

Pelvic retroperitoneal angioleiomyoma mimicking a uterine mass

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ABSTRACT

Angioleiomyoma (vascular myoma) is a rare type of leiomyoma originating from smooth muscle cells and containing thick-walled vessels. There are only a few cases of retroperitoneal angioleiomyoma reported in the literature. Herein, we present the ultrasonography, magnetic resonance imaging and histopathological findings of an asymptomatic patient with a pelvic retroperitoneal angioleiomyoma which radiologically mimicked a uterine mass.

Key words: • leiomyoma • angioleiomyoma • retroperitoneal space • magnetic resonance imaging

Angioleiomyoma (vascular myoma) is a rare type of leiomyoma originating from smooth muscle cells and containing thick-walled vessels (1). There are only a few cases of retroperitoneal angioleiomyoma reported in the literature (2–4). Therefore there is limited information about the radiological findings of retroperitoneal angioleiomyomas. Herein, we present the imaging findings of a patient with pelvic retroperitoneal angioleiomyoma which was confirmed histopathologically following surgery. Imaging findings of the tumor mimicked a uterine mass preoperatively.

Case report

A 55-year-old perimenopausal woman was referred to our radiology department for further evaluation of a pelvic mass that was first noticed during a transvaginal ultrasonography (TVUS) examination as part of her routine gynecologic control. She had no symptoms such as menorrhagia, pelvic pain or abdominal distension. The patient mentioned a TVUS examination which had been performed at another clinic two years earlier and reported as normal.

In our department, initially, a transabdominal pelvic US examination was performed (Fig. 1). A well-demarcated, heterogeneous, large mass measuring up to 14 x 11 cm was seen. The mass lesion contained anechoic/hypoechoic areas of cystic degeneration. It was located posterolateral to the uterus, slightly displacing the uterus anteriorly. With a suspicion of subserous uterine leiomyoma, pelvic magnetic resonance imaging (MRI) was performed on a 1 Tesla magnet (GE Medical Systems, Signa 1 T, Milwaukee, Wisconsin, USA) to better demonstrate the origin and the extent of the mass. MRI revealed a large pelvic mass which could not be distinguished from the uterine corpus and cervix (Fig. 2). The mass lesion was located between the uterus and the rectum, filling nearly the entire pelvis (Fig. 2a). On T2-weighted MR images, the tumor was well-demarcated, containing extensive high intensity structures presumably due to cystic degeneration, and curvilinear signal void areas pointing out an abnormally increased vascularization in the solid components (Fig. 2b). On postcontrast T1-weighted MR images, the tumor showed enhancement except for the areas of presumed cystic degeneration (Fig. 2c). The ovaries appeared to be normal. According to the sonographic and MRI findings, the mass lesion was thought to be a large degenerated subserous leiomyoma. However, a malignant lesion could not be ruled out. Therefore pelvic mass resection and total abdominal hysterectomy with bilateral salpingo-oophorectomy were performed. During surgery, it was noticed that the mass lesion was located in the retroperitoneal space and was separate from the uterus.

During histopathological examination, a 14 x 11 cm, circumscribed, white-gray, nodular tumor with focal hemorrhages was seen macroscop-

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Received 25 February 2008; revision requested 29 February 2008; revision received 2 March 2008; accepted 5 March 2008.

