

CASE REPORT

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Cystic endometrial stromal sarcoma mimicking a cystic ovarian tumor : an unusual presentation

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Abstract

Low grade endometrial stromal sarcoma (LGESS) typically presents as a solid intracavitary or intramural uterine mass. On imaging, LGESS is usually seen as an endometrial or myometrial solid mass. Cystic change is unusual and may lead to a differential diagnosis of an ovarian mass as was seen in the present case. Here we present a case of a 38 year old woman who presented with a cystic ovarian mass clinically and radiologically. Per-operatively, it was found to be a multiloculated cystic mass, filled with serosanguinous fluid, in the uterine fundus. Histopathological examination showed a tumor mass composed of mainly spindle cells arranged in fascicular pattern showing marked myxoid degeneration and cystic areas. On immunohistochemistry(IHC), tumor cells showed positivity for CD 10, ER, PR, SMA and desmin while HMB45 was negative. Based on histopathological and IHC, a diagnosis of LGESS was made. Most cases of cystic uterine masses have a benign course but, LGESS exhibits a relatively poorer outcome and a risk of metastasis. Hence, we present this case for its unusual presentation which mimics an ovarian mass but has worse prognosis.

Keywords Endometrial stromal sarcoma, Cystic, Ovarian mass

Background

Low grade endometrial stromal sarcoma (LGESS) constitutes less than 1% of all uterine malignancies (Lee et al. 2019). This rare form of cancer typically manifests as abnormal uterine bleeding or abdominal discomfort. Upon gross examination, LGESS may appear as an intracavitary polypoid or an intramural solid mass. These masses often exhibit indistinct borders and infiltrate the myometrium (Lee et al. 2019).

A seldom reported morphological characteristic of primary LGESS is cystic change (Dionigi et al. 2002;

Perez-Montiel et al. 2004). In the case we encountered, the patient presented with a cystic adnexal mass clinically and radiologically, thus, a provisional diagnosis of ovarian tumor was made. However, histopathological examination showed it to be a LGESS of uterus with cystic change – an unusual presentation and an important entity as it closely mimics adnexal cystic mass.

Case presentation

A 38-year-old female presented with complaints of lower abdominal pain of 12 weeks duration. She had six live births and one abortion. Physical examination suggested a firm immobile pelvic mass. On laboratory workup, Carcinoembryonic Antigen was 2.55 ng/mL (normal < 5 ng/mL), Carbohydrate Antigen-19.9 was 4.11 U/mL (normal < 37.0 U/mL), Alpha Fetoprotein was 3.73 ng/mL

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(normal 0.89–8.78 ng/mL), Cancer Antigen-125 was 28.5 U/mL (normal <35.0 U/mL).

On ultrasonography and computed tomography (CT) scan (Fig. 1a), uterus and right sided adnexa could not be visualized separately. A multiseptated well-defined cystic mass measuring 142.0 mm X 136.5 mm with anechoic/hypo-attenuated content was seen in the right adnexa, with normal uterus and cervix. A clinico-radiological opinion of right ovarian/pelvic cyst was given.

A total abdominal hysterectomy with bilateral salpingo-oophorectomy was done. Differential diagnoses of fibroid with degenerative changes and adenomyosis with cystic degeneration were considered. Grossly, uterus with cervix with bilateral fallopian tubes and ovaries was received measuring 14.5 cm X 10.5 cm X 6.5 cm. The uterine serosal surface was smooth with a large mass in the fundal region measuring 8.2 cm X 8.0 cm X 7.0 cm consisting of multiple fluid-filled cystic spaces. The cut surface showed an intramural poorly demarcated multiloculated cystic mass with few solid areas. On cutting the cysts, serosanguinous fluid came out. The cyst wall thickness varied

from papery thin to 0.4 cm (Fig. 1b). Areas of hemorrhage were seen in the cystic mass. Rest of the myometrium was unremarkable. Endometrial cavity and cervical canal were normal (Fig. 1b). Grossly, both sided adnexa were unremarkable.

