

Atypical leiomyoma of the uterus: A case report

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ABSTRACT

Introduction: Leiomyoma of the uterus is the most common tumor of the female reproductive tract. While most leiomyomas on histology are usual myomas, sometimes rare variants may be encountered. **Case Report:** We report a case of an atypical uterine leiomyoma in a 49-year-old multipara, who presented with continuous bleeding per vaginum of one month duration and discuss the pathophysiology, salient clinical features and management of this rare condition. **Conclusion:** Atypical leiomyoma is a rare variant of uterine smooth muscle tumors, which mimics leiomyosarcomas, and may even be its precursor lesion.

Keywords: Atypical leiomyoma, Leiomyosarcoma, Uterine smooth muscle tumours

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INTRODUCTION

Leiomyomas of the uterus are the most common tumors of the female reproductive tract. While on histology most leiomyomas are usual myomas, sometimes rare variants of uterine smooth muscle tumors may be encountered [1]. These tumours pose difficulty in diagnosis, management and prognostication. We report a rare case of an atypical leiomyoma of the uterus, where the diagnosis was made postoperatively, and discuss the pathophysiology, management options and prognosis. The patient provided consent for writing this report.

CASE REPORT

A 49-year-old female, para 2, presented with continuous bleeding per vaginum since last one month. She had two full term normal deliveries before, her last childbirth being 23 years back and had been sterilized. Her previous menstrual cycles were regular with average flow. Her past history was significant for intermittent retention of urine four times in the last three years, for which she had been catheterized at a local hospital. The patient was advised to attend a higher centre for further evaluation, which she did not adhere to. She was a known hypertensive on treatment since the last two years. She had been treated with tablet tranexamic acid 500 mg three times daily for four days from the same hospital before coming to our tertiary centre.

On examination, she was of average built with mild pallor. Her pulse rate was regular at 80 beats per minute and blood pressure measured 110/70 mmHg in the upper extremity; no other abnormality was detected. Per abdominal examination revealed an enlarged uterus approximately 18 weeks in size, firm in consistency with restricted mobility. On per speculum examination, her

cervix was found to be healthy. Per vaginum examination revealed uterus to be enlarged to 18 weeks of gestation with a large anterior fibroid; bilateral fornices were free. Her hemoglobin was 10.5 g/dL and other biochemical parameters were within normal limits.

Abdominal and pelvic ultrasonography as well as plain computed tomography (CT) scan showed enlarged uterus measuring 12.11 x 6.78 x 4.67 cm (Figures 1 and 2A). A large rounded hypoechoic lesion of size 9x7 cm was present in the anterior myometrium displacing the endometrium posteriorly suggestive of intramural myoma (Figure 1). A hypoechoic lesion of size 3.4x4 cm was also seen in the left lobe of liver, suggestive of hepatic cavernous venous malformation (hemangioma). This was confirmed on CT (Figure 2B); conservative management was advised for this incidental finding.

With a provisional diagnosis of fibroid uterus (myoma) with intermittent retention of urine and heavy menstrual bleeding, the patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy (Figure 3) under regional anesthesia. On gross examination, the uterus with cervix and bilateral appendages measured 13x16x8 cm (Figure 4). Cut section showed a large intramural fibroid in the fundus and body of uterus measuring 8.5x6 cm, which was greyish white, firm, solid with whorling.

Histopathology revealed a well-circumscribed tumor comprising of spindle cells in long fascicles and whorls (Figure 5), with the cells exhibiting marked degree of pleomorphism and high nuclear-cytoplasmic ratio with prominent nucleoli. Tumor giant cells and scanty mitotic figures of 1–2 mitosis /10 high power fields were also seen without any evidence of coagulative necrosis (Figures 6 and 7). These histological features were consistent with atypical leiomyoma.

The patient was put on follow-up and is currently six months postoperative with no signs of recurrence and no increase in size of the hepatic hemangioma.

DISCUSSION

Atypical leiomyoma, also known as symplastic, bizarre or pleomorphic leiomyoma is a rare variant of uterine smooth muscle tumors that include at least six major histologically defined tumor types: leiomyoma, mitotically active leiomyoma, cellular leiomyoma, atypical leiomyoma, smooth muscle tumor of uncertain malignant potential and leiomyosarcoma [1].

