

Low-Grade Endometrial Stromal Sarcoma with Intravenous and Intracardiac Extension: A Case Report

Xiaoshan Chai

Central South University

Hui Ding

Central South University

Peng Zhou

Central South University

Xilong Mei

Central South University

XiaoXue Li

Central South University

Guangshi Tao (✉ taoguangshi@csu.edu.cn)

The Second Xiangya Hospital of Central South University

Case Report

Keywords: Endometrial stromal sarcoma, Intravenous and Intracardiac Extension, CD10, Case report

Posted Date: September 9th, 2020

DOI: <https://doi.org/10.21203/rs.3.rs-69379/v1>

License:  This work is licensed under a Creative Commons Attribution 4.0 International License. [Read Full License](#)

Abstract

Background: Low-grade endometrial stromal sarcoma (ESS) is rare mesenchymal neoplasm, which has an indolent history with late recurrences. ESS usually spread through the lymph nodes and venous system but very seldom involve large vessels or the heart.

Case presentation: A 38-year-old Chinese woman was admitted to our department due to pelvic mass found on physical examination. The superior and inferior vena cava CT angiography (CTA) showed an enlarged uterine as well as low density image in the left internal iliac vein, the left common iliac vein, the inferior vena cava, the left renal vein adjacent to the heart and the right atrium, with a range of 110*16mm. The filling defect of right atrium was about 30*14mm. The three-dimensional computed tomography reconstruction showed that the mass originated from the uterine and invaded into the reproductive vein, subsequently extended along the inferior vena cava to the right atrium. Needle biopsy of the pelvic mass was performed and the tissue indicates smooth muscle. The preoperative diagnosis was intravascular leiomyomatosis and the patient underwent radical resection: thrombectomy and total hysterectomy with bilateral salpingo-oophorectomy. Postoperative histopathology revealed low grade endometrial stromal sarcoma. Microscopically, the tumors in both original uterine lesions and intravascular and intracardiac metastases shared morphologic features characterized by neoplastic cells similar to proliferative-phase endometrial stromal cells, in which small spiral artery differentiation was recognized and tumor tissue showed invasive growth pattern by inserting into the surrounding smooth muscle. Immunohistochemistry showed tumor cells were reactive to Estrogen Receptor, Progesterone Receptor, CD10. Primary uterine foci showed cyclin D1(5%+) and Ki-67(20%+), whereas metastatic lesions of the intracardiac and the intravascular component identified cyclin D1(negative) and Ki-67(2%+). The patient is alive without evidence of recurrence 3 months after surgery.

Conclusions: Distant metastasis of low-grade endometrial stromal sarcoma is rare, especially involving large vessels or the heart. This case demonstrates that malignant tumor metastasis should be considered as a differential diagnosis of intracardiac and intravascular masses. The treatment rely on multidisciplinary cooperation.

Background

In the differential diagnosis of lesions in inferior vena cava and the heart, in addition to thrombosis, lesions caused by the invasion of malignant tumor should be considered. Common tumors that may transfer to the inferior vena cava and the heart including renal cancer, liver cancer, uterine leiomyomatosis and nephroblastoma[1]. Uterine fibroids invading the inferior vena cava and the right atrium is also known as intravascular leiomyomatosis[2]. The case described in this paper was admitted to our department due to pelvic mass found on physical examination, other positive findings including lesions extending from the iliac vein to the inferior vena cava and finally reaching the right atrium. The preoperative PET-CT suggested the possibility of uterine malignancy, and thrombosis was considered of the lesions in the blood vessels and atria. Needle biopsy of the pelvic mass was performed and the tissue indicates smooth muscle. Combined with the patient's medical history, physical examination, preoperative imaging and tumor puncture pathological results, the clinical diagnosis was intended to be intravascular leiomyomatosis. Nevertheless the postoperative pathological results revealed low grade endometrial stromal sarcoma.

