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Angiolipoleiomyoma [ALLM] of uterus: A rare case report

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Abstract
Angiolipoleiomyoma (ALLM) is a rare variant of benign neoplasm of the uterus. It has a prevalence of only 0.06% with only a handful of cases. Although renal angiomyolipoma's are more prevalent, extrarenal manifestation especially that which is found in the uterus is still a rare entity. We present the case of ALLM of uterus in a 41 year old female due to its rarity.

Keywords: Angiolipoleiomyoma, Hamartoma, Benign, Uterus

Introduction
Leiomyomas are frequently diagnosed in women of reproductive age and are commonly referred to as fibroids of the uterus. Rarely discovered variants include those with abnormal blood vessels (Angio), smooth muscle (MYO), and/or mature adipose tissue (Lipo). Due to the presence of a mixture of tissues, this finding can also be referred to as a hamartoma. ALLM is a very rare lesion of the uterus. To the best of our knowledge, only 20 cases have been described in the literature. Microscopically, it consists of a variable admixture of smooth muscle, adipose tissue, and blood vessels. There are only a handful of cases of ALLM of the uterus reported which currently has a prevalence of 0.06% among benign uterine lesions [1].
These benign tumours have a higher incidence in women in their fourth to sixth decades of their lives [2]. The incidence of ALLM has been reported 0.2% in women in general population without tuberous sclerosis [3]. We report a case of an angiolipoleiomyoma of the uterus in a 41 year old postmenopausal woman.

Case report
A 41years old female, presented with heavy menstrual bleeding for 1 year. Total abdominal hysterectomy with bilateral salphingo-opherectomy was done. On gross, both the fallopian tubes and ovaries were unremarkable, and uterus of size 8 x 8 x 5 cm was received with cervical length of 4 cm. Cut section of uterus [Fig.1] was grayish black and revealed slit like endometrial cavity with myometrial thickness of 3.5 cm with small cystic, hemorrhagic, and small slit like areas.
Microscopy [Fig. 2, 3, 4] revealed a well circumscribed and nodular lesion composed of variable cellularity of spindle cells admixed with blood vessels of both arterial and venous calibre and variable amount of mature adipose tissue.
The spindle cells appeared to be smooth muscle cells with eosinophilic cytoplasm and cigar shaped nuclei. Heterologous element like osteoid formation along with smooth muscle proliferation in the vessel walls and occasional thrombus formation was further observed. No visible nucleocytoplasmic atypia was observed. Mitotic count was 1 to 2 mitoses based on the assessment of 10 high power fields [40X]. The margins of the tumour were well circumscribed with no evidence of necrosis or increased mitotic activity in the tumour cells revealing no malignant potential of the tumour.
On IHC [Immunohistochemistry] [Fig.5], this lesion exhibited positivity for [SMA] smooth muscle actin and desmin, while negative for CD10 and HMB45. This pathological examination resulted in a diagnosis of ALLM of uterus.

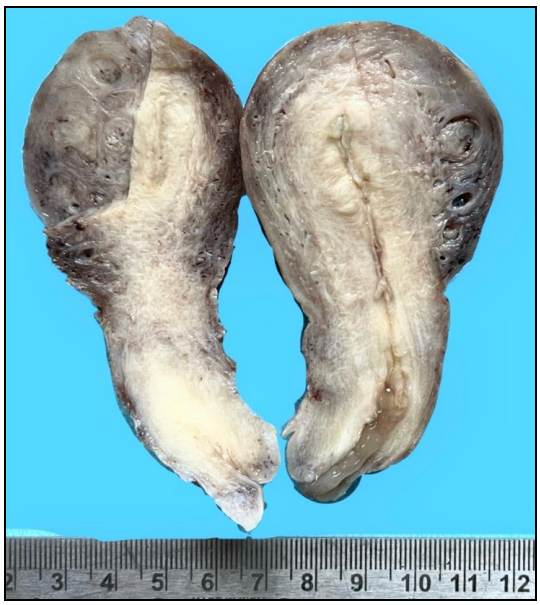

Fig 1: Gross: Cut section of hysterectomy specimen showing haemorrhagic and cystic nodules in the myometrium

