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Case Report

Uterine angiomyolipoma: A case report of an unusual entity

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ABSTRACT

Angiomyolipoma (AML) is a rare benign mesenchymal neoplasm composed of a variable mixture of smooth muscle cells, adipose tissue and anomalous blood vessels. It belongs to the family of perivascular epithelioid cell tumors (PEComas). It is quite common in kidney but is rare at the extrarenal sites. In the uterus, only a few cases have been reported. We describe a case of uterine AML without evidence of tuberous sclerosis (TS). It was clinicoradiologically mistaken for fibroid. The clinical presentation of uterine AML is similar to more common uterine leiomyomas. AML can be suspected on the imaging but histopathology establishes a confirmed diagnosis. The case is presented for its rarity and likelihood of mistaking it for some other mesenchymal tumors. Overall, just 36 cases of Uterine AML have been reported in the English literature prior to this and this is only the second Indian case.

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1. Introduction

Angiomyolipoma (AML) is a rare mesenchymal neoplasm composed of a variable mixture of smooth muscle cells, adipose tissue and blood vessels.¹ It is commonly seen in the kidney, but has also been described in extrarenal sites such as the liver, retroperitoneum, alimentary tract, nasal cavity and female reproductive system, including uterus, vagina and salpinx.²⁻⁴ Unlike leiomyoma, AML is invariably benign, and usually occur in women older than 40 years.⁵ It is well known that renal AML are often associated with tuberous sclerosis (TS). However, only 7% of patients with uterine AML have TS.^{1,2} We describe a case of uterine AML in a premenopausal lady without evidence of TS.

2. Case History

A 48-year-old premenopausal woman, presented with a history of lower abdominal pain and discomfort and

history of menorrhagia of 4 months duration, unresponsive to treatment. She did not have any prior episodes of abnormal vaginal bleeding. On physical examination, an immobile mass was observed in the lower abdomen. The abdomino-pelvic ultrasonography (USG) examination revealed a 7.5x7 cm mass originating from the posterior wall of body of uterus. No adnexal pathology was observed. The CT examination confirmed the USG findings and suggested a possibility of leiomyoma. With a clinical diagnosis of uterine fibroid, she underwent total abdominal hysterectomy. Macroscopically, a circumscribed solid, greyish yellow mass measuring 7.5x7x6 cm was seen in the posterior wall of body of uterus (Figure 1). Microscopically, the uterus showed a circumscribed tumor composed of many thick-walled large blood vessels surrounded by smooth muscle fibers and admixed with adipose tissue. No nuclear atypia, mitoses or necrosis were seen (Figures 2 and 3). On immunohistochemistry (IHC), spindle cells expressed smooth muscle markers desmin and smooth muscle actin (Figure 4). No immunoreactivity was seen with S-100,

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HMB-45 and Melan A (Figure 5). Hence, a diagnosis of extra-renal AML was made. Further clinical evaluation did not reveal any evidence of TS. The patient is symptom free 10 months after the surgery.



Fig. 1: Gross photograph showing a circumscribed greyish yellow mass in the posterior wall of body of uterus

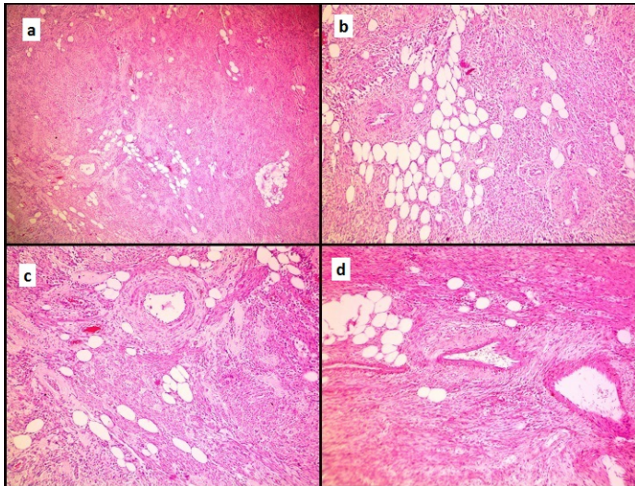


Fig. 2: H & E images showing (a) to (d) a tumor composed of mixture smooth muscle cells, adipose tissue and anomalous large blood vessels. [(a)x40, (b)&(c)x100, (d)x400]