In the current WHO classification published in 2014, Endometrial stromal tumors were classified into three types according to cell morphology and mitosis: benign endometrial stromal nodules (ESN), Endometrial stromal sarcoma (ESS), and Undifferentiated uterine sarcoma(UUS)[3]. ESS was classified into high grade(HG-ESS) and low grade(LG-ESS). ESS is a very rare tumor entity, represent only around 0.2% of all uterine malignancies, but they make up approximately 10% of uterine sarcomas[4]. Preoperative diagnosis of LG-ESS is difficult as it is often confused with uterine fibroids[5]. ESS usually spread through the lymph nodes and venous system but very seldom involve large vessels or the heart. Cardiac ESS metastases are extremely rare, and their treatment is very complex[6, 7]. Patients with heart involvements are often recurrent cases who had a past history of surgery for endometrial stromal sarcoma[7–11]. To our knowledge, this is the fifth patient involving the inferior vena cava and right atrium at initial diagnosis.

Case Presentation

A 38-year-old woman (gravida 3, para2) was admitted to the Second XiangYa Hospital due to pelvic mass found on physical examination. Further abdominal and pelvic ultrasound as well as the cardiac echocardiography showed inferior vena cava and right atrium lumps. The past history included fallopian tube ligation 12 years ago and hysteroscopic submucosal myomectomy 7 years ago. She had no family history of cancer. Blood testing showed hyperlipemia, cholesterol 6.28 mmol/L(normal range: 2.9–5.2 mmol/L) and triglycerides 1.79 mmol/L(normal range: <1.7 mmol/L). Blood testing also showed slightly elevated levels of trioxypurine 366 U/L(normal range: 155–357 U/L) and cancer antigen (CA)125 and CA 19–9 level was within normal limits. The pelvic examination and transvaginal ultrasonography revealed a three months pregnant-sized uterine without abnormalities in other reproductive organs. The cervical cytology was normal. The superior and inferior vena cava CTA showed an enlarged uterine as well as low density image in the left internal iliac vein, the left common iliac vein, the inferior vena cava, the left renal vein adjacent to the heart and the right atrium, with a range of 110*16 mm. The filling defect of right atrium was about 30*14 mm. The tumor grows along the veins, and the venous system shows low-density shadows (Fig. 1A-F). The three-dimensional computed tomography reconstruction showed that the mass originated from the uterine and invaded into the reproductive vein, subsequently extended along the inferior vena cava to the right atrium(Fig. 2A-D). FDG-PET CT demonstrated maximum standardized uptake value of 14.5 confined to the uterine masses in contrast to elevated uptake value of 8.5 of the intravascular and intracardiac metastatic tumor masses. Segmental curettage was performed to rule out endometrial lesions, In an attempt to clarify the nature of the tumor, needle biopsy of the pelvic mass was performed and the tissue indicates smooth muscle. Immunohistochemistry showed tumor cells were reactive to Ki-67(2%+), Vim, S100, SMA, Desmin, HHF35 and were inactive to CD117, ALK, Dog-1. Preoperative diagnosis of intravascular leiomyomatosis was made. The patient underwent radical resection: thrombectomy and total hysterectomy with bilateral salpingo-oophorectomy. Intraoperative transesophageal echocardiography showed that the atrial tumor was completely removed and the tumors in the vena cava and the left common iliac vein, internal iliac vein, and left renal vein were continued removed. The operation was successful.

Gross specimens showed an enlarged uterus, with multiple fibroids-like nodules, partially fused in subserosal layer, with unclear boundaries and vortex-like structures. The longest tumor segment (from the inferior vena cava and right atrium) is 14.5 cm (Fig. 3A-C). Postoperative histological morphology showed a group of small and consistent round-oval cells, little-medium eosinophilic cytoplasm, 1 mitosis /HPF, small spiral artery differentiation was recognized and tumor tissue showed invasive growth pattern by inserting into the surrounding smooth muscle. Immunohistochemistry showed ER, PR, CD10 positive (Fig. 4A-H). The primary uterine foci showed cyclin D1(5%+) and Ki-67(20%+), whereas metastatic lesions of the intracardiac and the intravascular component identified cyclin D1(< 1%) and Ki-67(2%+) (Fig. 5A-D). Overall histomorphology and immunohistochemistry confirmed the diagnosis of a low-grade endometrial stromal sarcoma. The patient was discharged 12 days after surgery. Treatment with aromatase inhibitor letrozole 2.5 mg everyday. 3 months after the operation, the patient has fully recovered from the operation and can engage in normal activities. MRI showed no obvious signs of tumor recurrence 3 months after surgery.

