Uterine Angioleiomyoma: Case Report

Uterin Anjiyoleiomiyom

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ABSTRACT Angioleiomyoma is a rare form of uterine smooth muscle tumors consist of muscular and vascular components. A 31-year-old woman presented with a 3-month history of irregular vaginal bleeding and pelvic pain. Transabdominal ultrasonographic evaluation showed an intramural mass and it was excised. Microscopically, the tumor was composed of spindle, smooth muscle cells which arranged in short thick bundles intermingled with a large number of capillary vessels like cavernous and capillary hemangioma. Cellular atypia, necrosis and mitosis were not identified. Spindle cell components were immunoreactive for smooth muscle actin (SMA) and the vascular component was immunoreactive for CD34. The final histopathological diagnosis was uterine angioleiomyoma, mixed cavernous and capillary type.

Key Words: Angiomyoma; uterus

ÖZET Anjiyoleiomiyom uterin düz kas tümörlerinin, damar komponenti içeren nadir bir formudur. 31 yaşında kadın hasta 3 aydır düzensiz vajinal kanama ve pelvik ağrı şikayetiyle başvurdu. Transabdominal USG'de intramural kitle saptandı ve eksize edildi. Mikroskopik incelemede tümör kısa kalın demetler yapan iğsi düz kas hücreler ile iç içe geçmiş, kavernoz ve kapiller hemanjiyomu anımsatan, çok sayıda kapiller damardan oluşmaktaydı. Hücresel atipi, nekroz ve mitoz görülmedi. İğsi hücreli komponent düz kas aktin (SMA) ile pozitifken, vasküler komponentte CD34 pozitifliği izlendi. Sonuç olarak olgu mikst kavernöz ve kapiller tipte uterin anjiyoleiomiyom tanısı aldı.

Anahtar Kelimeler: Anjiyomiyom; uterus

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Angioleiomyoma (angiomyoma, vascular leiomyoma) is one of the rare forms of uterine smooth muscle tumors. There have been only few cases of uterine angioleiomyoma reported in the literature. We present a case of mixed cavernous and capillary type angioleiomyoma of uterus in a young woman who underwent myomectomy.

CASE REPORT

A 31-year-old gravida 5, para 5, woman presented with a 3-month history of irregular vaginal bleeding and pelvic pain. She did not have any medical

or surgical illness. There was no family history of malignancy. Gynecological examination revealed an uterus with 12 gestational weeks in size, irregular, and nontender. There was no adnexial tenderness or masses appreciated. Transabdominal ultrasonographic evaluation showed an intramural mass, measuring 62× 67 × 63 mm (Figure 1). Both the ovaries were normal. Papanicolaou smear test was negative in terms of malignancy, and proliferative endometrium was reported in fractional curettage. After discussing all the available options, she opted for a laparotomy with a diagnosis of a large intramural uterine fibroid. An exploratory laparotomy was performed and it revealed a solid mass at the uterine fundus and corpus, with normal right and left ovaries. No other gross abnormal finding was observed in the exploration of the whole abdominal cavity. Myomectomy was carried out.

On gross examination, the resected material was 8 cm in diameter, firm, well-circumscribed and unencapsulated. The cut section was reddish-brown and showed whorled pattern of fibers (Figure 2). Microscopically, the tumor was composed of spindle, smooth muscle cells which arranged in short thick bundles intermingled with a large number of capillary vessels like cavernous and capillary hemangioma (Figure 3). Cellular atypia, necrosis and mitosis were not identified. Spindle cell components were immunoreactive for smooth muscle actin (SMA) and the vascular component was immunoreactive for CD34 (Figure 4, 5). The final histopathological diagnosis was uterine angioleiomyoma (vascular leiomyoma), mixed cavernous and capillary type.

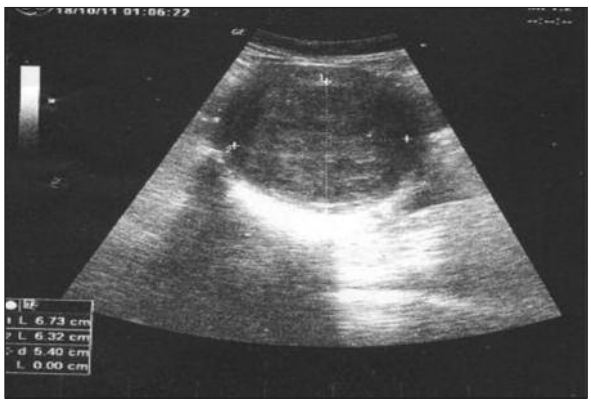

FIGURE 1: Sagital transabdominal view of intramural myoma demarcated by the calipers.

