

World Journal of *Clinical Cases*

World J Clin Cases 2022 August 16; 10(23): 8057-8431



OPINION REVIEW

- 8057** Invasive intervention timing for infected necrotizing pancreatitis: Late invasive intervention is not late for collection
Xiao NJ, Cui TT, Liu F, Li W
- 8063** Clinical utility of left atrial strain in predicting atrial fibrillation recurrence after catheter ablation: An up-to-date review
Yu ZX, Yang W, Yin WS, Peng KX, Pan YL, Chen WW, Du BB, He YQ, Yang P

MINIREVIEWS

- 8076** Gut microbiota and COVID-19: An intriguing pediatric perspective
Valentino MS, Esposito C, Colosimo S, Caprio AM, Puzone S, Guarino S, Marzuillo P, Miraglia del Giudice E, Di Sessa A
- 8088** Beta receptor blocker therapy for the elderly in the COVID-19 era
Santillo E, Migale M

ORIGINAL ARTICLE**Retrospective Cohort Study**

- 8097** Nonselective beta-blocker use is associated with increased hepatic encephalopathy-related readmissions in cirrhosis
Fallahzadeh MA, Asrani SK, Tapper EB, Saracino G, Rahimi RS

Retrospective Study

- 8107** Different squatting positions after total knee arthroplasty: A retrospective study
Li TJ, Sun JY, Du YQ, Shen JM, Zhang BH, Zhou YG
- 8115** Outcomes of seromuscular bladder augmentation compared with standard bladder augmentation in the treatment of children with neurogenic bladder
Sun XG, Li YX, Ji LF, Xu JL, Chen WX, Wang RY
- 8124** Distinctive clinical features of spontaneous pneumoperitoneum in neonates: A retrospective analysis
Kim SH, Cho YH, Kim HY
- 8133** Cognitive training for elderly patients with early Alzheimer's disease in the Qinghai-Tibet Plateau: A pilot study
Wang XH, Luo MQ
- 8141** Diagnostic value of elevated serum carbohydrate antigen 125 level in sarcoidosis
Zhang Q, Jing XY, Yang XY, Xu ZJ

- 8152** Evaluation of progressive early rehabilitation training mode in intensive care unit patients with mechanical ventilation

Qie XJ, Liu ZH, Guo LM

- 8161** Comparison of demographic features and laboratory parameters between COVID-19 deceased patients and surviving severe and critically ill cases

Wang L, Gao Y, Zhang ZJ, Pan CK, Wang Y, Zhu YC, Qi YP, Xie FJ, Du X, Li NN, Chen PF, Yue CS, Wu JH, Wang XT, Tang YJ, Lai QQ, Kang K

Clinical Trials Study

- 8170** Role of H₂receptor blocker famotidine over the clinical recovery of COVID-19 patients: A randomized controlled trial

Mohiuddin Chowdhury ATM, Kamal A, Abbas MKU, Karim MR, Ali MA, Talukder S, Hamidullah Mehedi H, Hassan H, Shahin AH, Li Y, He S

Observational Study

- 8186** Short-term prognostic factors for hepatitis B virus-related acute-on-chronic liver failure

Ye QX, Huang JF, Xu ZJ, Yan YY, Yan Y, Liu LG

- 8196** Three-dimensional psychological guidance combined with evidence-based health intervention in patients with liver abscess treated with ultrasound

Shan YN, Yu Y, Zhao YH, Tang LL, Chen XM

- 8205** Role of serum β 2-microglobulin, glycosylated hemoglobin, and vascular endothelial growth factor levels in diabetic nephropathy

Yang B, Zhao XH, Ma GB

SYSTEMATIC REVIEWS

- 8212** Gallbladder neuroendocrine carcinoma diagnosis, treatment and prognosis based on the SEER database: A literature review

Cai XC, Wu SD

CASE REPORT

- 8224** Sepsis complicated with secondary hemophagocytic syndrome induced by giant gouty tophi rupture: A case report

Lai B, Pang ZH

- 8232** Spontaneous remission of autoimmune pancreatitis: Four case reports

Zhang BB, Huo JW, Yang ZH, Wang ZC, Jin EH

- 8242** Epstein-Barr-virus-associated hepatitis with aplastic anemia: A case report

Zhang WJ, Wu LQ, Wang J, Lin SY, Wang B

- 8249** Aspiration as the first-choice procedure for airway management in an infant with large epiglottic cysts: A case report

