The Journal of Obstetrics and Gynecology of India (November–December 2022) 72 (S2):S418–S420
https://doi.org/10.1007/s13224-021-01607-6

CASE REPORT

Angiomyolipoma of Uterus: A Case Report

Indrani Roy[1] · Punam Jain[1] · Harveen Kaur[2]

Received: 23 June 2021 / Accepted: 1 December 2021 / Published online: 6 April 2022

Introduction

Angiomyolipoma (AML) is a benign mesenchymal neoplasm consisting of smooth muscle cells, mixture of adipose tissue and blood vessels. The commonest site though being the kidneys, it has also been reported from extra renal sites such as the nasal cavity, liver, alimentary tracts, retroperitoneal area and female reproductive system, including uterus, vagina, cervix and salpinx. The association of epithelioid AML in the absence of tuberous sclerosis in the female genital tract is a rare entity [1]. Only a few cases have been reported of uterine mesenchymal neoplasm. The diagnosis of uterine angiomyolipoma is not clearly appointed, because uterine AML is not officially listed in the World Health Organization's histological typing of female genital tract tumours [2].

Case

P6L6A1, 49-year-old lady presented with complaints of intermittent abdominal pain and swelling of her lower abdomen for 6 months. On general examination, all parameters were normal except that she was hypertensive. Per abdominal examination revealed a mass of approximately 16-week size palpable per abdomen with restricted mobility. On per speculum examination, a fibroid polyp was seen arising from the cavity at the os, with cervix flushed with vagina. On bimanual examination, 18-week size firm mass with restricted mobility was noted. Ultrasound findings were suggestive of a complex POD cyst, and further investigations including CT scan abdomen were advised to rule out underlying malignant aetiology. Total laparoscopic hysterectomy was done. Intraoperatively, a large posterior myoma was diagnosed with extensive degenerative and cystic changes. B/L tubes and ovaries were normal. The patient had an uneventful post-operative period during her hospital stay. The HPE report of the specimen was reported as angiomyolipoma of the uterus. The patient was followed up in opd at 1-month and 6-month intervals and had no complaints or post-operative complications.

Investigations

Radiological Findings

CT scan showed a heterogenous solid–cystic lesion in POD adherent to posterior wall of uterus, with enhancing septae suggestive of degenerated myoma/exophytic ovarian neoplasm of 11.3*7.5 cm. The uterus was bulky. The lesion was abutting the rectum and rectosigmoid junction. Right ovary was seen with prominent follicles. Left ovary was normal. No ascites or lymphadenopathy was noted.

Papanicolaou smear showed benign cellular changes. D/C showed proliferative endometrium. Serum CA 125, carcinoembryonic antigen (CEA), lactate dehydrogenase (LDH), serum beta HCG and alpha-fetoprotein (AFP) were within normal limits.

The patient was posted for total laparoscopic hysterectomy. Intraoperative finding showed a large posterior wall fibroid in the uterocervical junction with extensive degenerative and cystic changes. Both tubes and ovaries were normal. Total laparoscopic hysterectomy was done, and the specimen was retrieved vaginally using an Endopouch bag.

Histopathological examination: Hysterectomy specimen was approximately 15 × 14x7cm with irregular nodular firm surface.

Dr. Punam Jain is a DNB (T), Department of Obstetrics and Gynecology, Nazareth hospital, Shillong, Meghalaya, India; Dr. Harveen Kaur is a MD Pathology, Department of Pathology, Nazareth Hospital, Shillong, Meghalaya, India.

✉ Indrani Roy
indraniroydeb@gmail.com

[1] Department of Obstetrics and Gynecology, Nazareth Hospital, Shillong, Meghalaya, India

[2] Department of Pathology, Nazareth Hospital, Shillong, Meghalaya, India

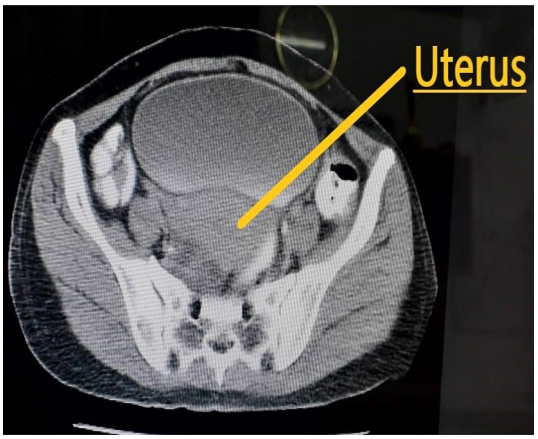

Fig. 1 CT scan of abdomen showing tumour over uterus

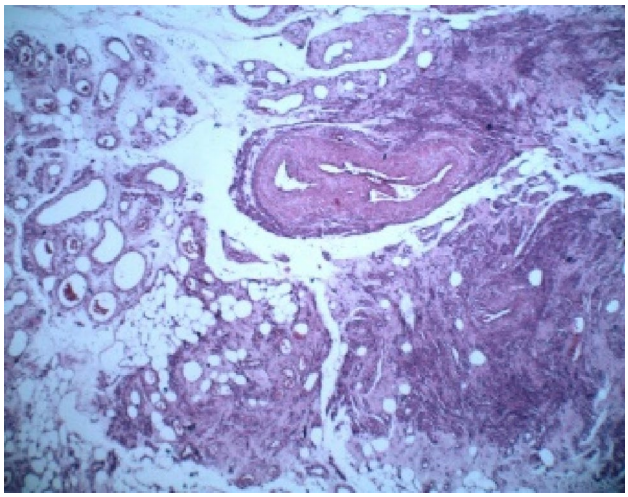

Fig. 2 Thick-walled vessels surrounded by smooth muscle fibres, adipocytes and numerous congested vessels

