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Case Report

Lipoleiomyosarcoma with pleomorphic liposarcoma of the uterus: Computed tomography findings with pathological correlation. A case report and review of literature ☆,☆☆

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ABSTRACT

Uterine lipoleiomyosarcomas (L-LMS) are rare malignant tumors with only few cases described in literature. As well, liposarcomas (LPS) arising from uterine corpus are extremely rare since fat tissue is commonly poor or absent in uterus. We report a case of L-LMS of the uterine corpus with an associated component of pleomorphic LPS. As for other female pelvic malignancies, these neoplasms are more common in postmenopausal women and clinical findings are nonspecific. Most frequent signs and symptoms are metrorrhagia and abdominal pain often associated with pelvic distension/mass. We describe diagnostic and therapeutic steps of our patient from acceptation to conclusive diagnosis. In this case, contrast-enhanced computed tomography (CECT) depicted pivotal diagnostic findings and showed a clear distinction between different tumor components. Therefore, our goal with this essay is to stress the role of CECT imaging for diagnosis of these rare neoplasms. We also perform a review of current literature about liposarcomas of the uterine corpus.

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Introduction

Uterine sarcomas represent a very rare category of gynecologic tumors, representing 3%-7% of all uterine malignancies [1]. Leiomyosarcoma (LMS) is the most common variant, attributed to the rich muscular component of the uterus. Liposarcoma (LPS) arising from uterine corpus is exceptionally rare with few cases reported to date.

In this essay, we present a case of a rare mixed tumor of the uterine corpus, predominantly comprising lipoleiomyosarcoma (L-LMS) with a minor pleomorphic component. We outline diagnostic steps of our patient, detailing pathological features and highlighting role of contrast enhanced computed tomography (CECT) for an accurate diagnosis. Additionally, we provide a review of the current literature on LPSs of the uterine corpus.

Epidemiology and clinical presentation

Uterine sarcomas primarily affect postmenopausal patients, with an estimated incidence rate of about 0.36-0.64 cases per 100,000 women [2]. Key risk factors include prior pelvic radiation exposure and prolonged use of tamoxifen [3].

Uterine sarcomas present typically with nonspecific signs and symptoms, including abnormal uterine bleeding, which is the most common symptom and may present as menorrhagia, metrorrhagia, or postmenopausal bleeding. Groin pain, often referred as dull and persistent, is another common symptom. A pelvic mass may be palpable during physical examination. Additionally, symptoms such as urinary or bowel difficulties can arise due to tumor growth and compression of surrounding structures.

Imaging

Role of imaging techniques is critical in the accurate diagnosis and management of uterine sarcomas. Among these, contrast enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) are particularly valuable. These imaging modalities provide complementary information that increase diagnostic accuracy.

In CT imaging, tumors of liposarcoma series exhibit fat-density areas intermixed with soft tissue components, appearing variable hypodense. Dedifferentiated forms have tendency to show heterogenous contrast enhancement and variable presence of necrosis and calcifications.

In MRI, LPSs are generally hyperintense on T1-weighted images due to fat content with low-signal on fat-suppressed sequences [4]. Dedifferentiated forms show a heterogeneous signal on T2-weighted images with regions of soft tissue and possible necrosis. Pleomorphic and myxoid LPS exhibit complex signal patterns reflecting the mixed tissue components, with nonhomogeneous enhancement. In CT, leiomyosarcomas appear as large, heterogeneous masses with areas of necrosis and hemorrhage. They show variable contrast enhancement and may have areas calcifications and hemorrhage.

In MRI, LMSs exhibit heterogeneous signal intensity on both T1 and T2-weighted images. Areas of necrosis and hemorrhage appear hyperintense on T2-weighted images. Strong enhancement of viable tumor portions is observed and restriction in diffusion-weighted sequences (DWI) may be seen in high-grade tumors [5]. Mixed tumors like L-LMSs show intermediate features between LMS and LPS with a heterogeneous enhancement pattern and common areas of necrosis and hemorrhage.

