DOI: 10.7860/JCDR/2023/57875.17329

Case Report

Obstetrics and Gynaecology Section

Uterine Fibroid and Extensive Myxoid Degeneration- Is it Benign or Malignant?

SWATI SUGANDHA[1], GAZAL MAHAJAN[2], MONIKA JINDAL[3], SANTOSH MINHAS[4]

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ABSTRACT

Uterine leiomyomas are the most common benign uterine tumours that may undergo various types of degenerative changes. Rarely, myxoid degeneration can occur in giant leiomyomas that may pose difficulty in diagnosis and may be confused with choriocarcinoma, sarcomatous transformation or even hydatid cyst. This manuscript reports a large leiomyoma weighing approximately 4.5 kg, arising from the posterior wall of the uterus with myxoid degeneration in a 39-year-old female patient. She underwent a total abdominal hysterectomy with bilateral salpingectomy and left oophorectomy. Malignancy was ruled out after a histopathological examination. Through this case, the authors wish to highlight and learn about this rare pathology and understand the diagnostic challenges, thereby helping future gynaecologists to be familiar with the same.

Keywords: Giant leiomyoma, Hysterectomy, Salpingectomy

CASE REPORT

A 39-year-old P4L4 female patient presented with complaints of abdominal heaviness, decreased appetite, easy fatiguability, menorrhagia, and dysmenorrhoea which developed over a period of three to four months. The patient complained of fullness in her lower abdomen and tiredness, while performing activities of daily living. She denied any weight loss or change in bladder and bowel habits. She had all previous vaginal births and delivered her youngest child 21-years ago. She was using a barrier method of contraception. No other significant history was there, except that of type 2 Diabetes Mellitus for three years controlled on oral hypoglycaemic drugs. She attained her menarche at 13 years of age and had regular and normal cycles with the moderate flow (5 days/28-30 days/3-4 pads per day) till she developed the above-mentioned symptoms. Present cycles were of 7-8 days/21-28 days. She used 7-8 pads per day with the passage of clots. The pads were moderate to fully soaked. She experienced pain during her menstruation which was partially relieved using anti-spasmodic medication. There was no history of intermenstrual, postcoital bleeding, or foul-smelling vaginal discharge that excludes cervical malignancy to some extent.

On clinical examination, she was moderately built haemodynamically stable having pallor 2+, but had no oedema or lymphadenopathy. Abdominal examination revealed a 20-22-week pregnant uterus size; non tender mass appearing to be arising from the pelvis, having a uniform smooth surface with regular margins and side-to-side mobility. The lower pole of the mass could not be reached. On per speculum examination, cervix and vagina were healthy. On per vaginal examination, the same mass was felt through both the fornices, but non tender. There was transmitted mobility from the cervix to the abdominal mass. Per rectal examination confirmed the above findings.

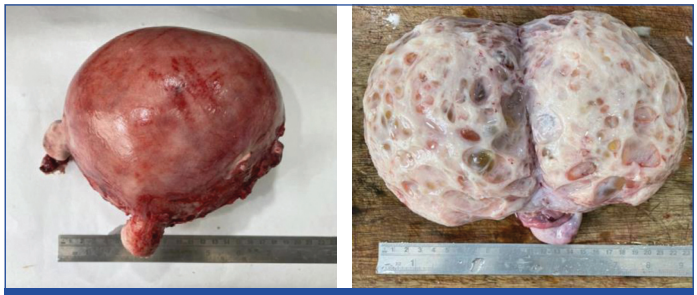
Blood investigation showed a haemoglobin level of 7 gm/dL with normal total and differential blood counts, platelets count, renal function test, liver function test, and blood sugar levels. HbA1c level was 6.8. Other blood investigations including tumour markers (Cancer Antigen (CA)-125, Beta-hCG (human Chorionic Gonadotropin), Lactate Dehydrogenase (LDH), Alpha-Foetoprotein (AFP), Carcinoembryonic Antigen (CEA), CA 19-9) were normal. Her urine pregnancy test was negative. Transabdominal sonography showed a bulky uterus of size 11.8×12.8 cm with a large heterogenous hypoechoic lesion in posterior myometrium 10.8×14.3 cm with multiple cystic areas in it suggesting cystic degeneration of fibroid, hydatid cyst or a malignant transformation.

Magnetic Resonance Imaging (MRI) revealed high signal intensity on T2-weighted images, narrowing the diagnosis to degeneration within the fibroid. However, malignancy could not be ruled out. The patient was enrolled for surgery after obtaining consent and preanaesthetic evaluation. She underwent exploratory laparotomy followed by peritoneal cytology, omental and peritoneal biopsy. There were no palpable pelvic and para-aortic lymph nodes.

