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Case Report

Spindle cell sarcoma of uterus-a rare tale

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ABSTRACT

Uterine sarcoma are a heterologous group of are malignancies and account for one-tenth of uterine malignancies. Presentation of uterine sarcomas are usually very non-specific and varies with the histological sub type. Here we discuss a rare case of spindle cell neoplasm which presented to us with massive abdominal enlargement.she had no pressure symptoms. Decision to perform an exploratory laparotomy was taken given the high suspicion of ovarian tumour. Diagnosis was confirmed after histopathological report. Patient is doing well on follow up.

Keywords: Spindle cell, Uterine sarcoma, Benign, Abdominopelvic mass

INTRODUCTION

With the advent of modern technology and advances in imaging techniques, diagnosis and treatment of abdominopelvic masses can be done at an early stage and thus provide good prognosis. Most common causes of huge abdominal -pelvic masses are pregnancy, adnexal tumors, fibroids, ovarian cysts and neoplasms. Pelvic masses of reproductive age group are mostly benign. Uterine sarcoma are a heterologous group of are malignancies and account for one-tenth of uterine malignancies.[1] Malignant mesenchymal tumors (sarcoma) of uterus are a rare entity, but leioyosarcoma and endometrial sarcoma are its commonly found sub types.

Presentation of uterine sarcomas are usually very non-specific and varies with the histological sub type but all of them present as rapidly growing pelvic mass extending to the abdomen along with pain in abdomen, distension or heaviness. Rhabdomyosarcoma is a type of mixed mesenchymal tumour and spindle cell variant is a rare morphological variant of RMS.

We present here one of such rare case of spindle cell neoplasm which grew very rapidly but due to its benign nature had a favourable outcome.

CASE REPORT

A patient presented to our out-patient department in Max Smart hospital with massive abdominal enlargement. She is 48 years old P3L3 female with complaints of heaviness in abdomen associated with abdominal distension since 2 years accompanied with mild pain on and off. She also had oligomennorhea since past 2 years. Pain was dull aching in nature and non-radiating type. Patient explained that initially she had heaviness in abdomen, which was later associated with uniform increase in size of abdomen, to an extent that it hindered her daily activities. She had no pressure symptoms. No history of anorexia, weight loss, constipation or any urinary complaints during this period. Her menstrual cycles were irregular with scanty flow. There was no history of dysmenorrhea, menorraghia or inter menstrual bleeding. Past and family history was not significant and she had no commodities. On general Examination, her weight was 55 kg, had mild pallor and no other positive findings. Her physical examination revealed significantly distended abdomen with no tenderness, guarding or rigidity. A huge lump uniformly globular, firm to medium in consistency, fixed and occupying whole abdomen including epigastrium and bulging in both lateral sides of abdomen was seen. On per speculum examination, cervix could not be visualized and was pulled up due to the large mass. Per vaginal

examination, revealed that mass was occupying all fornixes and a posterior fornix bulge was seen due to the mass. On per rectal examination, rectal mucosa was free, mass palpable.

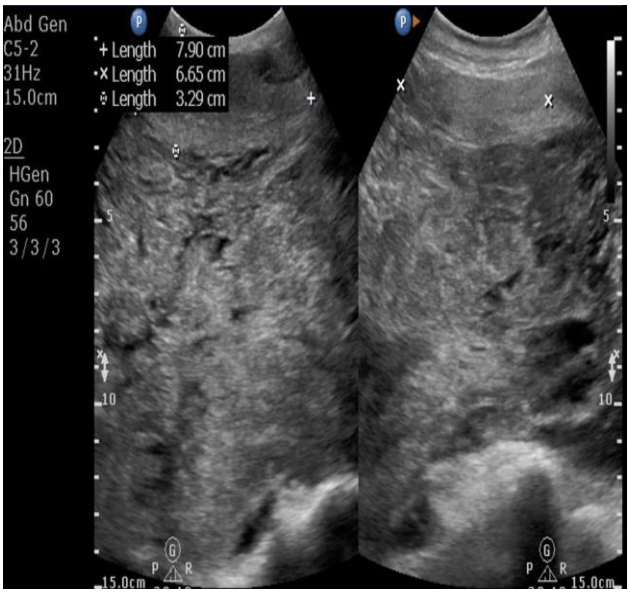

Figure 1: USG whole abdomen of large pelvi-abdominal heterogenous mass.