Published online 5 October 2009
DOI 10.4261/1305-3825.DIR.1603-07.2

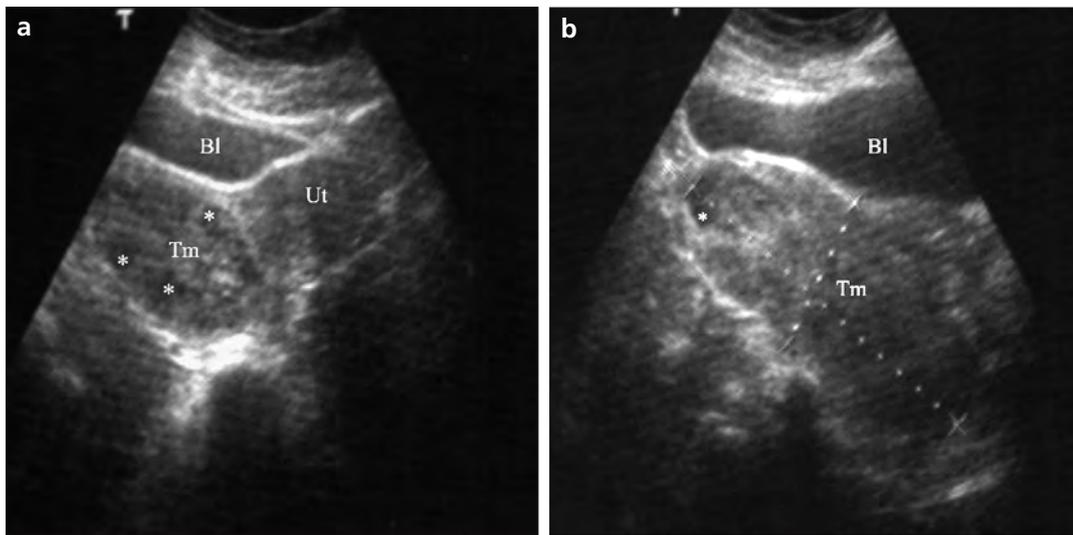


Figure 1. a, b. On transverse (a) and longitudinal (b) transabdominal pelvic US images, a well-demarcated, heterogeneous, large pelvic mass (Tm) measuring 14 x 11 cm is seen. The mass lesion includes anechoic/hypoechoic areas of cystic degeneration (*asterisks*). It is located posterolateral to the uterus (Ut), slightly displacing the uterus anteriorly (Bl, bladder).

