 3: Intraoperative picture.

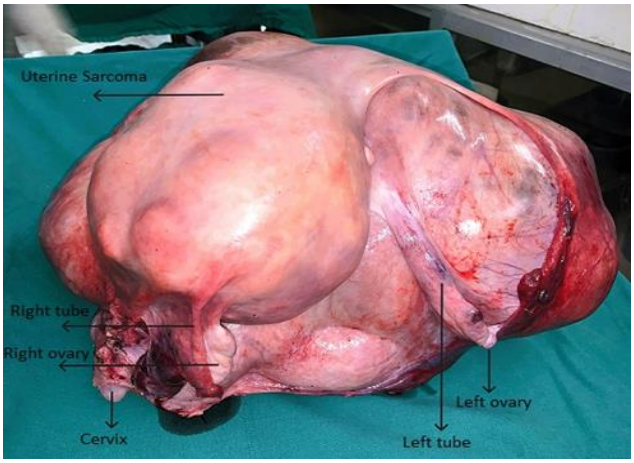

Figure 4: Postoperative picture

DISCUSSION

uLMS are usually of the spindle cell (conventional) type, but less common variants are myxoid and epitheliod. MLMS is a very rare and aggressive neoplasm. Only 26 cases have been reported in the literature since it was first described by King et al.[3] Because of its rarity, there has always been difficulty in preoperative diagnosis by clinicians and in confirming the diagnosis by histopathology.

They usually present as large abdominopelvic masses, usually as large as 8 cm with a mean diameter of 8 cm (2-5). In our case, the tumor filled the entire abdominal cavity and weighed 12 kg, extending 40 cm in diameter. This is thus the largest reported size of MLMS so far.

Occurring in a wide age range (20-86 years, with a median of 57 years), a majority of patients present with uterine bleeding, uterine mass, or abdominal pain.[12]

A particular characteristic of uterine MLMS is their gelatinous consistency and well-circumscribed borders. They usually have typical histologic and architectural features consisting of spindle-shaped cells with little or no atypia, a low mitotic count, and an abundance of intercellular myxoid tissue; an infiltrative growth pattern is the major indicator of malignancy.[3,12]

Until now, there is no laboratory test or imaging study that can reliably diagnose uterine LMS preoperatively. Though some patients will have elevated lactate dehydrogenase (LDH) and/or CA125 levels these markers are nonspecific, and the data for their use as a reliable predictor of malignancy is lacking. Endometrial sampling in an office or operative setting may yield a pathologic diagnosis; however, a negative biopsy does not preclude the diagnosis until the complete hysterectomy specimen is examined.[17,18]

The standard treatment for leiyomyosarcoma is primary debulking surgery.

The combined evaluation makes the diagnosis of uterine LMS of three factors: tumor cell necrosis (TCN), the number of cell divisions evaluated on 10 high-power fields (HPFs), and the presence of cellular atypia (cell pleomorphism). These criteria must be adapted according to the tumor subtype, myxoid and epithelioid tumors of the myometrium.[13]

MLMS causes irregular myometrial invasion with or without lymphovascular invasion. Cell mitosis is rare, and cytology may be unclear due to pleomorphism. The minimum number of mitotic divisions (MDs) to diagnose MLMS varies between 0-4/10 HPFs.[13] Because of rarity of tumor, treatment, prognosis, and follow-up are unclear.

The histopathological (HP) diagnosis of MLMS is made when there is a myxoid extracellular component with a cut-off of 30-50%.[14] Atkins et al used a 30% cut-off in terms of the density of the myxoid ECM, and Burch and Tavassoli suggested a 60% cut-off for the myxoid matrix.[15,16]

From an IHC point of view, MLMS showed positive staining for alpha-smooth muscle actin (SMA), desmin, and vimentin.[17] The classical HP examination and immunohistochemistry confirm the phenogenic type and the malignancy indicator. In a study by Mittal et al Ki67 contributed to the indication of tumor malignancy (mitotic index).

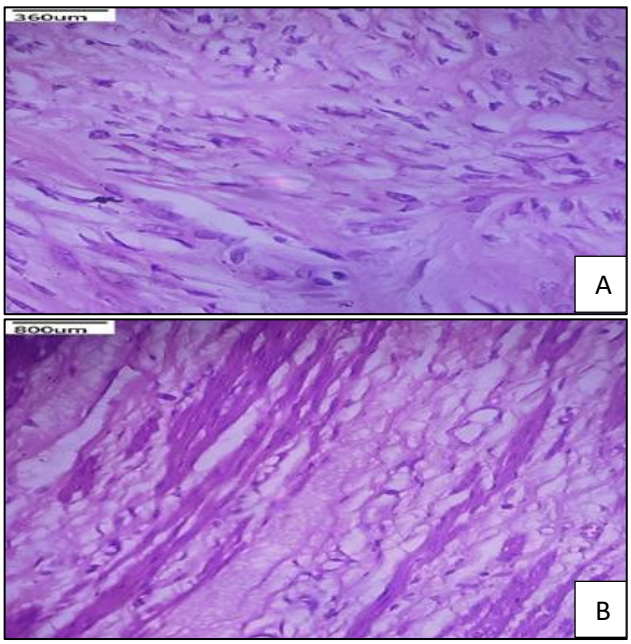

Figure 5 (A and B): Histpopathology images of our patient showing myometrial neoplasm composed of atypical spindle to stellate cells disposed as loose sheets within an abundant myxoid stroma. The cells show hyperchromatic nuclei with conspicuous nuclei; mitosis is about 2/10 HPF.

In our case, clinical examination and imaging findings were more towards a malignant ovarian tumor; hence, there was no suspicion of a sarcoma preoperatively. Intraoperatively it was found that the mass was arising from the uterus, which looked like a degenerative fibroid. As malignancy is very rare in the case of a uterine mass, its possibility was in doubt. Hence, grossly, it was difficult to conclude whether it was an ovarian mass/uterine benign or malignant mass. Postoperatively, we were able to confirm it was a FIGO stage II A MLMS (adnexal involvement) by histopathological examination, which revealed myometrial neoplasm composed of atypical spindle to stellate cells disposed as loose sheets within an abundant myxoid stroma. The cells showed hyperchromatic nuclei with conspicuous nuclei; mitosis is about 2/10 HPF. Immunohistochemically, SMA and Ki-67 were positive, which clinched the diagnosis. Ki-67 was 4% positive, which indicates the aggressiveness of the tumor.

From pre-op to post-op, there was much difficulty in diagnosis; histopathological examination and immunohistochemistry helped in decoding this puzzle.

In completely resected stage II and III tumors, it is appropriate to consider adjuvant systemic therapy and/or EBRT because of the increased risk profile. Observation can be considered for patients with completely resected tumors with negative margins.[20]

The recurrence rate is high in uLMS (50%-70%). Local recurrences are classified as recurrence in the vagina/pelvis with imaging that is negative for distant metastatic disease. Studies have shown that MLMS frequently metastasizes to the lungs, liver, and brain.[3]

Hence, once the diagnosis was arrived, plan was for 6 cycles of chemotherapy, which was decided in the tumor board meeting based on the size of the tumor. (Nuclei mitosis is about 2/10 HPF; Ki-67 is 4% positive, which indicates the aggressiveness of the tumor).

CONCLUSION

MLMS is a very rare tumor, and as such, very few cases are reported in the literature. Preoperative diagnosis with imaging modalities is still difficult. They are confirmed only postoperatively with histopathology and immunohistochemistry. Hence, adjuvant treatment has to be individualized and discussed in multidisciplinary tumour board meetings to give a tailored treatment for the patient.

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