



## Case Report

### Angiomyolipoma of the cervix – report of a rare entity

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**ABSTRACT:** Angiomyolipoma (AML) is a mesenchymal neoplasm seen usually in the kidney. Few cases of extra renal AML have been documented in various organs including the female genital tract, where the uterus is the most common site. To the best of our knowledge, only 4 cases of AML in the cervix have been reported in the literature. Association of AML with tuberous sclerosis is well known. Presently AML is included in the spectrum of disease entities called PEComa. We report a case of AML without tuberous sclerosis arising from the uterine cervix, which has to be differentiated from lipoleiomyoma.

**KEY WORDS:** *Angiomyolipoma; Uterine cervix; PEComas; Uterine tumor*

#### INTRODUCTION

Angiomyolipoma (AML) occurs most frequently in the kidney, where it is closely related to tuberous sclerosis complex (TSC)<sup>1,2</sup>, occasionally in other organs, most commonly the liver, but occurrence at other sites is extremely rare<sup>3</sup>. Irrespective of the type of organ involved, the diagnosis can be established by the presence of a marker of melanogenesis, such as human melanoma black 45 (HMB-45) and positive smooth muscle cells<sup>4</sup>. Angiomyolipoma (AML) has undergone a remarkable transformation in recent years, from a rare and rather pedestrian tumor type restricted to kidney, to a biologically fascinating and morphologically heterogenous entity which can occur in a wide variety of sites and which has been given the collective name of PEComa<sup>5</sup>. Typically, AML is benign, but malignant AMLs of the kidney and liver have been described in exceptional cases<sup>6,7</sup>. Regarding uterine AMLs, in most cases AMLs originate from the body of the uterus but cases have been reported of cervical

localization<sup>8</sup>, and mostly in females over 40<sup>9</sup>. The occurrence of AML in the cervix without concurrent incidence in the kidney is extremely rare, and only four cases have been reported in the scientific literature<sup>10</sup>. We are reporting a case of uterine cervix AML without tuberous sclerosis because of its rarity.

#### CASE DETAILS

##### Clinical Summary

A 30-year-old female patient presented with bleeding per vagina for the last two weeks. Clinical examination revealed that she was mentally retarded. She was not cooperative for complete clinical examination. The patient did not have a noteworthy family history of any disease. The patient's attendant gave history of seizures in the patient. Per speculum examination revealed a firm polypoidal mass of about 4 x 5cms size with thick pedicle arising from the uterine cervix. Ultrasonography disclosed normal kidneys, mildly cystic ovaries and a 3 x 4cms sized homogenous well-circumscribed lesion with high vascularity arising from the anterior part of the cervix and extending into the myometrium. Clinical diagnosis of endocervical polyp/cervical fibroid was made. Hysterectomy was performed, and sent for histopathological examination.

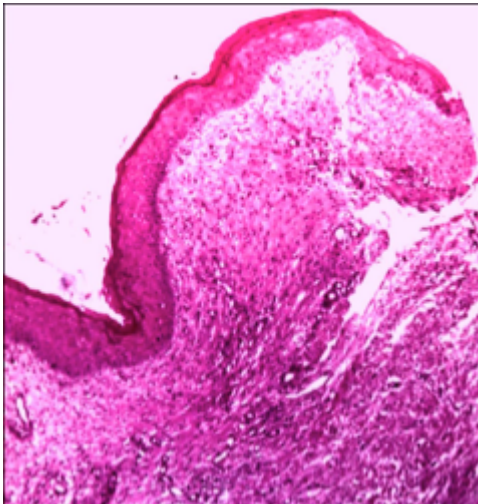
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### Pathological Findings

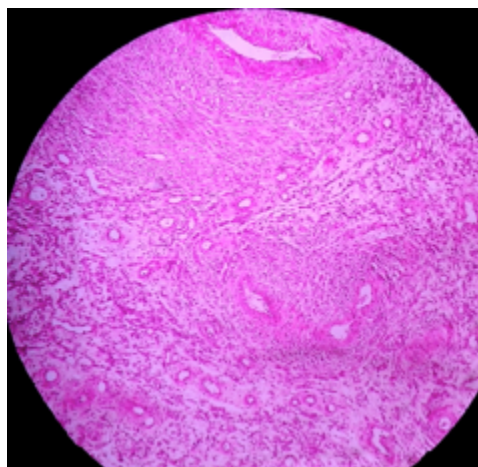
Macroscopically, the hysterectomy specimen contained a polypoidal growth arising from the anterior lip of the cervix. The growth measured 3 x 4 x 6cms, and the cut surface was a uniform greyish brown color. On microscopic examination, the cervical growth consisted of blood vessels and smooth muscle fibers in the cervical stroma covered by stratified squamous epithelium (**Figure 1**). In addition, the focal clusters of adipocytes and the plump spindle cells surrounding small or medium sized thick walled blood vessels were also seen (**Figure 2**). There was no atypia or increased mitotic activity in the tumor.