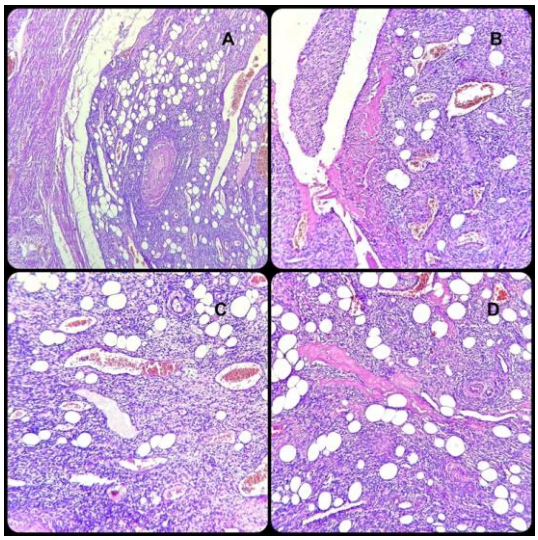

Fig 2: Showing well circumscribed tumor tissue composed of spindle cells admixed with mature adipose tissue and plenty of blood vessels [H&E;10X]

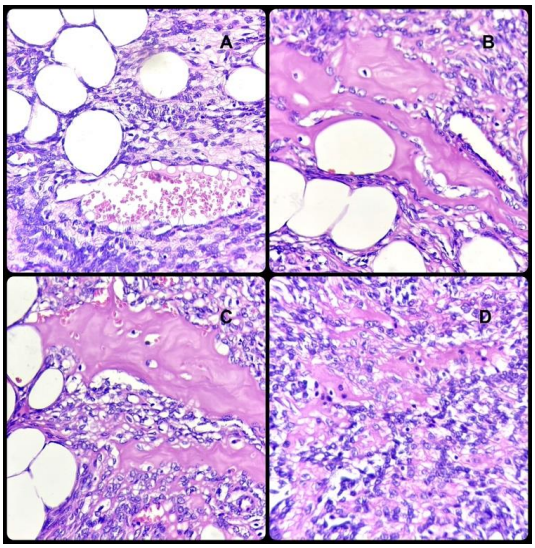

Fig 3: IHC: Showing tumour tissue [A, B]: composed of smooth muscle cells strongly positive for SMA and vimentin respectively; [C]: Giant cells positive for CD68. [IHC: 40X]

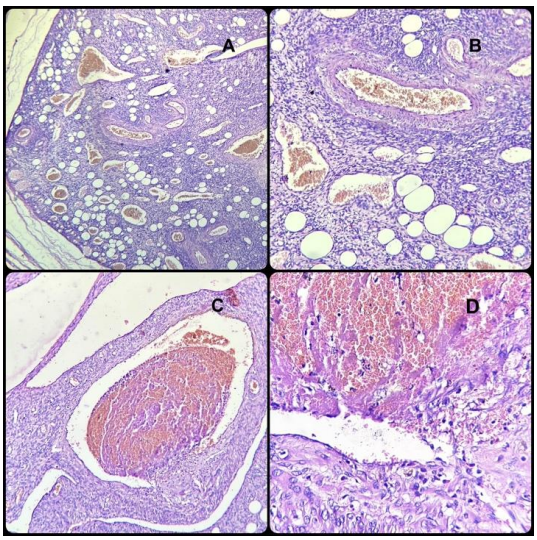

Fig 4: [A, B]: Showing tumor tissue composed of spindle cells admixed with mature adipose tissue and plenty of blood vessels [H&E;10 X] and few areas showing dilated vascular spaces lined by endothelial cells and thrombus formation [H&E: C:10X; D:40X]

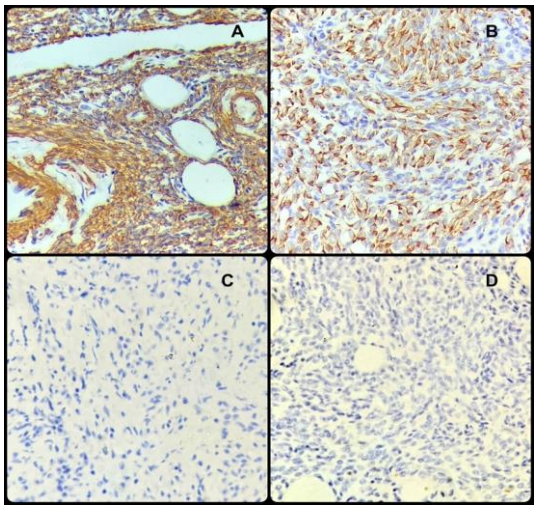

Fig 5: IHC: Showing tumor tissue [A, B]: strongly positive for SMA and desmin respectively and [C, D]: negative for CD10 and HMB45 [IHC: 40X]

Discussion

Uterine ALLM is a rare benign tumour, with an estimated incidence of 0.06% of the benign uterine lesions [1, 4, 5]. To the best of our knowledge, the first case was described by McKeithen *et al*. in 1964 [6].