Pathology

According to the latest WHO Classification, liposarcomas are categorized into 5 main types: well-differentiated, dedifferentiated, pleomorphic, myxoid, and myxoid pleomorphic [6]. Pathological examination is necessary for conclusive diagnosis uterine sarcomas. Biopsy and histopathological analysis are pivotal to confirm the type and grade of the tumor. Immunohistochemical staining and molecular genetic studies further assist in differentiating between various subtypes of sarcomas. Key pathological features for liposarcoma include the presence of lipoblasts with varying degrees of fat differentiation [7]. Leiomyosarcoma is typically characterized by spindle cells with significant mitotic activity and atypia [8]. Mixed tumors, such as L-LMS, are identified by the presence of distinct LPS and LMS components.

Treatment and prognosis

Prognosis of uterine sarcomas is widely variable and main treatment strategy is surgical resection, typically involving a hysterectomy and possibly bilateral salpingo-oophorectomy [9]. Adjuvant therapies such as radiation and chemotherapy may be used based on tumor type, grade, and stage. Emerging treatments include less aggressive therapies such as targeted and hormonal therapies. The 5-year survival rate is 50%-55% for patients with early uterine sarcoma and 8%-12% for advanced cases [10]. Well-differentiated LPSs have a better prognosis, while dedifferentiated, pleomorphic, and myxoid subtypes are more aggressive. LMSs generally have a high risk of recurrence and metastasis, depending on the stage. Mixed tumors are treated as high-grade sarcomas with a similar prognosis to LMSs.

Case report

In February 2022, we admitted to our hospital a 62-year-old woman in menopause with recent history of metrorrhagia and low abdominal pain. She stated of being suffering those symptoms for several months without similar issues in the past. She did not refer either significant previous surgeries or history of tumors in her family. In addition, there was no history of hormonal drugs assumption. After an accurate gynecological examination, our patient underwent a triple-phase protocol contrast-enhanced CT (CECT) scan. Imaging study revealed a large neoplasm (9.6 × 5 × 5 cm) arising from uterine corpus and protruding into the cervix with nonuniform density owing to solid areas mixed with fat-density tissue. Cranial portions of the tumor showed prevalent adipose component with solely contrast enhancing septa (Fig. 1A), while most dependent

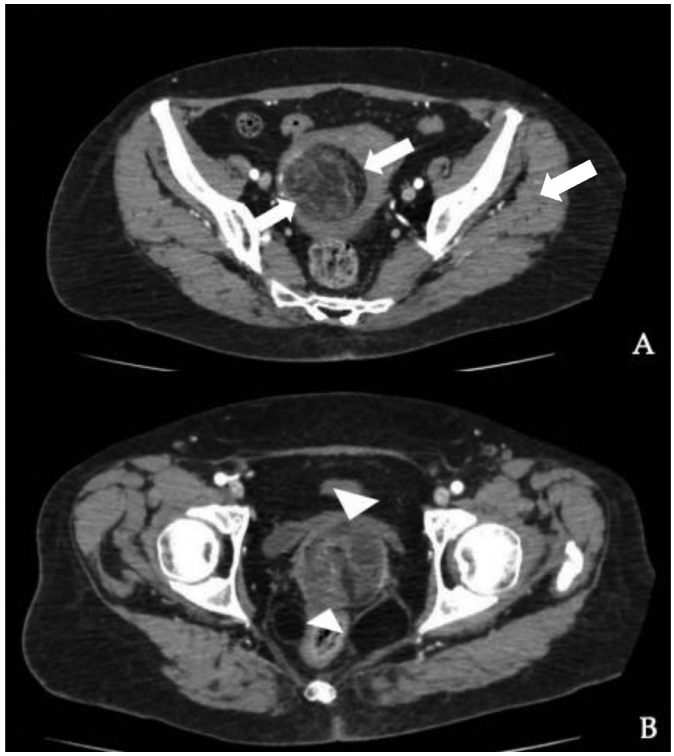

Fig. 1 – Axial Contrast enhanced CT scan: (A) neoplasm of the uterine corpus (cranial portion) with prevalent adipose component with multiple enhancing septa contextually due to pleomorphic LPS (arrows). (B) tumor tissue in the cervix (caudal portion) with contrast enhancing solid fraction and minor fat component due to L-LMS (arrowheads).

