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Case Report

Extrarenal angiomyolipoma in uterine cervix: rare presentation in unusual site

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ABSTRACT

Angiomyolipoma is a benign neoplasm composed of variable admixture of blood vessels, smooth muscle cells and adipose tissue. Cervical angiomyolipoma are extremely rare and to the best of our knowledge only five cases of angiomyolipoma in cervix have been reported in the literature till date. Authors are presenting a case of angiomyolipoma arising from the uterine cervix. 43 years old female presented with mass descending per vagina for 6 months. This case had no association with tuberous sclerosis. Microscopic examination showed an ill-defined polypoidal, non-encapsulated lesion covered by keratinized stratified epithelium. The lesion is made up of three components, predominantly by fascicles of spindle shaped cells, varying sized blood vessels and multiple foci of mature adipocytes with no evidence of atypia or increased mitotic activity. Smooth muscle component showed strong immunoreactivity to SMA and absence of elastic fibres in the blood vessels were confirmed by histochemistry. Non-vascular smooth muscle cells were negative for HMB-45 in contrast to renal and other extra-renal angiomyolipoma in which HMB-45 immunoreactivity in seen in these cells. To conclude, the differential diagnosis of lower abdominal mass and dysfunctional uterine bleeding should include the angiomyolipoma, even though the uterine cervix is an extremely rare location where they occur.

Keywords: Angiomyolipoma, Female genital tract, Histochemistry, Immunohistochemistry

INTRODUCTION

Angiomyolipoma is a benign neoplasm composed of variable admixture of blood vessels, smooth muscle cells and adipose tissue. It occurs most frequently in the kidney where it is closely associated with tuberous sclerosis and occasionally in extrarenal sites, most commonly liver, but occurrence at other sites is extremely rare.[1-3] Many cases of extra-renal angiomyolipoma have been reported in various organs like the liver, pelvic region, retroperitoneum (not connected to kidney), uterus, somatic soft tissue, large bowel, nasal cavity and bone. Few cases of extra-renal angiomyolipoma have been reported in the female genital tract among which uterus is the most common site. Cervical angiomyolipoma without concurrent incidence in the kidney is extremely rare and only four cases of angiomyolipoma in cervix have been reported in the literature till date.[4]

Herein author report a case of angiomyolipoma, occurring in uterine cervix in a 43 years old female.

CASE REPORT

A 43 years old female presented with complaints of mass descending per vagina for 6 months in the gynaecology OPD. The patient was P2L2, sterilized, last childbirth was

12 years back and had a regular history of menstrual cycles. The patient was not on any medications for some other disease. No history of tuberous sclerosis was present.

Per speculum examination showed a pink globular firm mass arising from cervix, hanging outside the vagina. External os was not visible (Figure 1). Ultrasonography showed uterus was normal in size and contour and bilateral ovaries appeared normal. Bilateral kidneys were also normal. The patient was taken up for excision of the mass.

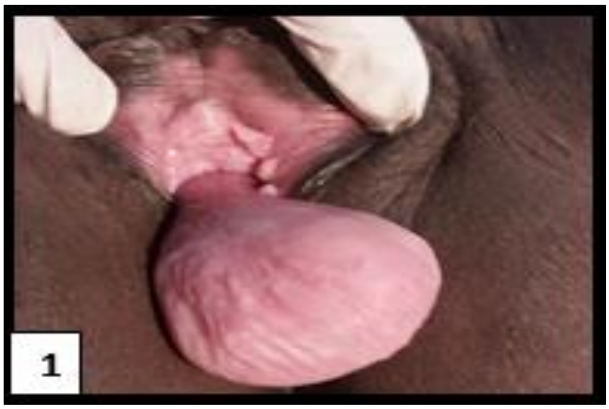

Figure 1: Clinical picture showing mass descending per vagina. external os not visible.

Intra operative findings showed a 6 x 4 cm globular mass arising from cervix with a pinpoint external os. Mass was clamped at base and cut and ligated with catgut. Post-operative period was uneventful. The mass was sent for histopathological examination.

Grossly, author received a polypoidal grey white soft tissue mass measuring 6 x 4 x 2.5 cm. External surface was grey white with few congested areas. Cut surface was grey white with few congested areas and focal whorled appearance (Figure 2).