Discussion

As a very rare malignant gynecological neoplasm, the annual incidence of ESS is 0.19 per 100,000 women [8]. Tumor stage is the most important prognostic factor in LG-ESS [12], which are staged along with uterine leiomyosarcomas in accordance with the FIGO and TNM classifications [13, 14]. In patients with tumor stage I–II, the 5-year survival rate is over 90%, while with stages III–IV it is around 50% [15]. LG-ESS have an indolent history with recurrence rate of 10–20% that often happen many years after the diagnosis and initial surgery [6, 16]. Therefore, they have better prognosis than other uterine sarcomas [17]. In most cases, the recurrence period varied from 3 months to 23 years, with a median interval of 3 years [18]. Long-term follow-up is critical due to the high long-term recurrence rate.

Preoperative diagnosis of LG-ESS can be difficult and is often confused with uterine fibroids on account of the absence of specific clinical manifestations [5]. Imageological procedures such as ultrasound, computed tomography and magnetic resonance imaging are not able to display any specific characteristics of LG-ESS [19]. In contrast to carcinomas of the endometrium, a diagnosis of LG-ESS as a mesenchymal tumor cannot be securely established using hysteroscopy and fractional curettage, in our case no abnormal endometrium was observed during preoperative curettage. Pathologically, a clear distinction from benign ESN can only be reliably made after histological analysis of the tumor's entire interface with the neighboring myometrium [20]. Therefore most diagnosis of LG-ESS were made after surgery. According to the literature, the most frequently reported sites of metastases of the LG-ESS are the vagina, pelvis, and peritoneal cavity. Cardiac metastases are rare because ESS commonly spreads through the lymph nodes and venous system [6]. To our knowledge, 18 LG-ESS patients with heart involvement have been reported to date, and their clinical characteristics are summarized as shown in Table 1.

The age of the 18 patients with heart involvement ranged from 24 years old to 71 years old (average age 46 years), 66.7% had a history of surgery for LG-ESS and only 33.3% without definite history and were diagnosed for the first time. Our patient had a history of "submucosal myomectomy" 7 years ago, the diagnosis of ESN was determined, when reassessing the pathological sections from the last operation, ESN was still considered due to the limited amount of specimens. It is not possible to know the fact back then. In this case we predict it is likely that the patient already had LG-ESS 7 years ago, and the tumor relapsed this time. Therefore, whether surgery is a cause of LG-ESS invasion into the great vessels and the heart remains to be further explored. For patients with a history of LG-ESS, it is not difficult to predicate the recurrence. However, for patients with no relevant history, when tumors in the atrium and inferior vena cava were discovered for the first time, the differential diagnosis of uterine malignant tumor metastasis should also be considered in addition to thrombi and venous leiomyomatosis. The clinical symptoms of LG-ESS involving the heart and great vessels vary from asymptomatic to obvious dyspnea and right heart failure. CT and MRI examinations are of value for early detection of cardiac and vascular involvement. In our case, the three-dimensional CT reconstruction imaging was used to simulate tumor route and scope, which was of great significance for comprehensive evaluation of the range of the lesion and complete resection of the tumors. Several cases pointed out that transesophageal ultrasound (TEE) is useful for diagnosis of cardiac involvement before operation and can also guide the doctor to determine whether the cardiac mass is removed completely during the surgery [21–22]. Moreover there were patients whose intracardiac lesions were detected accidentally by TEE [23], suggesting that for patients with possible great vascular metastasis, TEE should be stressed and applied as the important means.

