Multilocular cystic leiomyoma of the anterolateral abdominal wall
A case report and literature review

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Abstract
Rationale: Leiomyomas arising from the anterolateral abdominal wall are uncommon, and their pathogenesis remains unknown. We present the 15th case of such a tumor, having this unique tumor morphology, followed by a detailed discussion on disease pathogenesis.

Patient concerns: A 48-year-old, asymptomatic perimenopausal, multiparous Japanese woman presented with a left-sided pelvic mass. She had no history of previous surgeries or uterine leiomyomas. Although a transabdominal ultrasonogram raised suspicions of an ovarian tumor, a transvaginal ultrasonogram confirmed normal ovaries. Radiological images showed a multilocular cystic mass with enhanced solid lesions connected to the uterus. Retrospective radiological evaluation showed that the mass was largely connected to the peritoneum of the anterolateral abdominal wall.

Interventions: Intraoperatively, the mass appeared as a dome-like protrusion from the left lower quadrant of the abdominal wall, without connection to the uterus, ovaries, or the left round ligament. No other peritoneal masses were seen. The mass was easily enucleated from the abdominal wall. Pathology confirmed that the mass was a leiomyoma with hydropic and myxoid degeneration. No striated muscle tissues were noted between the tumor and resection margin, but a thin smooth muscle layer, positive for hormone receptors, was present at the periphery, suggesting the origin of the tumor.

Lessons: Benign leiomyomas of the anterolateral abdominal wall likely originate from Müllerian-like smooth muscle remnants in this region. They should be considered in the differential diagnosis of solid and cystic masses and be distinguished from uterine and ovarian masses on imaging to avoid unnecessary organ resection.

Abbreviations: AR = androgen receptor, CT = computed tomography, ER = estrogen receptor, MRI = magnetic resonance imaging, PgR = progesterone receptor, US = ultrasonography.

Keywords: abdominal wall, leiomyoma, pathogenesis

1. Introduction
Leiomyomas of the anterolateral abdominal wall are rare, with only 14 cases reported, to date. [1-13] Herein, we present a case of an anterolateral abdominal wall leiomyoma, as well as a literature review describing the clinicopathological characteristics of these lesions. This is the first case showing an ovarian tumor-like multilocular cystic morphology in conjunction with a discussion of differential diagnoses and tumorigenesis.

2. Case presentation
2.1. Patient information
A 48-year-old, asymptomatic, Japanese woman was referred to our hospital for further investigation of a left pelvic cavity tumor identified on transabdominal ultrasonography (US) and initially thought to represent an ovarian tumor. She had a history of atopic dermatitis, but no history of surgery or treatment of uterine leiomyomas. She had given birth to 2 healthy children by vaginal delivery. Her menstrual cycle was regular, with no history of dysmenorrhea.

2.2. Clinical findings
The patient’s vital signs were unremarkable. On clinical examination, the mass was smooth, firm, well demarcated, and nontender. A transvaginal US confirmed a 93 × 90 × 62-mm multilocular mass, which appeared connected to the posterior wall of the uterus. Her bilateral ovaries were normal, and there was no associated ascites. Based on these features, a uterine
tumor was suspected. Endoscopy did not reveal any abnormality of the upper and lower intestinal tracts. Various tumor markers, including carcinoembryonic antigen, cancer antigen 72–4, carbohydrate antigen 19–9, and cancer antigen 125 were within normal ranges. Computed tomography (CT) showed a low-density mass located mainly in the left pelvic cavity, and contrast enhanced CT revealed heterogeneous enhancement of the tumor (Fig. 1A and B). Nonenhanced and enhanced magnetic resonance imaging (MRI) (Fig. 1C–E) revealed that the mass was a multilocular cystic lesion containing solid portions and fluid-filled cavities. A subserosal uterine tumor was suspected, but the presence of a left ovarian tumor could not be excluded because the left ovary was not visible on CT and MRI images. Due to suspicion of malignancy, surgical resection was planned. During surgery, the mass was found adherent to the left lower quadrant of the anterolateral abdominal wall and showed dome-like protrusions towards the peritoneal cavity (Fig. 2A and B). The intestinal tract, uterus, bilateral uterine adnexa (Fig. 2A), and left round ligament were not adherent to the tumor, and they appeared normal. No other peritoneal masses or signs of disseminated leiomyomatosis peritonealis were present. The cystic lesion was easily exfoliated from the abdominal wall, without damage to the abdominal wall muscle layers. The feeding vessel was an inferior epigastric artery. The patient was discharged 6 days, postoperatively, without sequelae; she remained disease-free at the 4-month follow-up. Retrospective