### Immunohistochemical Findings

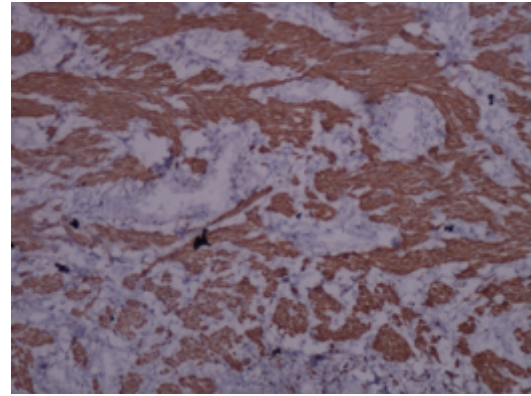
The tumor showed positive staining for Desmin (**Figure 3**), and negative for HMB-45 and S-100.



**Figure 1: Scanner view of tumor consisting of blood vessels and smooth muscle fibers in the cervical stroma covered by stratified squamous epithelium. H & E X 5**



**Figure 2: Tumor consisting of thick walled blood vessels surrounded by plump spindle cells. Foci of adipocytes and smooth muscle fibers are also seen. H & E x 20.**



**Figure 3: Smooth muscle fibers showing desmin positivity by immunostain X 20**

### DISCUSSION

Angiomyolipomas (AMLs) are benign mesenchymal neoplasms composed of a variable mixture of adipose tissue and smooth muscle cells with anomalous vascular component. Intrauterine AMLs are extremely rare, and are not officially listed in the WHO Classification of female reproductive system tumors<sup>11</sup>. According to a summary out of 22 cases of reported uterine angiomyolipomatous lesions<sup>10</sup>, only 4 cases arose in the uterine cervix.

The clinical presentations of uterine AMLs are non-specific and variable, and the ultrasound image depends on the biological features of the tumor. Therefore, neither the clinical presentation nor the ultrasound findings are sufficient to make a preoperative diagnosis<sup>12</sup>. The diagnosis can be confirmed by FNAC supplemented by immunohistochemistry. It is of interest that non-vascular smooth muscle cells were negative for HMB-45, in contrast to renal and other extra-renal AML in which HMB-45 immunoreactivity has been demonstrated in these cells<sup>13</sup>. In our case, the non-vascular smooth muscle cells were negative for HMB-45 and S-100. Aung *et al*<sup>14</sup> reported that 20% of angiomyolipomas are negative for HMB-45. This is the major point to consider when using HMB-45 antigen alone for the diagnosis of uterine angiomyolipoma. HMB-45 antigen staining is useful for diagnosing uterine smooth muscle tumors, but it is not a definitive indicator of uterine angiomyolipoma.

The association of angiomyolipoma with tuberous sclerosis (TS) patients is well recognized. Some literature has suggested that patients with TS are at increased risk of renal neoplasm. It is well known that AMLs are associated with TS in 5% to 50% of cases<sup>15</sup>. However, our case was not associated with TS.

Angiomyolipomas may mimic lipoleiomyoma, degenerated myoma and vascular leiomyoma with a fat component<sup>16</sup>. Lipoleiomyomas have a significant percentage of fat between the muscle

cells and the fat cell component may arise from perivascular immature proliferating mesenchymal cells or from direct transformation of smooth muscle cells into adipocytes by means of progressive intracellular storage of lipids<sup>14</sup>. In lipoleiomyoma the vascular prominence is not significant whereas in the AML the thick walled blood vessels are the most prominent morphological finding. Degenerated myomas can be differentiated from AML by echogenicity, without shadowing and irregular margins.

## CONCLUSION

To conclude, the differential diagnosis of a lower abdominal mass and dysfunctional uterine bleeding should include the AMLs even though the uterine cervix is an extremely rare location for these tumors. We also believe that angioleiomyomas should be recognized as a distinct entity and included in the World Health Organization classification of tumors of the female genital tract.

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