The primary treatment for LG-ESS is surgery with total hysterectomy (without morcellation) and bilateral salpingo-oophorectomy [18]. It has been shown that LG-ESS are hormone-dependent, it is not clear whether the ovaries can be preserved in young, premenopausal women [24]. There is no evidence that cytoreduction and lymphadenectomy are beneficial for long-term survival [25]. All 18 patients with cardiac involvement were stage IV LG-ESS and radical resection were performed in 14 cases, the perioperative mortality rate was zero. All patients recovered uneventfully after the operation. Except for one case, all 13 patients showed no signs of recurrence by the end of the follow-up. Although the long-term effects of surgery on patients cannot be proved, the surgery can prevent patients from developing into severe complications such as heart failure and pulmonary embolism even sudden death.

The final diagnosis of this disease mainly depends on histopathology, the pathological diagnosis of our case is certain. Interestingly, immunohistochemistry of cyclin D1 and Ki-67 in the primary uterine foci and the metastatic lesions of the intracardiac and the intravascular component are different. This result is consistent with the study of Koto Fujiishi et al [26]. In their study, a patient with LG-ESS whose uterine lesions composed of three components, all of which meet the criterion of LG-ESS. However, PET-CT indicated high uptake of fluorodeoxyglucose in 2 lesions (subserosal tumors with SUV value 13.28) and no uptake of fluorodeoxyglucose in 1 lesion (intramural tumor with SUV value 0) before surgery. Postoperative nuclear cyclin D1 immunostaining was identified 50% of neoplastic cells in the two subserosal tumors whereas < 1% positive cells in the intramural component. From another perspective, the heterogeneity of tumor cells in the same disease existed. In our case, the preoperative PET-CT SUVmax of primary lesion was about 14.5 and SUVmax of the metastasis was about 8.5. The immunohistochemical result of CyclinD1 and Ki-67 corresponds with the SUVmax of PET-CT. We assumed that there was heterogeneity in LG-ESS tumor cells. The two cell cycle proteins CyclinD1 and Ki-67 of the same tumor showed different levels of expression in different lesions and this may explain the difference in response at different regions in patients with LG-ESS when treated with chemotherapeutic agents targeting for cell division. Some masses can shrink by nearly 80%, while others have no significant change [27]. There are no valid data to show that adjuvant chemotherapy leads to any

improvement in survival in patients with LG-ESS[18]. Postoperative radiotherapy in patients with ESS only appears to improve locoregional control, so that the medium-term and long-term side effects of pelvic irradiation need to be weighed carefully against what is in any case a good prognosis in relation to locoregional recurrences [28]. The expression of steroid receptors and aromatases in LG-ESS suggests that adjuvant therapy with gestagens, GnRH analogues, or aromatase inhibitors should be effective[18]. Whether PET-CT as well as the immunohistochemical results of CyclinD1 and KI-67 can also guide subsequent treatment, such as chemotherapy and radiotherapy, remains to be further explored.

Conclusions

Despite its well-known good prognostic nature, low-grade ESS may behave as an aggressive malignancy. The differential diagnosis of malignant tumor metastasis should be considered in the lump of great vessels and the heart. Complete resection of tumor requires multidisciplinary cooperation. The perioperative mortality is low and the risk of fatal complications such as heart failure and pulmonary embolism can be reduced.

Abbreviations

ESS
endometrial stromal sarcoma
CTA
CT angiography
ESN
endometrial stromal nodules
UUS
Undifferentiated uterine sarcoma
CA
cancer antigen
TEE
transesophageal ultrasound

Declarations

Ethics approval and consent to participate

The experimental protocol was established, according to the ethical guidelines of the Helsinki Declaration and was approved by the Human Ethics Committee of The Second Xiangya Hospital of Central South University (No.2019(218)). Written informed consent was obtained from all participants.

Consent for publication

Written consent was obtained from the patient to publish this case report.

Availability of data and materials

All data generated or analysed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests.

Funding

Not applicable.

Authors' contributions

XC, GT conceived and designed the experiments; XC, HD, XM, PZ carried out the experiments and analyzed the patient data; XC, ST, XL wrote the manuscript. All authors read and approved the final manuscript.

