

Giant cystic leiomyoma of the uterus occupying the retroperitoneal space

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ABSTRACT

A 31-year-old nulliparous woman visited our hospital complaining of abdominal distension. Abdominal ultrasonography and computed tomography revealed a 40 × 40 × 30-cm, multilocular cystic mass extending from the upper abdomen to the pelvis. Magnetic resonance imaging (MRI) revealed a cystic tumor that was hypointense on T1-weighted images and was heterogeneously hyperintense on T2-weighted images. The final diagnosis was an 8 kg leiomyoma with cystic degeneration. Uterine leiomyomas are common benign tumors in females of reproductive age. However, subserosal leiomyomas with complete cystic degeneration of the retroperitoneal space are rare, and they are difficult to accurately diagnose without pathological examination.

CASE REPORT

CASE REPORT

A 31-year-old nulliparous woman visited our hospital with a history of abdominal distension and weight gain for two months. She did not have abdominal pain or any gastrointestinal symptoms. Her menstrual cycle was regular, and she did not have menstrual disorders such as hypermenorrhea or severe dysmenorrhea. Her abdomen was fully distended, and a soft, nontender mass was palpable. Chest X-ray and routine blood investigation results were normal. The levels of carcinoembryonic antigen, alpha-fetoprotein, and cancer antigen 125 were also within normal limits.

Abdominal ultrasonography revealed a large, multilocular cystic mass occupying the abdomen from the pelvis to the upper abdominal cavity. The cystic parts of the tumor were almost anechoic, suggesting serous fluid collection (Figure 1). Abdominopelvic computed tomography (CT) revealed a huge cystic mass. The tumor growth cranially compressed the small intestine and sigmoid colon and laterally compressed the rectum and uterus (Figure 2). Magnetic resonance imaging

(MRI) of the pelvis revealed a 40 × 40 × 30-cm, well-defined, multilocular, cystic mass that was hypointense on T1-weighted images and was heterogeneously hyperintense on T2-weighted images. Gadolinium-enhanced MRI revealed an enhanced tumor rim and mild soft tissue density. The mass was mildly hyperintense on diffusion-weighted imaging (Figures 3A-3E).

A provisional diagnosis of an abdominal tumor of unknown origin was made. Tumor resection via median laparotomy was attempted. On opening the abdomen, we observed a large, well-defined, retroperitoneal tumor that was occupying the pelvis and growing cranially. The tumor was easily dissected without massive bleeding in the upper abdomen because there was no infiltration or invasion into the surrounding tissues in this region; however, it was firmly fixed to the left isthmus of the uterus. Gross resection was performed with left salpingo-oophorectomy because the left ovary was originally suspected of being the primary organ (Figure 4).

The resected mass weighed 8 kg and was composed of solid and cystic components. The cystic portions contained clear, straw-colored serous fluid. Examination of a cross-section revealed a soft whitish-gray mass with myxoid or cystic changes (Figure 5). Microscopic examination revealed that the tumor was not connected to the left ovary.

Hematoxylin and eosin staining revealed the presence of spindle cells distributed in a myxoid or edematous stroma. In some areas, the spindle cells either encircled the vessels and progressed between dilated vessels or showed a cord-like arrangement. The spindle cell density was not high and cellular atypia was low (Figures 6A-6C). The spindle cells were immunoreactive for desmin and alpha-smooth muscle actin (α -SMA) and negative for S-100 and CD34, suggesting that the tumor originated from smooth muscle.

On the basis of surgical and histopathological findings, the tumor was diagnosed as a giant subserous leiomyoma with severe cystic degeneration that originated from the normal-sized uterus near the left-side of the broad ligament.

The patient's postoperative course was uneventful. At 1 year after surgery, she showed no signs of recurrence and had a normal menstrual cycle.

DISCUSSION

The retroperitoneal space is the region between the peritoneum and posterior parietal wall of the abdominal cavity, and it extends from the pelvic floor to the diaphragm. This space contains urogenital organs, adrenal glands, blood vessels, nerve fibers and ganglia, lymph nodes, lymph channels, and adipose and connective tissues. Cysts arising in the retroperitoneal space are exceedingly rare [1, 2]. Retroperitoneal cysts are classified as neoplastic cysts, which include cystic lymphangioma, mucinous cystadenoma, cystic mesothelioma, müllerian cysts, and non-neoplastic cysts, including urinomas and lymphoceles [1-3]. Some types of solid retroperitoneal tumors can also undergo cystic degeneration. Chemotherapy has been reported to induce central tumor necrosis and cystic degeneration; moreover, large solitary tumors sometime present a partially cystic appearance [4]. Retroperitoneal cysts can produce obstructive and compressive symptoms depending on their location and size; however, these symptoms are often vague and nonspecific [2, 5-6].

