Pathologic Quiz Case

A 40-Year-Old Woman With an Unusual Uterine Tumor

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A 40-year-old nullipara woman presented with a long history of low back and pelvic pain. Magnetic resonance imaging (MRI) studies demonstrated a 5.0 × 4.0-cm, well-circumscribed, heterogeneous, cystlike mass in the right side of the pelvis (Figure 1). The mass showed a low signal on a T_1-weighted image and a high signal on a T_2-weighted image. Chemical shift artifact suggested fat content. Because of the uncertain nature of the mass, laparoscopy was performed, and a large uterine mass was found. The patient underwent a hysterectomy. Gross examination showed a 5.0 × 4.5 × 4.0-cm, well-circumscribed, yellow/tan, subserosal mass in the right fundus of the uterus. The cut surface showed a cauliflower-like appearance (Figure 2). The tumor had extended into the myometrium and focally into the endometrium. Microscopically, the tumor sections showed many thick-walled blood vessels surrounded by smooth muscle fibers and admixed with adipose tissue (Figure 3). Focally, there was a 3-mm focus of atypical smooth muscle cell proliferation containing many pleomorphic giant cells with multinucleated and multilobulated nuclei. No mitoses or necrosis were seen (Figure 4). Immunohistochemical stains, including smooth muscle actin, desmin, and HMB-45 (Dako Corporation, Carpinteria, Calif), were performed on a representative block of formalin-fixed, paraffin-embedded tissue using the avidin-biotin immunoalkaline phosphatase method. Smooth muscle actin and desmin were strongly positive in many spindle cells and atypical pleomorphic cells. No immunoreactivity was seen with antibodies directed against HMB-45.

What is your diagnosis?

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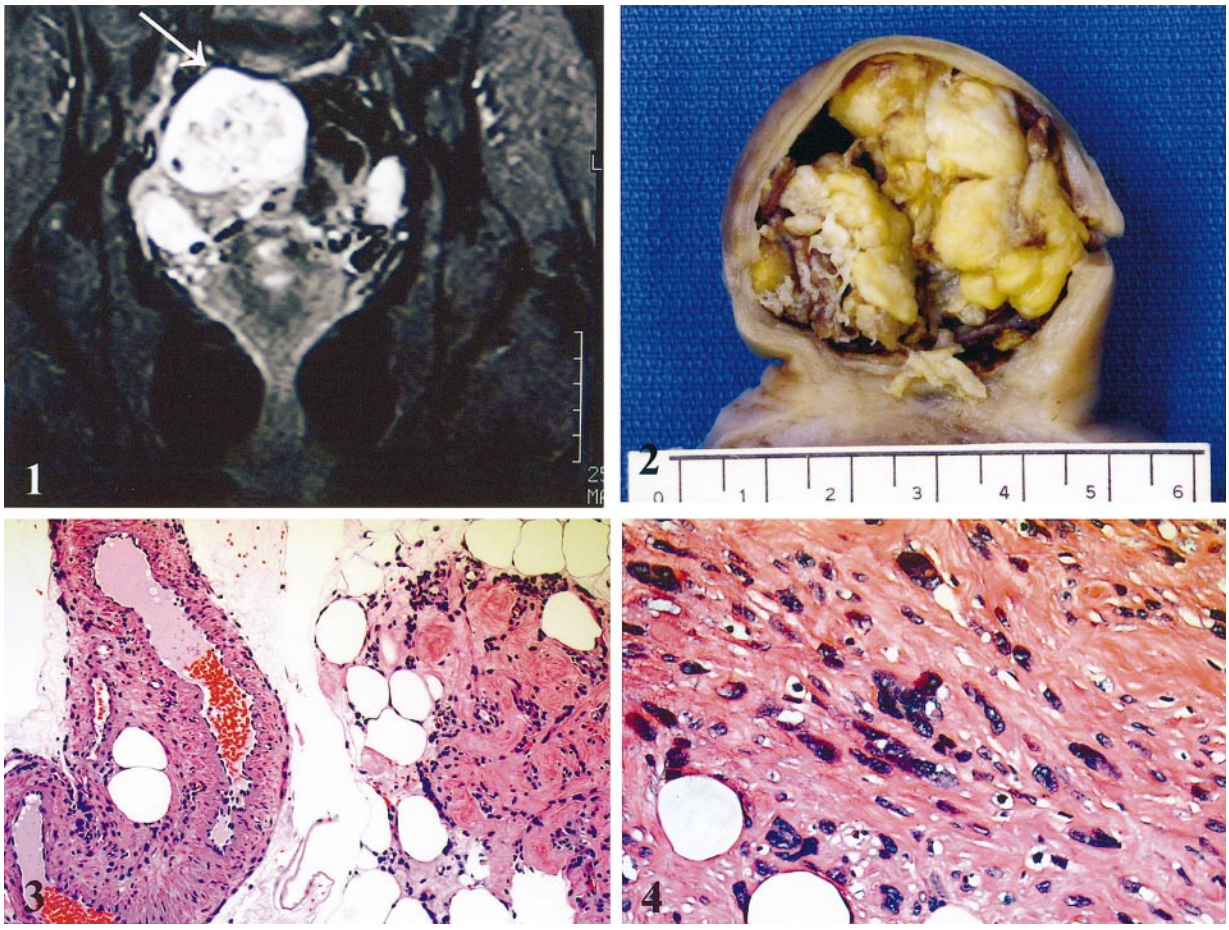