Acknowledgements

Not Applicable.

Authors' information

Affiliations

Department of Obstetrics & Gynecology, The Second Xiangya Hospital of Central South University, Changsha, china, 410011.

Xiaoshan Chai& Hui Ding& Xiaoxue Li& Guangshi Tao

Department of pathology, The Second Xiangya Hospital of Central South University, Changsha, china,410011.

Peng Zhou

Department of radiology, The Second Xiangya Hospital of Central South University, Changsha, china, 410011.

Xilong Mei

Footnotes

Not Applicable.

References

1. Leitman M, Rahanani E, Wassermann I, Rosenblatt S, Peleg E, Krakover R, Zuckermann B, Fuchs T, Theodorovich N, Vered Z. Unusual Right-Sided Cardiac Masses. *Echocardiography*. 2010;27:1151–5.
2. Durck H. Ueber ien kontinuierlich durch die entere hohlvene in das herz vorwachsendes: Fibromyom des uterus. *Munch Med Wochenschr*. 1907;54:1154. (In German).
3. Ali RH, Rouzbahman M. Endometrial stromal tumors revisited:an update based on the 2014 WHO classification. *J Clin Pathol*. 2015;68:325–32.
4. Koss LG, Spiro RH, Brunschwig A. Endometrial stromal sarcoma. *SurgGynecol Obstet*. 1965;121:531.
5. Amant F, Moerman P, Cadron I, Neven P, Berteloot P, Vergote I. The diagnostic problem of endometrial stromal sarcoma: report on six cases. *Gynecol Oncol*. 2003;90(1):37–43.
6. Marijana T, Evgeny B, Cesare C,Burkert P, Sabine H. Right heart masses in a patient with endometrial stromal sarcoma. *J Clin Ultrasound*. 2020;48(2):117–20.
7. Nathan D, Szeto W, Gutsche J, Min H, Kalapatapu V. Metastaic endometrial sarcoma with inferior vena caval and cardiac involvement: a combined surgical approach. *Vasc Endovascular Surg*. 2014;48:267–70.
8. Scher D, Nghiem W, Aziz S, et al. Endometrial stromal sarcoma metastatic from the uterus to the inferior vena cava and right atrium. *Heart Inst J*. 2015;42:558–60.
9. Gabal S, Ashour Z, Hamada G, et al. Low-grade endometrial stromal sarcoma with intravenous extension to the heart. *Medscape J Med*. 2009;11:23.
10. Kudaka W, Inafuku H, Iraha Y, et al. Low-grade endometrial stromal sarcoma with intravenous and intracardiac extension: a multidisciplinary approach. *Case Rep Obstet Gynecol*. 2016;2016:3467849.
11. Kuo PL, Lee TY, Wu MT, Wu FZ. Endometrial stromal sarcoma with intravenous and cardiac extension. *Heart Lung Circ*. 2015;24:e220–1.
12. Chew I, Oliva E. Endometrial stromal sarcomas: a review of potential prognostic factors. *Adv Anat Pathol*. 2010;17:113–21.