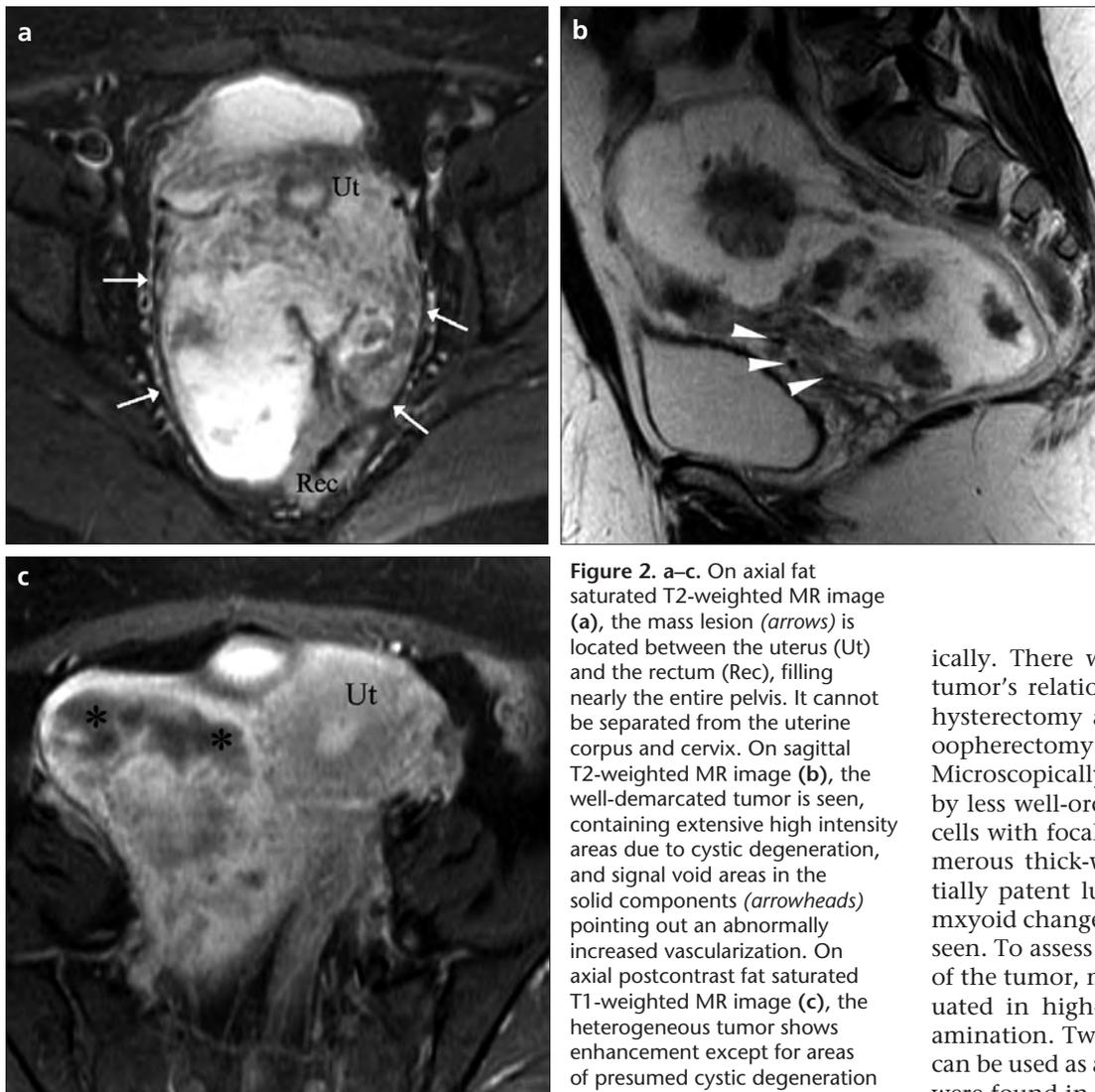


Figure 2. a–c. On axial fat saturated T2-weighted MR image (a), the mass lesion (*arrows*) is located between the uterus (Ut) and the rectum (Rec), filling nearly the entire pelvis. It cannot be separated from the uterine corpus and cervix. On sagittal T2-weighted MR image (b), the well-demarcated tumor is seen, containing extensive high intensity areas due to cystic degeneration, and signal void areas in the solid components (*arrowheads*) pointing out an abnormally increased vascularization. On axial postcontrast fat saturated T1-weighted MR image (c), the heterogeneous tumor shows enhancement except for areas of presumed cystic degeneration (*asterisks*).

ically. There was no evidence of the tumor's relation with the uterus, and hysterectomy and bilateral salpingo-oophorectomy specimen was normal. Microscopically, the tumor was formed by less well-ordered, partially fusiform cells with focal nuclear atypia and numerous thick-walled vessels with partially patent lumens (Fig. 3). Areas of myxoid change and hyalinization were seen. To assess the neoplastic potential of the tumor, mitotic activity was evaluated in high-power microscopic examination. Two mitotic figures, which can be used as a pointer of benignancy, were found in 50 high-power fields. In immunohistochemical studies, smooth

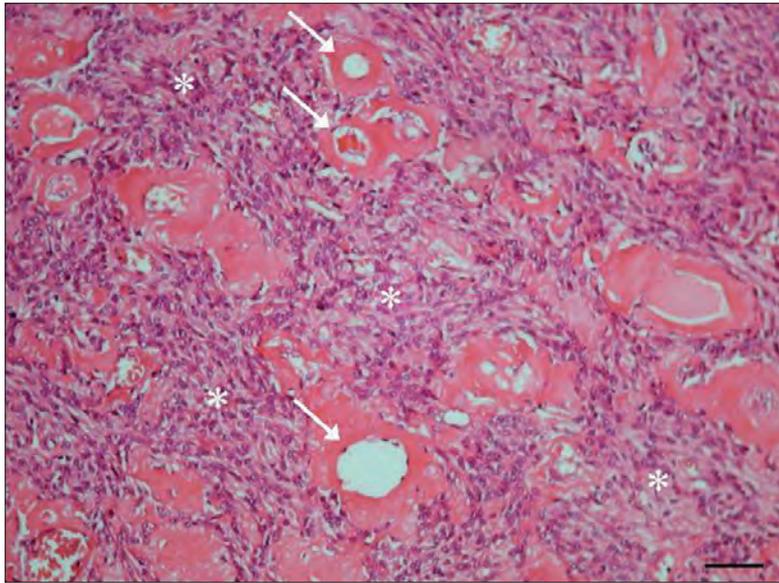


Figure 3. Microscopically, numerous thick-walled vessels (*arrows*) blend with less well-ordered smooth muscle tissue (*asterisks*) of the tumor (hematoxylin and eosin, x200, bar indicates 0.05 mm).

muscle antigens (SMA), desmin, S-100, CD34, cytokeratin, CD117, and Ki-67 were used by means of streptavidin-biotin peroxidase method. Tumor cells expressed SMA and desmin in a diffused manner while CD117, cytokeratin and S100 were negative. CD117, cytokeratin and S-100 negativity exclude the diagnosis of gastrointestinal stromal tumors (GIST), epithelial neoplasia, and peripheral nerve sheath tumors, respectively. Most of the vessels were stained with CD34 antibody, which is constitutively expressed on endothelial cells. Ki-67 positivity was detected in 8% of the tumor cells. Based on these results, the tumor was diagnosed as angioleiomyoma of the retroperitoneal space.

At one year follow-up, there was no evidence of recurrence on pelvic MRI.