Zheng JQ, Du L, Zhang WY

- 8255** Sequential multidisciplinary minimally invasive therapeutic strategy for heart failure caused by four diseases: A case report
Zhao CZ, Yan Y, Cui Y, Zhu N, Ding XY
- 8262** Primary ascending colon cancer accompanying skip metastases in left shoulder skin and left neck lymph node: A case report
Zhou JC, Wang JJ, Liu T, Tong Q, Fang YJ, Wu ZQ, Hong Q
- 8271** Clinical and genetic study of ataxia with vitamin E deficiency: A case report
Zhang LW, Liu B, Peng DT
- 8277** Complete resection of large-cell neuroendocrine and hepatocellular carcinoma of the liver: A case report
Noh BG, Seo HI, Park YM, Kim S, Hong SB, Lee SJ
- 8284** Immunotherapy combined with antiangiogenic agents in patients with advanced malignant pleural mesothelioma: A case report
Xuan TT, Li GY, Meng SB, Wang ZM, Qu LL
- 8291** Bladder malacoplakia: A case report
Wang HK, Hang G, Wang YY, Wen Q, Chen B
- 8298** Delayed inflammatory response evoked in nasal alloplastic implants after COVID-19 vaccination: A case report
Seo MG, Choi EK, Chung KJ
- 8304** Phosphoglyceride crystal deposition disease requiring differential diagnosis from malignant tumors and confirmed by Raman spectroscopy: A case report
Ohkura Y, Uruga H, Shiiba M, Ito S, Shimoyama H, Ishihara M, Ueno M, Udagawa H
- 8312** Vulvovaginal myeloid sarcoma with massive pelvic floor infiltration: A case report and review of literature
Wang JX, Zhang H, Ning G, Bao L
- 8323** Femoral neck stress fracture and medial tibial stress syndrome following high intensity interval training: A case report and review of literature
Tan DS, Cheung FM, Ng D, Cheung TLA
- 8330** Periosteal chondroma of the rib: A case report
Gao Y, Wang JG, Liu H, Gao CP
- 8336** Papillary thyroid carcinoma occurring with undifferentiated pleomorphic sarcoma: A case report
Lee YL, Cheng YQ, Zhu CF, Huo HZ
- 8344** Laparoscopic treatment of bilateral duplex kidney and ectopic ureter: A case report
Wang SB, Wan L, Wang Y, Yi ZJ, Xiao C, Cao JZ, Liu XY, Tang RP, Luo Y
- 8352** Incontinentia pigmenti with intracranial arachnoid cyst: A case report
Li WC, Li ML, Ding JW, Wang L, Wang SR, Wang YY, Xiao LF, Sun T

- 8360** Relapsing polychondritis causing breathlessness: Two case reports
Zhai SY, Zhang YH, Guo RY, Hao JW, Wen SX
- 8367** Endodontic management of a fused left maxillary second molar and two paramolars using cone beam computed tomography: A case report
Mei XH, Liu J, Wang W, Zhang QX, Hong T, Bai SZ, Cheng XG, Tian Y, Jiang WK
- 8375** Infant biliary cirrhosis secondary to a biliary inflammatory myofibroblastic tumor: A case report and review of literature
Huang Y, Shu SN, Zhou H, Liu LL, Fang F
- 8384** Metastatic low-grade endometrial stromal sarcoma with variable morphologies in the ovaries and mesentery: A case report
Yu HY, Jin YL
- 8392** Bronchogenic cysts with infection in the chest wall skin of a 64-year-old asymptomatic patient: A case report
Ma B, Fu KW, Xie XD, Cheng Y, Wang SQ
- 8400** Incidental accumulation of Technetium-99m pertechnetate in subacute cerebral infarction: A case report
Han YH, Jeong HJ, Kang HG, Lim ST
- 8406** Metal stent combined with ileus drainage tube for the treatment of delayed rectal perforation: A case report
Cheng SL, Xie L, Wu HW, Zhang XF, Lou LL, Shen HZ
- 8417** Using ketamine in a patient with a near-occlusion tracheal tumor undergoing tracheal resection and reconstruction: A case report
Xu XH, Gao H, Chen XM, Ma HB, Huang YG

LETTER TO THE EDITOR

- 8422** Reflections on the prevalence of human leukocyte antigen-B27 and human leukocyte antigen-B51 co-occurrence in patients with spondylarthritis
Gonçalves Júnior J, Sampaio-Barros PD, Shinjo SK
- 8425** Comment on "Disease exacerbation is common in inflammatory bowel disease patients treated with immune checkpoint inhibitors for malignancy"
Argyriou K, Kotsakis A
- 8428** Intranasal sufentanil combined with intranasal dexmedetomidine: A promising method for non-anesthesiologist sedation during endoscopic ultrasonography
Wang Y, Ge ZJ, Han C

ABOUT COVER

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RESPONSIBLE EDITORS FOR THIS ISSUE

Production Editor: *Hua-Ge Yu*; Production Department Director: *Xiang Li*; Editorial Office Director: *Jin-Lei Wang*.