![Figure 1](image1.png)

**Figure 1.** Radiographic images of the tumor. (A and B) Enhanced computed tomography images. A heterogeneously enhanced nodular lesion is present mainly in the left pelvic cavity (A). The tumor appears broadly connected to the anterolateral abdominal wall (B). (C–E) Magnetic resonance images of the sagittal planes. The tumor appears to connect to the posterior wall of the uterus and to the anterior abdominal wall. The tumor had low signal intensity on a T1-weighted image (C). High-signal-intensity areas with low-intensity lesions are seen on a T2-weighted image (D). A multilocular enhancement pattern with enhanced solid lesions is seen on a contrast-enhanced, fat-suppressed T1-weighted image (E). The white bars represent 5 cm.

![Figure 2](image2.png)

**Figure 2.** Macroscopic images during the surgery. (A) The tumor (arrow) attaches to the left lower quadrant of the anterolateral abdominal wall and does not connect to the bilateral ovaries (arrowheads) or to the uterus that is grasped with forceps. (B) The tumor shows dome-like protrusions towards the peritoneal cavity and does not attach other peritoneal organs.
evaluation of the radiological images confirmed that the mass was largely connected to the anterolateral abdominal wall, confirming the intra-operative findings (Fig. 1B–E).

2.3. Pathological findings

Macroscopically, a well-demarcated mass measuring 110 × 85 × 65 mm and weighing 300 g was seen. The cut surface was solid, with several cavities containing gelatinous materials (Fig. 3A). Microscopically, the solid and cystic portions of the mass were composed of fascicular and interlacing bland spindle cells (Fig. 3B–E). The various-sized cavities were likely due to edematous changes (Fig. 3C); mucinous degenerations, highlighted by periodic acid-Schiff and Alcian Blue, were identified. There was no evidence of mitoses or coagulation necrosis, and no features of endometriosis. A leiomyoma was suspected, with possible differential diagnoses including a solitary fibrous tumor, endometrial stromal tumor, extragastrointestinal stromal tumor, perivascular epithelioid cell tumor, and schwannoma. Immunohistochemically, the spindle cells were diffusely positive for α-smooth muscle actin (1A4, DAKO, Glostrup, Denmark), desmin (D33, DAKO, Fig. 3G), estrogen receptor (ER; 1D5, DAKO, Fig. 3H), and partly positive for h-caldesmon (h-CD, DAKO), progesterone receptor (PgR; PgR636, DAKO), androgen receptor (AR; AR27, Novocastra Laboratories, Newcastle, UK), WT-1 (6F-H2, DAKO, Fig. 3I). They were negative for S-100 protein (polyclonal, DAKO), melanosome (HMB45, DAKO), melan-A (A103, Novocastra Laboratories), CD10 (56C6, Novocastra Laboratories), CD34 (My10, Becton Dickinson, San Diego, CA), and Bcl-2 protein (AR27, Novocastra Laboratories). The Ki-67 labeling index was <1% (Fig. 3J). These findings were consistent with a final diagnosis of a leiomyoma with mucinous and hydropic degeneration. Between the tumor and the resection margin, a layer of nonatypical spindle cells (Fig. 3B, D, F–J) was seen extending throughout the whole mount preparation of the resected tissue. The spindle cell layer was diffusely positive for smooth muscle markers (Fig. 3G), ER (Fig. 3H), and PgR; largely negative for Bcl-2 protein (Fig. 3I), AR, and WT-1; and almost negative for Ki-67 (Fig. 3J). Striated
Anterolateral abdominal wall leiomyomas tend to be located either above, within, or under the rectus abdominis. Those that are located beneath the rectus abdominis muscle may result in a delayed diagnosis, possibly because they are grossly invisible or poorly palpable.