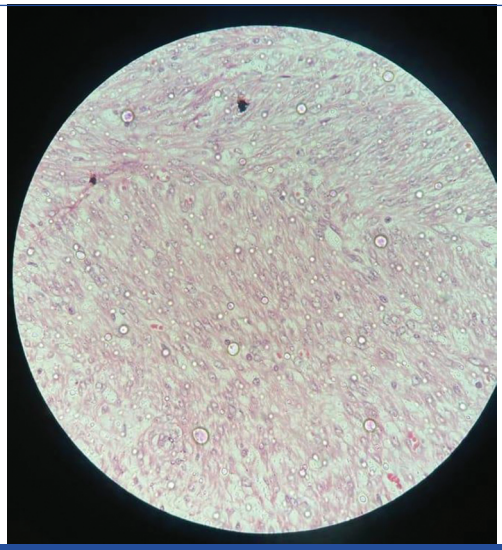
No other evidence of macroscopic metastatic deposits was found in the abdomen and pelvis after a thorough clockwise exploration. The left ovary was enlarged and cystic, measuring 5×3 cm on gross examination. The capsule of the left ovary was intact. The right ovary was grossly normal. There was increased vascularity with engorgement of the feeding vessels of the fibroid. Hence, a total abdominal hysterectomy with bilateral salpingectomy and left oophorectomy was done.

The specimen was sent for the frozen section which ruled out malignancy. Thereafter, one ovary was preserved given her age. Peroperative, findings revealed a grossly enlarged uterus of 22-24 weeks weighing 4.5 kilograms with a smooth surface (21×18×16 cm) and soft consistency, bilateral fallopian tubes were grossly normal (5-5.5 cm) [Table/Fig-1]. The left ovary was bulky and cystic (5×3×5.1 cm) and hence removed. Cut-section revealed the presence of 150 mL of sero-mucinous fluid [Table/Fig-2]. Large fibroid was present in the posterior myometrium and fundus of the uterus of size 12×10 cm with a honeycomb appearance suggestive of myxoid degeneration. The endometrial cavity was distorted, without any intracavitary lesion. The endometrial lining was thin. The cervical canal (3.5 cm) and cervix were normal. On histopathological examination, the endometrium was secretory. The myometrium had a well-circumscribed, encapsulated tumour with cigar-shaped nuclei, fine nuclear chromatin, and small nucleoli, with abundant, eosinophilic, and fibrillary cytoplasm. Within the mass, multiple cystic spaces with mucin were found [Table/Fig-3]. Numerous thin to thick-walled dilated staghorn-shaped blood vessels were

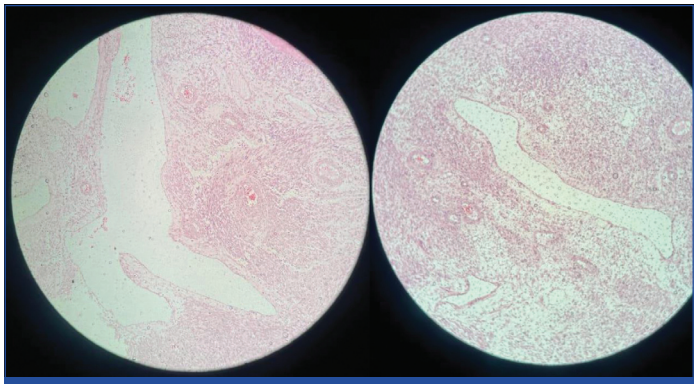
seen in the background. A 1-2 mitosis/10 High-Power Field (HPF) were seen in the myometrium. No tumour necrosis, nuclear atypia, vascular invasion were noted [Table/Fig-4,5].



[Table/Fig-1]: Enlarged uterus.
[Table/Fig-2]: Cut section showing myxoid degeneration of the leiomyoma. Cysts filled with mucinous fluid can be seen. (Images from left to right)



[Table/Fig-3]: Cells with abundant eosinophilic cytoplasm and cigar shaped nuclei at places showing nucleoli-Haematoxylin & Eosin (H&E) stain (High power view).



[Table/Fig-4,5]: Numerous variable sized branched thin and thick walled blood vessels (Staghorn cells) in low power view (H&E stain). (Images from left to right)

Features were suggestive of extensive myxoid degeneration of leiomyoma and the possibility of malignant transformation was ruled out. Postoperatively, the patient recovered well and was discharged on the 7th postoperative day in satisfactory condition.