She had a Hb of 11 gm. Her routine labs were normal. Although her CA-125 level was slightly raised (40 mIU) but other tumour markers were within normal range. Initial imaging of ultrasound abdomen suggested a large pelviabdominal heterogeneous mass of about 28×30×12 cm with small cystic components displacing the abdominal viscera causing bilateral hydrouretonephrosis. Uterus and ovaries could not be visualized separately. Further evaluation of MRI pelvis with contrast showed a huge abdominopelvic mass of 31×11×22 cm cantered in left adnexal region bulging towards peritoneal cavity, displacing bowel loops. Mass showed hypo to isointense signal in T2 weighted image with solid and septated cystic area showing restricted diffusion in DWI with post contrast heterogeneous enhancement of solid part and non-enhancing cystic part. Uterus was displaced towards right iliac fossa showing both solid and septate cystic areas. Bilateral ovaries could not be visualized.

Decision to perform an exploratory laparotomy was taken given the high suspicion of ovarian tumour. Abdomen was opened via a vertical mid line incision from supra-pubic region to the epigastrium. No free fluid was observed. Intra operatively, a smooth walled 30×28 cm large highly vascular abdominopelvic mass was seen occupying whole of the abdomen and pouch of Douglas. Mass was globular, uniformly enlarged with smooth exterior surface. This mass was adherent to the posterior wall of the uterus, left infundibulo-pelvic ligament and large bowel. While identifying the ureters above adhesions were separated. Total abdominal hysterectomy with bilateral Salphingo-oophorectomy with removal of large mass was done. Second opinion of an oncologist was also taken. No deposits were noted bowel, under the diaphragmatic surface, para colic gutters. No enlarged lymph nodes were noted. Mass weighing of 9.5 kgs was removed intact and sent for frozen section. Cut section showed multi lobulated cystic lesion with myxomatous contents inside along with small cystic changes. The report was suggestive of spindle cell sarcoma. Two units of blood was given to the patient intra operatively. A drain was kept in. The POD and removed on the 4th post operative day. Rest of the post operative period was uneventful. HPE report was suggested of spindle cell sarcoma with extensive areas of cystic degeneration, necrosis all over.no increase in mitosis was observed. Fascicles of spindle cell were seen with normal nuclear-cytoplasmic ratio. On follow up visit, patient is doing well.

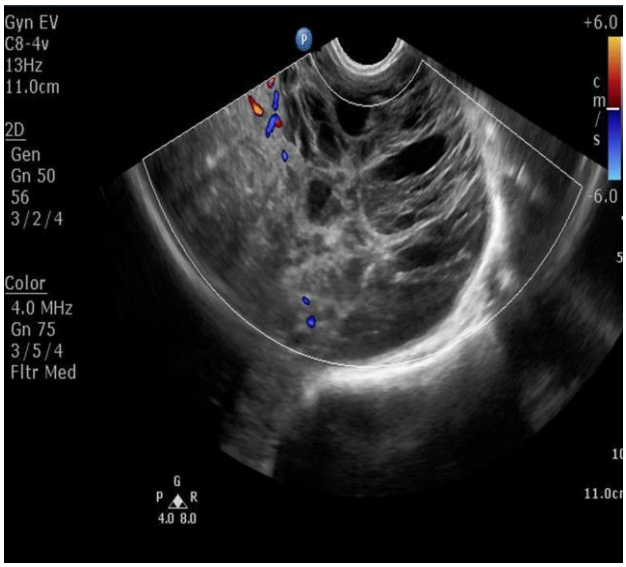

Figure 2: TVS scan showing huge mass of vascularity with cystic changes.