These variants of leiomyomas are important to recognize as they not only closely mimic the malignant counterpart, i.e., leiomyosarcoma, which is an aggressive tumor, but also because atypical leiomyoma may be a precursor lesion of leiomyosarcoma [1]. Atypical leiomyoma is characterized by the presence of bizarre shaped, multilobated or multinucleated hyperchromatic nuclei, with a low mitotic count, sometimes up to 7/10 high power field by the highest count method. However,

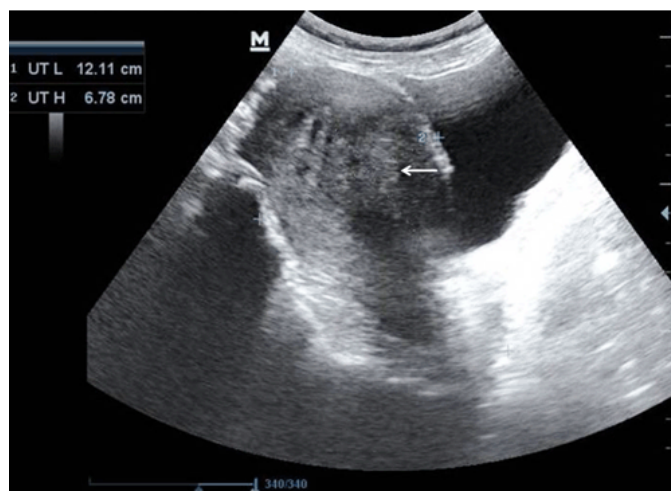


Figure 1: Ultrasonography of abdomen and pelvis showing enlarged uterus measuring 12.11x6.78 cm, with a large anterior myoma (arrow) measuring 9x7 cm.

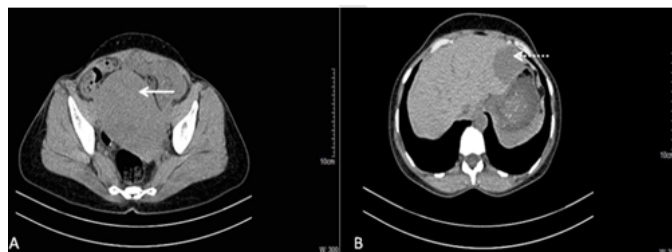


Figure 2: Plain computed tomography abdomen showing (A) enlarged uterus (arrow), (B) Space occupying lesion in left lobe of liver, measuring 3.4x4 cm (dashed arrow).

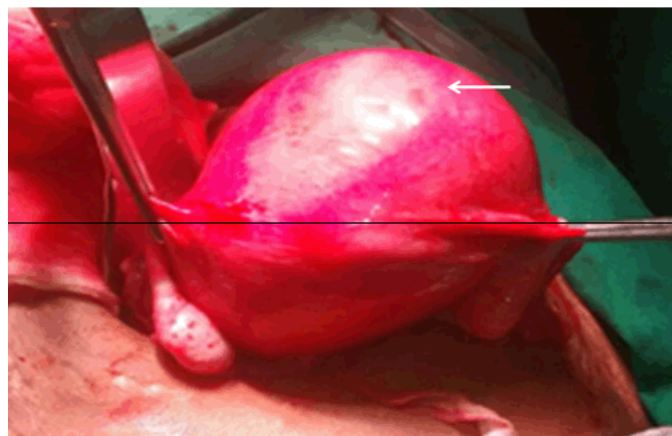


Figure 3: Intra-operative picture showing the large myoma (arrow) bulging anteriorly.

it is differentiated from leiomyosarcoma by the absence of tumor cell necrosis and mitotic counts <10/10 high power field [2].

The exact incidence of atypical leiomyoma is not known. In an analysis of the peritoneal dissemination complicating power morcellation of uterine mesenchymal neoplasms, Seidman et al. [3], found that out of 1091 provisionally diagnosed cases of clinical fibroids which

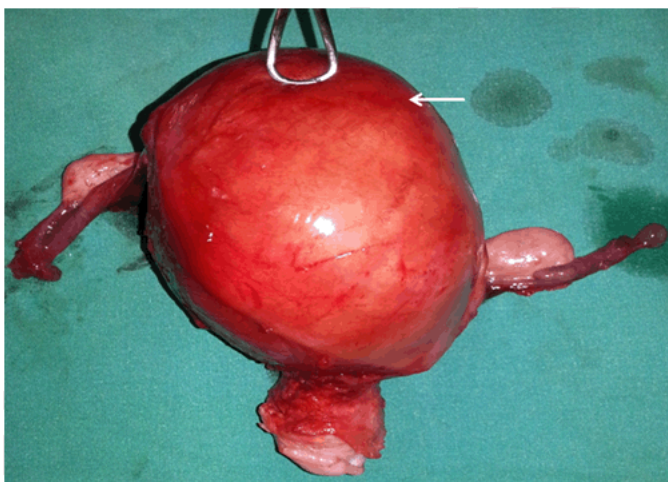


Figure 4: Surgical specimen showing the enlarged uterus with myoma (arrow) and bilateral ovaries and fallopian tube.