Microscopic examination showed a tumor composed of densely cellular areas admixed with paucicellular areas showing marked myxoid degeneration. Cystic areas were also seen. The cellular areas showed mainly spindle cells arranged in fascicular pattern (Fig. 1c). At places, tongue-like projections of these spindle cells were seen invaginating into the surrounding myometrium. These projections extended >3 mm into the myometrium (Fig. 1d). These tumor cells showed moderate atypia with hyperchromatic to vesicular nuclei, inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm. Mitotic count was 1–2 per mm² (Fig. 1e). Numerous capillaries with peri-arteriolar whorling of spindle cells were also seen (Fig. 1f). Necrosis was absent. Differential diagnoses of low-grade-endometrial-stromal-sarcoma, multicystic leiomyosarcoma, leiomyoma with cystic

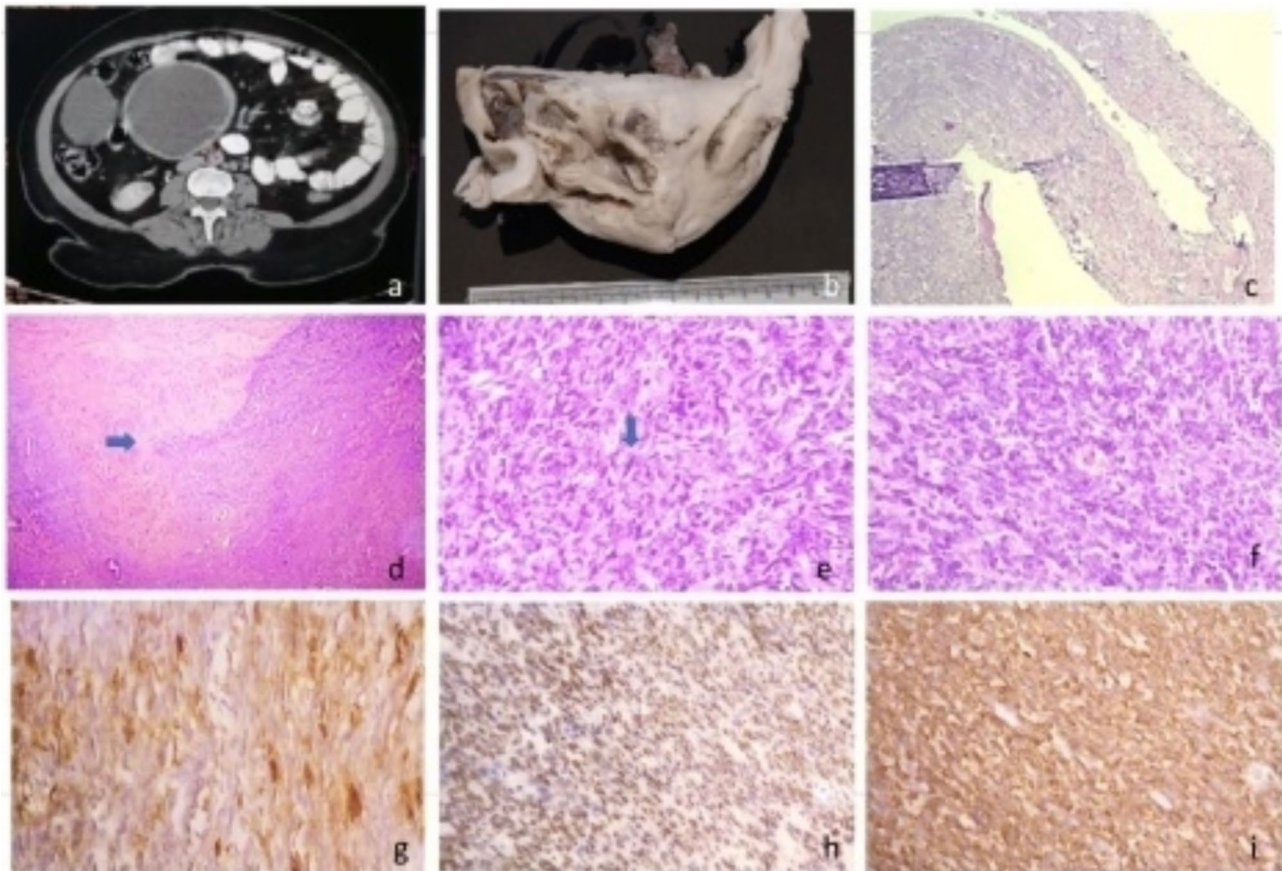


Fig. 1 **a**. CT findings suggested an adnexal mass with multiseptate cystic mass with hypoattenuated **b**. Gross Specimen – cut section of uterus shows a multiloculated solid-cystic mass in the fundus. Histological and IHC findings of multicystic LGESS **c**. The cyst wall composed of monotonous spindle cells (10X; H&E) **d**. infiltrative mass composed of spindle cells invading into the myometrium in tongue like fashion (arrow) (4X; H&E) **e**. spindle cells showing moderate atypia; eosinophilic cytoplasm with low mitotic activity (arrow) (40X; H&E) **f**. tumor cells showing peri-arteriolar whorling (40X; H&E). Tumor cells showing positivity for (40X) **g**. CD10 **h**. ER **i**. SMA

degeneration and perivascular-epithelioid-cell-tumor (PEComa) were kept.

Immunohistochemical (IHC) panel to confirm the final diagnosis was done (Table 1) (Folkins et al. 2019, Oliva et al. 2019).

Tumor cells showed positivity for Cluster of Differentiation 10 (CD 10) (Fig. 1g), ER (Fig. 1h), PR, SMA (Fig. 1i) and Desmin while Human Melanoma Black 45 (HMB45) was negative.

Based on histopathological and IHC findings, a final diagnosis of low-grade-endometrial-stromal-sarcoma was made.

On follow-up patient is alive and well with no residual disease.

Discussion and conclusion