Gross examination revealed a solid–cystic lesion attached to the right posterior wall of the uterus, extending from the right lower part of the uterine wall to cervix. The lesion measured 6.0 × 4.0 × 3.0 cm. The overlying serosa appeared unremarkable. On the cut surface, the lesion on the posterior surface was solid–cystic and filled with clear fluid. Solid areas appeared greyish yellow.

Sections from the posterior solid–cystic lesion showed a well-circumscribed lesion composed predominantly of spindle cells arranged in a fascicular pattern, at the periphery. The central part of the lesion showed abundant myxoid stroma separated by smooth muscle bundles with focal epithelioid morphology. Intervening areas showed the presence of adipocytes as well as numerous variable-sized blood vessels. There was no evidence of malignancy in the sections examined (Figs. 1, 2).

Discussion

AML had been regarded as a hamartoma rather than as a true neoplasm because it is composed of an admixture of mature adipose tissue, thick-walled vessels, smooth muscle and perivascular spindle cells of varying proportions within the tumour. Primary extrarenal AML is distinctly uncommon and usually develops in the liver, nasal cavity, abdominal wall and fallopian tube [1].

The World Health Organization has defined angiolipomas as a family of neoplasms termed as perivascular epithelioid cell tumours (PEComas), which can include renal or extrarenal AML, lymphangioleiomyomatosis and clear cell tumours of various sites. AML can also be classified as classical or regular and epithelioid. The epithelioid variety can be further classified as epithelioid with atypia and without atypia. Classic AML is a benign lesion characterized by the presence of mature fat tissue, smooth muscle cells and thick-walled vessels with recurrence potential but without any metastasis, whereas epithelioid AML commonly exhibits the proliferation of predominantly epithelioid cells with malignant potential [3]. Most of the cases of extrarenal AML are of the classic type and lack epithelioid components.

MRI is also an important diagnostic tool for AMLs, as the proportion of fat within the lesion is suitable for imaging. High signal intensity on T1 and T2 sequence imaging is associated with high sensitivity in the detection of extrarenal AML. The various immunohistochemical markers which aid in the diagnosis of AML are smooth muscle actin (SMA), desmin, S100, HMB-45 and Melan-A. Positive immunoreactivity for HMB-45 in non-vascular smooth muscle cells is characteristic of AML reported at sites like kidney, colon, parametria and lymph node, but not uterine AML [4]. In our case, none of the immunohistochemical markers were used and the diagnosis was made based on the histopathological examination of the specimen.

The spectrum of clinical presentation in cases of uterine AML is broad and depends on the tumour location and size. Symptoms can include mass per abdomen, abdominal distension, back pain, lower abdominal pain and bleeding per vaginum. Angiomyolipomas are associated with tuberous sclerosis, an autosomal dominant syndrome. Patients with tuberous sclerosis have an increased risk for renal tumours. In the present case, the patient was not associated with tuberous sclerosis which was also the case in most of the reported uterine angiomyolipomas.

Conclusion

We recommend that radiologists should be aware of the imaging characteristics of AML. To conclude, the differential diagnosis of a lower abdominal mass and dysfunctional uterine bleeding should include the AMLs even though the uterus is a rare location for these tumours. The high index of suspicion and classical morphology can help us to clinch the correct histopathological diagnosis in unusual anatomical locations like uterus.

Funding No funding sources were involved.

Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Human and Animal Rights This article does not contain any studies with human participants or animals performed by any of the authors.

References

1. Li W, Wang Y, Wu X, et al. Epithelioid angiomyolipoma of the uterus: a case report and review of literature. Biomed Res (0970–938X). 2019;30(6):1.
2. Monteiro R, Sharma S, Gupta S, et al. Extrarenal angiomyolipoma in uterine cervix: rare presentation in unusual site. Indian J Res Med Sci. 2019;8(1):367.
3. Dutta S, Marbaniang E, Dey B et al. Angiomyolipoma of the broad ligament. Autopsy and Case Reports. 2020; 10(3).
4. Walke V, Gaikwad A, Ratnaparkhi S et al. Angiomyolipoma of the Cervix: a case report.

About the Author

Dr. Indrani Roy MD, FICOG, FIAOG Senior Consultant and HOD Department of obstetrics and gynaecology Post MD more than 18 yrs experience Trained in Laparoscopy, Hysteroscopy, USG and Infertility DNB teacher, NBE faculty. Faculty in several National and Regional Conference and Post graduate training program Thesis guide of several DNB thesis Past President of Shillong OBGYN Society (2017 to 2019) Organizing Chairperson of East Zone Yuva Fogsi 2019 East Zone Cordinator Breast Committee Fogsi Special interest High risk obstetrics , PCOS State Co-ordinator Lakshya project Meghalaya State Co-ordinator for Cervical Cancer and breast cancer Screening from FOGSI.