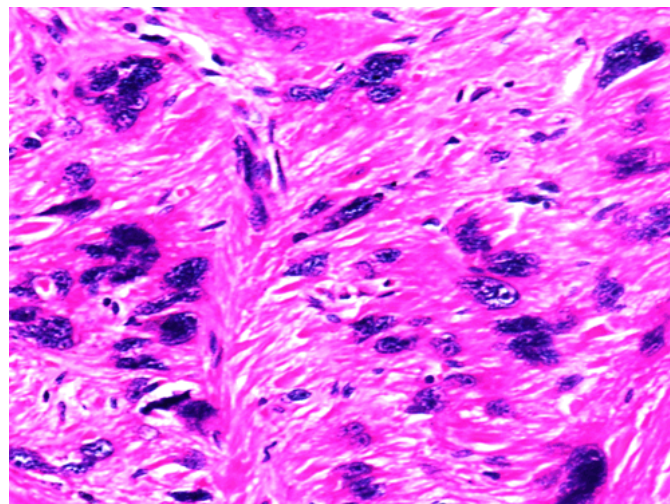


Figure 7: Pleomorphic spindle cells with high nuclear-cytoplasmic ratio with prominent nucleoli and scanty mitotic figures (H&E stain, x400).

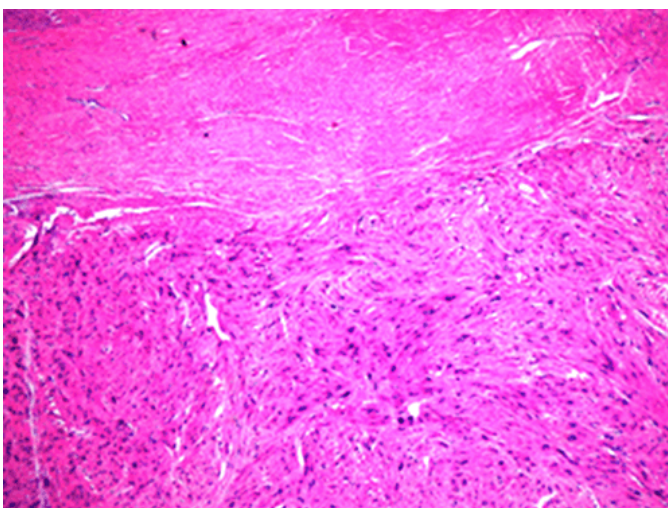


Figure 5: Spindle cells in fascicles with marked nuclear atypia (H&E stain, x40).

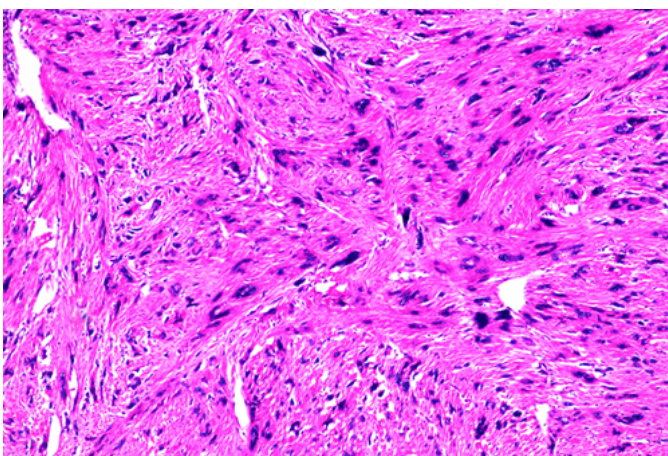


Figure 6: Pleomorphic spindle cells with no necrosis and scattered mitosis (H&E stain, x100).

underwent morcellation, unexpected diagnoses of leiomyoma variants or atypical and malignant smooth muscle tumors occurred in 1.2% of cases, while atypical leiomyoma was detected in only 6 cases. Similarly,

Abraham et al. [4] studied 200 consecutive cases of leiomyomas over a two-year period and found only two cases of atypical leiomyomas, giving an incidence of 1%.