Uterine leiomyoma is a common uterine tumor that occurs in >20% of women aged >30-years. Typical uterine leiomyoma is well recognized in MRI with intermediate and low signal intensities on T1- and T2-weighted images, but it is not easy to diagnose with CT. Uterine leiomyomas exhibit various growth patterns [7-9]. Common symptoms include heavy bleeding, dysmenorrhea, pelvic pain, and tumor bulk-related signs. Leiomyomas are classified into three subgroups on the basis of their location relative to uterine structures: intramural, submucosal, or subserosal. Subserosal uterine leiomyomas are relatively asymptomatic; therefore, they

sometimes become very large before the patient becomes aware of them. These tumors sometimes demonstrate retroperitoneal growth, usually within the broad ligament [10-11].

Smooth muscle tumors sometimes undergo strong secondary changes, such as hyaline, myxoid, red, and cystic degeneration, depending on their location or size [8-10]. Blood supply of leiomyomas is less than that of the normal myometrium; therefore, large leiomyomas may degenerate because of insufficient blood supply [10]. Hyaline degeneration is the most common type of degeneration and is hypointense on T2-weighted MRI. Myxoid changes can occur in both uterine leiomyoma and leiomyosarcoma. Red degeneration is an ischemic change that is most commonly observed during pregnancy. Cystic degeneration may be considered as an extreme sequel of edema, and it is reportedly observed in approximately 4% of all uterine leiomyomas [7, 10]. However, a high degree (11.8%) of cystic degeneration has been reported in uterine leiomyomas extending to the retroperitoneal space [6]. The cystic spaces usually contain fluid, and they appear hypointense and hyperintense on T1- and T2-weighted images, respectively. When submucosal uterine leiomyomas with cystic degeneration show retroperitoneal growth, it is difficult to differentiate them from ovarian cysts or other retroperitoneal cysts via any imaging modality. For example, most retroperitoneal cysts are hypointense and hyperintense on T1- and T2-weighted MRI, respectively [12]. The wall and septum of cystic leiomyomas are rather thick [13], whereas those of ovarian cysts and most retroperitoneal cysts are thin [3, 12].

In the present case, histopathological findings confirmed a benign smooth muscle tumor with cystic and myxoid components. Immunopathological results indicated that the tumor originated from smooth muscle and not the nervous system, because the tumor cells were positive for desmin and α -SMA and negative for S-100. A solitary fibrous tumor was not considered because it was negative for CD34 [4, 11].

Most retroperitoneal tumors are malignant [11, 12]. The incidence of leiomyomas among primary retroperitoneal tumors is 0.5-1.2% [6, 11]. There are several theories regarding the origin of retroperitoneal leiomyomas. Pedunculated retroperitoneal uterine leiomyomas that detach from the uterus and feed from adjacent vascular supplies are called parasitic leiomyomas [5, 7-8]. Uterine leiomyomas that originate from the broad ligament may grow into the retroperitoneal cavity. In such cases, the attachment of the leiomyoma to the uterus becomes restricted, and it may transform into a parasitic leiomyoma. This hypothesis gives one explanation for the high degree of cystic degeneration observed with retroperitoneal uterine leiomyomas. However, it does not provide a reasonable explanation for male cases; a study has reported 5 male cases among 56 retroperitoneal leiomyoma cases [11]. In addition, retroperitoneal leiomyomas may originate from embryonal remnants or local vessel musculature [14].

More than 40% of patients affected by retroperitoneal leiomyomas are reported to have a concurrent uterine

leiomyoma or a remote history of hysterectomy for uterine leiomyoma treatment [11]. Moreover, 73.1% of retroperitoneal leiomyomas are found in the pelvis [6]. On the basis of this knowledge, some retroperitoneal leiomyomas could arise from the uterus. The tumor described here was completely embedded in the retroperitoneal space but partially connected to the uterus, although there was no concurrent uterine leiomyoma in this case.

We encountered a rare giant cystic leiomyoma that originated from the uterus near the left side of the broad ligament and intruded into the retroperitoneal space. Our experience with this case should aid in the differential diagnosis of a retroperitoneal cystic mass in young women.

TEACHING POINT

Uterine leiomyoma can grow into the retroperitoneal space and sometimes degenerate like primary cystic tumors. Because uterine leiomyoma is a common tumor in reproductive-age females, it should be considered as a differential diagnosis of a retroperitoneal cystic mass in young women.

