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Case Report

Myxoid degeneration of leiomyoma-a masquerader

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ABSTRACT

Uterine leiomyomas are one of the most common benign lesions in the uterus. The lesion can vary in size from a few millimetres to centimetres, location, its presentation and degeneration. They are easy to diagnose clinically with the help of available imaging techniques. Myxoid degeneration is an uncommon degeneration and has to be kept as a differential diagnosis as it mimics uterine sarcoma. Histopathology and Immunohistochemical analysis help in confirming the diagnosis. Here is a patient in reproductive age group who presented with rapidly growing abdominal mass in a span of 1 year with a prior surgical history of myomectomy. Pre-operative imaging was inconclusive for benign etiology and possibility of uterine leiomyosarcoma could not be ruled out. She was hence counselled for a laparotomy with a frozen section to prevent extensive surgery. Myxoid degeneration is not a common degeneration of leiomyoma with a reported incidence of 10% while the incidence of uterine leiomyosarcoma is less than 1% in reproductive age group. Even though rare, myxoid degeneration should be considered as a differential diagnosis of rapidly growing uterine mass with inconclusive pre-operative imaging.

Keywords: Leiomyoma, Myxoid degeneration, Sarcoma

INTRODUCTION

Leiomyoma is the most common benign lesion of the uterus. Twenty percentages (20%) of the women would have got leiomyoma by the age of 30 years but most of them remain asymptomatic. With increase in size of the leiomyoma, they outgrow the blood supply resulting in various types of degeneration like hyaline, cystic, red, calcific and myxoid degeneration.[1]

Hyaline degeneration is the most common degeneration sixty percentages (60%) and rarest among them is myxoid degeneration. Myxoid degeneration presents as a rapidly growing mass mimicking clinical presentation of leiomyosarcoma. This case is reported to highlight the importance of comprehensive history, pre-operative imaging and frozen section to prevent extensive surgical morbidity in young patients.

CASE REPORT

We present a case of 42-year-old para 1 live 1 lady who presented to the out-patient department with heaviness and mass per abdomen since one year. She observed sudden increase in size over 2 months. She had no pain abdomen, no menstrual irregularities. She had no bladder or bowel symptoms and no loss of weight or appetite. She underwent open myomectomy 5 years back for fibroid uterus and had a caesarean delivery 13 years back. She had no past history of hormonal therapy or malignancy. She attained menarche at the age of 12 years and had regular menstrual cycles.

Physical examination revealed a large, palpable, painless abdomino-pelvic mass reaching up to the xiphisternum. The lower margins of the mass could not be well made out. Per-vaginal examination showed uterus enlarged and both

fornices were full. Laboratory tests including serum LDH were within normal limits.

Ultrasonography revealed a bulky uterus measuring 87×54×40 mm and showed two hypoechoic lesions, one in the anterior wall of fundus measuring 205×140 mm suggestive of a large subserosal anterior wall fundal fibroid and other in the posterior wall of the body measuring 27×24 mm suggestive of posterior intra-mural fibroid. Both ovaries were normal in size and echotexture.

MR Imaging revealed uterus to be grossly enlarged with a large heterogeneous mass measuring 180×200 mm extending from the anterior serosal border and filling the abdominal cavity. The lesion showed areas of restricted diffusion and T2 hyper intensity with linear septations and soft tissue component. Endometrial thickness was 9.5 mm and no fluid in the cavity. The final impression was a large uterine mass with soft tissue, myxoid component and septations filling the abdominal cavity causing mass effect on the bowel, uterine Sarcoma (Figure 1 and 2).

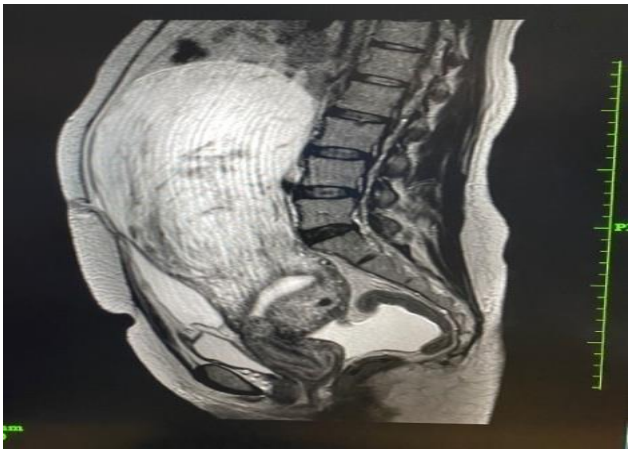

Figure 1: Sagittal section of T2 weighted image of the uterus.

