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Giant Angioleiomyoma of Uterus: A Rare Case with its Management.

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Abstract

Introduction: Uterine angioleiomyoma is one of the rarest forms of leiomyoma of the uterus. Very few cases have been reported so far. We have come across an unusually large angioleiomyoma of the uterus in a 45-year-old female which was diagnosed on histopathologic examination.

Case report: We report a rare case of a 45-year-old woman detected with giant angioleiomyoma of uterus along with endometrioma of right ovary and grade 4 endometriosis. She had come to us with gradual distension of abdomen and irregular menstrual periods and associated pain abdomen for nearly one year. On abdominal examination the uterus was 36 weeks size. She was taken up for hysterectomy in view of large uterine fibroid. On histopathology she was diagnosed with uterine angioleiomyoma.

Conclusion: The definitive diagnosis of angioleiomyoma can only be made postoperatively with the histopathology and immunohistochemistry report. The management of such a huge mass is best possible with open surgery as compared to laparoscopy.

Key words: Giant angioleiomyoma, Abdominal mass, uterine fibroids.

Introduction

Angioleiomyoma (AL) is a rare variant of fibroidscontaining thick-walled vessels and arising from smooth muscle cells of the blood vessels **[**1**]**. It is usually encountered in the subcutaneous tissue, mostly in the lower limbs **[**1**]**, occasionally it can be found in the submandibular gland **[**2**]**, in the head and neck region **[**3**]** and retroperitoneum **[**4**]**. This unusual pathology is rarely seen in the female urogenital tract. It accounts for 0.34%-0.40% of uterine fibroids **[**5**]**. Only a few cases have been reported in literature **[**6-18**]** Most cases were diagnosed postoperatively, and most of them were small in size. Ovary **[**19**]**, Cervix **[**20**]** and the broad ligaments are the other occasional sites of presentation of AL. We came across an unusually large angioleiomyoma of the uterus in a 45-year-old female which was diagnosed on histopathologic examination.

Aims and Objectives:

1. To present the diagnosis and management of an unusual case of angioleiomyoma of the uterus.

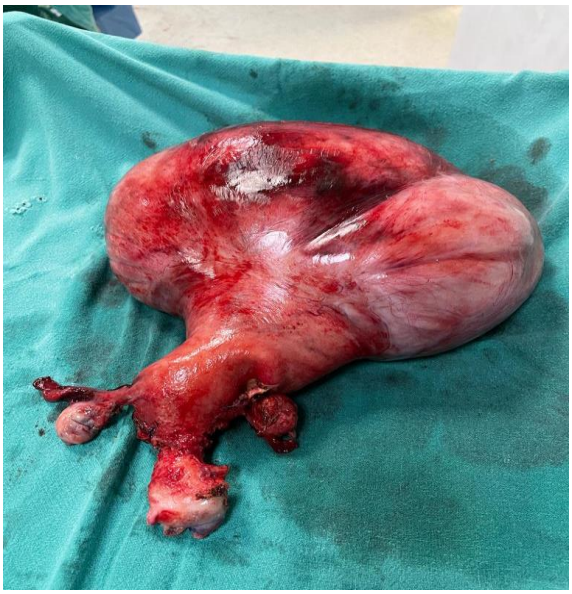
2. To review the available literature on angioleiomyoma of the uterus.

Material and Methods

Here we retrospectively present our experience on in managing an unusual case of giant Angioleiomyoma of the uterus and also analysis of accessible literature research reports on uterine angioleiomyoma, a variant of leiomyoma.

Case Presentation

We present a case report of a 45-year-old Para2Live3 with 5 previous Abortions lady who presented to the gynecology out-patient department with irregular menstrual bleeding and abdominal discomfort along with abdominal distension. The general physical examination of the patient was within the normal limits. On per abdominal examination the uterus was 36-week size with fullness of flanks. The per speculum examination revealed hypertrophy of the cervix. Chest X-ray and all the laboratory parameters were within normal limits. Ultrasound of the pelvis showed a large well defined homogeneously iso-hypoechoic focal lesion seen originating from the fundal subserosal region predominantly on the right side and extending cranially into the peritoneal cavity in the midline, measuring approximately 22.0X 18.4X 15.0 cm. (volume: 3959cc), the uterus was displaced posteriorly. The endometrial thickness measuring 9.6mm and cervix appeared normal in size. The Pap smear of cervix was negative for intraepithelial lesion or malignancy. The endometrial biopsy showed proliferative endometrium. The patient was taken up for total abdominal hysterectomy with bilateral salpingoophorectomy. Intraoperative the uterus was enlarged up to 36-week size, bowels were adhered on the fundus and the



posterior surface of the uterus- adhesions removed, right ovarian endometrioma present, Both the ovaries were stuck with each other and with the posterior surface of the uterus (Figure1).