NAME OF JOURNAL

World Journal of Clinical Cases

ISSN

ISSN 2307-8960 (online)

LAUNCH DATE

April 16, 2013

FREQUENCY

Thrice Monthly

EDITORS-IN-CHIEF

Bao-Gan Peng, Jerzy Tadeusz Chudek, George Kontogeorgos, Maurizio Serati, Ja Hyeon Ku

EDITORIAL BOARD MEMBERS

<https://www.wjgnet.com/2307-8960/editorialboard.htm>

PUBLICATION DATE

August 16, 2022

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INSTRUCTIONS TO AUTHORS

<https://www.wjgnet.com/bpg/gerinfo/204>

GUIDELINES FOR ETHICS DOCUMENTS

<https://www.wjgnet.com/bpg/GerInfo/287>

GUIDELINES FOR NON-NATIVE SPEAKERS OF ENGLISH

<https://www.wjgnet.com/bpg/gerinfo/240>

PUBLICATION ETHICS

<https://www.wjgnet.com/bpg/GerInfo/288>

PUBLICATION MISCONDUCT

<https://www.wjgnet.com/bpg/gerinfo/208>

ARTICLE PROCESSING CHARGE

<https://www.wjgnet.com/bpg/gerinfo/242>

STEPS FOR SUBMITTING MANUSCRIPTS

<https://www.wjgnet.com/bpg/GerInfo/239>

ONLINE SUBMISSION

<https://www.f6publishing.com>

Metastatic low-grade endometrial stromal sarcoma with variable morphologies in the ovaries and mesentery: A case report

Hai-Yun Yu, Yu-Lan Jin

Specialty type: Medicine, research and experimental

Provenance and peer review:

Unsolicited article; Externally peer reviewed.

Peer-review model: Single blind

Peer-review report's scientific quality classification

Grade A (Excellent): 0

Grade B (Very good): 0

Grade C (Good): C, C, C, C

Grade D (Fair): 0

Grade E (Poor): 0

P-Reviewer: Al-Ani RM, Iraq; Tajiri K, Japan

Received: April 3, 2022

Peer-review started: April 3, 2022

First decision: June 16, 2022

Revised: June 22, 2022

Accepted: July 11, 2022

Article in press: July 11, 2022

Published online: August 16, 2022



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Abstract

BACKGROUND

Low-grade endometrial stromal sarcoma (LGESS) classically exhibits a proliferative morphology. However, morphological variation of extrauterine tumors presents a diagnostic challenge.

CASE SUMMARY

We report the case of a 76-year-old female patient with extensive extrauterine and abdominal neoplastic lesions. Computed tomography showed massive pleural and ascitic fluid, and there was an increase in serum cancer antigen 125. She underwent bilateral adnexectomy and tumor resection. The right ovary had been replaced by a multinodular mass that was 8.5 cm × 4.5 cm × 3.5 cm in size. In addition, there was a 24 cm × 15 cm × 13 cm mesenteric mass, which was also multinodular, with local invasion of the intestinal serosa and underlying muscle. Under the microscope, the tumors in different places exhibited two different patterns, thus presenting great challenges to diagnosis and treatment. Thorough pathological assessment eliminated all differential diagnoses in favor of metastatic LGESS derived from a 20-year-old primary tumor initially misdiagnosed as leiomyosarcoma.

CONCLUSION

LGESS morphology varies according to tumor location. Accurate diagnosis is critical for appropriate treatment and improved prognosis and patient care.

Key Words: Gastrointestinal stromal tumor; Low-grade endometrial stromal sarcoma; Mesentery; Ovary; Tumor; Case report

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Core Tip: Low-grade endometrial stromal sarcoma (LGESS) could have a morphological variation of extrauterine tumors. These variations include smooth muscle differentiation, fibroma-like stromal pattern, and glandular differentiation. Misdiagnosis is common and may impact patient outcomes because of differences in the treatment approach for LGESS compared to that of other tumor types. Greater knowledge of LGESS morphological variability is critical in establishing a clear diagnosis and guiding patient care.

