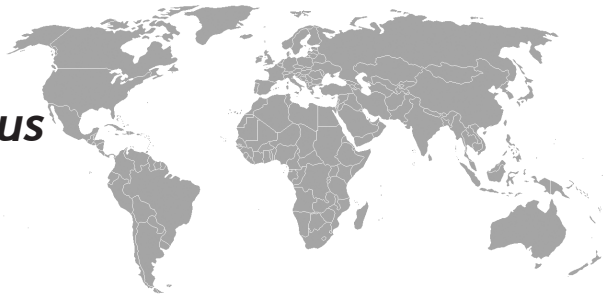
Angiomyolipoma of the Uterus



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ABSTRACT

Angiomyolipoma (AML) is a mesenchymal neoplasm composed of a variable mixture of adipose tissue, smooth muscle cells, and anomalous blood vessels. In the uterus, only a few cases have been reported. We described a uterine AML without evidence of tuberous sclerosus (TS). The clinical presentations of uterine angiomyomatous lesions are nonspecific. The diagnosis of angiomyolipoma can be established with ultrasound and/or computed tomography (CT). The patients whose lower abdomen mass in the abdominopelvic ultrasound and dysfunctional uterine bleeding should be considered AML in the differential diagnosis.

Key words: *Angiomyolipoma, uterus, mesenchymal neoplasm*

Uterusun Anjiomiyolipomu

ÖZET

Anjiyomiyolipom (AML) yağ dokusu, düz kas hücreleri ve anormal kan damarlarının değişken karışımından oluşan bir mezenkimal tümördür. Uterusta, sadece birkaç vaka bildirilmiştir. Biz tuberoz skleroz (TS) olmadan bir uterin AML tanımladık. Uterus anjiomiyomatöz lezyonların kliniği spesifik değildir. Anjiyomiyolipom tanısı ultrasonografik ve / veya bilgisayarlı tomografi (BT) ile konulabilir. Abdominopelvik ultrasonda alt karında kitle ve disfonksiyonel uterin kanaması olan hastaların ayırıcı tanısında AML dikkate alınmalıdır.

Anahtar kelimeler: *Anjiyomiyolipom, uterus, mezankimal tümör*

INTRODUCTION

Angiomyolipoma (AML) is a mesenchymal neoplasm composed of a variable mixture of adipose tissue, smooth muscle cells, and anomalous blood vessels. It often arises in the kidney, but has also been described in extra renal areas such as the liver, nasal cavity, retroperitoneal area, alimentary tracts and female reproductive system, including uterus, vagina and salpinx. Only a few cases have been reported uterine mesenchymal neoplasm. The adipose tissue in an AML may be contained within well circumscribed foci or distributed diffusely throughout the mass. AML share some histopathologic features of leiomyoma but also include blood vessels and fat components. Most AML are located within the walls of the uterine corpus. Unlike leiomyoma, AML is invariably benign, and usually occur in women older than 40 years. It is well know that renal AML are often associated with tuberous sclerosis (TS). The association of TS and AML varies from 5 to 80% of cases. However, it is now well known that AML may form in the absence of TS. In fact, 80% of the patients who have AML do not have TS. We described a uterine AML without evidence of TS.

CASE

A 44-year-old premenopausal woman, gravida 4, parity 4, presented with a lower abdominal mass with a 4 month history of lower abdominal distension and 6 month history of menorrhagia unresponsive to treatment. The patient had not any prior episodes of abnor-

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mal vaginal bleeding and another physical symptom. She wasn't a smoker and drink alcohol. In the physical examination, immobile mass filling the pelvis up to umbilicus was observed. The abdominopelvic and transvaginal US noticed 16x10 cm mass originating from uterine corpus. No adnexial pathology was observed. The MRI correlated the USG findings. The mass had heterogeneous solid and cystic components in T1AG and T2AG. These findings were suggestive of degenerated myoma. The CA125, CA19-9, CEA and AFP values were normal. Both of ovaries and fallopian tubes were grossly normal at laparotomy. Macroscopically, solid- mass with about 16 cm in diameter arising from the posterior uterine wall was seen. Intraoperatively diagnosis was myoma uteri. Hysterectomy was performed and operative material sent to pathologic examination. Postoperative recovery was uneventful. Microscopically, the tumor sections showed many thick-walled blood vessels surrounded by smooth muscle fibers and admixed with adipose tissue. No mitoses or necrosis were seen (Figure 1). Immunohistochemical stains, Smooth muscle actin and desmin were strongly positive in spindle cells. Adipocytes were immunoreactive with the anti-S100 protein antibody. No immunoreactivity was seen with HMB-45.