Discussion

Although leiomyomas are very common tumors of the uterus, they can very rarely occur in the retroperitoneal space (2, 5). Retroperitoneal leiomyomas occur almost exclusively in women (6). Most of these tumors resemble uterine leiomyomas by histology and positive hormone receptors. They seem to have a good prognosis, with a small potential for local recurrence (2). Although radiological appearances of uterine leiomyomas are well-known, there are no series describing the radiological findings of retroperitoneal lei-

omyomas in the literature, except for a few case reports that provide limited information about their imaging findings.

The most important differential diagnoses for retroperitoneal leiomyomas include leiomyosarcomas that have a relatively higher incidence, considering this location. In a review by McLeod et al. including 118 patients with leiomyosarcomas, 23 of the tumors were retroperitoneal (7). On CT imaging, these tumors were found to be quite large in size with extensive internal necrotic or cystic changes, whereas calcification was not observed. The investigators stressed that although CT appearances of leiomyosarcomas were not specific, other findings of tumor spread, including metastases or lymphadenopathy, could suggest the diagnosis of leiomyosarcoma (7). In another article by Lane et al. including the CT findings of 10 retroperitoneal leiomyosarcomas, extensive necrotic regions were also depicted in 7 of the tumors (8). Unfortunately, imaging is generally not helpful in distinguishing leiomyomas from leiomyosarcomas, as they have non-specific radiological findings (5). Thus, histology appears to be the only choice in differentiating these two tumors. Mitotic rate, presence of cellular atypia, and coagulative tumor necrosis have been used as criteria. Tumors showing such features have a high risk of recurrence and metastasis. Also neg-

ativity for estrogen and progesterone receptors favors the diagnosis of leiomyosarcoma (2).

As seen in the presented case, it is not always possible to determine the origin of large pelvic masses using MRI, especially when the tumor exceeds 5 cm in diameter (9). The majority of large pelvic masses in female patients are uterine leiomyomas, ovarian cysts, and benign or malignant ovarian tumors. Uncommon pelvic masses include mesothelioma, sarcomas, desmoid tumors, etc. (9). Leiomyomas are well-defined and have low signal intensity on T2-weighted MR images, but can show variable appearances depending on the presence of cystic degeneration, necrosis, hemorrhage, or cellular type leiomyoma. They are isointense compared to the myometrium on T1-weighted images, and show enhancement following administration of intravenous contrast material. Unfortunately, MRI features of uterine sarcomas are similar to that of other pelvic masses. Sarcomas are isointense on T1-weighted images, and can be hyperintense or with variable intensity on T2-weighted images. They enhance heterogeneously on post-contrast images (9). In our case, both ovaries were intact and visualized separately from the mass. However, the mass was inseparable from the uterus. Because the endometrial cavity was intact on MRI, we concluded that this tumor could represent a myometrial mass such as a subserosal leiomyoma. However, given the atypical imaging features of the mass on MRI for a leiomyoma, we also considered the possibility of a malignant neoplasm such as a uterine sarcoma.

Angioleiomyoma is an uncommon type of leiomyoma that originates from smooth muscle cells, and contains thick-walled vessels (1). It can undergo degenerative changes with large cavernous deformation of the vascular spaces (10). Angioleiomyoma usually occurs in the subcutaneous tissue, most often in the lower extremities (1). It can very rarely be located in the head and neck region (11), in the submandibular gland (12), or in the uterus (10, 13–16). There are only a few cases of retroperitoneal angioleiomyoma reported in the literature (2–4). Paal and Miettinen reported a large series of retroperitoneal tumors including 56 patients; only one of them had prominent vascular pattern, that of a solid variant of angioleiomyoma (2).

Since they are very uncommon, there is not enough information about the MRI appearance of retroperitoneal angioleiomyomas. However, previous reports in the literature about uterine angioleiomyomas (10, 13–16) agree that these tumors show areas of cystic degeneration. Hsieh et al. mentioned prominent tortuous vascular-like enhancing structures in uterine angioleiomyomas on CT examination (14). In our patient, we too observed both cystic areas and abnormally increased number of vessels in the solid parts of the lesion indicating a hypervascular tumor.

In conclusion, preoperative diagnosis of a retroperitoneal leiomyoma is rarely possible and it usually cannot be differentiated from malignant tumors. Differential diagnosis on the basis of radiological features is difficult and histopathological examination is essential. Angioleiomyoma, which is an uncommon type of leiomyoma, also called vascular myoma, should be included in the differential diagnosis of retroperitoneal mass lesions having cystic components and prominent vascularization.

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