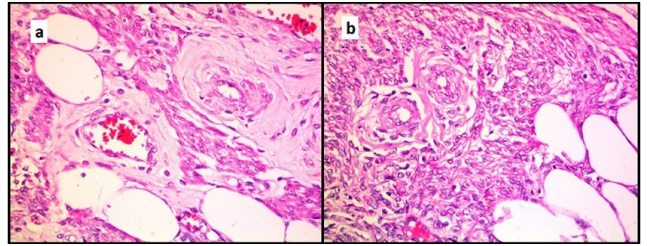


Fig. 3: H & E images showing (a) & (b) a tumor composed of adipose tissue and spindle to oval smooth muscle cells surrounding anomalous blood vessels. [(a&b)x400]

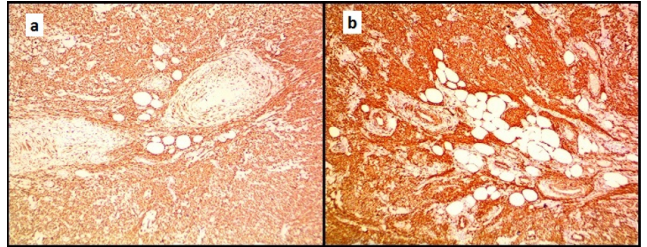


Fig. 4: IHC images showing smooth muscle spindle cells expressing (a) desmin and (b) SMA. [(a) & (b) x100]

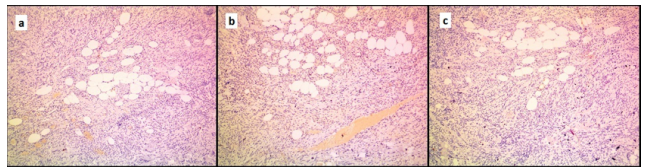


Fig. 5: IHC images showing lack of expression of melanocytic markers by spindle cells. (a) S-100, (b) HMB-45 & (c) Melan A. [(a) to (c) x100]

3. Discussion

Angiomyolipoma (AML) is a rare mesenchymal neoplasm originating from perivascular epithelioid cells (PECs) and is characterized by mixture of smooth muscle cells, adipose tissue and blood vessels in varying proportions. It is commonly seen in the kidney and the liver and less common in the female reproductive system.¹ AMLs constitute 0.06% of all the benign uterine tumors.² In the female genital tract, uterine corpus is the most common site for AMLs followed by cervix, vagina, ovary, fallopian tube, broad ligament and parametria.^{2,3}

As per definition of World Health Organization (WHO), perivascular epithelioid cell tumors (PEComas) is a family of neoplasms, which includes renal or extrarenal AML, lymphangiomyomatosis and clear cell tumors of diverse sites.^{3,5} AML may be categorized as sporadic and syndromic (accompanying TS) types. The sporadic type of AML is the most common (80-90% of cases) and usually develops in older patients.⁵⁻⁷

On extensive review of literature, till date only 36 cases of uterine AML have been reported in the English literature, most of them being single case reports.^{1,2} Mulchandani et al reported the only Indian case of uterine AML and did a comprehensive review of 28 cases of uterine AML reported till then. Wang et al recently reported the largest series of 8 cases of uterine AML.¹ Laffargue et al reported an interesting case of uterine AML associated with pregnancy in a 20 year-old primigravida.⁸