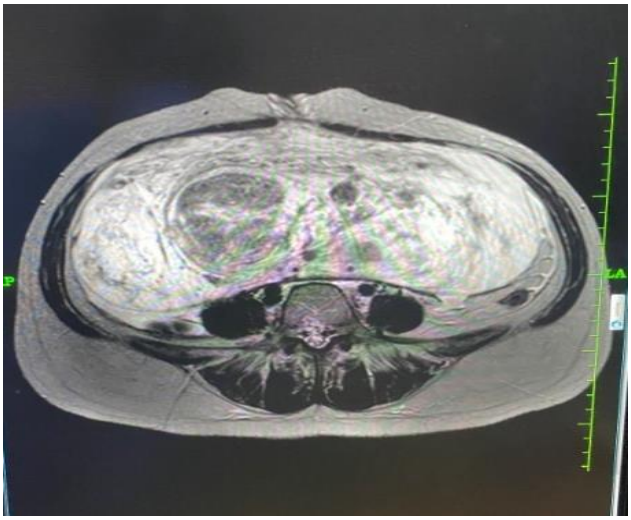

Figure 2: Coronal section of T2 weighted image of the mass.

Intra-operative findings revealed large, cystic, irregular mass, looking like an ovarian cyst with large vessels running on the surface. On thorough examination, the mass was seen arising from the fundus of the uterus measuring around 20×20 cm and occupying whole of the abdominal cavity and looked like a fibroid with cystic degeneration rather than a sarcoma (Figure 3-5). The mass weighed 2 kg. Bilateral tubes and left ovary were normal. Right ovary had a small hemorrhagic cyst.

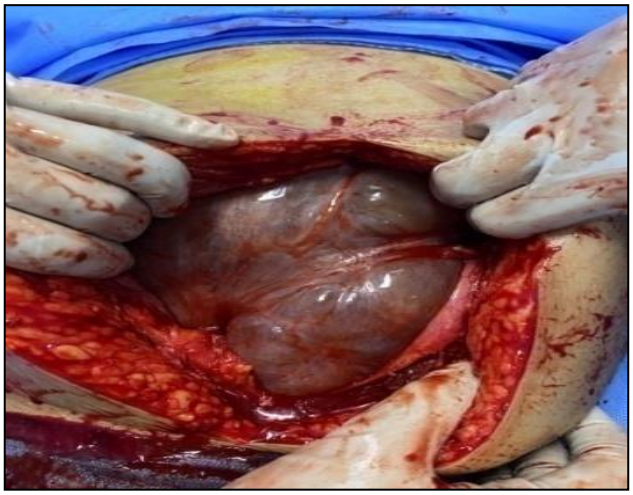

Figure 3: Mass occupying the abdominal cavity which looked cystic.

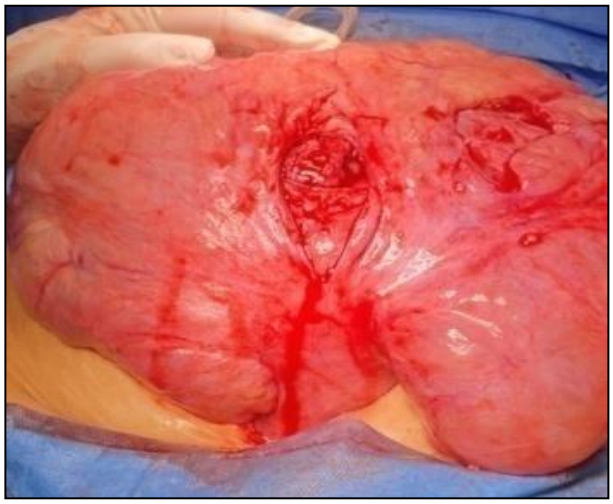

Figure 4: On decompressing and removing the mass outside the abdominal cavity.

