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Angioleiomyoma of the uterus: report of a distinctive benign leiomyoma variant

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Summary

Angioleiomyoma is a relatively rare type of leiomyoma of the uterus that originates from smooth muscle cells and contains thick-walled vessels. Angioleiomyoma is usually found in the skin of the lower extremities. Uterine angioleiomyoma has similar morphological features to that of the skin. The authors present a case of a 50-year-old woman who was admitted to the present hospital with the complaint of lower abdominal pain. On clinical examination, she was found to have a palpable lower central abdominal mass. Pelvic ultrasound revealed uterine enlargement, multiple small leiomyomas, and a large mass in the myometrium. The patient underwent total hysterectomy and bilateral salpingo-oophorectomy. On histological examination, the mass was diagnosed as angioleiomyoma. Hemangioma, angiofibroma or angiomyofibroblastoma were also included in the differential diagnosis. The treatment of choice for angioleiomyoma is surgical excision, and either angiomyomectomy or simple hysterectomy are proven to be equally effective; the decision depends on the patient's symptoms and her desire to preserve fertility.

Key words: Angioleiomyoma; Uterus; Immunohistochemistry.

Introduction

Angioleiomyoma or vascular leiomyoma is a benign mesenchymal neoplasm that is composed of smooth muscle cells and thick-walled vessels [1]. Angioleiomyomas are usually found in the subcutis of the lower extremities, head and trunk, and to the best of our knowledge, only a few cases of uterine angioleiomyomas have been reported so far [2-5]. Clinical diagnosis may be difficult but microscopically it can be easy recognized as a specific type of leiomyoma. The authors describe a case of angioleiomyoma of the uterus presenting with lower abdominal pain.

Case Report

A 50-year-old multipara premenopausal woman presented to the gynecology outpatient department with a one-month history of lower abdominal pain and abnormal uterine bleeding for the last three weeks. She mentioned that she was on medication with ramipril/hydrochlorothiazide for hypertension and that she had underwent right saphenectomy 20 years ago. Clinical examination revealed an irregularly enlarged uterus but no other masses in the pelvis. Pelvic ultrasound scan (US) revealed uterine enlargement, multiple uterine leiomyomas, and a large mass in the myometrium (Figure 1). The latter exhibited abnormally increased vascularization in the color Doppler ultrasonography. The patient underwent through extensive laboratory examinations and all results were within normal limits. Tumor markers, including CEA, CA125, CA19-9, and CA15-3, were normal as well.

The patient underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy and the surgical specimen was sent for histological examination. Grossly, the uterus weighted 850 grams and was lobulated. Multiple leiomyomas and a well-circumscribed blood-filled multicystic tumoral mass measuring 6.8 cm in greatest diameter were found in the myometrium. The cut surface of the latter was whitish-yellow in color and had a sponge-like appearance (Figure 2). The endometrium appeared unremarkable. Both ovaries and fallopian tubes were normal. Histological examination of the mass showed a moderately cellular neoplasm composed of interlacing smooth muscle bundles with interspersed abundant thick-walled vessels (Figures 3a and 3b). Mitoses were absent and neither pleomorphism nor necrosis was observed. In the immunohistochemical study, the spindle cell component showed positivity for smooth muscle actin (SMA) (Figure 4a) and vimentin while the vascular component was immunoreactive for CD34 (Figure 4b). No reactivity to desmin and S100 was found. These findings led to the diagnosis of angioleiomyoma. Histological examination of the endometrium, bilateral tubes, and ovaries revealed no significant pathology. Multiple leiomyomas and foci of adenomyosis were found in the myometrium.

During follow-up, the patient has had no complications or further symptoms and remains disease-free 2.5 years after surgery.

Discussion

Angioleiomyomas are benign, relatively common neoplasms described in the lower extremities, head, and trunk [1]. They differ from other types of leiomyomas in that they are encapsulated and contain numerous vessels. The numerous veins that are present vary in size and have muscular walls of varying thickness. On this basis, three histological subtypes have been recognized: solid, cav-

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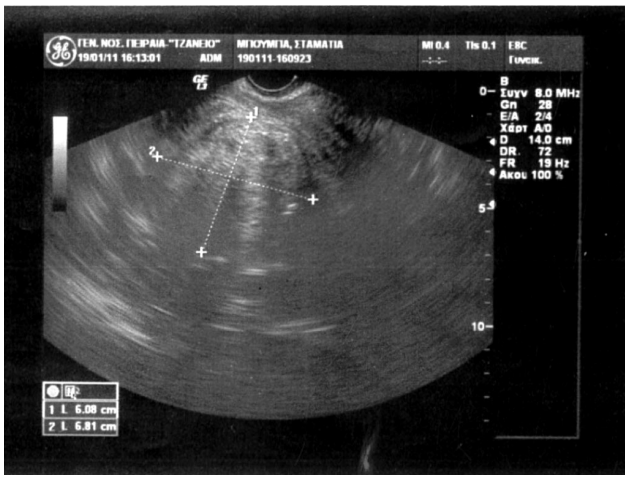

Figure 1. — US revealing a large mass in the myometrium.