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FIGURES

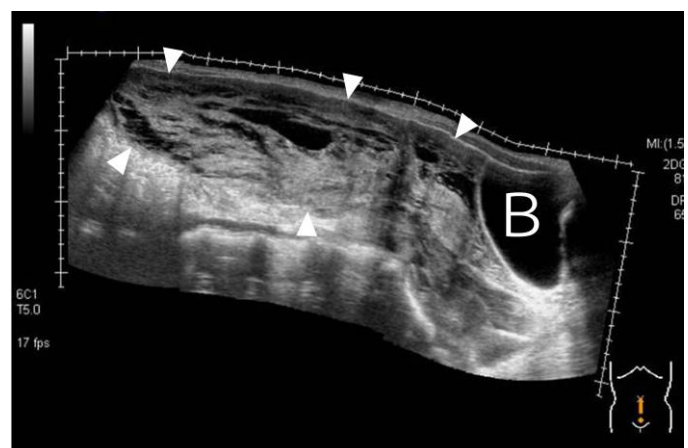


Figure 1: Abdominal ultrasonography of a 31-year-old female with retroperitoneal giant cystic leiomyoma. A large, multilocular cystic mass occupies the abdominal space from the upper abdomen to the pelvis. The wall and septum are thick. The cystic parts of the tumor are almost anechoic, suggesting serous fluid collection. (TOSHIBA, AplioMX; convex probe, 3.5MHz) B, bladder; arrowhead, cystic tumor.

Figure 2 (right): Computed Tomography of a 31-year-old female with retroperitoneal giant cystic leiomyoma. Axial CT showing a huge mass occupying the entire pelvis that extends up to the upper abdomen, displacing the rectum laterally (arrow) and surrounding the uterus (arrowhead). (GE LightSpeed, 16-Slice Scanner; Protocol: 370mA, 120kV, 10mm slice thickness, no contrast).

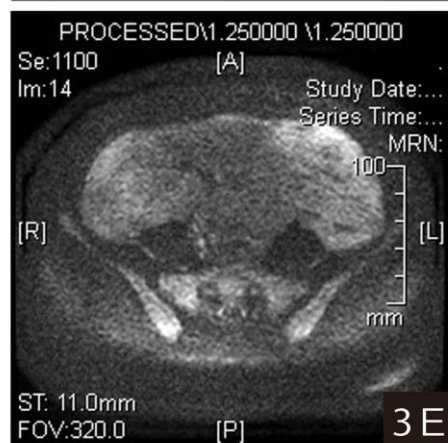
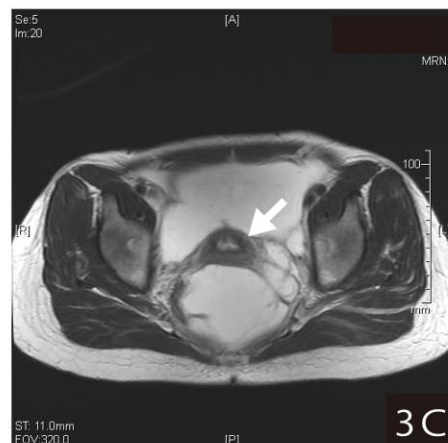
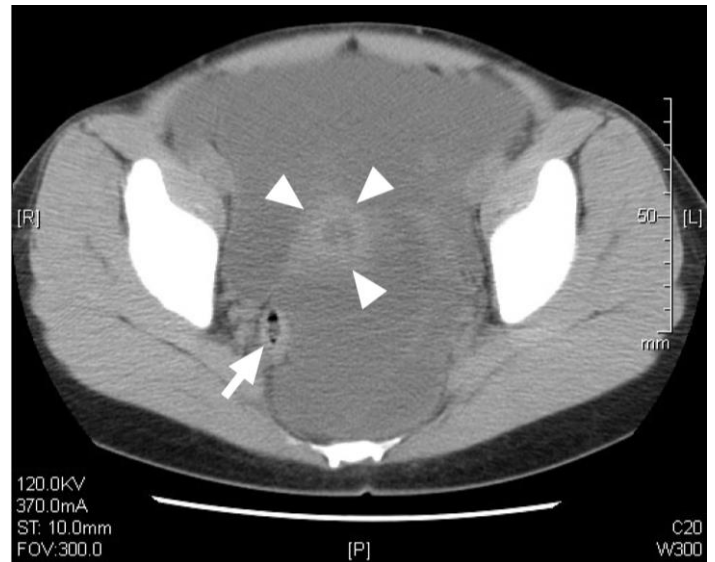


Figure 3 (left): Magnetic resonance imaging of a 31-year-old female with retroperitoneal giant cystic leiomyoma.

