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Case Report

Obstetrics and Gynaecology Section

A Perplexing Case of Cellular Leiomyoma with Excessive Myxoid Degeneration

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ABSTRACT

Uterine leiomyomas are the most common benign lesions of the uterus. This tumour can vary in size, location, presentation and degenerations. In most cases, they are easy to diagnose clinically with help of imaging. We present a case of a fibroid with excessive myxoid degeneration where the preoperative diagnosis was haemangio/angiosarcoma. Whereas, cellular leiomyomas are a variant of leiomyoma with increased cellular component, myxoid degeneration in leiomyoma is uncommon and has to be differentiated from leiomyosarcoma. Histopathology and immunohistochemical analysis is confirmatory in arriving at this diagnosis.

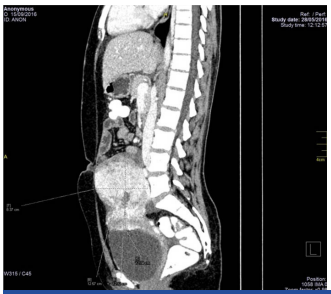
Keywords: Fibroid, Sarcoma, Uterine masses

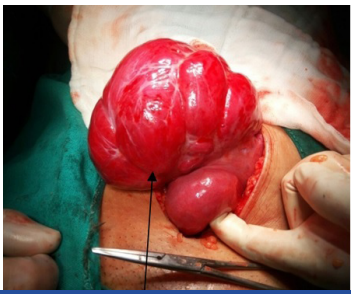
CASE REPORT

A 25-year-old unmarried girl presented to the out-patient department with complaints of mild to moderate pain in lower abdomen for two months which was gradual in onset and not associated with urinary or gastrointestinal symptoms. She attained menarche at 13 years of age, with regular menstrual cycles with mild spasmodic dysmenorrhoea. She was thin built with Body Mass Index (BMI) 19.9 kg/m^2. Abdominal examination revealed an abdomino-pelvic lump of 16 week size (gravid uterus size), 6 x 6 cm above symphysis pubis, with mild tenderness, firm consistency having smooth margins but restricted mobility.

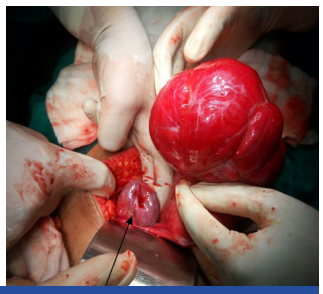
Per abdominal ultrasonography reported a bulky uterus with distorted contours with a highly vascular fibroid like growth of 10 x 8 cm from posterior wall of uterus with degeneration. Bilateral ovaries were normal and mild ascites was present. A previous sonography had reported presence of adnexal pathology with ascites in view of which ovarian tumour markers (B Human Chorionic Gonadotrophin, Lactate Dehydrogenase, AlfaFeto Protein, CA-125 markers) were done and found to be normal except a mildly raised CA-125 value of 122U/ml (normal 0-35 U/ml).

Contrast Enhanced Computed Tomography (CECT) [Table/Fig-1] finding suggested a large exophytic subserosal uterine mass with intense heterogenous enhancement, small nodules and cysts are seen in right adnexa with moderate ascites. In view of soft tissue mass, MRI was done and a lobulated hyper intense enhancing mass lesion was seen involving posterior wall of uterus with marked vascular signals seen in it. Multiple tortuous voids in uterus were seen suggesting areas of necrosis and oedema. Mass in close proximity of uterus appears T2 hyperintense. Differential diagnoses were uterine haemangiosarcoma, chronic angioma or vascular leiomyoma. A second review of MRI suggested the findings to represent fibroid with sarcomatous change and haemorrhage with possibility of metastasis in right adnexa/broad ligament area.

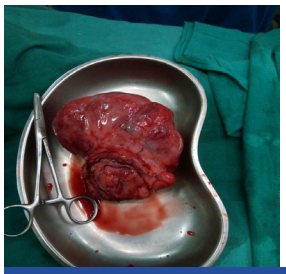


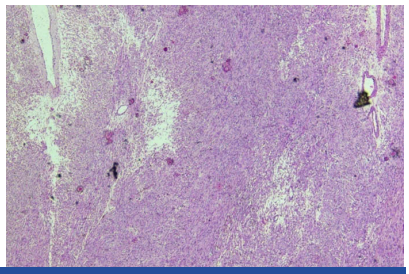


[Table/Fig-1]: CT scan image mass from uterus. **[Table/Fig-2]:** Uterus with mass arising from posterior surface. (Images from left to right)



[Table/Fig-3]: Normal left ovary. **[Table/Fig-4]:** Haemorrhagic right ovary. (Images from left to right)





[Table/Fig-5]: Excised vascular mass. **[Table/Fig-6]:** Microphotograph show hypercellular area of leiomyoma with extensive myxoid degeneration (H&E, 40x). (Images from left to right)

Fine Needle Aspiration Cytology of the mass was done earlier in some other center which reported a benign vascular lesion.

So, clinically an abdomino-pelvic lump gave a varied and confusing picture after imaging reports. Patient was taken for exploratory laparotomy in view of imaging diagnosis of sarcomatous growth from uterus with plan of hysterectomy and exploration of abdomen with full preoperative workup.

On laparotomy, 150ml of straw coloured fluid was present in cul-de sac. Uterine mass [Table/Fig-2] was arising from posterior & left lateral wall,15x12 cm in size, firm to soft in consistency, red, angry looking, vascular and bosselated. Both fallopian tubes and left ovary were apparently normal [Table/Fig-3]. A 2 x 2 cm haemorrhagic cyst was seen in right ovary [Table/Fig-4].