Citation: Yu HY, Jin YL. Metastatic low-grade endometrial stromal sarcoma with variable morphologies in the ovaries and mesentery: A case report. *World J Clin Cases* 2022; 10(23): 8384-8391

URL: <https://www.wjgnet.com/2307-8960/full/v10/i23/8384.htm>

DOI: <https://dx.doi.org/10.12998/wjcc.v10.i23.8384>

INTRODUCTION

Low-grade endometrial stromal sarcoma (LGESS) is rare, accounting for only 1% of all malignant uterine tumors and 10% of uterine sarcomas[1-3]. The tumor is persistent and may metastasize years after initial resection[4,5]. Classically, LGESS exhibits a proliferative morphology. However, metastases may present with variable forms of differentiation including smooth muscle differentiation, fibroma-like stromal pattern, glandular differentiation, sex cord differentiation, or prominent myxoid change within the tumor[6-8]. Thus, the range of differential diagnoses is wide and includes both benign and malignant tumors. LGESS with fibroblastic features and extensive metastasis has rarely been reported. To the best of our knowledge, our case is the first to describe metastatic LGESS with variable morphologies occurring in both ovaries and the mesentery.

CASE PRESENTATION

Chief complaints

A 76-year-old female patient presented to our hospital with abdominal distension and a chief complaint of constipation.

History of present illness

The patient's constipation had lasted for two months, and it was worse after eating. There was no vaginal bleeding, fluid production, or obvious increase in the frequency of urination.

History of past illness

Upon presentation, review of the patient's medical history revealed a vaginal hysterectomy 20 years prior in response to a mass that was then diagnosed as leiomyomasarcoma.

Personal and family history

She denied any family history.

Physical examination

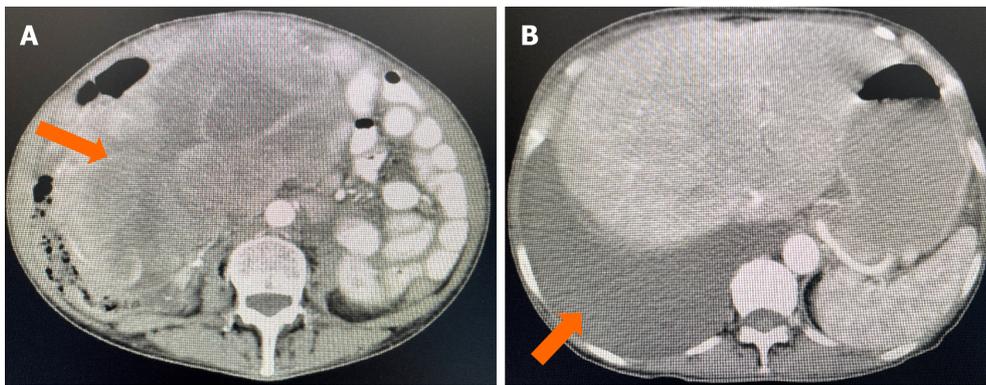
The patient was observed to be short of breath with nominal exercise. The shape of her chest was generally normal. Breath sounds from the right lung were reduced, while those from the left were normal. She had abdominal distention and a palpably large mass, the upper edge of which reached two transverse fingers above the umbilicus. Vulva and vagina revealed no obvious abnormalities.

Laboratory examinations

Laboratory findings measured the levels of the tumor marker cancer antigen 125 at 420.3 U/mL. No tumor cells were found in the pleural or ascitic fluids.

Imaging examinations

Computed tomography (CT) of the abdomen and pelvic area revealed a large, cystic, and solid mass with unclear bilateral attachments (Figure 1A). After assessment by gynecologists, the mass was suspected to be a malignant tumor originating from the appendix that had undergone peritoneal metastasis. Multiple small cysts were identified in the liver accompanied by evident ascites (Figure 1B), and another small cyst was found in the left kidney. Chest CT revealed a massive right pleural effusion.



DOI: 10.12998/wjcc.v10.i23.8384 Copyright ©The Author(s) 2022.

Figure 1 Computed tomography of the whole abdomen and pelvis. A: A huge cystic and solid mass in the pelvic and abdominal cavity; B: There was also a large amount of fluid in the pelvic and abdominal cavity.