13. Oliva E, Carcangiu ML, Carinelli SG, et al: Mesenchymal tumors of the uterus; in Kurman RJ, Carcangiu ML, Herrington CS, Young RH, editors: WHO Classification of Tumours of Female Reproductive Organs.Lyon, IARC Press, 2014, pp. 135–147.
14. Wittekind C, Meyer HJ: TNM-Klassifikation Maligner Tumoren. Weinheim, Wiley-VHC Verlag, 2010.
15. Horng HC, Wen KC, Wang PH, et al. Uterine sarcoma part II-uterine endometrial stromal sarcoma: the TAG systematic review. *Taiwan J Obstet Gynecol*. 2016;55:472–9.
16. Lee CH, Marino-Enriquez A, Ou W, et al. The clinicopathologic features of YWHAЕ-FAM22 endometrial stromal sarcomas: a histologically high-grade and clinically aggressive tumor. *Am J Surg Pathol*. 2012;36:641–53.
17. Alaaraj R, Lataifeh I, Elsaadi J, Salameh M, Sughayer M. Primary,low-grade endometrioid stromal sarcoma of the ovary with multiple omental metastases. *Gulf Med J*. 2016;5:69–73.
18. Thiel FC, Halmen S. Low-Grade Endometrial Stromal Sarcoma - a Review. *Oncol Res Treat*. 2018;41:687–92.
19. Amant F, Coosemans A, Debiec-Rychter M, Timmerman D, Vergote I. Clinical management of uterine sarcomas. *Lancet Oncol*. 2009;10:1188–98.
20. Hoang L, Chiang S, Lee CH. Endometrial stromal sarcomas and related neoplasms: new developments and diagnostic considerations. *Pathology*. 2018;50:162–77.
21. Daniel Scher W, Nghiem S, Aziz R, Rahbar W, Banks. Anthony Venbrux, Shawn Sarin: Endometrial Stromal Sarcoma Metastatic from the Uterus to the Inferior Vena Cava and Right Atrium. *Tex Heart Inst J*. 2015;42(6):558–60.
22. Derek Nathan W, Szeto J, Gutsche H, Min. Venkat Kalapatapu:Metastatic endometrial sarcoma with inferior vena caval and cardiac involvement: a combined surgical approach. *Vasc Endovascular Surg*. 2014;48(3):267–70.
23. Cristina L, Wood J. Sederberg 2nd, Paul Russ, Tamas Seres:Cystic appearance of low-grade endometrial stromal sarcoma in the right atrium: case report. *Cardiovasc Ultrasound*. 2011;9:23.
24. Einstein MH, Barakat RR, Chi DS, et al. Management of uterine malignancy found incidentally after supracervical hysterectomy or uterine morcellation for presumed benign disease. *Int J Gynecol Cancer*. 2008;18:1065–70.
25. Chan JK, Kawar NM, Shin JY, et al. Endometrial stromal sarcoma: a population-based analysis. *Br J Cancer*. 2008;99:1210–5.
26. Koto Fujiishi S, Nagata R, Kano C, Kubo M, Shirayanagi M, Ozaki T, Yamamoto K, Nakanishi. Shoji Kamiura, Shin-Ichi Nakatsuka: JAZF1-SUZ12 endometrial stromal sarcoma forming subserosal masses with extraordinary uptake of fluorodeoxyglucose on positron emission tomography: a case