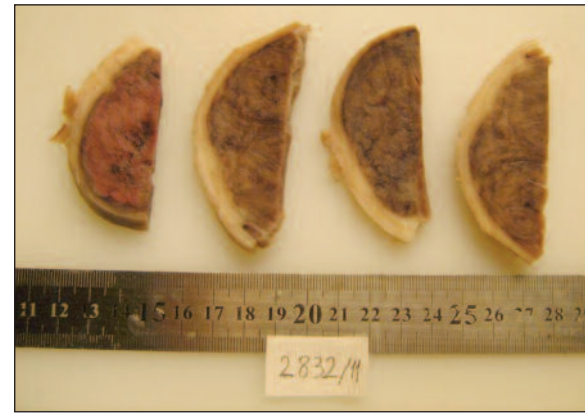

FIGURE 2: On gross examination, the cut section of the tumor is reddish-brown and shows whorled pattern of fibers.
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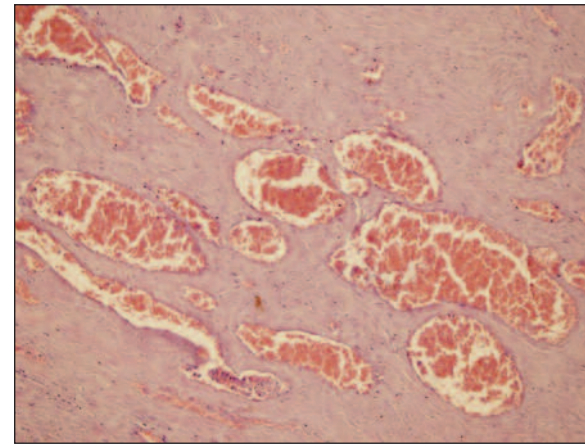

FIGURE 3: Microscopically, the tumor is composed of spindle, smooth muscle cells which arranged in short thick bundles intermingled with a large number of dilated capillary vessels (HEX200).
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Informed consent was verbally obtained.

DISCUSSION

Leiomyomas are the most common uterine neoplasms. Angioleiomyoma is a rare form of uterine smooth muscle tumors consist of muscular and vascular components. In 2009, Hakverdi et al. reported that the number of uterine angioleiomyomas was

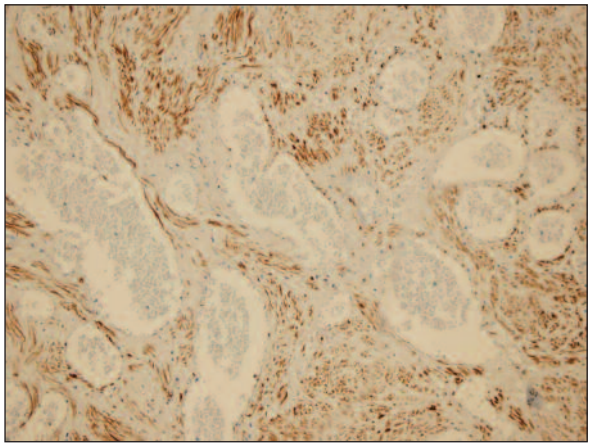

FIGURE 4: Smooth muscle actin (SMA) positivity in smooth muscle component (X200).
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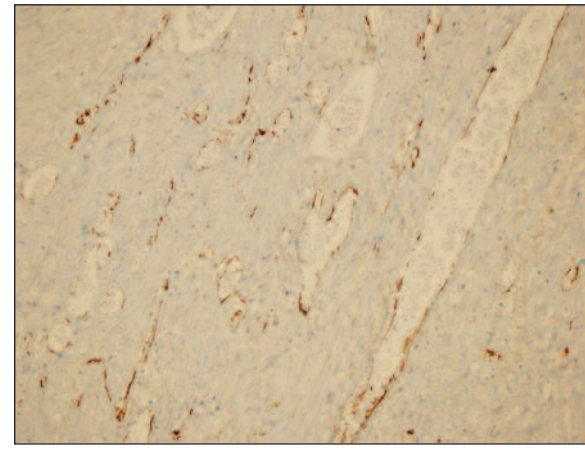

FIGURE 5: Vascular component was immunoreactive for CD34 (X200).
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eleven in English literature from 1996 to 2007.[1] So, we present the thirteenth case to our knowledge.