Surgical exploration

After evaluation, clinicians decided to proceed with surgery. Gynecologists and general surgeons participated in the operation. Intraoperative exploration uncovered bilateral ovarian, mesenteric, and abdominal wall tumors. The right ovary had been replaced by a multinodular mass that was 8.5 cm × 4.5 cm × 3.5 cm in size. The cut surface was grayish and powdery, and some areas were slightly fragile, while others were solid. The tumor had grown as a sheet containing thick collagen fibers; some areas were edematous, and some areas were densely packed. The 24 cm × 15 cm × 13 cm mesenteric mass was also multinodular with local invasion of the intestinal serosa and underlying muscle. The cut surface was yellowish-gray, powdery, and solid. The mass showed signs of edema and degeneration.

Pathological investigations

Resected samples from the ovarian and mesenteric tumors were sent for frozen section biopsy. Cells from the bilateral ovarian lesions were oval-shaped or short and spindle-shaped; the cells were slightly atypical, but any mitotic figures were unobvious. In some areas, capillary involvement was evident (Figure 2A and B). In contrast, cells of the mesenteric tumor had grown in sheets, bundles, or fishbone-like arrangements. The cells were oval-shaped or short and fusiform. Nuclear atypia was unapparent, and mitotic figures were clearly visible (10-20 cells/10 high-power fields). Tumor involvement in the intestinal smooth muscle was evident, and regions of flaky necrosis were observed (Figure 2C and D).

Considering the patient's medical history and clinical and histopathological findings, we entertained a diagnosis of Meigs syndrome. We initially believed the right ovarian tumor to be a sex cord-stromal cell tumor (*e.g.*, fibroid or follicular membrane fibroma). The mesenteric mass was initially believed to be a gastrointestinal stromal tumor (GIST). However, to narrow the differential diagnoses, we opted to perform further immunohistochemistry (IHC) and molecular analyses.

IHC revealed diffuse expressions of estrogen receptor (ER), progesterone receptor (PR), cluster of differentiation (CD) 10 (CD10), and Wilms' tumor 1 (WT1) in both the ovarian and mesenteric lesions (Figure 3). Calretinin, α -inhibin, desmin, smooth muscle actin, Cytokeratin, epithelial membrane antigen (EMA), CD117, Dog-1, and CD34 were not detected. Approximately 30% of cells were Ki-67+, and wild-type P53 was ubiquitously expressed (Table 1). We further performed fluorescence in situ hybridization (FISH) analysis on the mesenteric neoplastic tissue. By our analysis, variants of the following genes were not detected: BCL6 corepressor (BCOR), 14-3-3 protein epsilon (YWHAE), and JAZF zinc finger 1 (JAZF1). All three variants are common diagnostic indicators of ESS despite morphologic variation. We also performed next-generation sequencing (NGS) on two specimens separately. Mutations in five cancer-related genes (PTEN, TEK, CDK4, MDM2, and CDKN1C) were detected on the mesenteric mass. The ovarian tumor had mutations in the CHD [c.735G>C (p.E245D)], TSHR [c.928C>T (p.R310C)], and MC1R [c.219G>A (p.M73I)] genes, which have not been reported to cause disease.