These tumours present with symptoms like leiomyomas of the uterus. Although usually asymptomatic with just the presence of a pelvic mass, frequent symptoms comprise of menstrual cycle disturbances [menometrorrhagia] and urinary frequency [pressure symptoms] along with mild abdominal pain. Size of the tumour range from 2-16 cm with a median of 8.4 cm [1, 4]. Usually the margins of the tumour are circumscribed and encapsulated with no evidence of necrosis, therefore, this is congruent with the case presented. Most commonly, the tumour is localized in the uterine corpus. Other sites include the cervix and lower uterine segment. The growth pattern is intramural or subserosal. Macroscopically, the tumour appears grey, white, or pink on the cut surface, and it is usually encapsulated. Microscopically, the tumour consists of an admixture of three components in various proportions: adipose tissue,

smooth muscle cells, and blood vessels. The vascular component usually consists of a proliferation of large thick-wall blood vessels sometimes tortuous and aggregated and fusing with the stroma [4, 5, 7]. The fascicles penetrate between the blood vessels and adipose tissue and surround them [4]. Although these tumours are usually surrounded by a pseudo capsule they can demonstrate an infiltrative growing pattern too [8].

The histogenesis of the ALLM is controversial and not really investigated [9]. In this case, its origin is either a benign hamartoma with adipose metaplasia, or a variant of a Mullerian mixed tumour [10, 11].

Over the last two decades, these tumours have been broadly classified as Perivascular Epithelioid Cell tumours [PEComas]. Tumours included in this group include variants of angiomyolipomas (ALM), lymphangioleiomyomatosis, clear cell/sugar tumour of the lung and myelomelanocytic tumour of the falciform ligament [3]. The kidney is an organ where ALMs have been frequently diagnosed having a strong association with tuberous sclerosis. Due to its similarity to renal angiomyolipoma, it is referred to as uterine ALLM, however, it is not associated with tuberous sclerosis [4].

Given that most cases of ALLM were described 40 years ago, limited information is available on their immunohistochemical profile [4, 6]. However, when immunohistochemistry was performed it revealed strong cytoplasmic positivity for smooth muscle actin [SMA], desmin, and calponin in the smooth muscle component, but no positivity for HMB45 in any of the 3 components [1, 4, 5, 7].

The differential diagnosis of uterine ALLM depends upon the histopathological picture. In our case it was confused with endometrial stromal tumour [EST] because of strikingly similar arrangement of blood vessels. CD-10 negativity along with diffuse SMA positivity confirmed the diagnosis of ALLM, as CD-10 positivity is usually strong in EST but negative or weak in ALLM, as in our case.

Another differential diagnosis is PEComas. The angiomyolipoma is composed of epithelioid perivascular cells, and it is classified as PEComas. These tumoral cells are also positive for the muscle markers SMA, Desmin, Caldesmon but they are especially immunoreactive for HMB45 while the ALLM is not [4, 12].

Vascular leiomyoma can be reasonably distinguished by its myometrium-like capillaries and few arterioles unlike thick sheathed blood vessels of AL. Fibrin deposition in the vessel walls is also an important feature of ALLM [as seen in our cases too] but unusual in vascular leiomyomas. Mitosis is sometimes seen in ALLM but is usually < 2 per 10 HPF.

Endometrial or cervical polyps are sometimes replete with proliferating vascular channels, with spindly compact stroma. Misdiagnosis of ALLM in such cases can be avoided by SMA staining which is positive in vascular walls but negative in stroma, unlike ALLM where both tissues are strongly positive.

Conclusion

The aim of this case report is to introduce this infrequently seen uterine tumour to gynaecologist's attention, hereby this rare tumour can be kept in mind while making a differential diagnosis of uterine masses.

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Conflicts of interest: There are no conflicts of interest.

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