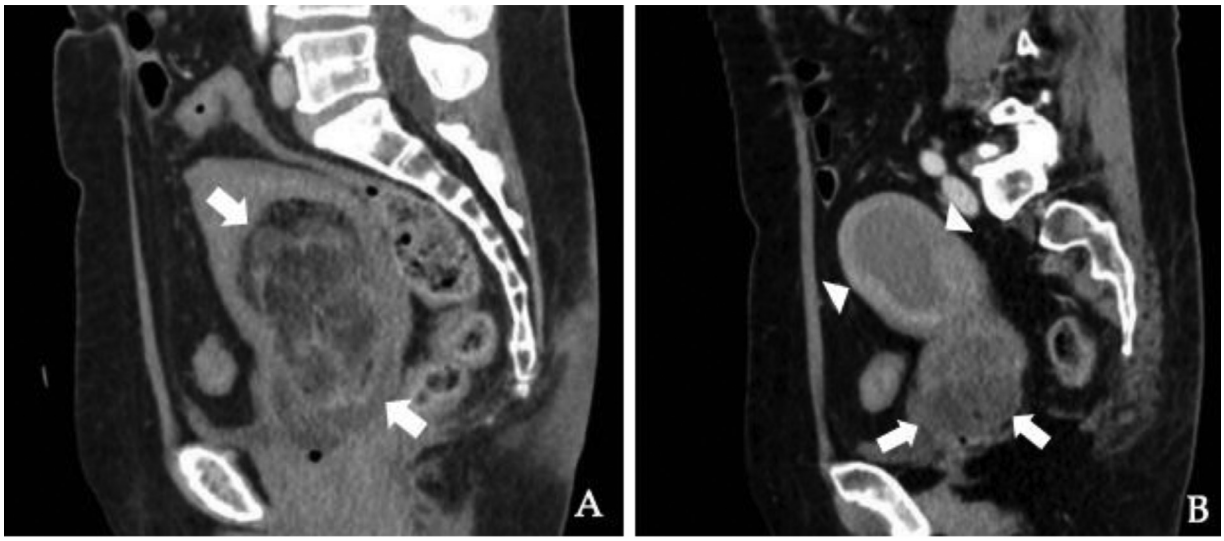

Fig. 2 – Sagittal contrast-enhanced CT Multiplanar (MPR) reconstruction: (A) well depicted extension of pleomorphic LPS of the uterine corpus (black arrows). (B) Lipoleiomyosarcoma arising from the cervix (white arrows) with distension of above endometrial cavity (white arrowheads).

parts were mostly solid with avid contrast enhancement and poor adipose tissue (Fig. 1B). Multi-planar reconstruction (MPR) (Figs. 2 and 3) highlighted even more the presence of these 2 different portions. Furthermore, a blood-like collection (32 HU) was detected in the uterine fundus. Above-mentioned CT findings were highly suggestive for a tumor of the liposarcoma series. No significant lymphadenopathies or metastases were found. Therefore, our patient underwent hysterectomy with bilateral adnexectomy with further examination of surgical samplings. Macroscopical analysis revealed an enlarged uterus (8.5 × 7 × 5.5) cm due to an intramural nodular mass with dimensions according to CT findings (maximum diame-