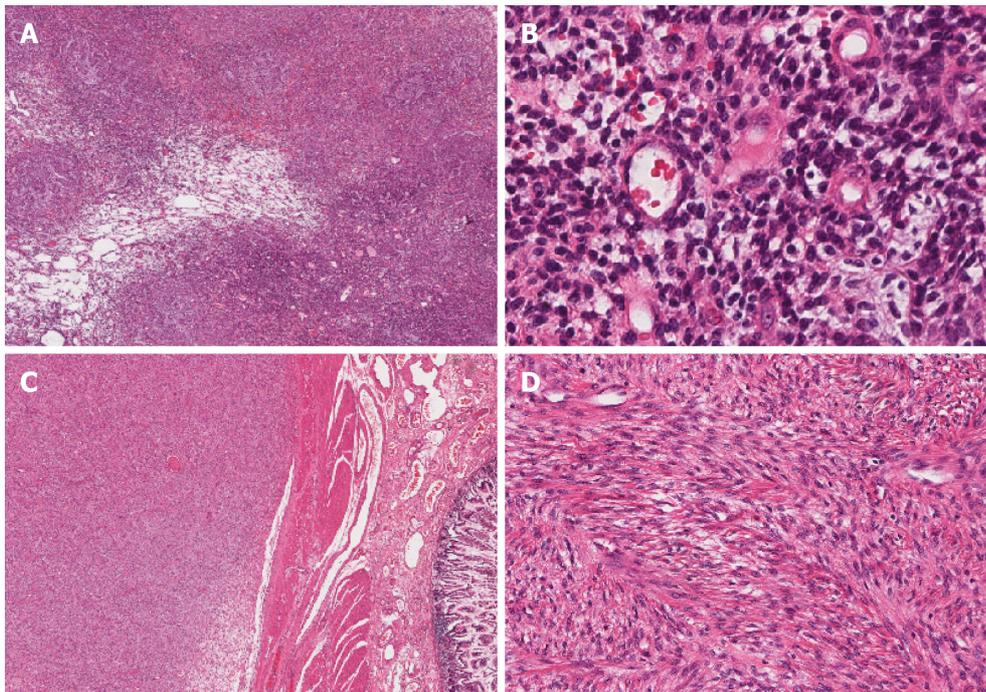
FINAL DIAGNOSIS

Our analyses confirmed a diagnosis of LGESS for all neoplasms. To distinguish whether the tumors were primary extrauterine ESSs or metastases, we reviewed the patient's uterine tumor biopsy material from the prior vaginal hysterectomy. Using standard light microscopy at low magnification, the tumor was visible in the uterine smooth muscle as an infiltrative nodular mass (Figure 4A). The cellular growth pattern resembled that of the current ovarian tumor (Figure 4B). We believe that the patient was previously misdiagnosed and that the correct diagnosis should have been LGESS. Therefore, the current

Table 1 The immunohistochemical profiles of the ovarian tumor and the mesenteric tumor

The ovarian tumor		The mesenteric tumor	
Positive	Negative	Positive	Negative
ER	CD117	ER	CD117
PR	Dog-1	PR	Dog-1
CD10	CD34	CD10	CD34
WT-1	P53	WT-1	P53
Bcl2	BCOR	Bcl2	Desmin
Ki-67 (30%)	S-100	Ki-67 (30%)	SMA
SMA (focal)	CK		Calretinin
Desmin (focal)	Pax-8		Inhibin- α
CD99 (focal)	EMA		S-100
Inhibin- α (focal)	ALK		CK
SF-1 (focal)	CyclinD1		ALK
			CyclinD1

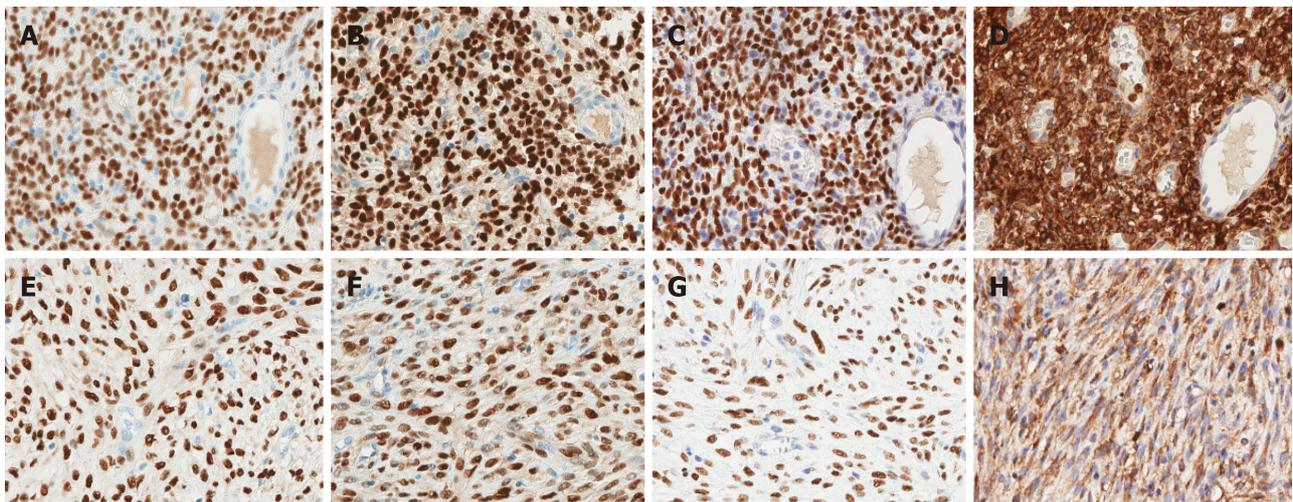
ER: Estrogen receptor; PR: Progesterone receptor; CD10: Cluster of differentiation 10; WT-1: Wilms' tumor 1; Bcl-2: B-cell leukemia/lymphoma 2 gene; SMA: Alpha-Smooth Muscle Actin; CD99: Cluster Of Differentiation 99; SF-1: Adrenal 4-binding protein; CD117: Cluster Of Differentiation 117; Dog-1: Discovered on GIST-1; CD34: Cluster Of Differentiation 34; P53: Tumor Protein p53; BCOR: B-cell leukemia/lymphoma 6 gen corepressor; S-100: Soluble protein-100; CK: Cytokeratin; Pax-8: Paired box protein 8; EMA: Epithelial membrane antigen; ALK: Anaplastic lymphoma kinase.