Ly et al. [5] in a clinicopathological study of 51 cases of atypical leiomyoma reported average tumor size of 6.8 cm and an average patient age of 42.5 years. Deodhar et al. [6] reported 21 cases of atypical leiomyoma/ smooth muscle tumor of uncertain malignant potential over nearly a seven-year-period with 12/21 patients fitting a diagnosis of atypical leiomyoma, atypical leiomyoma with a low risk of recurrence or symplastic leiomyomas. They found the mean patient age of 45 years and median tumor size of 10.9 cm. Our case was also similar to those reported in literature with tumor size of 8.5x6 cm and patient age of 49 years.

While the diagnosis of atypical leiomyoma is traditionally made by histopathology, attempts have been made to establish a preoperative diagnosis. In a retrospective analysis of 24 cases of atypical leiomyoma, Jiang et al. [7] reported that on preoperative magnetic resonance imaging (MRI), atypical leiomyoma was seen as a solid tumor mass surrounded by cystic degeneration, pseudotumors, or as a solid mass with homogeneous signal intensity. All but 2 of the 24 cases had lesions appearing as solid cystic mass, whose solid part showed hypo- or iso-intense signals on T1-weighted imaging and moderate hyper intense signals on T2-weighted imaging, with heterogeneous enhancement after contrast agent injection. The authors concluded that evaluation of the relationship between the solid mass and cystic portion and observation for the presence of low signal on T2-weighted imaging may help in the preoperative diagnosis of atypical leiomyoma.

Matsuda et al. [8] reported that the preoperative diagnosis of smooth muscle tumor of uncertain malignant potential may be improved by using a combination of immunohistochemical (IHC) and clinical findings. They

stated that since treatment of symptomatic fibroids includes uterus preserving options like gonadotropin releasing hormone analogues, uterine artery embolization and MRI guided focused ultrasound surgery which may not yield surgical specimens for histological diagnosis, pretreatment differentiation of these smooth muscle tumors of uncertain malignant potential is important. While imaging modalities like MRI scan and fluorodeoxyglucose positron emission tomography (FDG-PET) scan have been found to be useful, they are not accurate. The authors performed transcervical needle biopsy with IHC in over 600 patients to improve the preoperative diagnosis of smooth muscle tumors of uncertain malignant potential.

In a 13-year retrospective analysis of cases between 2000 and 2013, they found 34 cases of smooth muscle tumors of uncertain malignant potential with seven cases of atypical leiomyoma [8]. IHC findings of low molecular mass polypeptide 2 (LMP2) and Ki-67 score and clinical findings (menopause, serum lactate dehydrogenase level) were significantly different between usual leiomyomas and atypical leiomyomas, between usual leiomyomas and leiomyosarcomas and between atypical leiomyomas and leiomyosarcomas. The degree of malignancy associated with atypical leiomyoma is unknown [8]. However, atypical leiomyoma was found to have a low rate of intra-abdominal, extra uterine recurrence (<2%) and a negligible risk for distant metastasis [5].

None of the patients with atypical leiomyoma in Seidman's series had disseminated disease after power morcellation, unlike smooth muscle tumor of uncertain malignant potential or leiomyosarcoma [3]. Therefore, in patients desiring future fertility, treatment by myomectomy alone may be performed and patients should be monitored for local intrauterine residual/recurrent disease [5].

Our case is unique because the patient gave a history of intermittent retention of urine for the last three years with continuous bleeding per vaginum only for the last one month. Therefore, whether our patient initially had a usual leiomyoma which had an atypical transformation can at best be only speculative. The second important question is if left untreated, would it degenerate into a leiomyosarcoma? This is important, because Zhang et al. [1] reported that the six types of uterine smooth muscle tumors have different gene mutation fingerprints and atypical leiomyoma shares many molecular alterations with leiomyosarcoma like P53 mutations and PTEN deletions.

A thorough literature search did not reveal any association of benign hepatic hemangioma with atypical leiomyomas of the uterus; hence it is probably represents an incidental finding.

CONCLUSION

Diagnosis is usually made postoperatively on myomas of relatively larger size, occurring in women in their

forties. Preoperative diagnosis can be aided by MRI, FDG-PET, transcervical needle biopsy with IHC and clinical findings. Hysterectomy is the usual mode of treatment to rule out leiomyosarcoma, although myomectomy may be performed in women desiring future fertility. Follow-up without adjuvant therapy is recommended.

Author Contributions

Sweta Singh – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Monalisha Naik – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Jagadish Chandra Behera – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Pritinanda Mishra – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mamita Nayak – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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