Our literature review revealed that clinical differential diagnoses of anterolateral abdominal wall leiomyomas include ovarian tumors,[4,8] uterine leiomyomas,[4,8] and desmoid tumors.[6] The preoperative diagnosis of a degenerated abdominal wall leiomyoma is difficult when the tumor is located close to the uterus or uterine adnexa. Clinicians should pay attention to the positional relationship between a pelvic cavity tumor, uterus, uterine adnexa, and the abdominal wall. If the pelvic cavity tumor broadly attaches to the abdominal wall, as in the present case, clinicians should be mindful of the possible differential diagnosis of an abdominal wall tumor. Parasitic leiomyomas can grow in abdominal wall incisions after laparoscopic myomectomy,[15] therefore, obtaining a patient’s past medical history and carefully examining the abdomen are important. Pathological exploration is required to rule out a desmoid tumor because more than two-thirds of abdominal wall desmoid tumors show well-defined borders.[16]

The multilocular, partially cystic morphology of the present tumor was likely due to marked hydropic and myxoid...
degereation. Degenerative or regressive changes, such as fibrosis, calcification, and cystic and myxoid changes, are frequently seen in leiomyomas of the deep soft tissue; however, a multilocular cystic tumor has not been previously reported in published cases of leiomyomas of the deep soft tissue or abdominal wall. One case of an anterior abdominal wall lipoleiomyoma was clinically diagnosed as a dermoid tumor due to the fatty tumor component rather than due to cystic degeneration. Degenerative or regressive changes are also frequently found in uterine leiomyomas, and multilocular uterine leiomyomas mimicking ovarian tumors have been reported. Hydropic and mucinous degeneration can contribute to the cystic morphology of uterine leiomyomas. Leiomyomas of the deep soft tissue are considered analogs of uterine leiomyomas. The cause of degeneration is unknown. Kamat et al suggested that cystic degeneration of uterine leiomyomas represents a late stage of hyaline degeneration; however, no hyaline degeneration was observed in our case.

The histological diagnosis of our case was uncomplicated due to the presence of smooth muscle differentiation features, confirmed by diffuse immunoreactivity for multiple smooth muscle markers. An adenomyoma or uterus-like mass occurring in the peritoneal wall was excluded due to the absence of endometrial tissue. Interestingly (worrisome), the tumor cells were positive for CD34, Bcl-2, and CD10. Spindle cell tumor immunoreactivity for these 3 markers leads to possible differential diagnoses that include extragastrointestinal stromal tumor, solitary fibrous tumor, and endometrial stromal tumor; all of which may occur in the abdominal wall. Similarly, abdominal wall recurrence of gastrointestinal stromal tumors is possible after laparoscopic surgery. We excluded extragastrointestinal stromal and parasitic gastrointestinal stromal tumors because our case was negative for CD117 and DOG1. A solitary fibrous tumor was also excluded because our case did not have a staghorn-like vascular structure and was negative for STAT6. Other potential differentials included an endometrial stromal nodule and a low-grade endometrial stromal sarcoma because the tumor cells of the present case were positive for CD10, ER, and PgR; however, these diagnoses were excluded due to the lack of endometrial spiral arteriole-like vessels, diffuse immunoreactivity for CD10, and endometriosis. Endometrial stromal nodules/sarcomas can show smooth muscle differentiation that is not usually diffuse.