She was planned for exploratory laparotomy with frozen section biopsy. Consent was taken for total abdominal hysterectomy and Staging laparotomy if frozen section reported as sarcoma. Patient was also counselled for a second definitive surgery if frozen section was inconclusive. Vertical midline incision was given and abdomen opened. Peritoneal lavage was sent for cytology and malignant cells. Incision made on the lesion and lesion excised and sent for frozen Biopsy. Frozen section reported as myxoid lesion with the possibility of sarcoma

could not be ruled out on frozen section. Hence proceeded with total abdominal hysterectomy with bilateral salpingectomy.

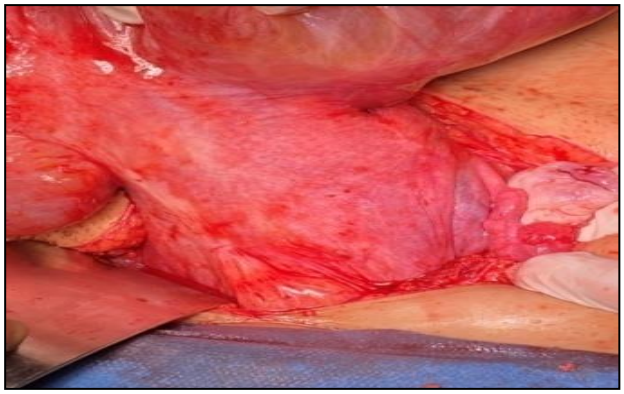

Figure 5: Leiomyoma seen arising from the fundus of the uterus.

In final histopathology, endometrium showed round to tubular glands lined by columnar cells with nuclear pseudo stratification and an oedematous stroma. Section from separate fibroid showed fascicles of smooth muscle cells with oval nuclei and fine chromatin, sections from the raw areas showed small focus of myxoid area with loosely arranged smooth muscle cells. There was no evidence of infiltrative margins or atypia. Peritoneal lavage revealed hemorrhagic clusters of mesothelial cells and negative for malignancy.

DISCUSSION

Leiomyoma is a benign smooth muscle tumor of the uterus. They undergo different types of degeneration like cystic, hyaline, myxoid, dystrophic calcification and red degeneration.[1] Degeneration in leiomyoma has been described in 65% of the cases. Hyaline degeneration is the most common accounting 60%. Myxoid is a rare condition composed primarily of smooth muscle cells with significant accumulation of cellular rich acid mucin. Large thick-walled vessels are left out after this degeneration which is visualised as high vascularity on CT/ MRI imaging.[2] Clinical diagnosis of myxoid degeneration is difficult as it is often asymptomatic. It usually presents as an abdomino-pelvic mass and pelvic pain and poses a diagnostic dilemma. Pre-operative imaging is a must to differentiate leiomyoma from leiomyosarcoma to decrease the risk of upstaging the disease and to reduce surgical morbidity.[3]

MR imaging is well established for diagnosis of leiomyoma but may pose challenges in differentiating degenerations from leiomyosarcoma. A classical leiomyoma without degeneration appears as a rounded or focal mass with well circumscribed margins and homogenously decreased signal intensity on T2 weighted images compared with the outer myometrium. Areas of myxoid degeneration will appear as heterogeneous and markedly increased signal intensity on T2 weighted images with progressive enhancement after contrast administration. The similar findings in myxoid degeneration and sarcoma is lack of cellular and nuclear atypia and presence of mitotic figures in less than two fields out of ten field on microscopy.[4-6] Myxoid leiomyoma is characterized by absence of mitotic activity and the presence of myogenic phenotype.[7]

CONCLUSION

Myxoid degeneration is a rare degeneration of leiomyoma and difficult to diagnose clinically. Pre-operative imaging is a must to differentiate this leiomyoma from leiomyosarcoma to decrease the risk of upstaging the disease and to reduce surgical morbidity. It should be considered as a differential diagnosis in patients of reproductive age group, presenting with rapidly growing fibroids.

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