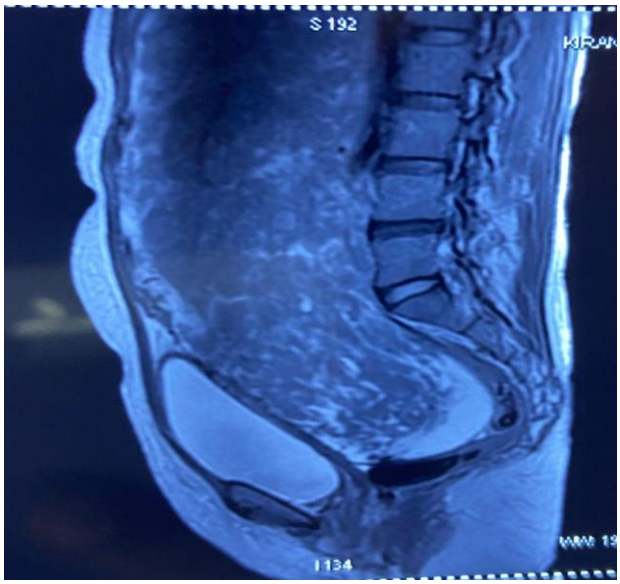

Figure 3: Sagital section of MRI.

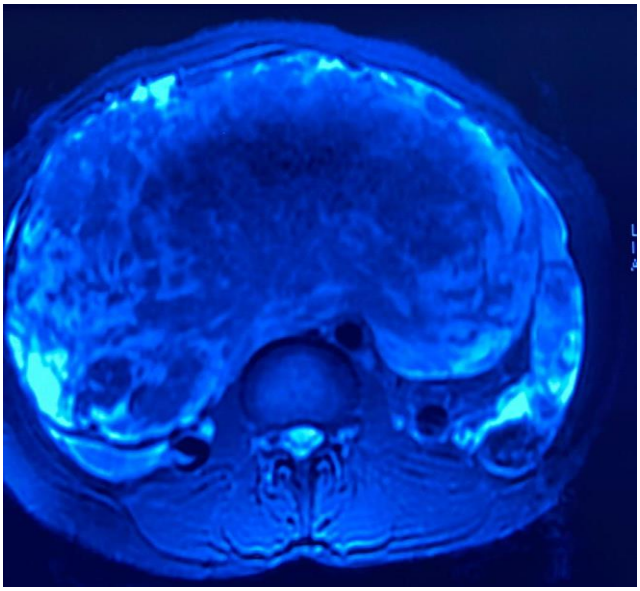

Figure 4: Axial section of MRI.

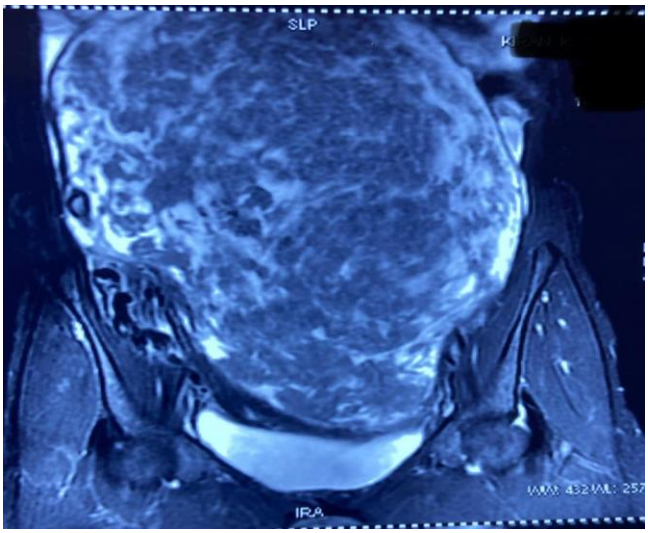

Figure 5: MRI showing mass.

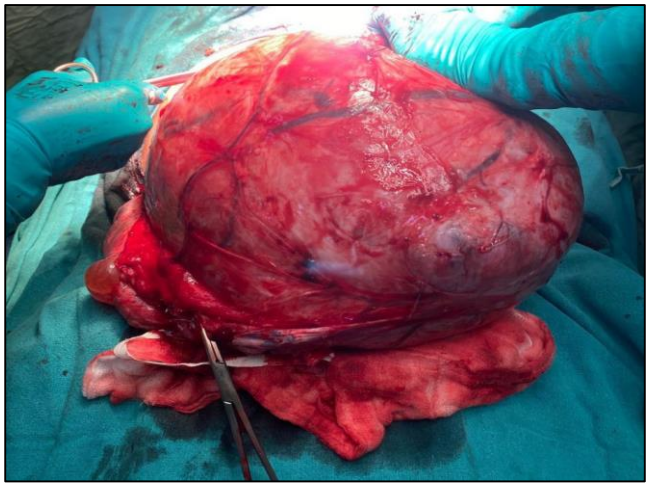

Figure 6: Intra-operative huge mass seen.