DISCUSSION

Perivascular epithelioid cell tumors (PEComas) are rare neoplasms probably arise from perivascular epithelioid cells. Perivascular epithelioid cell tumors include AML, clear cell/sugar tumor of the lung, lymphangioleiomyomatosis (LAM), and myelomelanocytic tumor of ligamentum teres/falciform ligament. Perivascular epithelioid cell tumors have been described in a variety of locations such as soft tissues, visceral organs, oral mucosa, skin, orbit and base of skull (1). Forty-four uterine/cervical PEComas have been reported in the literature (2), including 4 in the uterine cervix and 40 in the uterine corpus. Angiomyolipoma (AML) is a very rare disease in the uterus. Twenty of all uterine/cervical PEComa cases are angiomyolipomatous lesions. Renal angiomyolipoma has a close association with tuberous sclerosis (TS), but none of the reported uterine angiomyolipoma cases is associated with TS (3). Association of TS and renal AML varies from 5 to 50% of cases (4,5). TS is a genetic multisystem disorder with an estimated prevelance of 1:10000 (6). 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. The affected genes are TSC1 (on chromosome 9q34) and TSC2 (on chromosome 16p13), encoding hamartin and tuberin respectively. TS is an autosomal dominant syndrome, but most cases appear to represent spontaneous mutations. The diagnosis of TS (not otherwise specified) is based on clinical, radiological and histopathological findings (6). The diagnosis of uterine angiomyolipoma is not clearly appointed, because uterine AML is not officially listed in the World Health Organization's (WHO) histological typing of female genital tract tumors (1). Morphological similar lesions such as lipoleiomyoma, angiolipoleiomyoma, lipoleiomyomatous tumor, benign mixed mesodermal tumor, and benign lipomatous lesion composed of blood vessels, smooth muscle, and adipose tissue (3). Adding four cases (Ren et al. (7), Darai et al. (8), Kajo et al.(9) and the peresent case) to 18 cases of uterin angiomatous lesions in Cil's report, it was found that patients ranged in age from 20 to 77 years (9) (Table 1). The most of the uterin cases of angiomyolipomatous lesion arose in the uterine body; only four arose in the uterin cervix. The clinical presentations of uterine angiomyomatous lesions are nonspesific. They are often similar to those of typical leiomyomas and include abdominal or genital symptoms. Patients may be fully asymptom-

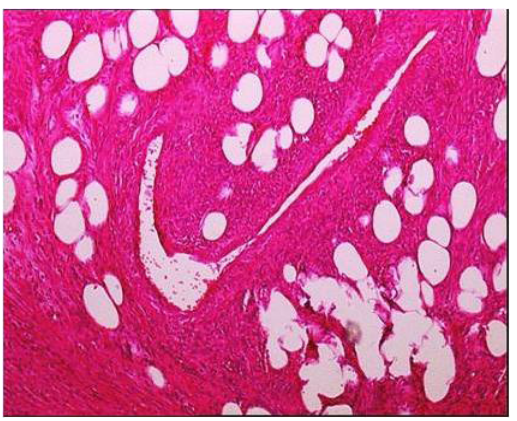

Figure 1. *The tumor sections showed many thick-walled blood vessels surrounded by smooth muscle fibers and admixed with adipose tissue (hematoxylin-eosin stain X200)*

***Table 1**. A summary of reported uterine angiomyolipomatous lesions*

Reference	*Year*	*Age*	*Location*	*Size(cm)*	*Symptom*
McKeithen et al	*1964*	*62*	*corpus*	*2*	*pelvic pressure*
Jacobs et al.	*1965*	*32*	*corpus*	*8*	*bleeding*
	35		*corpus*	*7*	*bleeding*
	77		*corpus*	*15*	*abdominal mass*
Demopoulos et al.	*1973*	*60*	*corpus*	*12*	*abdominal heaviness*
		49	*corpus*	*14*	*menometrorrhagia*
		39	*cervix*	*4*	*menometrorrhagia*
		44	*Uterus*	*16*	*menometrorrhagia*
Lo Re et al.	*1987*	*47*	*corpus*	*5*	*menometrorrhagia*
Sieinski et al.	*1989*	*52*	*corpus*	*6*	*menometrorrhagia*
		52	*cervix*	*16*	*menometrorrhagia*
		57	*cervix*	*9*	*menometrorrhagia*
Laffargue et al.	*1993*	*20*	*corpus*	*20*	*menometrorrhagia*
Shintaku et al.	*1996*	*67*	*corpus*	*7*	*uterine prolapsus*
Huang et	*2000*	*34*	*cervix*	*5*	*menometrorrhagia*
Yaegahsi et al.	*2001*	*40*	*corpus*	*12*	*abdominal distension*
Braun et al.	*2002*	*51*	*corpus*	*2*	*postmenopausal bleeding*
Cil et al.	*2003*	*32*	*corpus*	*2*	*menometrorrhagia*
Ren et al.	*2003*	*40*	*corpus*	*5*	*low back and pelvic pain*
Darai et al.	*2004*	*41*	*corpus*	*8*	*menometrorrhagia*
Kajo et al.	*2010*	*53*	*multiple*		*menometrorrhagia*
Present case	*2011*	*44*	*corpus*	*16*	*abdominal distension and menorrhagia*

atic; thus tumors may be determined incidentally. There were no reported cases with aggressive behavior so it is believed to be of benign nature. The diagnosis of angiomyolipoma can be established with ultrasound and/or computed tomography (CT). Ultrasound alone can be sufficient for diagnosis as findings are strongly suggestive, with the mass typically appearing as hyperechoic, although the precise imaging appearance depends on the amounts of fat contained in the lesion (10). While these findings were suggestive of degenerated myoma, echogenicity without shadowing and irregular margins were distinguished from leiomyoma (9). Uterine lesions, that non-classical appearance of leiomyoma on abdominopelvic ultrasound such as uterine AML must be considered differential diagnosis of the lower abdominal mass and dysfunctional uterine bleeding.

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