Pathologic Diagnosis: Uterine Angiolipoleiomyoma With Focal Atypical Leiomyoma

The nomenclature of a uterine tumor with mature adipose tissue, smooth muscle, and significant vascular components is quite controversial. Hamartoma,[1] lipoleiomyoma,[2–5] and angiolipoleiomyoma[6,7] have been used in the literature. Because this tumor has histologic features similar to those of renal angiomyolipoma, some authors have also named it uterine angiomyolipoma.[8–10] However, unlike a renal angiomyolipoma, the uterine angiomyolipoma is not associated with tuberous sclerosis and is HMB-45 negative. To avoid confusion, we prefer to use the term *angiolipoleiomyoma of the uterus.*

Angiolipoleiomyomas of the uterus are relatively rare tumors. The estimated incidence of angiolipoleiomyomas is 0.06% of all benign uterine tumors.[5] Only 17 cases of the uterine angiolipoleiomyoma have been reported to date.[1–10] The clinical presentations of uterine angiolipoleiomyomas are nonspecific. They are often similar to those of typical leiomyomas and include chronic abdominal and/or pelvic pain, menometrorrhagia, vaginal bleeding, urinary frequency and incontinence, and prolapses of pelvic organs. Patients may be completely asymptomatic; tumors may therefore be identified incidentally. Imaging studies including ultrasonography, computed tomography, and MRI have been used in the preoperative evaluation of uterine masses. Ultrasonography is the most commonly used image study in the reported cases. Computed tomography and MRI may be able to identify the tumor components more accurately than ultrasonography. Therefore, when used in combination with sonography, computed tomography and MRI may assist in the preoperative diagnosis of angiolipoleiomyomas.[7]

A review of the literature reveals that most angiolipoleiomyomas are located in the corpus uteri, followed by the cervix and the lower uterine segment as subserosal or intramural growths. The tumor size ranges from 2 to 16 cm with a median of 8.4 cm. Usually, the tumors are well defined with a pseudocapsule, but occasional tumors demonstrate infiltrative growth.[10] The tumors are either soft or firm, as a consequence of the amount of smooth muscle, adipose tissue, and vascular components. On the cut surface, the tumors show a gray, pink/tan, and variegated appearance. Necrosis and hemorrhage are very rare.

Microscopically, the tumors consist of smooth muscle cells, mature adipose tissue, and prominent blood vessels in varying proportions. The variability is almost as great in different parts of one tumor as among the various tumors.[3] Currently, there are no criteria about the proportions (percentage) of the 3 components for the diagnosis of angiolipoleiomyoma. The smooth muscle cells are present in both thin and thick fascicles coursing through adipose and connective tissue. They could circumferentially arrange into round nodules mimicking microscopic leiomyomas. In most cases, the smooth muscle cells are uniform and moderately cellular, without anaplasia or mitoses. However, in our case, a 3-mm focus of atypical smooth muscle cell proliferation with many pleomorphic giant cells was observed. No mitoses or necrosis was noted; therefore, we called this focus an atypical or symplastic leiomyoma. The adipose tissue consisted of mature adult-type lipocytes with unilocular, clear cytoplasm and peripheral nuclei. No nuclear atypia, pleomorphism, or immature lipoblasts were noted. There was no precise definition for so-called prominent blood vessels. Generally speaking, however, many anomalous, tortuous, small or medium-sized arteries with variably thickened walls are required for the diagnosis of an angiolipoleiomyoma. The walls of the vessels may be smudged, hyalinized, and merged imperceptibly with the surrounding fibromuscular tissue of the stroma. The capillary network is also relatively well developed. The architecture of these blood vessels differs from that of normal blood vessels. Some of the vessels are redundant, aggregated, and virtually back to back throughout the tumor.

There is little information regarding the immunohistochemical staining pattern of uterine angiolipoleiomyomas in the literature. Smooth muscle cells of uterine angiolipoleiomyomas show strong cytoplasmic positivity with antibodies against α-smooth muscle actin and desmin.[5,9,10] On the contrary, smooth muscle cells in renal angiomyolipomas demonstrate only scattered and weak desmin immunoreactivity.[9] The HMB-45–positive epithelioid cells, usually arranged around blood vessels frequently seen in the renal angiomyolipoma, have not, to our knowledge, been reported in the uterine angiolipoleiomyoma. In 2 cases of the uterine angiolipoleiomyomas reported, neither demonstrated HMB-45 immunoreactivity.[9,10]

In conclusion, uterine angiolipoleiomyoma is a relatively rare benign tumor. It usually occurs in women 40 years or older. A combination of ultrasonography, computed tomography, and MRI may assist in the preoperative diagnosis. Most tumors are located in the uterine body with subserosal or intramural growths. Demonstrations of adipose tissue, smooth muscle cells, and medium-sized thick-walled blood vessels are required for diagnosis. Immunohistochemical stains show the tumor to have strong positivity to desmin and α-smooth muscle actin but negativity to HMB-45. Atypical leiomyomatous proliferation with anaplastic, pleomorphic giant cells can arise from this tumor. Complete resection of the uterus by hysterectomy is the treatment of choice.

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