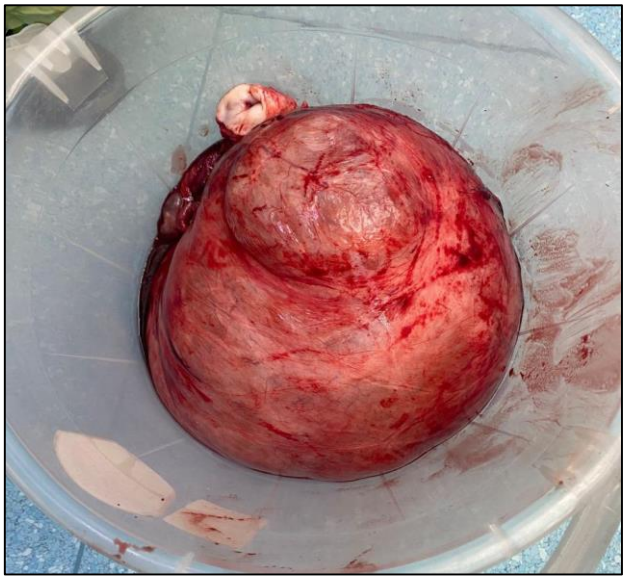

Figure 7: Uterus with cervix and mass along with fluid drained.

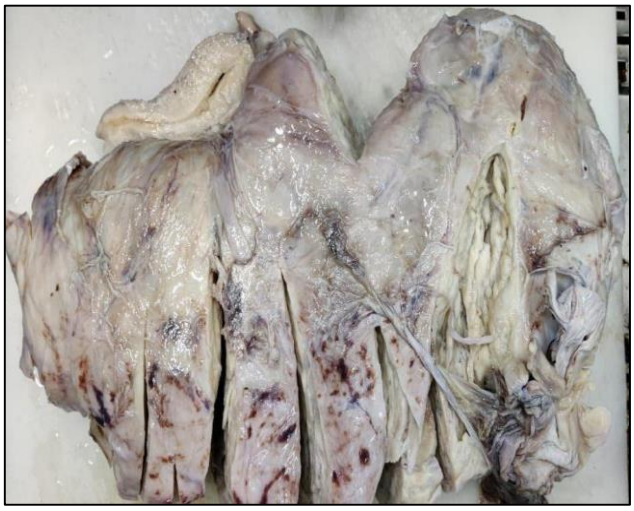

Figure 8: Cut section of the mass.

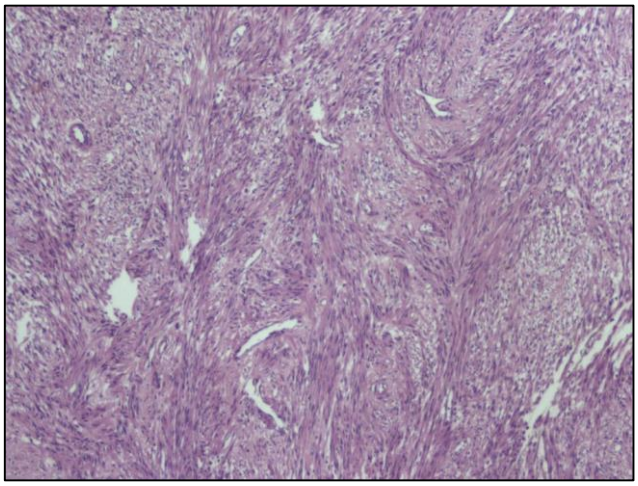

Figure 9: HPE image showing spindle cells.