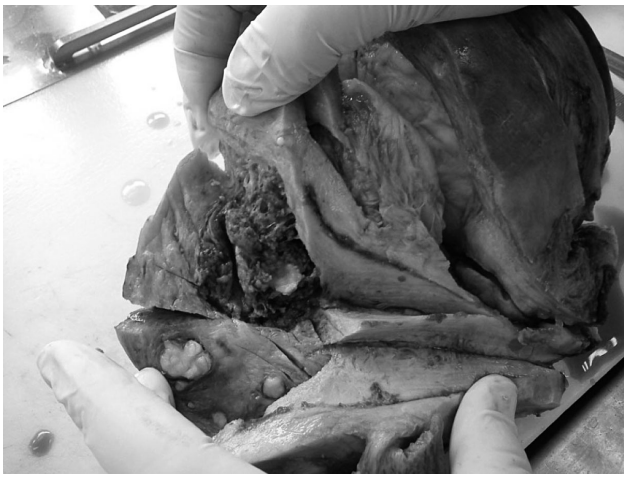

Figure 2. — Gross appearance of a large hemorrhagic mass in the myometrium.

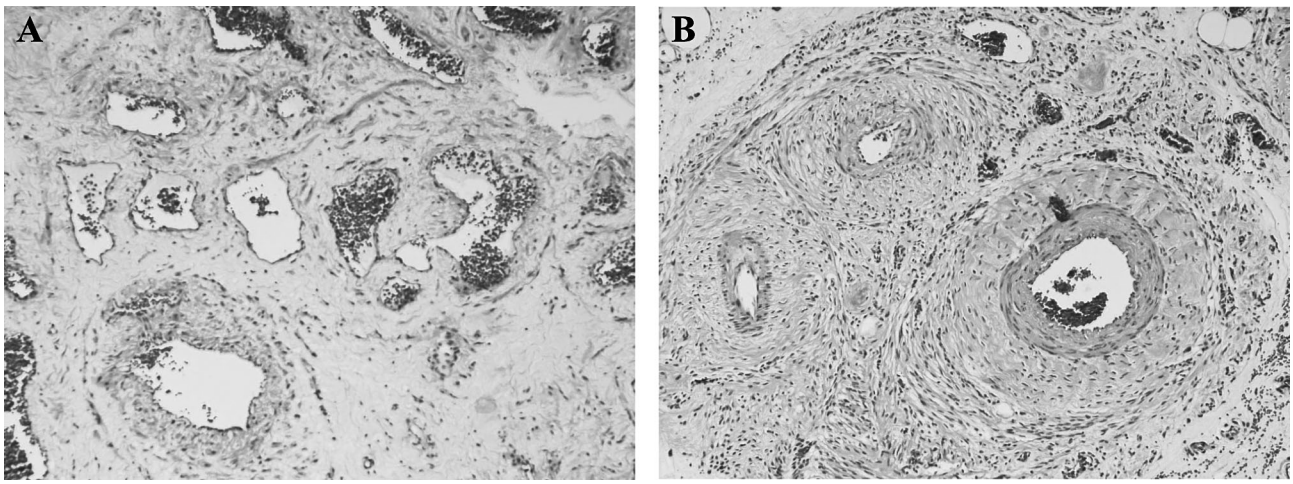

Figure 3. — A) Dilated vascular channels with small amounts of smooth muscle (H-E x100). B): Thick-walled vessels interspersed between smooth muscle cells (H-E x400).

