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## Case Report

# Uterine myxoid leiomyosarcoma: a diagnostic challenge

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### ABSTRACT

Uterine sarcomas are histologically diverse tumours which comprise about 3-7% of uterine malignancies. Myxoid leiomyosarcoma (MLMS) is an extremely uncommon variant of uterine leiomyosarcoma. This report is of a 35 year old woman who presented with urinary retention and abnormal vaginal bleeding, who underwent a total abdominal hysterectomy for cervical fibroid. A diagnosis of leiomyoma with myxomatous degeneration was made. The woman subsequently developed a recurrence within 5 months of the surgery and was diagnosed with MLMS following biopsy. This case reiterates the uncertainties associated with the diagnosis of MLMS and the need for heightened vigilance in women with myxomatous degeneration of leiomyoma.

**Keywords:** Myxomatous degeneration, Uterine myxoid leiomyosarcoma, Uterine sarcomas

### INTRODUCTION

Uterine sarcomas are histologically diverse tumours which comprise about 3-7% of uterine malignancies.<sup>1</sup> Myxoid leiomyosarcoma (MLMS) is an extremely uncommon variant of uterine leiomyosarcoma (LMS). Its diagnosis is extremely challenging owing to the clinical and histological disparity; an aggressive disease course with seemingly innocuous and bland cellularity. Diagnosing MLMS accurately is critical; unlike other malignancies, mitotic figures and abundant cellularity are not characteristic. We herein present a case of a 35 year old multiparous woman who underwent a total abdominal hysterectomy for a cervical fibroid. This case is important due to initial diagnosis of a benign leiomyoma with myxomatous degeneration, but subsequent clinical deterioration owing to a recurrence and a final diagnosis of myxoid leiomyosarcoma.

### CASE REPORT

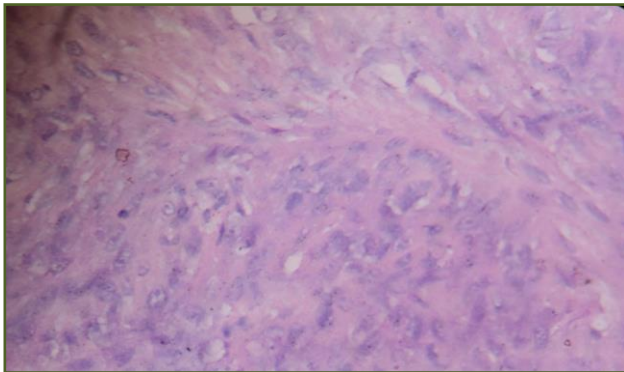
A 35 year old multiparous woman presented to the emergency with acute urinary retention. This was

preceded by difficulty in voiding for three days. She also had episodes of prolonged and heavy menstrual bleeding associated with intermenstrual spotting for 3 months. Prior to this, her menstrual cycles were regular with moderate flow. She had three spontaneous vaginal deliveries and her last child was 3 years old.

Examination revealed a moderate degree of pallor and dehydration, with otherwise stable vitals. On abdominal examination, bladder distension was noted upto the umbilicus and was tender on palpation, which subsided following catheterisation. On speculum examination, a huge smooth mass was noted occupying the entire vagina. Bimanual examination revealed a firm mass obliterating the vagina and the cervical lips could not be delineated. Uterine size could not be appreciated.

On investigating, a urinary tract infection with klebsiella pneumonia was detected for which appropriate antibiotics were initiated. Trans-abdominal ultrasound revealed a large cervical fibroid and an endometrial thickness of 10mm. The ovaries could not be visualised. A diagnosis of cervical fibroid was made.

After correction of anemia with packed cell transfusions, the woman underwent a total abdominal hysterectomy. Intraoperatively, a large degenerated cervical fibroid measuring 10x8x6 cm was noted extending to the isthmus and broad ligament laterally. Both the tubes and ovaries were normal. The patient had an uneventful postoperative period and was discharged on the tenth post operative day after suture removal. On microscopy, histopathology showed leiomyoma with myxoid changes and no associated atypia. The endometrium was proliferative with cervix exhibiting features of chronic cervicitis. The patient was doing well on follow up six weeks after surgery.



**Figure 1: Tumour cells arranged in vague fascicles comprising spindle to polygonal cells exhibiting moderate nuclear atypia, few cells with prominent nucleoli.**

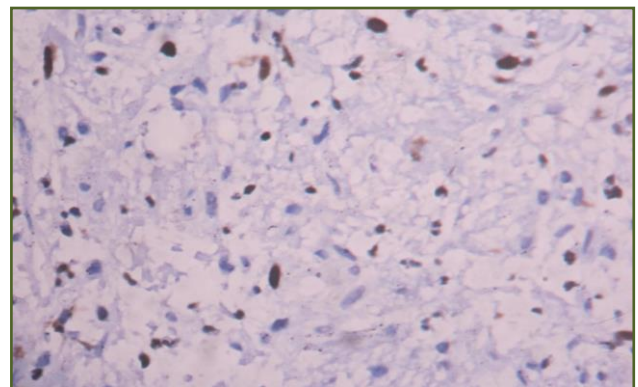
After a period of 5 months, she presented to the emergency with complaints of an abdominal mass for one week and continuous vaginal bleeding for three days. During the interim, the woman had experienced progressive loss of appetite and weight which went unreported.