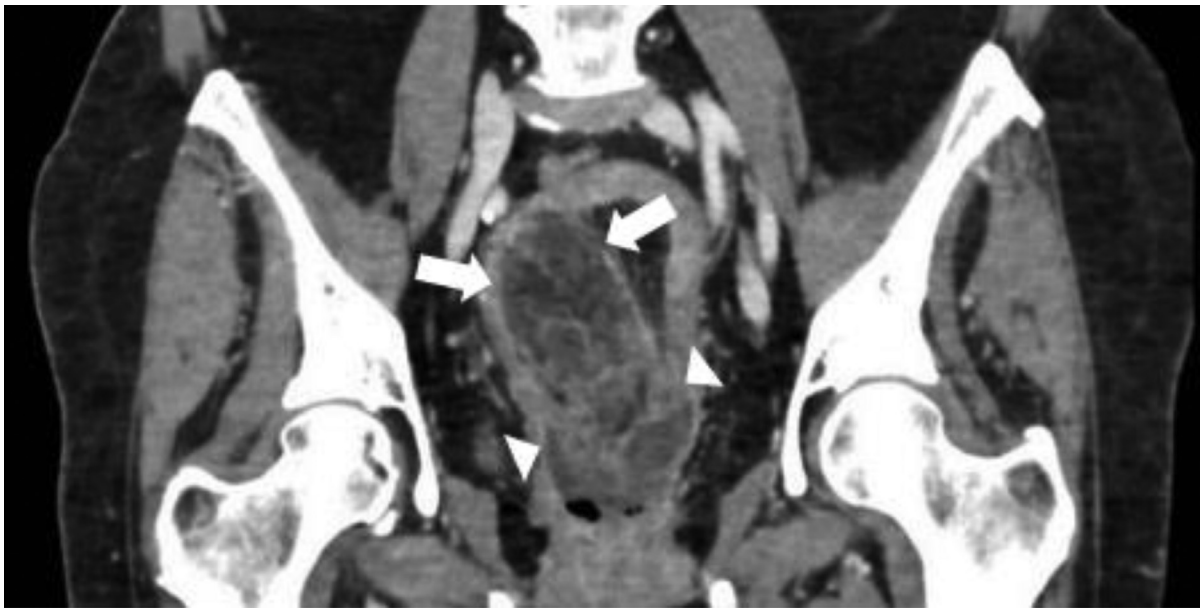

Fig. 3 – Coronal contrast enhanced CT MPR reconstruction: well depicted both endometrial (white arrows) and cervical (white arrowhead) component of the uterine mass.

ter 95 mm). Neoplasm revealed yellowish areas and extended necrosis. As seen with CECT scan, also microscopic analysis showed 2 different tumor components. Samplings from caudal portions were made of bundles of spindle cells with moderate/severe atypia and mature adipocytes, with occasional presence of lipoblasts (Figs. 4A and C). Instead, samplings from cranial parts corpus mainly showed lobules of markedly atypical lipoblasts and polygonal epithelioid cells (Fig. 4B). Therefore, conclusive diagnosis was of uterine L-LMS with an associated component of LPS.

Discussion

Among uterine tumors, sarcomas account only for 1% overall [11]. LMS is the most common uterine sarcoma, characterized by its aggressiveness and poor prognosis [12]. LPS represents up to 0.2% of uterine tumors, with only 9 cases of LPS arising from the uterine corpus described so far, due to its typically poor or absent adipose component [13]. L-LMS is a mixed sarcomatous tumor with intermediate features between LMS and LPS. However, while LPS and LMS have been well documented, only 8 cases of uterine L-LMS have reported in literature [14]. L-LMS mainly comprises 2 malignant cellular populations derived from smooth muscle and adipose tissue [15]. Adipose malignant cells, known as lipoblasts, may have a greater potential for evolution and could undergo further dedifferentiation. Therefore, the pleomorphic LPS of our case could likely originate from transformative lipoblasts of LMS component. The coexistence of pleomorphic LPS with another neoplasm is a rare occurrence, with only 3 documented cases to date. The mean age of patients in our literature review is 60.3 years, with a standard deviation (SD) of 7.9 years. The most commonly reported clinical findings were metrorrhagia and abdominal pain. Thus, age and symptoms of our patient are consistent with those reported in other cases in our review. To the best of our knowledge, imaging findings of these tumors have been poorly documented. CT findings may be similar for many uterine tumors, especially for pure sarcomas and carcinosarcomas, previously known as malignant mixed müllerian tumors (MMMT) [16]. CECT scan displays heterogeneous attenuation values, as haemorrhage and necrosis commonly develop among solid areas [17]. Solid components exhibit different density values depending on histology and degree of differentiation. LMS shows a soft tissue density (30-45 HU) in its solid parts with peripheric contrast enhancement; calcifications are uncommon. LPS typically presents fat attenuation values (-100 HU). The pleomorphic component demonstrates solid areas with weaker contrast enhancement, and calcifications are more common [4]. In our case, the most striking finding was the massive filling of the uterine cavity due to tumoral growth, with the presence of hemorrhage and necrosis highlighting its aggressiveness. The cranial portion arising from the uterine corpus, ultimately confirmed as pleomorphic LPS, showed nonuniform attenuation with multiple enhancing septa (Fig. 2A). Conversely, cervical fraction, confirmed as L-LMS, appeared more homogeneous (Fig. 2B). No evident calcifications were found. Based on our analysis, while pleomorphic LPS exhibited more distinctive features, uterine L-LMS showed nonspecific intermediate characteristics between LPS and LMS. Including our patient and considering cases from our review, mean maximum diameter is 12.7 cm with a SD of 4.5 cm. Therefore, tumors examined in this essay showed large dimensions at diagnosis, demonstrating strong aggressiveness. Hong et al. reported a case of a myxoid LPS arising in a uterine leyomioma, with CECT images showing a large, inhomogeneous neoplasm with massive filling of uterine cavity, as found in our patient [18]. Despite these features of aggressiveness, there are no imaging criteria to assess malig-