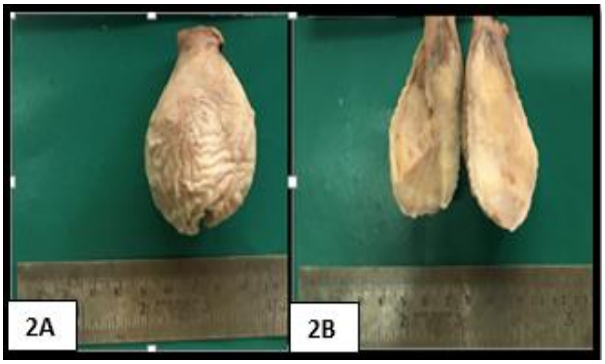

Figure 2: (A) Polypoidal grey white soft tissue mass measuring 6 x 4 x 2.5 cm, (B) cut surface is grey white with focal whorled appearance.

Microscopically, multiple sections studied showed an ill-defined polypoidal, non-encapsulated lesion covered by keratinized stratified squamous epithelium (Figure 3).

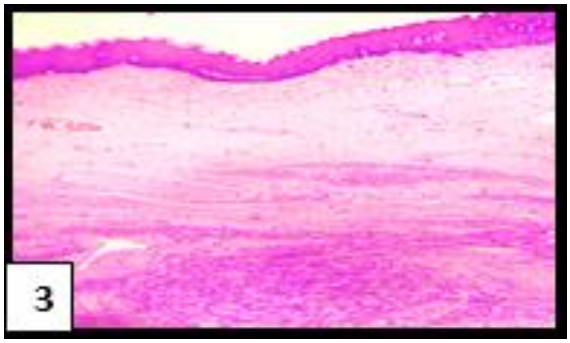

Figure 3: Non-encapsulated lesion covered by keratinized stratified squamous epithelium (H and E, 40x).

The lesion was made up of three components, predominantly by spindle shaped cells arranged in fascicles. The cells had plump ovoid vesicular nuclei with abundant bright eosinophilic cytoplasm. Many varying sized blood vessels, many of them were thick walled and hyalinized. Multiple small foci of mature adipocytes were also seen admixed with spindle shaped cells. There was no evidence of atypia or increased mitotic activity in the sections studied. The morphological impression given was 'Benign unencapsulated lesion with triphasic morphology, suggestive of Angiomyolipoma-cervix' (Figure 4).

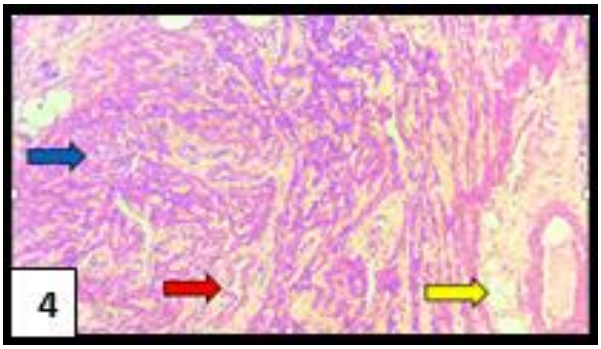

Figure 4: Lesion is composed of fascicles of spindle cells (blue arrow), varying sized blood vessels (red arrow), small foci of mature adipocytes (yellow arrow) (H and E 100x).

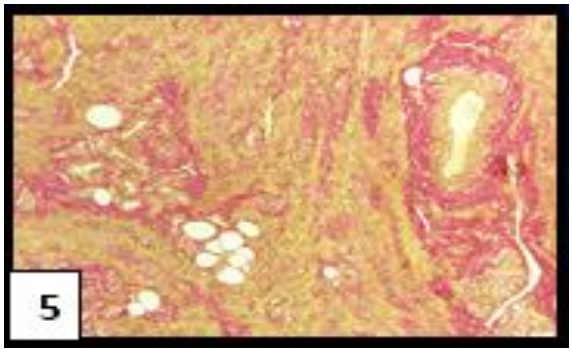

Figure 5: Lack of elastic fibres in blood vessels (Verhoeff Van Gieson, 100X).

This was confirmed by histochemistry and immunohistochemistry. Verhoeff Van Gieson stain was negative for elastic fibres in blood vessels (Figure 5). Immunohistochemistry for smooth muscle fibres showed positive staining for SMA (Figure 6) and non-vascular smooth muscle cells were negative for HMB-45 (Figure 7).