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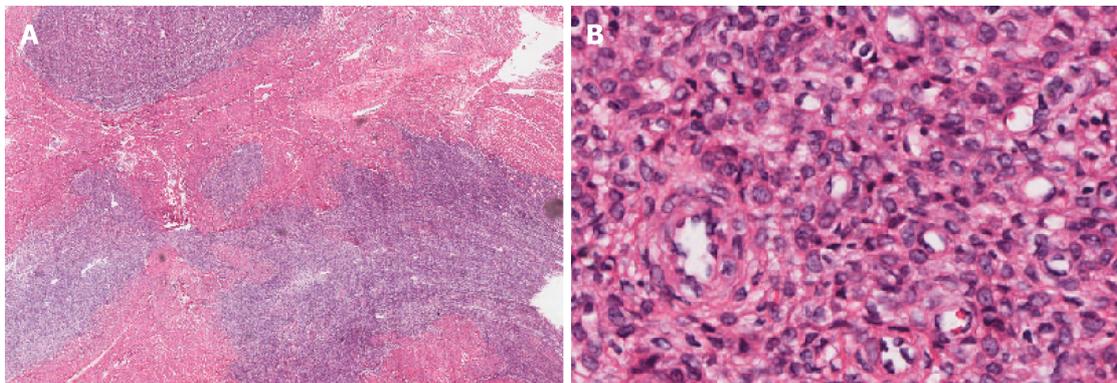
Figure 2 The right ovarian mass and the mesenteric mass in hematoxylin and eosin stain. A: The ovarian tumor has grown as a solid sheet (Original magnification: 40 \times ; scale bar: 100 μ m); B: In some areas, tumors cells have grown around small blood vessels (Original magnification: 200 \times ; scale bar: 100 μ m); C: The mesenteric tumor exhibits local invasion of the intestinal serosa and underlying muscle (Original magnification: 40 \times ; scale bar: 100 μ m); D: Cells have grown in a fishbone-like arrangement (Original magnification: 100 \times ; scale bar: 100 μ m).

tumors were diagnosed as bilateral ovarian and abdominal metastases of the primary tumor from 20 years ago.



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Figure 3 Immunohistochemistry of the right ovarian tumor tissue and the mesenteric tumor tissue. A and E: The tumor cells are diffusely and strongly positive for estrogen receptor; B and F: Progesterone receptor; C and G: Wilms' tumor 1; D and H: Cluster of differentiation 10. Original magnification: 200 ×; scale bar: 100 μm.



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Figure 4 Biopsy material from the primary intrauterine tumor 20 years ago. A: Light microscopy examination shows that the tumor had infiltrated the adjacent vaginal smooth muscular layer as tongues or islands (Original magnification: 40 ×; scale bar: 100 μm); B: Small oval to fusiform cells resemble the cells of proliferative endometrial stroma and whirl around spiral arteriole-like vessels (Original magnification: 400 ×; scale bar: 100 μm).

TREATMENT

The patient underwent bilateral adnexectomy + omentectomy + partial bowel wall resection + mesenteric mass excision + rectal anterior mass excision + abdominal wall tumor resection. After the surgery, she was treated with medications that included anti-inflammatory, anticoagulation, gastric protection, fluid supplementation, water and electrolyte balance. The patient was discharged on the sixth day after the operation and did not complain of any complications. Endocrine therapy was conducted with oral letrozole tablets.

OUTCOME AND FOLLOW-UP

The patient had been followed up for nine months after surgery. She had come back eight times for re-examination and prescriptions subsequent to hospital discharge. Imaging examination was not repeated. Abnormal liver function was detected with elevated alanine aminotransferase and aspartate aminotransferase.