A clear cut diagnosis of fibroid could not be made by looking at the mass. Considering young age and unmarried status and also lack of frozen section facility, a decision for fertility preservation was taken and mass was excised/separated from the uterus, with a clear plane between uterus and the mass which was highly vasulcar. Uterine cavity was not entered during excision of mass. Exploration of the abdomen did not reveal any intra-abdominal deposits or retroperitoneal lymphnodes. Right ovarian cystectomy was also performed.

On gross appearance, the uterine mass was 10x8 cm size, multilobulated, predominantly solid and fibrous with gelatinous material interspersed with some cystic spaces [Table/Fig-5]. The cytological examination of peritoneal fluid did not show malignant cells.

Histopathological findings were of cellular leiomyoma where proliferation of smooth cells showed the mitotic figure count of 2-3/high power field. There were areas in between muscle cells which were rich in acid mucin, excessive myxoid degeneration [Table/Fig-6]. Usually in myxoid degeneration, there is absence of cellular mitotic activity but presence in this case leads to diagnosis of cellular leiomyoma with myxoid degeneration. Right ovary had a haemorrhagic corpus luteal cyst.

Postoperative period of the patient was uneventful. Regular postoperative medications were prescribed, antibiotics, antacids and analgesics were prescribed for a period of five days. In view of pathological report she was advised monthly follow up for 6 months and then 6 monthly for 1 year. Follow up of patient clinically and by imaging was done and found to be normal at all follow up visits till one year after surgery.

DISCUSSION

Leiomyomas are generally firm and rubbery solid, benign smooth muscle tumours of uterus. They can undergo different types of degeneration including hyaline, cystic, myxoid degeneration, dystrophic calcification and red degeneration [1]. Myxoid degeneration of the leiomyoma shows extensive myxoid changes. It is characterized by absence of mitotic activity. Myxoid leiomyomas are histologically a subtype composed mainly of smooth muscle cells, with significant accumulation of a cellular material rich in acid mucins [2].

This form of leiomyoma is often asymptomatic, and is usually discovered by the presentation of an abdominal mass and pelvic pain. Clinical diagnosis of myxoid leiomyoma is difficult. Computed-Tomography (CT) imaging and Magnetic Resonance Imaging (MRI) can help in preoperative diagnosis. The myxoid component in the leiomyoma is heterogenous, high intensity on MRI T2 weighted images and low intensity in T1 weighted images, it seems in low intensity. 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 [3,4].

Cellular leiomyoma is a distinct histomorphologic variant of leiomyoma, defined by the World Health Organization as a leiomyoma with significantly greater cellularity than the surrounding myometrium, but do not have a distinct clinical presentation. They are benign, uncommon variants of the usual leiomyoma and have a good outcome. The treatment approach for patients with cellular leiomyoma depends on the clinical picture, histopathology, and the patient's desire to bear children.

Myxoid leiomyomas accumulate cellular material rich in acid mucins after undergoing degeneration in between the primary smooth muscle cells of the leiomyoma. Large thick-walled vessels are left after this degeneration which is seen on imaging as vascular masses. This case showed high vascularity on USG and CT raising suspicion of haemangio/angiosarcoma.

Malignancy is suspected when the margin of myxoid leiomyoma and its adjacent myometrium becomes infiltrated and indistinct and the cells are atypical [5]. The similar findings in both myxoid leiomyoma and leimyosarcoma are the lack of cellular and nuclear atypia. There is also presence of mitotic figures in less than two fields out of ten fields on microscopy. Whereas, the leiomyosarcomas arising from uterus have high p16, p53 and MIB1 expression, benign leiomyomas, leiomyoma variants and smooth muscle tumours of uncertain malignant potential do not [6].

A case reported by Kamra HT et al., on myxoid leiomyoma of cervix showed no cellular atypia but abundant mucin, oedema, inflammatory cells and blood vessels. The tissue was positive for vimentin and negative for CD34 which clinched the diagnosis as myxoid leiomyoma [7].

Myxoid leimyosarcoma has been described as early as 1998 by Chang E et al., as a rare variant of uterine sarcoma. They have less mitotic figures but have aggressive course where recurrence of disease has been seen. Osteoclast like giant cells also confers aggressiveness to this leimyosarcoma which was not seen in the case reported here [8].

Although MR imaging is well established for diagnosis of leiomyoma, different degenerations may pose challenge in differentiating between them. MR spectroscopy has been reported to distinguish benign from malignant lesions and underlying pathophysiology of uterine leimyomas [9].

It was a diagnostic dilemma as the probable pre-operative diagnosis ranged from benign to malignant causes with differential diagnosis of leiomyoma, leiomyoma with degeneration, ovarian malignancy, malignant vascular tumour of uterus based on history, examination and imaging studies. The speculations were put to rest by histopathology.

Considering that such myxoid degeneration usually occurs in very large fibroids and have been reported in perimenopausal multipara patients, this case is being discussed as it had a relatively small 10x 8 cm leiomyoma in a young 25-year-old girl.

CONCLUSION

This case highlights the need for increased knowledge amongst gynaecologists and radiologist for proper imaging techniques to come to a sound clinical diagnosis prior to operation so that a realistic plan for treatment can be made. Differentiating benign from malignant myxoid leiomyosarcoma is important for follow up management of patients and their prognosis.

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