Angioleiomyomas typically appears as a small, solitary, subcutaneous nodule, usually in the lower extremities. They can occur in various locations like trunk wall, head and neck.[1-4] However, the female genital tract is rarely involved.[1] In the largest series there was female predominance in the lower extremity and male predominance in the upper extremity.[4] These tumors usually develop between the fourth and sixth decades.[1-3] In reported cases of uterine angiomyoma, the age ranged from 32 to 69.[1-3,5-9] Two cases had multiple foci of angioleiomyoma in the uterine wall.[1, 2]

Abdominal pain is the dominant clinical feature of the uterine angioleiomyomas while they present as tenderness masses in head and neck.[2,3] The mechanism of pain is inconclusive; it may be attributable to local ischemia from vessel contraction.[3] Anyway uterine angioleiomyomas can cause severe menorrhagia.[2] Some authors suggest that dysregulation of some growth factors and/or their receptors which regulate the angiogenesis or have other effects on vascular structures in angioleiomyomatous uterus is responsible for the excessively heavy menses.[2] When there is heavy bleeding, clinically, sometimes it can be mistaken for ectopic pregnancy or a malignant gynecological tumor.[3]

Ultrasonography (USG) is the primary modality for evaluating these tumors. Generally USG appearance is typical but sonographic characteristics of tumors undergoing cystic degeneration may be misleading and resemble cystic ovarian tumors.[5] Uterine angioleiomyomas should be considered when prominent tortuous vascular like enhancing structures are noted on Computed Tomography (CT) examination of a well demarcated soft tissue mass arising from the uterus in pelvis.[2]

On gross examination, uterine angioleiomyomas are usually described as a well circumscribed, whitish, firm nodule that measures 1,5 cm to 22 cm.[1-3,5-9] Sometimes they can have dilated vessels that can be mistaken for multiloculated and multiseptated ovarian neoplasm.[1,5]

Microscopically these tumors composed of spindle, smooth muscle cells which arranged in short thick bundles intermingled with a large number of vessels. Three histological variants of angioleiomyomas have been recognized.[1-5] Classification is based on the variable relationship between smooth muscles and vascular cavities of different shapes.[1,2] The most common (67%) is the capillary and solid type, with large numbers of narrow vascular slits

surrounded by well- differentiated smooth muscle cells. The vessels are almost always small veins. The second one is the venous type (23%). The last one is the cavernous type which has wide vascular lumina resembling cavernous hemangioma, but the septal elements are composed of smooth muscle cells as in our case.[4] Areas of myxoid changes, hyalinization, calcification and adipose tissue may be seen.[1,3] Mitotic figures and necrosis are infrequent.[1, 3] Karyotypic evaluation did not fit any of the previously described classical cytogenetic subgroups of ordinary uterine leiomyomas.[6] Sakai et al. reported a case of epithelioid vascular leiomyoma of the uterus mimicking glomangiomyoma.[7]

Jameson reported a case of uterine angiomyoma in a patient with tuberous sclerosis.[8] However, he avoided to term this tumor as angiomyolipoma because of the absence of a significant fatty component. He said that these lesions were regarded as hamartomatous growth and adipose tissue was not normally present in the uterus, then the uterine tumor would be expected to be an angiomyoma.[8] Mc Gluggage and Boyde reported three cases of uterine angiomyoma with no fat and showed negative staining for HMB45 which is positive in angiomyolipomas.[9]

The main differential diagnosis is likely to be with other benign uterine leiomyomas. Leiomyomas that exhibit perinodular hidropic degeneration may contain prominent blood vessels and submucosal polypoid leiomyomas may contain numerous thick walled blood vessels on the surface, secondary to ulceration.[9]

The study on vascular system of intramural leiomyomas by Walocha et al. revealed that usual leiomyomas contained vascular network with density similar to or lower than that of normal myometrium.[10] These are predominantly capillaries along with a few arterioles and small arteries. In contrast angioleiomyomas have abundant thick walled vessels with intersecting smooth muscle bundles. On Color Doppler ultrasound examination a mass with tumor vascularity was seen in patients with angioleiomyoma.[2] So that, transabdo- minal or transvaginal Color Doppler ultrasound examination may help in the differential diagnosis.

It can be difficult to distinguish angioleiomyoma from a hemangioma or an arterivenous malformation if the vascular component predominates. Angioleiomyomas are well circumscribed and contain at least foci of typical spindle smooth muscle cells. Hemangiomas are rare in the uterus and tend to be poorly defined grossly and microscopically.[1,3] Special stains for smooth muscle cells like SMA and vessels marker as CD34 and CD31 are necessary to differentiate angioleiomyoma from other neoplasms such as angiofibroma, fibroma and angiomyofibroblastoma.[1,3]

The main treatment is complete excision. Either angiomyomectomy or hysterectomy for women who do not wish to have more children proved to be an effective treatment and resulted in a good recovery and a satisfactory outcome.[1-3] Hysterectomy was performed in the reported of ten cases with uterine angioleiomyoma and myomectomy was performed in two patients as in our case.

In summary, angioleiomyomas (vascular leiomyoma) are extremely rare benign tumors of the uterus. It should be kept in mind when there is heavy bleeding. Cystic degenerated angioleiomyomas should be included in the differential diagnosis of a multicystic mass located in the pelvis, because it can mimic an ovarian neoplasm. Both myomectomy and simple hysterectomy are the effective treatments in these cases.

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