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Angiomyolipoma of the Cervix: A Case Report

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Abstract
Angiomyolipoma (AML) is a benign mesenchymal neoplasm seen mostly in kidney. It was previously classified as Hamartomas, however now it belong to a family of tumors collectively referred to as PEComas. Very Few cases of extra renal AML arising from uterine cervix have been reported in the literature. Angiomyolipomais usually associated with tuberous sclerosis. We report a case of Angiomyolipoma occurring at a rare site of uterine cervix, without evidence of Tuberous Sclerosis.
Keywords: *Angiomyolipoma; PEComas; Tuberous sclerosis.*

Introduction

Angiomyolipoma (AML) is a mesenchymal neoplasm composed of a variable mixture of adipose tissue, smooth muscle cells, and anomalous blood vessels.(1)

Extrarenal angiomyolipomas (AML) have been reported at various anatomical sites, but infrequent in the female genital tract. In the uterus, only few cases have been described which originate from the body of uterus but its occurrence in cervix is unusual. AML of cervix without its concurrence in kidney is extremely rare, and only five cases have been reported in the scientific literature. We hereby discuss a case Angiomyolipoma presenting at unusual site of uterine cervix.

Case Report

A 35 year nulliparous female, presented with dysfunctional uterine bleeding of 2 months duration without any other associated symptoms. The physical examination and per abdominal examination was within normal limits. Per speculum examination revealed a firm mass of about 4x3cm size arising from the uterine cervix. Ultrasonography of abdomen revealed normal sized kidneys with a heterogeneously hyperechoic circumferential anterior cervical wall thickening with increased vascularity.MRI suggested a vascular neoplasm of cervix. We received total abdominal hysterectomy specimen. Grossly it showed a diffuse mass arising from the anterior lip of the cervix. It measured 4 x4x4cms, cut

surface of which was yellowish- white with focal areas of hemorrhage. The histomorphology showed a well circumscribed, unencapsulated tumor beneath the endocervical epithelium. This triphasic tumor consisted of fascicles and bundles of spindle shaped cells, interspersed by lobules of mature adipose tissue and numerous variable sized thick walled dismorphic blood vessels. The spindled component was monomorphic with oval to elongated nuclei having uniform chromatin and inconspicuous nucleoli. The adipocytes were seen as small groups and lobules intermingled with other two components. The blood vessels form the important component and were of variable size and caliber. (Figure1) Most of them thick walled and hyalinized were well appreciated in Massons Trichrome Stain. (Figure 2) There was no evidence of mitosis, necrosis and invasion into the adjacent tissue. The Smooth muscle component was also highlighted well in Masson Trichrome stain. (Figure 2) All these features contribute to the Angiomyolipoma.

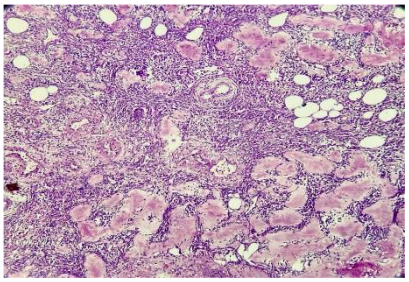

Fig 1: Tumor consisting of spindle cell smooth muscle component, thick walled blood vessels and mature adipocytes in clusters.(HE stain,20X)

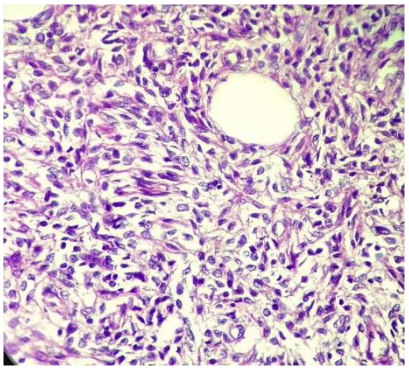

Fig 2: Spindle cell with oval to elongated nuclei having uniform chromatin and inconspicuous nucleoli. Cells are separated by little amount of collagen.(HE stain,40X)

Fig 3: Thick walled hyalinised blood vessels (green colour) surrounded by spindled smooth muscle (light red colour) and few clusters of mature adipocytes. (Massons Trichome Stain,20x)

Discussion

Angiomyolipomas (AMLs) are distinctive neoplasms composed of variable combinations of smooth muscle, adipose tissue, and vascular component.[2] Previously considered as hamartoma, however genetic studies have confirmed about its clonal neoplastic proliferation. These tumors are believed to be derived from perivascular epitheliod cells.[3] Perivascular epithelioid cell tumors (PEComas), include a group of mesenchymal tumors containing perivascular epithelioid cells (PECs) that usually show association with blood vessel walls and coexpress immunophenotypic features of both smooth muscle and melanocytic cells. PEComa family of tumors includes angiomyolipoma (AML), lymph angioleiomymatosis (LAM), clear cell sugar tumors (CCST), and a large number of myomelanocytic tumors described under varied terminologies in a wide variety of locations.[4]

Angiomyolipomais usually observed in the kidney of patients with Tuberous Sclerosis (TS), a disease caused by loss-of-functional mutations in TSC1 or TSC2 tumor suppressor genes in about 5-50% cases. TS is characterised by widespread hamartomas in several organs, including the brain (cortical tubers), heart (rhabdomyomas), eyes (phakomas or hamartomas), skin (angiofibromas or adenoma sebaceum), bone (sclerotic lesions), kidney (angiomyolipoma and renal cysts), lung (lymphangiomyomas) and liver.[1] Extrarenal AML is uncommon and only few sporadic cases have been reported in liver, mediastinum, heart,

spermatic cord,vaginal wall, fallopian tube, oral cavity, pharynx, nasal cavity, soft tissue, and skin.[5] Intrauterine AMLs are extremely rare and in most cases originate from the uterine body but cases have been reported of cervical localization mostly in females over 40 year of age.[6] According to a summary report of 22 cases of uterine angiomyolipomatous, only 4 cases arose in the uterine cervix.[1] The clinical presentation of uterine AMLs is non-specific and is similar to leiomyoma with menorrhagia, presence of pelvic mass, abdominal pain or even lack of symptoms.[7] Ultrasound features are too not specific. Therefore clinical symptoms and ultrasound findings are not sufficient for preoperative diagnosis and the burden of diagnosis lies with the pathologist.

Angiomyolipoma morphologically may mimiclipoleiomyoma, degenerated myoma, vascular leiomyoma with fat component and benign lipomatous tumor.[8] Lipoleiomyoma has significant amount of muscle component and element of fat may arise directly from smooth muscle cells due to progressive intracellular accumulation of lipids.[9] The presence of thick walled dismorphic blood vessels a hallmark of AML is absent in lipoleiomyoma. Degenerated myoma can also be a differential; AML however, reveals the triphasic elements in the form of smooth muscle cells, mature adipose tissue and thick walled blood vessels.

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 is not typical of uterine AML.[6,8]

Angiomyolipomas (AMLs) tends to be benign; however they may be associated with haemorrhage and invasion of nearby organs. Hence recently they are regarded as slow-growing malignant tumors with potential to metastasize.[10]

Conclusion

Angiomyolipoma should also be included in the differential diagnosis of a lower abdominal mass and dysfunctional uterine bleeding. The high index of suspicion and classical morphology can help us to clinch the correct histopathological diagnosis in unusual anatomical locations like uterine cervix.

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