Case Report
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Spindle cell sarcoma – a rare diagnosis

Abstract

Leiomyosarcomas are the malignant tumors arising from the myometrium. Spindle cell sarcoma is a very rare variety of uterine sarcomas. We present one such rare case that was diagnosed only after histopathological examination. Clinical presentation and clinical course were also unusual, had grown to a huge size without any local or systemic spread and imaging studies did not help in making a diagnosis preoperatively. She has been on follow up for one and half years without any recurrence.

Keywords: Uterine tumor, Spindle cell sarcoma, Leiomyosarcoma

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Introduction

Uterine fibroids; also known as leiomyomas are commonest benign tumors of the uterus arising from the myometrium. They can grow to any size and the symptoms depending on their size, number or location. Leiomyosarcomas are the malignant tumors arising from the myometrium. Uterine sarcomas are relatively uncommon[1,2] with leiomyosarcoma as the most common histologic type. Uterine sarcomas are a heterologous group of rare malignancies and they account for one tenth of uterine malignancies.[3] Malignant mesenchymal tumours may be found in the uterus though very rarely; leiomyosarcoma and endometrial sarcoma are the common mesenchymal tumours. Rhabdomyosarcoma (RMS)[4] is sometimes seen as a component of malignant mixed epithelial and mesenchymal tumors. This is categorized as embryonal, alveolar and pleomorphic kinds. Spindle cell variant of RMS is a very rare morphologic subtype of embryonal RMS and usually occurs in the paratesticular region in children and has favorable outcome,[5] can occur very rarely in uterus. Spindle cell sarcomas are not only very rare but their behaviour is also varied. We present one such rare case of spindle cell sarcoma of uterus which had attained a huge size but had favorable outcome.

Case report

37 years old unmarried lady reported with complaints of swelling of abdomen for the last six to eight months. The swelling was slow to grow initially but had been enlarging rapidly for the previous two months. The swelling had become so huge (Figure 1) that it was difficult for her to lie down and move around. Turing in bed also was difficult as the lump had to be moved first before she could turn in the bed; could not sleep while lying flat, had to sleep propped up only. There was no pain abdomen. She was having regular cycles till one year back but was having excessive bleeding with passage of clots every month. There was no inter-menstrual bleeding or bleeding on straining. There were no bladder or bowel complaints. Her appetite was normal and there was no loss of weight. Past and family history was not relevant. On examination; she was comfortable while sitting in bed. Weight was 81 Kg, there was bilateral pedal edema (Figure 2) and rest of the general examination was normal except mild pallor. Abdominal examination showed markedly distended abdomen, more anteriorly than laterally. Whole of abdomen was filled with a huge lump which was hard to soft in consistency and mobility was restricted but was present, one could reach below the lump. Percussion note was dull all over and shifting dullness could not be elicited. External genitalia appearance was normal. Per speculum examination was not done as she was not willing and digital examination did not reveal any abnormality. On per rectal examination it was found that rectal mucosa was free and the lump was felt high in the pelvis. There was no nodularity felt anterior to rectal mucosa or laterally. A provisional diagnosis of lump abdomen probably arising from ovary; mucinous cystadenoma with malignant change or fibroid uterus with cystic degenerations was made. Routine investigations were normal except that her Hb was low (6.7 gm %), peripheral smear showed it to be microcytic hypochromic anemia. Imaging studies like Ultra Sonography (USG) (Figure 3) and Computed Tomography (Figure 4) were not very informative. The whole abdomen was occupied by a mixed echogenic growth; uterus and ovaries could not be visualized clearly. There was hydroureter above the pelvic brim and mild hydronephrosis on left side. Tumor markers were normal except mild rise in Cancer Antigen 125. She was advised to go to higher center but was unwilling and requested to be managed locally only. Preoperative diagnosis of mucinous cystadenoma (because of massive size and imaging) was made and the patient and relatives were counseled and the case was prepared for exploratory laparotomy after three blood transfusions and bowel preparation.