In this case, the lack of mitosis and coagulation necrosis is more in keeping with a benign tumor. Furthermore, the proportion of Ki-67-positive cells was very low. The presence of a leiomyoma with myxoid changes raised the suspicion of a myxoid leiomyosarcoma, but this was ruled out due to the focal myxoid degeneration and absence of mitoses and atypia. However, peritoneal and retroperitoneal leiomyomas can have mitoses up to 5 per 50 high power fields and show frequent degeneration, including myxoid changes, making diagnostic exclusion of myxoid leiomyosarcomas difficult, according to the uterine smooth muscle tumor criteria. Using the PubMed database, 3 cases of abdominal wall leiomyosarcomas have been reported. Two cases showed frank sarcomatous features, including nuclear pleomorphism, frequent mitoses, and tumor necrosis, whereas 1 case described tumor cells with nuclear pleomorphism, but a low mitotic index (4 per 50 high-power fields) and an unknown necrosis status. Given the lack of specific data on abdominal wall leiomyosarcomas, suspected cases should be approached with caution until further cases are available for review. The most important issues regarding this and similar cases is the recognition of their Müllerian (uterine-like) nature and distinguishing them from nonuterine-type smooth muscle tumors. Hence, the same malignancy histological criteria are applied as for their uterine counterparts. This approach should help to prevent the overdiagnosis of leiomyosarcomas at this unexpected location.

The pathogenesis of abdominal wall leiomyomas is poorly understood, but 6 hypotheses have been suggested. The secondary/parasitic theory, which presumes that uterine leiomyomas detach from their subserosal location and attach to other peritoneal sites, is plausible. Uterine leiomyomas can grow in the abdominal wall incisional site after gynecological surgeries, including laparoscopic procedures, implying the seeding and growth of uterine leiomyoma cells in the incisional scar. The parasitic theory might explain the pathogenesis of our case; however, the present patient did not demonstrate a concurrent uterine leiomyoma nor have a history of uterine leiomyomas or abdominal surgeries. Second, these tumors may arise from vascular smooth muscle cells, but there is limited published immunohistochemical evidence to confirm whether vascular leiomyomas/angiroleiomyomas frequently express ER and PgR, which are seen in leiomyomas of the deep soft tissue.

Third, de novo leiomyomas arising from abdominal wall surgical scars have been reported, but an incisional scar was absent in our case. Fourth, the rectus sheath may be the origin of anterolateral wall leiomyomas, but the present tumor was removed easily from the rectus sheath, making this unlikely. Fifth, leiomyomas can primarily occur in the inguinal region, and the round ligament is postulated to be the origin of leiomyomas in these locations. In particular, round ligament smooth muscle cells that are immunohistochemically positive for ER and PgR can be the origin of leiomyomas. The present tumor was radiologically close to the left round ligament (Fig. 1A) and was identified close to the inguinal canal at the point where it is transected by the round ligament. However, a gross evaluation during surgery indicated that the tumor was not connected to the left round ligament. Further, the localization of the present tumor differed from that of previously published cases of round ligament leiomyoma. Finally, a thin layer of nonatypical smooth muscle cells adjacent to the leiomyoma could be the origin of the present tumor because the smooth muscle layer was intimately associated with the tumor and the layer was ER- and PgR-positive. Interestingly, Bcl-2 protein expression differed between the tumor and the adjacent smooth muscle layer. As in the present case, uterine leiomyoma cells are prominently immunoreactive for Bcl-2, but very few of the background uterine smooth muscle cells are positive for Bcl-2 protein. Thus, the tumorigenesis of our case might be similar to that of uterine leiomyomas. AR expression in the present tumor has not been reported in previously reported abdominal wall leiomyomas, but AR expression in uterine leiomyomas has been. We firstly described that the ER- and PgR-positive smooth muscle layer was present in the left lower quadrant of the abdominal wall, and the localization was postulated to be between the abdominal wall skeletal muscle and the peritoneal membrane. The smooth muscle layer has not been previously reported as a component of the abdominal wall or in cases of abdominal wall leiomyoma, suggesting that the structure might be acquired later in life, possibly in the sense of a Müllerian metaplasia or representing embryological remnants.