Figure1: Large Subserosal Angioleiomyoma with Uterus and Bilateral Ovaries

The specimen for histopathologic examination (HPE) was submitted which revealed Angioleiomyoma of the uterus. The details of the HPE are as follows:

Pathological examination: Uterus with cervix measuring 10x8x6 cm and weighing 3.4 kgs. Cervix externally keratinized. Endocervical canal measuring 3.0 cm in length. Endometrial cavity measuring 4.5 cm in length. Endometrium measuring 0.2 cm. Endomyometrium measuring 2.8 cm. A large subserosal mass at fundus measuring 25x18x15 cm. Cut section solid grey white. Bilateral adnexa identified. Left side fallopian tube measuring 5.0 cm. Cut section lumen identified, attached ovary measuring 2.5x2.0x1.5 cm. Cut section shows a cyst measuring 0.5x0.5x0.5 cm. Right fallopian tube measuring 3.0 cm. Cut section lumen identified. Fimbrial end identified, attached ovary measuring 3.0x2.5x1.0 cm. Cut section shows a cyst measuring 1.5x1x1 cm. (Figure 2)

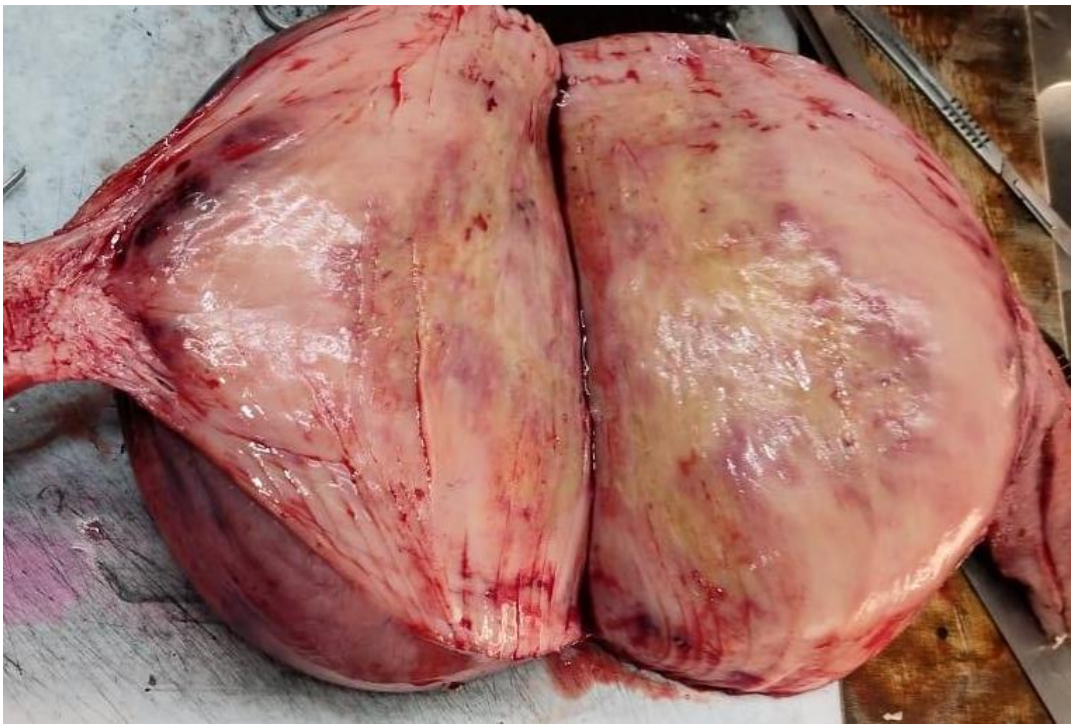


Figure 2: Large subserosal angioleiomyoma showing hemorrhagic areas and normal uterine cavity