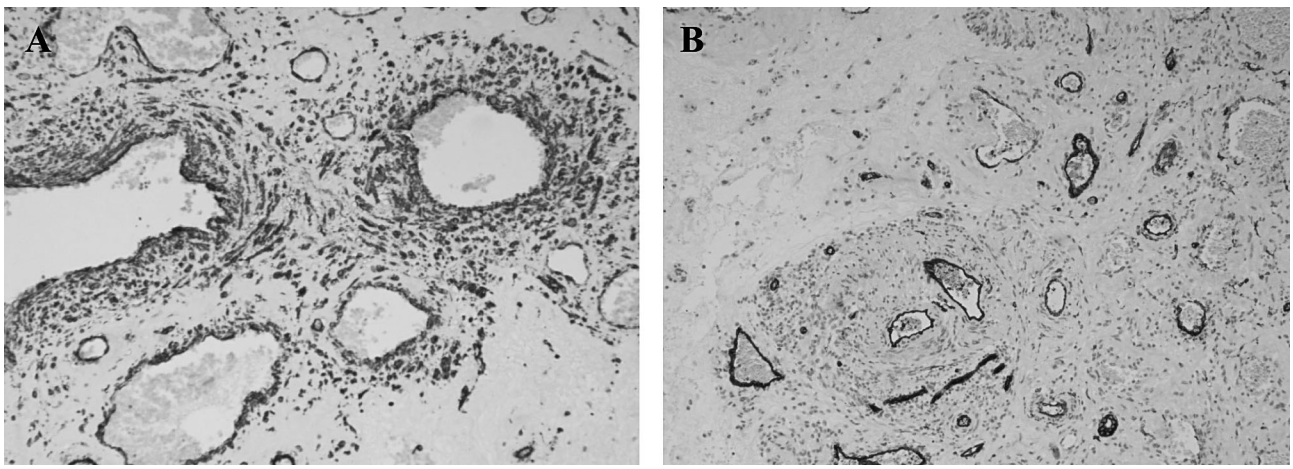

Figure 4. — A) Spindle cell component positive for smooth muscle actin (SMA x200). B) Vascular component immunoreactive for CD34 (CD34 x200).

ernous, and venous. In the solid type, the vascular channels are numerous but small while tumors of the cavernous type are composed of dilated vascular channels with small amounts of smooth muscle. Tumors of the venous type exhibit veins with thick muscular walls with smooth muscle cells that extend tangentially from the peripheries of the veins merging with the intravascular tumor substance [6].

Only very few cases of angioleiomyomas have been described at sites other than the extremities and head. These unusual sites include the oral cavity [7], the palatine tonsils [8], the scrotum [9], and the female genital tract [10]. According to Hsieh *et al.*, the number of uterine angioleiomyomas (UAL) reported in the literature was around six [2].

Similar to angioleiomyomas elsewhere, UALs are composed of smooth muscle bundles with prominent thick-walled vessels and represent a subtype of the uterine leiomyomas. 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 [11]. These are predominantly capillaries along with a few arterioles and small arteries. In contrast, UALs have abundant thick-walled vessels with intersecting smooth muscle bundles. They occur usually between the fourth and sixth decades of life and as in the presented case, pain is their dominant clinical feature. This symptom can be explained by local ischemia due to vessel contraction. However, the exact pathogenetic mechanism of the pain remains unclear. Uterine angioleiomyomas can reach a large size, presenting as an abdominal mass or can be multiple, resulting in severe menorrhagia [2]. Abnormal bleeding has also been attributed to local dysregulation of the vascular structures in the uterus.

Grossly, UALs present as circumscribed, gray-white nodules with blood-filled cystic spaces. Sometimes, the tumoral mass may contain dilated vessels that can be mistaken for multiloculated and multiseptated ovarian tumor or adenomyosis. Microscopic examination reveals abundant thick-walled vessels, separated by whorled, anastomosing fascicles of uniform, fusiform smooth muscle cells. Areas of myxoid changes, hyalinization, calcification and fat may be seen. As a rule, mitotic activity and coagulative necrosis are usually absent [3].

Clinical diagnosis may be difficult and the patients' age and symptoms very often lead to the suspicion of a malignant gynecological tumor, as occurred in the present case. Microscopically, when the vascular component predominates, angioleiomyoma needs to be differentiated from hemangioma or arteriovenous malformation. As a rule, angioleiomyomas are well-circumscribed neoplasms that conatin at least foci of typical spindled smooth muscle cells. Hemangiomas are rare in the uterus, and tend to be ill-demarcated grossly and microscopically. The differential diagnosis from other neoplasms, such as angiofibroma, fibroma, angiomyolipoma or angiofibroblastoma is based on immunohistochemical stains for smooth muscle cells (actin) and vessel markers (CD34, CD31).

Complete surgical excision is the treatment of choice, and either angiomyomyectomy or simple hysterectomy are both proven to be effective; the decision depends on the patient's symptoms and her desire to preserve fertility.

In conclusion, angioleiomyoma should be included in the differential diagnosis of a multicystic hypervascular mass located in the pelvis. However, clinical diagnosis of UAL is difficult and it seems impossible to be differentiated preoperatively from a malignant gynecological tumor . Surgical excision is the treatment of choice, either by angiomyomyectomy or by simple hysterectomy.

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