DISCUSSION

Uterine sarcomas are a rare heterogeneous group of tumors of mesenchymal origin, which in the past were thought to account for only 2 to 3% of all uterine tumors. However now they contribute to 8% of uterine malignancies.[2,3] This increased incidence may be result of improved diagnosis, as well as a true increase in an ageing population.[2]

According to the gynecologic oncology group, uterine sarcomas can be classified into two categories: non epithelial and mixed epithelial-non-epithelial, depending on the type of cancerous cell and its presumed tissue of origin.[4] Norris and Taylor in 1966 first described the characteristics of these rare tumors.[5] Non epithelial tumors can further be classified into endometrial stromal tumors comprising of endometrial stromal nodule, low grade stromal sarcoma, high grade stromal sarcoma and leiomyosarcoma. In 2009, the classification of uterine sarcomas changed modifying their staging and histologic classification, consequently their proportional distribution changed. Their rarity and histopathological diversity have contributed to the lack of consensus on risk factors for poor outcome and optimal treatment.[6]

Among all the sarcomas, leiomyosarcoma is the commonest seen histological sub type.[7] These tumors are most frequently seen in the 4th decade of life. One of the rare morphological variants of uterine leiomyosarcoma is uterine spindle cell leiomyosarcoma. Spindle cell sarcoma is considered as an uncommon variant/ sub type of RMS9 which mostly occurs in para testicular region in children. Thus, it occurring in uterus is very rare possibility.[8] Nascimento and Fletcher performed the largest review comparing the most common sites involved by adult spindle RMS and they reported head and neck region to be most frequent site followed by retro peritoneum and lower extremity.[9] Thus, making our case of spindle cell neoplasm (variant of RMS) very rare finding.

Leiomyosarcomas commonly present around the age of 40 years which was also found to be consistent with the case presented by Nagtode et al and Kathpalia et al.[8,10] There is a twofold incidence of leiomyosarcoma in African-American women. Long-term tamoxifen use and prior pelvic radiation also seem to be associated with a small increase in risk.[4] Some cases are also associated with a prior history of retinoblastoma (mutation of Rb gene).

As per past literature, clinical features of uterine sarcoma include abnormal vaginal bleeding (56%), palpable pelvic mass (54%) and pelvic pain (22%). Less frequently, they can present as hemoperitoneum (due to tumour rupture), or symptoms resulting from extra-uterine extension or metastases.[11] Our patient had a big palpable abdominal mass with oligomennorhea and no pressure symptoms seen clinically. However, ultrasound was suggestive of hydro-uretero nephrosis. Mbatani et al states that uterine sarcomas are usually bulky tumors with a mean diameter of 10 cm and above.[12] Cut surface is typically soft, bulging, fleshy, necrotic, hemorrhagic and lacks the usual whorled appearance. According to Rubin et al, the diagnosis of spindle cell myxoid neoplasm can be aided by extensive tissue sampling, meticulous microscopic examination and immunohistochemistry.[13] Preoperative distinction between benign leiomyomas and malignant leiomyosarcomas is very difficult (if not impossible) based solely on clinical features, and remains a challenge for clinicians.[2,11] On the contrary, it also seems quite important to have a preoperative diagnosis to avoid surgical resection of large areas. Compared with ultrasonography and computed tomography (CT), magnetic resonance imaging (MRI) with diffusion weighted imaging is best imaging modality for assessing uterine lesions.[14] According to Santos, leiomyosarcomas on MRI commonly manifest as large infiltrating myometrial mass of heterogeneous hyointensity on T1 weighted images, with irregular and ill-defined margins, on T2 weighted images they show intermediate to high signal intensity with central hyperintensity indicative of extensive necrosis.[7] After contrast administration, they present early heterogeneous enhancement, due to the aforementioned areas of necrosis and hemorrhage. All these findings were consistent with our patient's MRI expect our mass being benign had smooth, regular surface margins. We had also faced similar challenges during our diagnosis and were only able to reach a final verdict after histopathological results of the specimen.

According to Leon et al at present there is scarce evidence on the management of uterine sarcomas due to low incidence and histopathological diversity.[15] Furthermore, the recent change in their classification has given rise to the need for conducting new studies to evaluate the best treatment alternatives.

Mbatani et al states, treatment of leiomyosarcomas includes total abdominal hysterectomy and debulking of tumour if present outside the uterus.[12] Removal of ovaries and lymph node dissection remain controversial as metastases to these organs occur in only a small percentage of cases and are frequently associated with intraabdominal disease. Follow up of such patients also remain uneventful.

CONCLUSION

Spindle cell neoplasms in itself is a rare entity so its diagnosis and management require the expertise of a clinician, radiologist, oncosurgeon and pathologist. A multispecialty approach is mandatory.

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