Histologic examination

Uterus: Sections from large subserosal tumor showing tumor present in vague fascicles with edema. Tumor cells are having clear to eosinophilic cytoplasm with ill-defined cytoplasmic boundaries, oval to spindle nuclei. There are presence of numerous thin and thick walled vessels with focal area of infarct necrosis. No atypia / increased mitosis seen. (Figure 3 and 4)

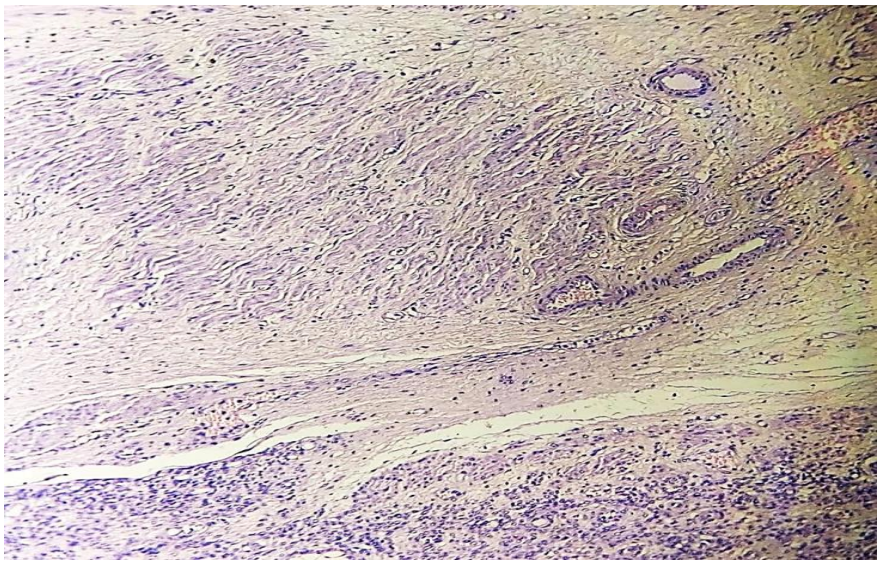


Figure 3: Angioleiomyoma with normal myometrium 40x; Angioleiomyoma and normal myometrium interface with no infiltration

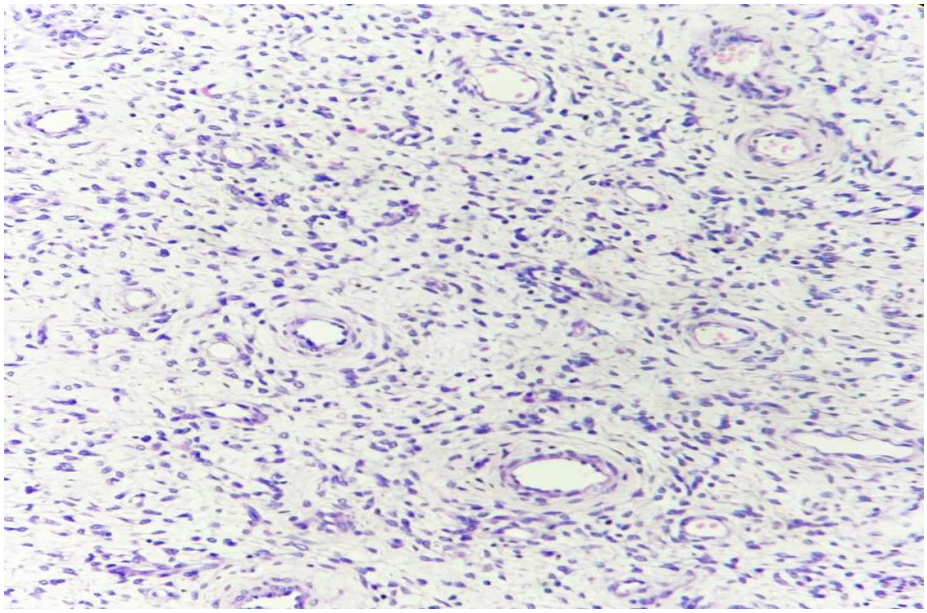


Figure 4: Angioleiomyoma at 100x; Tumor with bland spindle cells with no atypia and mitosis; Tumor with numerous interspersed vascular channel

On immunohistochemistry, tumor cells are positive for desmin(figure- 4) while negative for CD 10 (figure-5). However, CD 10 highlight the vessels.