Clinical examination revealed severe pallor. On abdominal examination, there was a hard mass corresponding to a 24 weeks gravid uterus, arising from the pelvis with restricted mobility. There was no other organomegaly or ascites. On speculum examination, an exophytic mass was noted protruding through the vault. On rectal examination, the rectal mucosa was free and a hard mass was felt in the POD. Investigations revealed moderate anemia (Hb-8gm%), with otherwise normal liver and renal parameters. Tumour markers CA-125 (19.5U/ml) and CEA (<0.5ng/ml) were within normal limits. Chest X-ray revealed a normal study. She was stabilized packed cell transfusions and tranexamic acid for antifibrinolytic milieu for cessation of ongoing blood loss.

Transabdominal ultrasound showed a left retroperitoneal mass compressing the left ureter, with few parietal wall deposits anterior to bladder with multiple deposits in the omentum and pelvis. There was no ascites. On CECT, a large abdominopelvic solid-cystic heterogenous mass

lesion was seen arising from pelvis and eroding the vault. Paraaortic lymphnodes were enlarged, largest of which was 10mm and omental deposits were noted. Ovaries could not be seen separately and liver appeared normal.

An FNAC was obtained from the mass during the radiological examination, which illustrated few scattered foamy macrophages and rare clusters of pleomorphic polygonal to spindle shaped cells exhibiting moderate amount of cytoplasm, ovoid to elongated vesicular nucleus and prominent nucleoli in an abundant myxoid background. No obvious mitotic figures were identified in the smears. Features were suggestive of a mesenchymal tumour with extensive myxoid changes. In view of significant pleomorphism, there was a strong suspicion of malignancy. A biopsy from the vault growth showed spindle cells in the background of myxoid change, moderate nuclear atypia and occasional mitotic figures (0-1/10 hpf) (Figure 1) Immunohistochemistry was positive for SMA, desmin, focally positive for S100 with Ki67 of 2% (Figure 2). It was diagnosed as a malignant tumour of smooth muscle origin, a myxoid leiomyosarcoma. Subsequently, chemotherapy with gemcitabine was initiated in concurrence with the medical oncologist.



**Figure 2: Immunohistochemistry with ki67 stain showing scattered positivity.**

## DISCUSSION

Carcinosarcomas have been redesignated as metaplastic carcinomas according to the recent WHO classification of uterine tumours, following which LMS has emerged the most common sarcoma.<sup>2</sup> Majority of LMS are sporadic and sarcomatous degeneration of a preexisting leiomyoma is unusual.<sup>3</sup> Myxoid MLMS is an extremely uncommon histological variant of LMS. The median age of women with LMS is 50-55 years, with only 15% of the women younger than 40 years.<sup>4</sup>

Clinical presentation includes abdominal mass, abnormal vaginal bleeding or pressure symptoms involving the bowel and bladder. LMS is an aggressive malignancy with poor prognosis and propensity for frequent recurrences.<sup>3</sup> Microscopically, MLMS consists of smooth

muscle cells widely dispersed in abundant myxoid stroma with the characteristic scarce cellularity and low mitotic index. Cellular pleiomorphism and nuclear atypia are commonly noted and there may be associated myometrial or vascular invasion.<sup>2</sup>

Our patient was a reproductive age woman who presented with abnormal bleeding and urinary retention. This is a case in point about the difficulties encountered in identification of MLMS. There were three plausible checkpoints to establish a diagnosis: during clinical evaluation, based on intraoperative findings or postoperative tissue evaluation. The initial clinical examination helped in establishing a baseline diagnosis of cervical fibroid, but it was not possible to differentiate a benign from malignant pathology. Intraoperatively, the gross appearance was that of a large cervical fibroid with degeneration. Histopathology was deemed conclusive in the diagnosis and was labeled as a leiomyoma with myxomatous degeneration. Despite the lack of initial pointers towards malignancy, following the recurrence in 5 months, a diagnosis of MLMS was made.

The initial tissue diagnosis of leiomyoma was made on obvious findings noted on microscopy, with little room for error. Despite the diagnostic expertise, it is not clear whether the sarcomatous transformation occurred subsequently or if the initial microscopic appearance was misleading. Nevertheless, it is important to emphasise the lack of cellular atypia in the initial specimen. This case has been presented to reiterate that a uterine myoma with myxomatous changes must be differentiated from MLMS. Myxoid degeneration in benign leiomyoma has been reported with an incidence as high as 15%, but myxoid transformation in leiomyosarcoma is extremely rare.<sup>5</sup> In retrospect, the first presentation may seem like a missed opportunity for accurate diagnosis. The fallacy lies in the presumption that myxoid leiomyosarcoma is a straight forward diagnosis. Owing to the rarity of these tumours, it is difficult to arrive at an absolute consensus on the diagnostic criteria further compounding the diagnostic dilemma. Under such circumstances, when leiomyoma with myxomatous degeneration is diagnosed,

based on clinical correlation, monitoring of such women may seem pertinent.

## CONCLUSION

Cases of MLMS reported worldwide are few and far between. Diagnosis of MLMS requires meticulous observation and careful differentiation from similar appearing benign conditions. Our experience reiterates the need for vigilance in indicated clinical scenarios and to acknowledge the possibility of MLMS in such cases.

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