LGESS is the second most common malignant mesenchymal tumor of uterus. It occurs over a wide age range with a mean of 52 years, but patients tend to be younger than those with other uterine sarcomas (Lee et al. 2019). Our patient was also of a younger age group. The findings on imaging match with those of epithelial or mesenchymal type of uterine tumors. On imaging, LGESS usually presents as a solid lesion but occasionally shows cystic degeneration (Rha et al. 2003). Radiologically, the present case was interpreted as a cystic ovarian mass which posed a diagnostic dilemma.

Grossly, LGESS usually presents with a yellow to tan fleshy cut surface, occasionally showing hemorrhage and/or necrosis. Size usually ranges from 5 to 10 cm. These may be intracavitary, polypoid or intramural masses, often with ill-defined but sometimes with well-defined margins and overt myometrial infiltration and/or intravascular plugs of tumor inside intramyometrial and parametrial veins (Lee et al. 2019). Our case presented as a predominantly cystic mass with few solid areas. No areas of necrosis were present.

The uterine lesions that may present as cystic masses are endometrial hyperplasia, endometrial polyps, leiomyoma with cystic change, endometrial-stromal-nodule, adenomyoma, adenofibroma, adenosarcoma, carcinosarcoma, adenomatoid tumor, adenomyomatous polyp,

leiomyosarcoma, low-grade-endometrial-stromal-sarcoma, high-grade-endometrial-stromal-sarcoma and PEComa. Among these, most entities have a benign course. However, adenocarcinoma, leiomyosarcoma, adenosarcoma, carcinosarcoma and endometrial-stromal-sarcoma present with a poorer outcome with high incidence of metastasis. Hence, accurate histopathological diagnosis is of utmost importance in lesions of uterus presenting as a cystic mass.

The microscopy in our case showed tongue-like infiltrative growth of spindle cells (>3 mm), along with no areas of necrosis and no foci of adenomyosis. IHC showed positivity for CD10. It was also ER and PR positive. Thus, various hormonal therapies for LGESS have been proposed over time but no consensus has been reached till date and needs further studies (Wang et al. 2018; Borella et al. 2022). Since, endometrial-stromal-sarcoma does not have a very specific IHC panel, a judicious combination of histopathological findings and IHC markers are helpful in making the diagnosis.