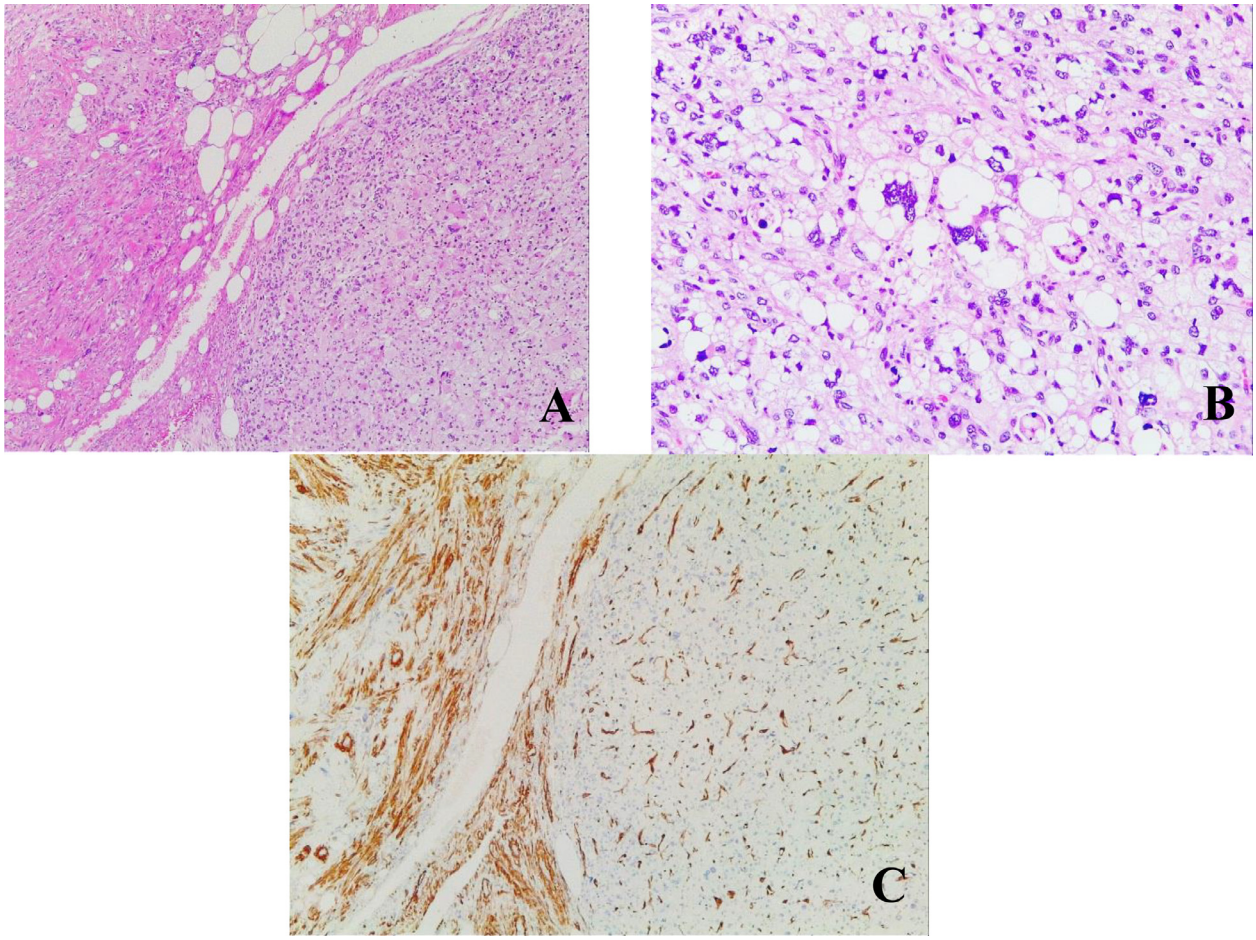

Fig. 4 – Histologic section of the mass showed 2 main components: (A, magnification 5x) spindle cells with moderate to severe atypia mixed with mature adipocytes (S100+); occasional lipoblasts with hypercromic nuclei and vacuolated cytoplasm were also found. (B, magnification 20x) markedly atypical lipoblasts and pleomorphic cells resembling an undifferentiated pleomorphic sarcoma. (C, magnification 5x) immunohistochemical stain showed only the spindle cell component to be strongly positive for smooth muscle actin (SMA).

nancy in a reliable manner [19]. In particular, the differential diagnosis of uterine sarcomas can be challenging, as benign uterine tumors can present features that mimic malignancy (e.g., hemorrhage, necrosis) [19,20]. Thus, pathological examination is necessary for a conclusive diagnosis. According to the experience of Hong et al. and Schoolmeester et al., microscopic examination is essential to demonstrate absence of epithelial component to rule out a diagnosis of MMMT [18,21]. Nonetheless, CECT findings from our patient were crucial for suspecting malignancy and for correct therapeutic management. Main results from our literary review are summarized in the Table 1 below.

Table 1 – Cases of liposarcoma of uterine corpus: Review of literature.

Year	Age	Clinics	Size (cm)	Histology	Outcome
1989 [22]	55	Metrorrhagia	12 × 7 × 6	Myxoid + Pleomorphic + LMS	12 m Recurrence
2006 [23]	71	Metrorrhagia	12.9 × 10.8	Pleomorphic	8 y No Recurrence
2008 [18]	48	Dysmenorrhea, abdominal mass	21 × 18	Myxoid	2 m No Recurrence
2011 [24]	62	Abdominal pain	7 × 6.3 × 4.5	Pleomorphic + LMS	2 m Recurrence