DISCUSSION

Uterine leiomyoma is the most common benign smooth muscle tumour of the uterus. These may outgrow their own blood supply, enlarge, and can result in various degenerations like hyaline (60%), cystic (4%), calcified (4%); red (3%), myxoid (1-3%); sarcomatous degeneration (0.1-0.8%) as per prevalence [1]. Mostly the sonographic appearance of uterine fibroids are well-defined, solid concentric, hypoechoic masses with a variable amount of acoustic shadowing, however, the sonographic appearance can be quite challenging in cases of degeneration, hence, posing a diagnostic dilemma [2].

Uterine leiomyoma occurs in about 20-30% of women in the reproductive age group, though no age is immune [3]. After the sarcomatous type, myxoid degeneration is the second-rarest degeneration that can occur in a uterine fibroid and is filled with mucinous gelatin-like material [4]. In the current case also, there was a large amount of gelatinous material filled in cystic spaces. Histologically, myxoid leiomyomas are a subtype composed mainly of smooth muscle cells, with a significant amount of cell material, rich in mucin [5]. Clinical diagnosis of myxoid leiomyoma is difficult owing to the rare incidence and sparsity of specific signs and symptoms pointing towards a benign pathology, like in this case.

Radiological investigations like ultrasonography and MRI can help in narrowing the differentials preoperatively, but tissue diagnosis remains the main stay [6]. In ultrasonography, degenerative leiomyoma is commonly hypoechoic and excessive degeneration may be recognised as a cystic pattern [7]. In this case, ultrasonography showed the presence of a large mass with solid and multi-cystic components in the uterus, which led to suspicion. While on MRI, the myxoid component in the leiomyoma is heterogenous, high intensity on MRI T2 weighted images and low intensity in T1 weighted images. Myxoid degeneration is very rare and shows unusual enhancement pattern, because the stroma has intense enhancement but there is no enhancement of the mucin areas [8-10]. It was difficult to diagnose this entity preoperatively as the differential diagnosis varied from benign to malignant pathology, like pregnancy, leiomyoma, sarcomatous degeneration, ovarian malignancy, malignant vascular tumour of genital tract, choriocarcinoma, or even hydatid cyst, based on history, examination and imaging studies.

These hypotheses were rested upon by histopathology which led to the definitive diagnosis of extensive myxoid degeneration. Tissue analysis determined the myxoid nature of the tumour and eliminated the fear of malignancy, especially the myxoid leiomyosarcoma of the uterus, although her LDH were normal. In the present case, during the initial clinical assessment, a baseline diagnosis of degenerated fibroid with malignant degeneration, ovarian cancer, choriocarcinoma, hydatid cyst was considered. Secondly, intraoperative diagnosis of sarcomatous change or extensive myxoid degeneration was made which was ruled out by histopathological findings and abdominopelvic exploration during laparotomy. There was however the absence of macroscopic disease pathology and metastatic deposits in rest of the pelvis and abdominal region. A conclusive diagnosis was made by histopathology which labelled it as leiomyoma with extensive myxomatous degeneration.

CONCLUSION(S)

Myxoid degeneration of fibroid is a rare entity with features ranging from a benign to a malignant pathology, it is of paramount importance to reach a definite diagnosis to avoid any delay in treatment. Although the final diagnosis is reached using histopathology, malignant changes in a fibroid should be kept on high suspicion. Awareness about malignant changes in the fibroid is of prime importance before labelling it as benign or malignant.

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PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Obstetrics and Gynaecology, Maharishi Markandeshwar Medical College and Hospital, Solan, Kumarhatti, Himachal Pradesh, India.
2. Postgraduate Resident, Department of Obstetrics and Gynaecology, Maharishi Markandeshwar Medical College and Hospital, Solan, Kumarhatti, Himachal Pradesh, India.
3. Professor, Department of Obstetrics and Gynaecology, Maharishi Markandeshwar Medical College and Hospital, Solan, Kumarhatti, Himachal Pradesh, India.
4. Professor, Department of Obstetrics and Gynaecology, Maharishi Markandeshwar Medical College and Hospital, Solan, Kumarhatti, Himachal Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Swati Sugandha,
House No: B-503, MMMCH Campus, Kumarhatti, Himachal Pradesh, India.
E-mail: swati.sugandha90@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 20, 2022
- Manual Googling: Oct 14, 2022
- iThenticate Software: Oct 19, 2022 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 18, 2022**
Date of Peer Review: **Jun 22, 2022**
Date of Acceptance: **Oct 20, 2022**
Date of Publishing: **Jan 01, 2023**