Endometrium: Endometrial hyperplasia without atypia.

Cervix: Acute on chronic cervicitis.

Bilateral ovaries: are showing focus of endometriosis.

Bilateral fallopian tubes: unremarkable.

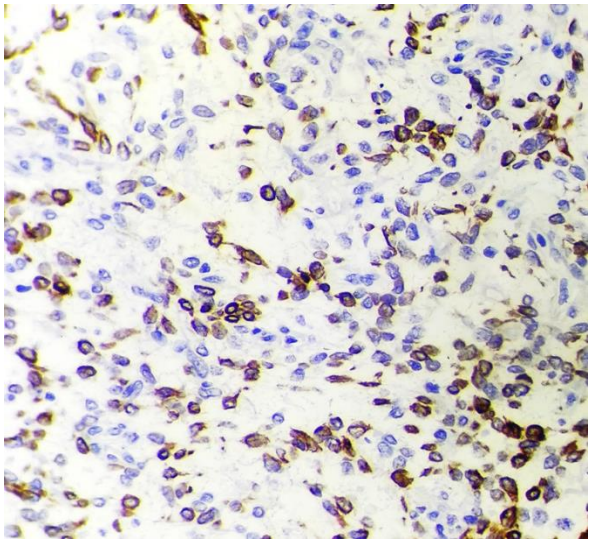


Figure 5: Desmin is positive in spindle tumor cells in angioleiomyoma

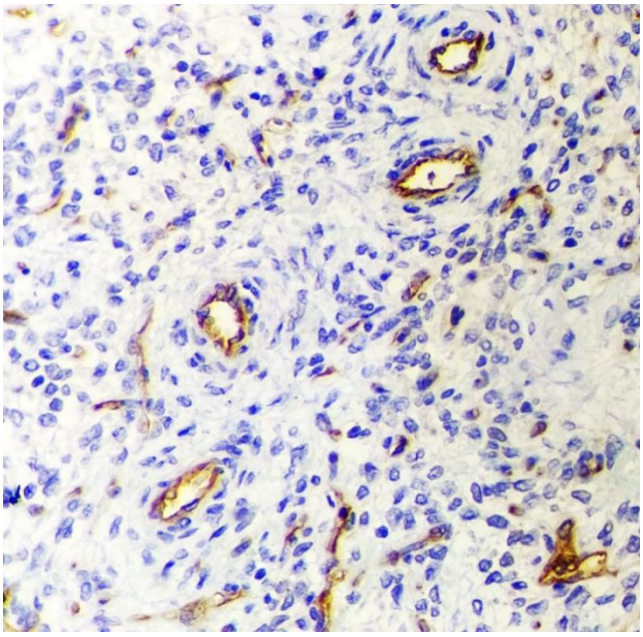


Figure 6: CD 10 highlights the endothelial cells in numerous vascular channels in angioleiomyoma

Anaesthesia consideration:

The main challanges in anesthetic management of such cases are the influence on respiration and circulation of tumor pressure on the great vessels and lungs, risk of aspiration at intubation, risk of massive bleeding, and occurrence of re expansion pulmonary oedema (RPE).Due to these risks, it has been recommended that the tumor mass is reduced preoperatively to prevent circulatory and respiratory depression. However, these cases involved cystic type tumors from which cystic fluid could be drained slowly. By contrast, our case had a solid mass tumor that could not be drained preoperatively. A further problem with giant abdominal tumors is the ventilatory management after administration of a muscle relaxant due to decreased lung and thoracic compliance caused by relaxation of the diaphragm and the enlarged abdomen, and the possibility of high airway pressures causing lung injury.Also, these giant abdominal masses that are sometimes very close to great vessels and may have large feeding vessels and adhesion to nearby structures, therefore, there is a risk of massive bleeding. In our case, there wasn't huge blood loss with resection of the tumor and extensive transfusion was not required. However, we prepared for blood loss and cardiac failure preoperatively by cannulation and preparation for transfusion, and we were able to maintain stable hemodynamics throughout the surgery.