2011 [25]	49	Abdominal mass	10.5	Pleomorphic	1 y No Recurrence
	58	Metrorrhagia	18	Myxoid + Pleomorphic	2 y No Recurrence
	70	Abdominal mass	10	Myxoid	20 y No Recurrence
2016 [21]	70	Dysuria	9 × 8 × 7.5	Pleomorphic	3 m Death
2018 [13]	58	Abdominal distension, metrorrhagia	16 × 17	Well differentiated	1 y No Recurrence
2022 (Present Case)	62	Metrorrhagia, abdominal pain	9.6 × 5 × 5	Pleomorphic + LMS	

Conclusions

In our experience, CECT scan has proven to be a reliable imaging technique for assessing tumor extension and suggesting possible malignancy. Specifically, the imaging findings indicated high tumor aggressiveness and supported the hypothesis of a mixed uterine sarcomatous tumor, providing valuable guidance for histological examination.

Institutional review board statement

Not applicable.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

CRediT authorship contribution statement

Valerio Arpaia: Data curation, Writing – original draft, Writing – review & editing, Supervision. **Luigi Pirolo:** Investigation, Resources. **Stefania Sfregola:** Resources. **Luana Licata:** Resources. **Maria Iovino:** Conceptualization, Validation. **Luca Sanduzzi:** Data curation, Writing – original draft, Writing – review & editing, Supervision. **Anna Ferrante:** Investigation. **Elisa Varriale:** Investigation. **Pietro Iacobelli:** Investigation, Resources. **Fabio Sandomenico:** Conceptualization, Methodology, Validation, Investigation, Resources, Data curation, Writing – original draft, Writing – review & editing, Supervision.

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