Abdomen was opened by midline vertical incision, there was no free fluid. The tumor could not be taken out due to its large size; which was reduced after sucking out fluid which was blood stained (Figure 5), with great difficulty the tumor was exteriorized and was found to be arising from uterus. There were no adhesions. Total abdominal hysterectomy with bilateral salpingo-oopherectomy was performed after ensuring the safety of ureters on both sides. She was transfused two packed red blood cells and made uneventful post-operative recovery. The inside of the tumor showed necrotic material (Figure 6). Approximate weight of the tumor was about 20 Kg. Histo Pathological Examination (HPE) showed extensive areas of cystic degeneration, hemorrhage and necrosis throughout the tumor. There were fascicles of spindle cells (Figure 7) with minimal cytoplasm and nuclear pleomorphism with mild mitosis. Ovaries, cervix and endometrium were normal and final diagnosis on HPE was 'Spindle Cell Sarcoma' of uterus. Patient was advised to go to an oncology center for second opinion but she could not go there due to financial constraints. Repeat USG showed disappearance of hydroureter and hydronephrosis. She has been on follow up for one and half year and has no symptoms and imaging studies have been normal.

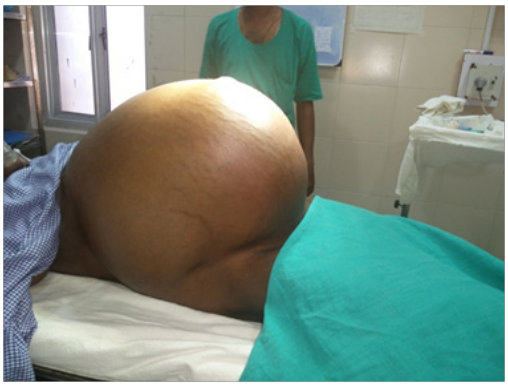

Figure 1 Hugely distended abdomen.

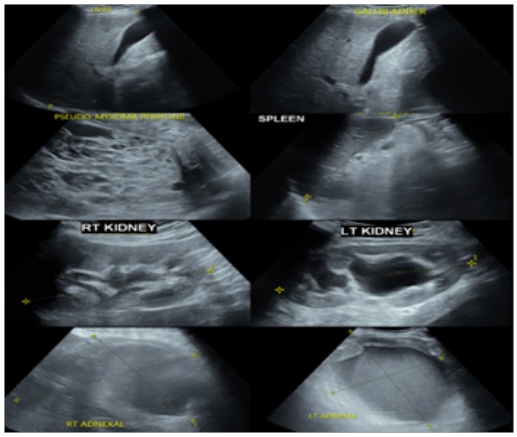

Figure 3 USG showing mixed echogenic mass

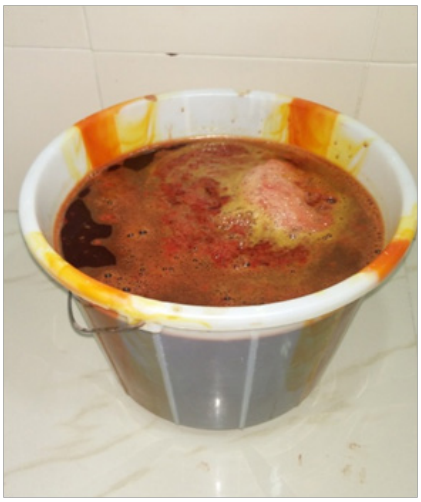

Figure 5 A large quantity of fluid mixed with blood drained out.

Discussion

Uterine sarcomas are rare malignancies accounting for 8-10% of all uterine malignancies[6] with leiomyosarcoma as the most common histologic type. They chiefly occur after the age of 40.[7] The symptoms of uterine sarcomas are similar to leiomyomas such as abnormal uterine bleeding, abdominal enlargement, abdominal pain/discomfort, and pressure symptoms of gastrointestinal or genitourinary tracts.