(A) Sagittal T1-weighted MRI showing a 40 × 40 × 30-cm well-defined cystic mass. It is hypointense, but it is hyperintense relative to the urinary bladder.

(B) Sagittal T2-weighted MRI showing a heterogeneously hyperintense cystic mass that is isointense relative to muscle. A normal-sized uterine body is visible in the middle of the pelvis.

(C) Axial T2-weighted MRI showing a cystic mass that occupied almost the entire pelvic space. A normal-sized uterus is shown in the middle of the pelvis.

(D) Coronal, contrast-enhanced, fat-saturated T1-weighted MRI showing that the mass has an enhanced rim and some soft tissue components.

(E) Axial diffusion-weighted MRI showing a large hyperintense tumor.

Arrows, uterine body.

(GE SIGNA, 1.5Tesla MR scanner; Protocol: Fasto Spin Echo (FSE) sequence, TR=4017, TE=103, with 10ml gadopentetate dimeglumine (Magnevist, Bayer Pharma, Japan) injection).

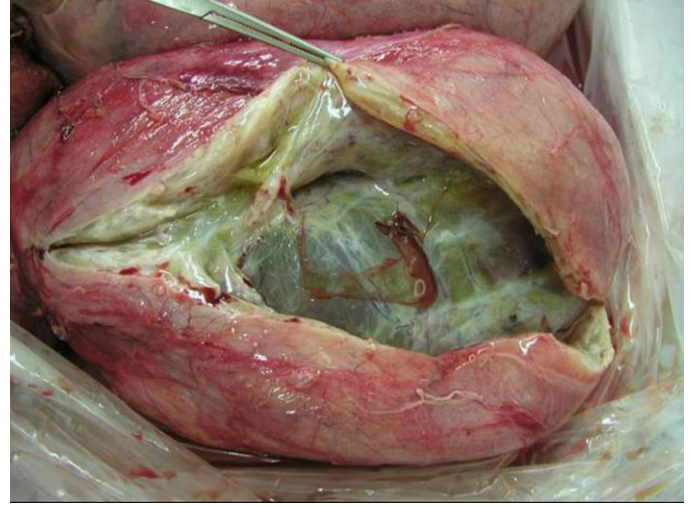
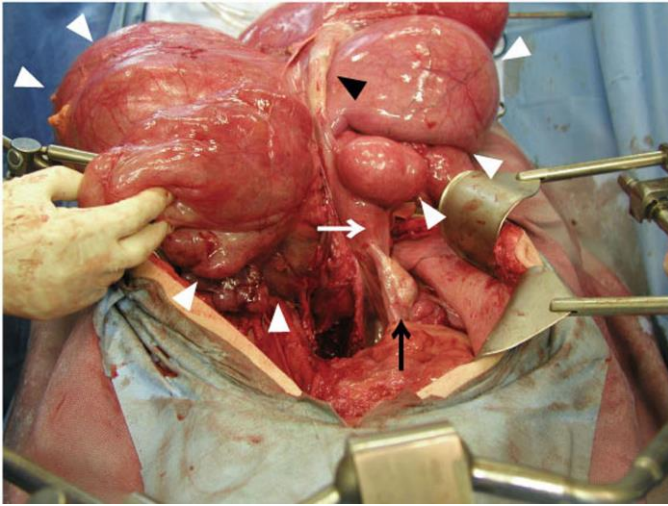


Figure 4: Macroscopic pathology of a 31-year-old female with retroperitoneal giant cystic leiomyoma.

A large well-defined mass occupies the retroperitoneal space. The tumor is shown to cranially compress the sigmoid colon and small intestine. The tumor has not infiltrated into or invaded the surrounding tissues; however, it is firmly fixed to the left isthmus of the uterus.

White arrow heads, lobulated cystic mass

Black arrow head, left ovary

Black arrow, right ovary

White arrow, uterus

Figure 5: Macroscopic pathology of a 31-year-old female with retroperitoneal giant cystic leiomyoma.

An 8-kg lobulated, multilocular, cystic mass with a massive solid component was removed. Cross-section shows a soft, whitish-gray mass with myxoid or cystic changes. The wall and septum are thick, and the cystic part contained clear, straw-colored, serous fluid.