After a thorough review of previous cases, Mulchandani et al have concluded that uterine AMLs have an age range of 20-83 years (mean=48 years) and are most commonly seen in the body, followed by the fundus and lower uterine segment with a mean size of 8 cm.² Our patient was a 48 years lady having mass of size 7.5 cm in the posterior wall of the body of uterus. The clinical presentation of uterine AMLs is nonspecific and resembles that of leiomyomas with most women presenting with menometrorrhagia, pelvic mass or pain in lower abdomen.² Our patient had almost similar complaints. Preoperative evaluation can be done by a transvaginal or abdominal sonography combined with computerized tomography (CT) and MRI studies. Uterine AMLs do not have the classical appearance of leiomyoma on ultrasound. The lipomatous component shows an increased echogenicity on ultrasonography which can be confirmed by CT or MRI. Most of the previously reported cases were labelled as teratoma/cystic neoplasms on radiological investigations.² Our case was clinically and radiologically misdiagnosed as uterine leiomyoma.

Grossly, cases of AML, like leiomyomas, endometrial stromal sarcoma and PEComa are identified as intramural or subserosal masses and may be solitary. AMLs are soft to firm with a grey to pink tan cut surface representing the variable smooth muscle, vascular and adipose tissue components in them.² Classically AMLs show a triphasic histology and are composed of a variable mixture of adipose tissue, spindled or epithelioid smooth muscle cells and anomalous blood vessels. No criteria define the percentage of these components needed for the diagnosis of AML. The mature adipocytes are lipid-distended PECs which appear unilocular with clear cytoplasm and peripherally pushed nuclei. The smooth muscle cells may be arranged in fascicles or haphazard pattern. The blood vessels can be small or medium sized with thick hyalinised walls, with or without tortuosity and focal areas showing back-to-back arrangement. Few normal-appearing blood vessels may be interspersed which represent the normal host blood vessels.²

The predominance of any one cell type in AML may mimic a number of uterine mesenchymal neoplasms such as leiomyoma, PEComa with spindle cell pattern, endometrial stromal sarcoma, epithelioid leiomyoma or PEComa with an epithelioid pattern, lipoleiomyoma, and angioleiomyomas.² The pathologists' awareness of the disease and reasonable selection of materials are important for the timely and

accurate diagnosis of AML.¹ On IHC, uterine AMLs show a strong cytoplasmic reactivity for desmin and SMA. The smooth muscle differentiation in leiomyoma, spindle cell pattern of PEComa, lipoleiomyoma, and angioleiomyomas is also stained by desmin and SMA immunostains. Therefore, it is important to note the morphologic appearance of all these lesions when interpreting IHC.² Tumor in our case exhibited all 3 elements and the spindle cells surrounding blood vessels were negative for melanocytic markers HMB-45 and Melan A.

PEComas in addition to smooth muscle markers are positive for melanocytic markers like HMB-45 and Melan-A while most spindle cells in most uterine AML are negative for melanocytic markers.^{1,2} Totev et al have suggested that an epithelioid AML is a kind of PEComa and hence, advised to do melanocytic markers in all uterine mesenchymal tumors with epithelioid appearance.^{9,10}

Surgical resection is the mainstay of treatment for uterine AML. Most cases are treated by total abdominal hysterectomy with or without salpingo-oophorectomy.^{1,2} However, Cil et al have suggested a follow-up of patients with smaller lesions than surgical management.³ Overall results show that uterine AML has a good prognosis and show a low recurrence rate, however, close follow-up is still recommended.¹

In conclusion, we report this case of extrarenal AML of the uterus for its rarity and to make pathologists aware about such an entity. Although, the clinical features of uterine AML are nonspecific and mimic leiomyoma, with modern imaging modalities; one can now suspect AML radiologically. A confirmed diagnosis still depends upon histopathological examination and IHC may be done when there is diagnostic dilemma.

4. Conflict of Interest

The authors declare that there is no conflict of interest.

5. Source of Funding

None.

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