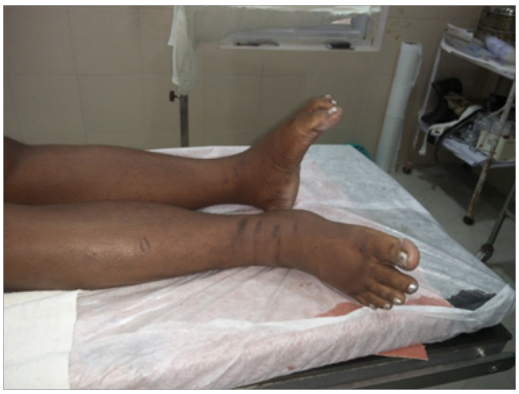

Figure 2 Pedal edema.

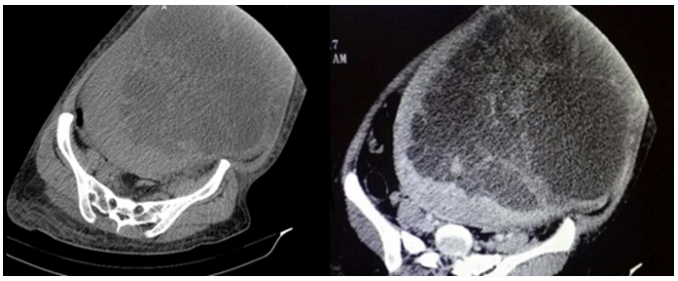

Figure 4 CT Scan.

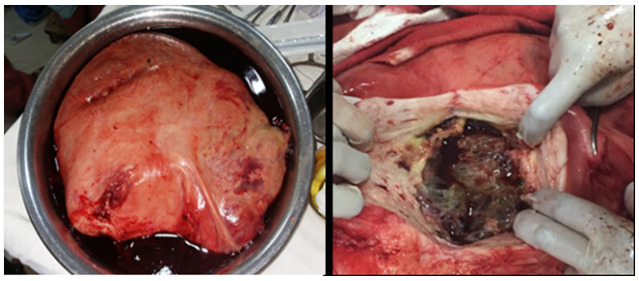

Figure 6 Cut open section showing necrotic material inside.

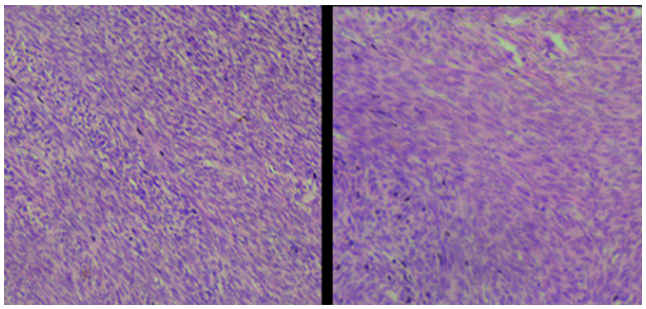

Figure 7 Fascicles of spindle cells in myxoid stroma.

Rupture of uterus and hemoperitoneum are extremely rare.[8] Uterine Spindle Cell Leiomyosarcoma is a morphological variant of uterine leiomyosarcoma. Spindle cell sarcoma is an uncommon variant or subtype of RMS[9] which mostly occurs in the paratesticular region in children.[4] It has very rarely been reported arising from uterus.[10,11] We have presented a rare case of 'Spindle Cell Sarcoma' of uterus which had grown to an unusually big size without involving any other tissue

which is normally unlikely. Our patient had favorable outcome which has sometimes been reported.[5] The diagnosis was not clear pre and per-operatively even after detailed imaging studies; the diagnosis was confirmed after HPE studies which showed fascicles of spindle cells in myxoid stroma (Figure 7). The role of imaging in the assessment of uterine sarcoma has not been well described given the rarity of the disease though some attempts have been made.[2,12,13] Most of these are diagnosed only after submitting the hysterectomy specimen for HPE.[2,14] Even HPE diagnosis is problematic due to many variants, subtle tumor characteristics and overlapping morphologic features that make differentiating these entities perplexing.[15,16] Endometrial biopsies and fine needle biopsies are insufficient and often mystified by sampling errors and bias.[3] Our case of spindle cell sarcoma of uterus is rare, was not suspected preoperatively and imaging studies were not of much help. Diagnosis was made only after HPE and the most unusual finding was purely local involvement and favorable outcome.

Acknowledgement

None.

Conflict of interest

None.

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