In conclusion, we presented a case of an anterolateral abdominal wall leiomyoma with cystic and myxoid degeneration. Although clinical modalities, including transvaginal US, CT, and MRI, failed to identify the abdominal wall origin of the tumor,
the broad attachment of the tumor to the abdominal wall was the key feature that raised suspicion of an abdominal wall tumor. Hydropic and mucinous degeneration of the tumor induced its key feature that raised suspicion of an abdominal wall tumor. We would like to thank Editage (www.editage.jp) for language editing.

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References

[1] Schindl M, Birner P, Losch A, et al. Preperitoneal lipoleiomyoma of the abdominal wall in a postmenopausal woman. Maturitas 2000;37:33–6.

[2] Lalor PF, Uribe A, Daum GS. De novo growth of a large preperitoneal lipoleiomyoma of the abdominal wall. Gynecol Oncol 2005;97:719–21.

[3] Schwarz EI, Ramach C, Mende KA, et al. Physiologic FDG uptake in the ovary together with an abdominal wall leiomyoma mimicking metastasizing ovarian cancer on PET/CT imaging. Clin Nucl Med 2009;34:249–50.

[4] Igberase GO, Mabiaku TO, Ebeigbe PN, et al. Solitary anterior abdominal wall leiomyoma in a 31-year-old multipara woman: a case report. Cases J 2009;2:113.

[5] Ono M, Inoue Y, Yokota M, et al. Abdominal wall leiomyoma in a reproductive age woman without antecedent pelvic surgery. Eur J Obstet Gynecol Reprod Biol 2010;151:225–6.

[6] Goyal N, Khurana N. Leiomyoma of rectus sheath: an uncommon entity: report of two cases. Indian J Pathol Microbiol 2010;53:391–2.

[7] Yesilkaya Y, Demirbas B, Kilincer A, et al. Primary anterior abdominal wall leiomyoma. Am Surg 2011;77:E208–9.

[8] Narayanaswamy M, Bhaskaran A, Kalyani R, et al. Anterior abdominal wall leiomyoma without antecedent pelvic surgery mimicking and ovarian tumour. J Clin Diomed Sci 2011;1:74–6.

[9] AF-Wadami HA. Anterior abdominal wall leiomyoma arising de novo in a perimenopausal woman. Oman Med J 2012;27:323–5.

[10] D’souza C, Bhat S, Purushothaman, et al. De novo growth of leiomyoma from rectus sheath: a rare presentation. Ann Trop Med Pub Health 2012;5:390–2.

[11] Miday M, Dewanda NK. Primary anterior abdominal wall leiomyoma—a diagnostic enigma. J Clin Diagn Res 2014;8:NJ01–2.

[12] Ernest Ong CW, Sow SL. Case report: Leiomyoma of the anterior abdominal wall. Med J Malaysia 2016;71:81–2.

[13] Cho JY, Woo JH, Hong HS, et al. Anterior abdominal wall leiomyoma arising de novo in a fertile women: a case report. J Korean Soc Radiol 2016;74:71–4.

[14] Mertinen M, Quade B, Fletcher C, Bridge J, Hogendoorn P, Mertens F. Leiomyoma of deep soft tissue. World Health Organization Classification of Tumours of Soft Tissue and Bone IARC Press, Lyon, France;2013:110–1.

[15] Moon HS, Koo JS, Park SH, et al. Parasitic leiomyoma in the abdominal wall after laparoscopic myomectomy. Fertil Steril 2008;90:1201e1201-1202.

[16] Einstein DM, Taglialue JR, Desai RK. Abdominal desmoids: CT findings in 25 patients. Am J Roentgenol 1991;157:275–9.

[17] Yorita K, Tanaka Y, Hirano K, et al. Subserosal, pedunculated, multilocular uterine leiomyoma with ovarian tumor-like morphology and histological architecture of adenomatoid tumors: a case report and review of the literature. J Med Case Rep 2016;10:352.