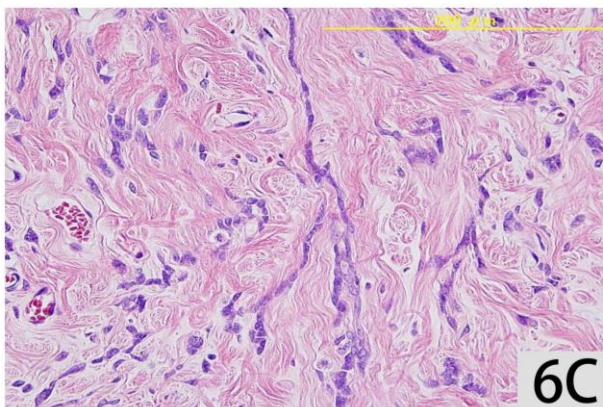
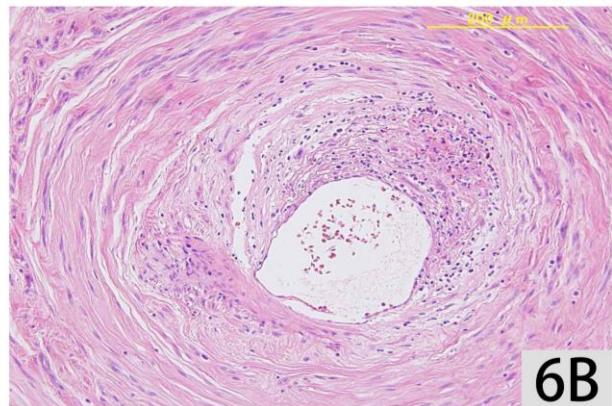
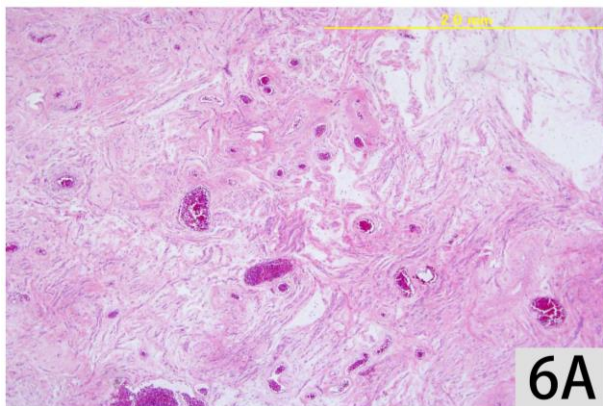


Figure 6: Microscopic findings (Hematoxylin and eosin staining) of a 31-year-old female with retroperitoneal giant cystic leiomyoma.

(A) Spindle cells are distributed in a myxoid or edematous stroma ($\times 40$).

(B) The spindle cells are shown to encircle the vessels ($\times 200$).

(C) The spindle cells that show a cord-like arrangement ($\times 400$).

Etiology	Unknown
Incidence	0.5-1.2% within primary retroperitoneal tumors
Gender ratio	female 10 : male 1
Age predilection	> 30 years
Risk factors	> 40% have concurrent uterine myoma or a remote history of uterine myoma
Symptoms	Vague abdominal, lumbar pain, gastric and intestinal disorders, nausea, pelvic mass palpation
Treatment	Surgical removal
Prognosis	Benign tumor. Good prognosis

Table 1: Summary table of retroperitoneal leiomyoma

Differential diagnosis	Non-imaging feature	CT	MRI
Retroperitoneal leiomyoma with cystic degeneration	Common with uterine myoma	Multilocular mass with thick walls or septa	T1-hypointense, T2-hyperintense
Cystic lymphangioma	Contains clear fluid or milky fluid and more common in men	Thin-walled unilocular or multilocular cyst with attenuation values ranging from that of fat to that of fluid	T1-hypointense, T2-hyperintense Chyle content can produce T1-hyperintense, T2-intermediate signals
Cystic mesothelioma	Contains watery fluid and is more common in females	Thin-walled uni- or multilocular cyst	T1-hypointense, T2-hyperintense
Müllerian cyst	Most often found in the pelvis	Thin-walled uni- or multilocular cyst	T1-hypointense, T2-hyperintense
Mucinous cystadenoma	Seen in women with normal ovaries	Thin-walled unilocular cyst, sometimes with calcification	A well-defined unilocular homogenous cyst
Urinoma	Hydronephrosis is found in most cases	Well-defined fluid collection	T1-hypointense, T2-hyperintense
Cystic solitary fibrous tumor	Mass with central necrosis	Soft-tissue attenuation mass with hemorrhage or necrosis	T1-hypointense or -isointense, T2-hypointense or -isointense mass with cystic part

Table 2: Differential diagnosis table for retroperitoneal leiomyoma

ABBREVIATIONS

α-SMA = alpha-smooth muscle actin
CT = Computed Tomography
MRI = Magnetic Resonance Imaging

KEYWORDS

retroperitoneal cyst; retroperitoneal leiomyoma; cystic degeneration

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