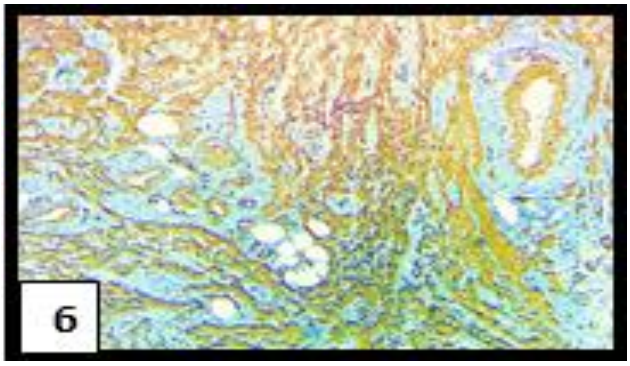

Figure 6: Positive for smooth muscle cells (SMA, 100X).

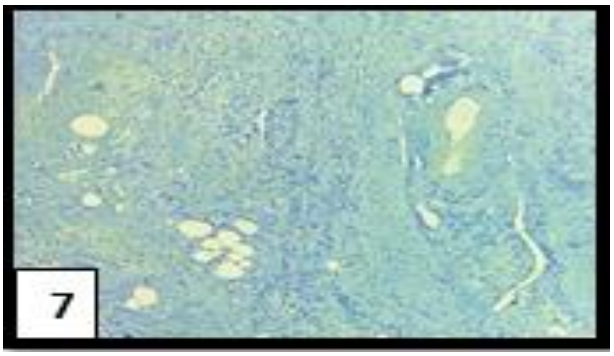

Figure 7: Negative for non- vascular smooth muscle cells (HMB-45, 100X).

DISCUSSION

Angiomyolipoma is a benign neoplasm composed of variable admixture of blood vessels, smooth muscle cells and adipose tissue. Angiomyolipomas rarely occur in the female genital tract and they are not even included in the World Health Organization (WHO) classification of female reproductive system tumors.[5] Only 4 cases of angiomyolipoma have occurred in cervical region out of 22 cases that have been reported in the female genital tract.[4]

Angiomyolipomas have variable and non-specific clinical presentation, while radiologically also the findings depend on the biological features of the tumor. Therefore, it's not sufficient to make a pre-operative diagnosis on the basis of clinical presentation and radiological findings and the diagnosis is depends on the combination of clinical presentation, radiological findings and histological examination.[6,7]

It is interesting to know that HMB-45 was negative for nonvascular smooth muscle cells in this case, in contrast to angiomyolipomas occurring in renal and other extra-renal sites where HMB-45 is positive in these cells.[8] According to Aung et al, 20% angiomyolipomas are negative for HMB-45. This should be kept in mind while using HMB-45 alone for diagnosing angiomyolipoma occurring in uterine.[9] HMB-45 is useful in diagnosing angiomyolipomas but is not the definitive marker that indicates angiomyolipoma.

Angiomyolipomas are associated with tuberous sclerosis. Patients with tuberous sclerosis have and increased risk for renal tumors. Literature has shown that patients with tuberous sclerosis have 5%-50% risk to develop angiomyolipomas.[8] In the present case, the patient was not associated with tuberous sclerosis.

There are few lesions that mimick angiomyolipoma and make the diagnosis difficult. Lipoleiomyomas show mature adipocytes along with muscle cells, but no prominent vascular elements as compared to angiomyolipoma, where there are prominent thick-walled blood vessels.[8] Degenerated myomas may also mimick angiomyolipomas but they can be distinguished from angiomyolipoma by echogenicity, without shadowing and irregular margins.[8]

Angiomyolipoma are usually benign, but they may be associated with haemorrhage and even show invasion of the surrounding organs. Therefore, recently angiomyolipoma is considered as a slow growing malignant neoplasm with ability to metastasize.[10]

CONCLUSION

In case of lower abdominal mass and dysfunctional uterine bleeding, angiomyolipoma should be considered as one of the differential diagnosis. Histopathological examination showing classical morphological features of angiomyolipoma can prompt pathologist to give appropriate diagnosis.

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