[18] Kamat NV, Telkar HB, Ramani SK, et al. Ruptured degenerated uterine fibroid diagnosed by imaging. Obstet Gynecol 2001;98(3 pt 2):961–3.

[19] Donnez O, Jadoul P, Squifflet J, et al. Iatrogenic peritoneal adenomyoma after laparoscopic subtotal hysterectomy and uterine morcellation. Fertil Steril 2006;86:1511–2.

[20] He J, Xu J, Zhou HY. Uterus-like mass: a very rare and elusive entity a case report. Medicine 2016;95:e4961.

[21] Allkhatib L, Altboush O, Bataineh N, et al. Extragastrointestinal stromal tumor (EGIST) in the abdominal wall: case report and literature review. Int J Surg Case Rep 2011;2:253–5.

[22] Huang HY, Sung MT, Eng HL, et al. Solitary fibrous tumor of the abdominal wall: a report of two cases immunohistochemical, flow cytometric, and ultrastructural studies and literature review. APMIS 2002;10:253–62.

[23] Masand RP, Euscher ED, Deavers MT, et al. Endometrioid stromal sarcoma: a clinicopathologic study of 63 cases. Am J Surg Pathol 2013;37:1635–47.

[24] Hachim H, Majbar AM, Alauoi M, et al. Abdominal wall recurrence of a gastrointestinal stromal tumor: case report. Springerplus 2013;4:429.

[25] Eken H, Karagul S, Topkul K, et al. Giant cutaneous leiomyosarcoma originating from the abdominal wall: a case report. Am J Case Rep 2016;17:35–8.

[26] Jena S, Bhattacharyya S, Roy S. Giant subcutaneous leiomyosarcoma of anterior abdominal wall. Case Rep Surg 2014;2014:308916.

[27] Gasparrini M, Virgilio E, Di Cesare T, et al. Primary leiomyosarcoma of the abdominal wall. Am Surg 2012;78:E249–250.

[28] Wada-Hizaike O, Yamamoto N, Osuga Y, et al. Aberrant implantation and growth of uterine leiomyoma in the abdominal wall after laparoscopically assisted myomectomy. Fertil Steril 2009;92:1747–1745.

[29] Orduzu Z, Dal Cin P, Chong WW, et al. Disseminated peritoneal leiomyomatosis after laparoscopic supracervical hysterectomy with characteristic molecular cytogenetic findings of uterine leiomyoma. Genes Chromosomes Cancer 2010;49:1152–60.

[30] Zhu G, Xiao D, Sun P. Expression of estrogen and progesterone receptors in angioleiomyoma of the nasal cavity of six patients. Onco Lett 2016;11:2359–64.

[31] Ishikawa S, Fujyama S, Kobayashi T, et al. Angioleiomyoma of the tongue: a case report and review of the literature. Odontology 2016;104:119–22.

[32] Terada T. Vascular leiomyoma of the lung arising from pulmonary artery. Int J Clin Exp Pathol 2013;6:97–9.

[33] Patil DT, Luskin WB, Fetsch JF, et al. Inguinal smooth muscle tumors in women—a dichotomous group consisting of Mullerian-type leiomyomas and soft tissue leiomyosarcomas: an analysis of 55 cases. Am J Surg Pathol 2011;35:315–24.

[34] Smith P, Heimer G, Norgren A, et al. The round ligament: a target organ for steroid hormones. Gynecol Endocrinol 1993;7:97–100.

[35] Bedir R, Yilmaz R, Sehitoglu I, et al. Round ligament leiomyoma developing during pregnancy: a case report and literature review. Iran J Pathol 2016;11:261–4.

[36] Matsuo H, Maruo T, Samoto T. Increased expression of Bcl-2 protein in human uterine leiomyoma and its up-regulation by progesterone. J Clin Endocrinol Metab 1997;82:293–9.

[37] Lottao MM, Soslow RA, Nonaka D, et al. Tissue microarray immunohistochemical expression of estrogen, progesterone, and androgen receptors in uterine leiomyomata and leiomyosarcoma. Cancer 2004;101:1455–62.