In our case, the lungs had been collapsed by the giant tumor for a long time and the patient had poor pulmonary function preoperatively. Therefore, she was a high-risk case for RPE and there was possibility of development of RPE during surgery. To prevent this occurrence, we chose to re-expand the collapsed

lungs very slowly and we maintained a relatively low TV after removal of the tumor, similar to that during spontaneous respiration preoperatively. Using this approach, we were able to manage the patient uneventfully during and after the operation.

Discussion

Leiomyomas are considered the most common neoplasms of the uterus. The leiomyomas have got different types such as epithelioid, cellular, lipoleiomyoma, myxoid etc., and amongst these variants AL which also known as vascular leiomyoma is an extremely rare variety.

AL is further classified into 3 histologic types on the basis of the relationship between vascular cavities and smooth muscles within the tumour: cavernous type, capillary or solid type and venous type **[**21**]**. These are well circumscribed tumours and usually are smaller than 2 cm in their dimensions, ranging from 0.2-4.3 cm **[**14**]**.

AL in the uterine corpus is usually presented in the middle-aged females **[**22,14**]**. The age range in the cases described is from 30 years to 69 years. Presenting symptoms in most of the cases are heavy menstrual bleeding, discomfort or pain in abdomen, distension of abdomen and/or abdominal mass **[**22**]**. In a case report presented by Handler et al. have described a case a large degenerated AL with secondary consumptive coagulopathy **[**8**]**. In some cases, it may be associated with Pseudo-Meig syndrome and increased serum CA125 levels and may simulate ovarian neoplasm **[**9**]**.

In most of the literature the common presentation of AL is a well-circumscribed mass arising from the body of uterus. Uterine AL in the largest dimension ranges from 4cm. to 30 cm. but only few cases have been reported of size more than 20cm. Only a few cases have reported multiple uterine AL **[**6,14**]**. These can be found in the subserosa, submucosa or can be intramural **[**6-18**]**. In the preoperative period the subserosal variety arising from posterior uterine wall,occupying pelvic cavity may confuse with primary ovarian neoplasm **[**6**]**.

Sikora-Szczęśniak DL. et al **[**23**]**. In their study reported total 9 cases of AL showed associated pathologies such as leiomyoma and endometriosis in 1 case, multiple leiomyomas in 1 case, endometriosis in 2 cases, erosion in 3 cases,thecoma of the right ovary – 1 case,paratubal serous cyst in 2 cases.

Usually there are no features suggestive of mitosis, nuclear atypia, pleomorphism, or necrosis on microscopy of AL cases. On the contrary in few cases authors have presented nuclear atypia in individual cases of AL **[**10,11,24**]**. Our case showed no atypia on microscopic examination.

Ischaemia leads to degeneration in angioleiomyoma and the type of degeneration varies with severity and duration of the vascular insufficiency**[**15**]**. Some cases have reported hyalinisation, mucoid areas,

fatty deposits and calcification [10,25,14,24]. Many cases have reported fibrin accumulation in the vasculature of AL [8,11,24,26].

Postoperative course was uneventful in all the reported cases and also symptoms were clinically improved without any recurrence. As AL of the uterus is a benign in nature, the main stay of the treatment remains its complete excision. In most of the literature the surgical treatment is hysterectomy with or without salpingoophorectomy; only in cases where fertility preservation is needed myomectomy can be considered if technically possible [23,11]. In our case the patient had completed her family and there were gross adhesions to the surrounding structures, associated endometriosis and the size of the tumour being big were the reasons behind the decisions for considering total abdominal hysterectomy with bilateral salpingoophorectomy.

The analysis of previously reported cases showed that final diagnosis of AL can only be made postoperatively with histopathological examination reports.

Conclusion

AL is an unusual variant of smooth muscle neoplasm of the uterus that shows unusual morphology and immunohistochemistry features. It is difficult to differentiate AL from other smooth muscle neoplasm because it has no specific imaging findings and hence pre-operative diagnosis is less likely. Hence, it is necessary for the gynaecologists and the pathologists to identify this unusual benign condition and differentiate it from its look alikes including leiomyosarcoma and endometrial stromal tumor by means of proper sampling and when necessary, using a correct immunohistochemistry testing.

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