First-line treatment of LGESS is total hysterectomy with bilateral salpingo-oophorectomy even in case of recurrence while lymphadenectomy, endocrine therapy and regional radiotherapy are debatable and needs a risk to benefit ratio analysis along with further studies (Thiel et al. 2018).

As already mentioned, cystic change in LGESS is an unusual morphologic finding (Dionigi et al. 2002). Extensive literature search showed only a few cases of this sarcoma presenting as cystic mass, of which most were metastatic lesions at extra-uterine sites (Kim et al. 2009, 2022; Efared et al. 2019; McCarthy et al. 2019; Moral et al. 2022; Yu et al. 2022). Only one case of primary uterine low-grade-endometrial-stromal-sarcoma with cystic change has been reported till date by Perez-Montiel et al. 2004. All other cases of this sarcoma presenting as a cystic mass are metastatic in nature, ovary being the most common site (Kim et al. 2009; Efared et al. 2019; Moral et al. 2022; Yu et al. 2022), other sites being mesentery (Yu et al. 2022), lung (Kim et al. 2022), tail of pancreas (McCarthy et al. 2019) and right atrium (Wood et al. 2011). Our

Table 1 Immunohistochemical (IHC) panel to confirm the diagnosis

IHC	Leiomyoma with cystic degeneration	Leiomyosarcoma	PEComa	LGESS	HGESS	Present case
CD10	-	+/-	-	+	+	+
ER	+	+	+/-	+	+	+
PR	+	+	+/-	+	+	+
Desmin	+	weak	+/-	+	+	+
SMA	+	weak	+/-	+	+	+
HMB45	-	-	+	-	-	-
Ki67	low	high	low	low	High	low

LGESS=low grade endometrial stromal sarcoma; HGESS=high grade endometrial stromal sarcoma; ER=estrogen receptor; PR=progesterone receptor; SMA=smooth muscle actin; += positive; -= negative; +/- = may be positive or negative

case is a case of primary uterine LGEES presenting as a cystic adnexal mass posing diagnostic dilemma.

Clinicoradiologically, cystic pelvic masses are almost always ovarian in origin but the possibility of a uterine mass presenting as a pelvic cyst should also be kept in mind. A detailed histopathological and immunohistochemical examination is essential for the correct diagnosis of such unusual presentations of low grade endometrial stromal sarcoma. Most cases of cystic uterine masses have a benign course but, LGEES exhibits a relatively poorer outcome and a risk of metastasis. Therefore, a possibility of LGEES should also be kept in the differential diagnoses of cystic uterine tumors to enable proper diagnosis and treatment.

Abbreviations

LGEES	Low grade endometrial stromal sarcoma
HGEES	High grade endometrial stromal sarcoma
ER	Estrogen receptor
PR	Progesterone receptor
SMA	Smooth muscle actin
IHC	Immunohistochemistry

Acknowledgements

None.

Author contributions

Author SK was responsible for the, acquisition of patient data, and drafting the manuscript. Author AS contributed to the writing of the Introduction and Case presentation sections. Author DS contributed to the conception of the case report, writing of the Discussion and critically revised the manuscript for important intellectual content. Author KV and author VM provided clinical expertise, contributed to the interpretation of the IHC results, and critically revised the manuscript for important intellectual content. All authors approved the final version of the manuscript and agreed to be accountable for all aspects of the work, ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Funding

The authors did not receive support from any organization for the submitted work.

Data availability

The clinical case data used to support the findings of this study are included within the article.

Declarations

Consent for publication

Informed consent was obtained from the patient. The participant has consented to the submission of the case report to the journal.

Conflict of interest

There was no conflict of interest.

Received: 22 April 2024 / Accepted: 6 August 2024

Published online: 23 August 2024

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