DISCUSSION

Endometrial stromal tumors mainly occur in the uterus and are categorized based on tumor morphology, immunophenotype, and molecular pathology as follows: endometrial stromal nodules, LGESS, high-grade ESS, and undifferentiated uterine sarcoma[9]. LGESS is a rare malignant tumor that shares morphologic resemblance with endometrial stroma in the proliferative phase. LGESS is capable of infiltrative growth with or without lymphovascular invasion.

At presentation, the patient had no known history of ESS. In the absence of uterine tissue, we initially considered a diagnosis of primary extrauterine ESS (EESS), a type of extrauterine ESS with no accompanying intrauterine lesions[10]. At present, the pathogenesis of EESS remains unclear, but the following two hypotheses have been proposed: (1) Malignant transformation of endometriosis; and (2) Malignant transformation of pluripotent body cavity epithelium established during embryonic development[11,12]. In support of the first hypothesis, multiple reports of EESS describe swollen endometriotic lesions located in tissue adjacent to the tumor. In other cases, EESS has been found in the fallopian tubes, ovaries, and pelvic peritoneum, or it has become widely distributed in the abdominal and pelvic cavities. In support of the second hypothesis, this may result from interstitial cells of the body cavity epithelium differentiating into Müllerian duct epithelium and stroma. However, more cases are required for further substantiation. We did not find endometriotic lesions in our patient, and after extensive analyses, a diagnosis of recurrent and metastatic LGESS was definitive.

The morphology of the bilateral ovarian tumors in our patient was indistinguishable from endometrial stroma in the proliferative stage. In contrast, the mesenteric lesions had a fibrous appearance. The morphologic variation of LGESS is known; however, variable forms are less frequently reported, possibly because misdiagnosis is common.

Previously, IHC has shown LGESS to be diffusely and strongly positive for ER, PR, CD10, and WT1, characteristics which serve to distinguish LGESS from other soft tissue tumors[2,3,13-15]. Specifically, WT1 shows diffuse nuclear positivity in LGESS, while cytoplasmic rather than nuclear staining is more common in other extrauterine soft tissue tumors (*e.g.*, GIST, solitary fibrous tumors, and leiomyomas) [16]. Smooth muscle markers like desmin and h-caldesmon are indicative of neoplasms with smooth muscle differentiation; neoplasms with sex cord-like differentiation may stain positive for α -inhibin, calretinin, and CD99. Therefore, the pattern of IHC positivity in our case supported the diagnosis of LGESS over other soft-tissue tumor types.

The common genetic mutation in LGESS is the fusion of JAZF1 with other genes[17-19]. However, neither our FISH nor our NGS results detected mutations in genes related to ESS such as JAZF1, BCOR, or YWHAE. The results did not refute our diagnosis because one-third of LGESSs do not harbor any genetic mutations[19]. Although the gene mutations in the two sites were not consistent with those found by NGS, it might be that they were somatic mutations in the process of tumor progression and that the tumors originated from the same tumor 20 years ago.

As LGESS usually has a favorable prognosis, the choice of surgical approach, especially for young female patients, is controversial. Surgery including hysterectomy and bilateral salpingo-oophorectomy is considered the initial standard management[20]. Bai *et al*[21] showed that although ovary preservation has a higher risk of recurrence, it does not affect the overall survival rate. Our patient underwent only a hysterectomy 20 years prior, and no further treatment was performed. For LGESS confined to the uterus, more studies are needed to confirm whether preserving ovarian function is feasible.

CONCLUSION

The morphology of LGESS may vary according to tumor location. LGESS with fibroblast differentiation is rare and may be easily misdiagnosed as GIST or leiomyosarcoma. Based on our findings, we suggest performing IHC or molecular analyses for LGESS diagnostic certainty. A clear and correct diagnosis is critical to support appropriate treatment, improve prognosis, and guide patient care.

ACKNOWLEDGEMENTS

The authors thank Hu QM of Geneseeq Biotechnology Incorporation for technical assistance.

FOOTNOTES

Author contributions: Jin YL provided writing ideas; Yu HY collected data and analyzed them; both authors read and approved the final manuscript.

Informed consent statement: Informed written consent was obtained from the patient for publication of this report

and any accompanying images.

Conflict-of-interest statement: All authors state that there are no conflicts of interest to disclose.

CARE Checklist (2016) statement: The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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Country/Territory of origin: China

ORCID number: Hai-Yun Yu 0000-0002-4261-7506; Yu-Lan Jin 0000-0002-9716-1948.

S-Editor: Wang LL

L-Editor: A

P-Editor: Wang LL

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