report. *Diagn Pathol.* 2019;14(1):110.

27. Hideki Mizunuma: Asymptomatic intracardiac metastasis from a low-grade endometrial

Yoshihito Yokoyama Y, Ono T, Sakamoto I, Fukuda. Hideki Mizunuma: Asymptomatic intracardiac metastasis from a low-grade endometrial.

28. stromal sarcoma with successful. surgical resection *Gynecol Oncol.* 2004;92(3):999–1001.

29. Denschlag D, Thiel FC, Ackermann S, et al.: Sarcoma of the uterus. Guideline of the DGGG (S2k-level, AWMF registry no. 015/074, August 2015). *Geburtshilfe Frauenheilkd* 2015; 75: 1028–1042.

Tables

Table 1

No.	Authors	Age	P/R	Symptoms	Heart involvement	Imaging diagnosis	Main surgery for recurrent tumor or advanced primary tumor	Additive treatment	Follow-up (years)	Follow-up (outcome)
1	STEPHEN P	50	R	dyspnea; edema of the lower extremities	Right atrium	n.a.	Subtotal resection of cardiac TU thrombus (R2)	Post-op CH, RX	1	Death
2	Michael R	52	R	edema, pain, tenderness.	Right ventricular	TEE MRI	Radical resection: thrombectomy, vena cava transected	n.a.	1.5	Alive and tumor-free
3	J. F. Val-Bernal	49	R	Edema ,dyspnea on exertion	Right ventricular; Pulmonary artery;	CT MRI	Radical resection: thrombectomy, tricuspid valve annuloplastia	Post-op CH, HO	0.4	Alive and tumor-free
4	Tsutomu Tabata	43	R	None	Left ventricle; Pulmonary artery	CT X ray	No operation, partial remission under CH, RX	CH, RX	14	Death (heart failure, TU within left ventricle)
5	Y. Yokoyama	52	R	abdominal pain and vomit	Right ventricular; Pulmonary artery	CT MRI	Radical resection: thrombectomy	CH	n.a.	Alive and tumor-free
6	P. Renzulli	47	R	hypertension, fatigue, shortness of breath and abdominal pain	Right atrium	CT MRI	Radical resection: thrombectomy, BSO	Post-op RX, HO	4.5	Alive and tumor-free
7	Cristina L Wood	71	R	fatigue	Right atrium	TEE CT MRI	Radical resection: thrombectomy, BSO	n.a.	n.a.	n.a.
8	D. Nathan	40	R	abdominal pain	Right atrium; Right ventricular	TEE CT	Radical resection: thrombectomy	n.a.	n.a.	n.a.
9	D.Scher	55	P	postmenopausal spotting	Right atrium; Pulmonary artery	TEE CT	Radical resection: thrombectomy,TAH,BSO	Post-op HO	1	Alive and tumor-free
10	Wataru Kudaka	40	P	None	Right atrium; Right ventricular	TEE CT MRI	Radical resection: thrombectomy,TAH,BSO	Post-op HO	2.4	Alive and tumor-free
11	Itzhak Kronzon	30	R	facial and lower extremity swelling, ascites	Right atrium; Right ventricular	TEE MRI	Radical resection: thrombectomy	n.a.	n.a.	n.a.
12	Yoshiki Mikami	29	R	right-sided cardiac failure	Right atrium;	MRI	Radical resection: thrombectomy	N	5	Asymptomatic with tumor
13	Hesham Alkady	30	R	abdominal discomfort , swelling	Right atrium;	CT	Radical resection: thrombectomy	Post-op CH	n.a.	n.a.
14	Marijana Tadic MD	61	R	fever , dyspnea	Right atrium; Right ventricular; RV outflow tract	CT	No operation	HO	0.5	Alive with tumor
15	Pei-Lun Kuo	51	P	lower pelvic pain; vaginal bleeding	Right atrium;	TEE,CT Dynamic cardiac CT angiography	No operation	HO	0.5	Alive with tumor

ESS: endometrial stromal sarcoma; P: primary tumor; R: recurrent tumor; TU: tumor; p.c.: present case; TAH: total abdominal

hysterectomy; BSO: bilateral salpingo-oophorectomy,USO: unilateral salpingo-oophorectomy, IVC:inferior vena cava; CH: chemotherapy; RX: radiotherapy; HO: hormone therapy; post-op: postoperative; peri-op: perioperative; n.a.: not available

No.	Authors	Age	P/R	Symptoms	Heart involvement	Imaging diagnosis	Main surgery for recurrent tumor or advanced primary tumor	Additive treatment	Follow-up (years)	Follow-up (outcome)
16	YUYA NOGAMI	58	p	asymptomatic	Right atrium;	CT MRI Echography	Radical resection: thrombectomy; TAH,BSO	Post-op HO	1	Alive and tumor-free
17	Samia Gabal,	24	P	irregular vaginal bleeding, swelling	Right atrium; Right ventricular	TEE MRI	Radical resection: thrombectomy; TAH,	Post-op CH, RX	1	Alive and tumor-free
18	Our case	38	P	asymptomatic	Right atrium;	TEE,CT	Radical resection: thrombectomy; TAH,BSO	Post-op HO	0.4	Alive and tumor-free

ESS: endometrial stromal sarcoma; P: primary tumor; R: recurrent tumor; TU: tumor; p.c.: present case; TAH: total abdominal hysterectomy; BSO: bilateral salpingo-oophorectomy,USO: unilateral salpingo-oophorectomy, IVC:inferior vena cava; CH: chemotherapy; RX: radiotherapy; HO: hormone therapy; post-op: postoperative; peri-op: perioperative; n.a.: not available

Figures

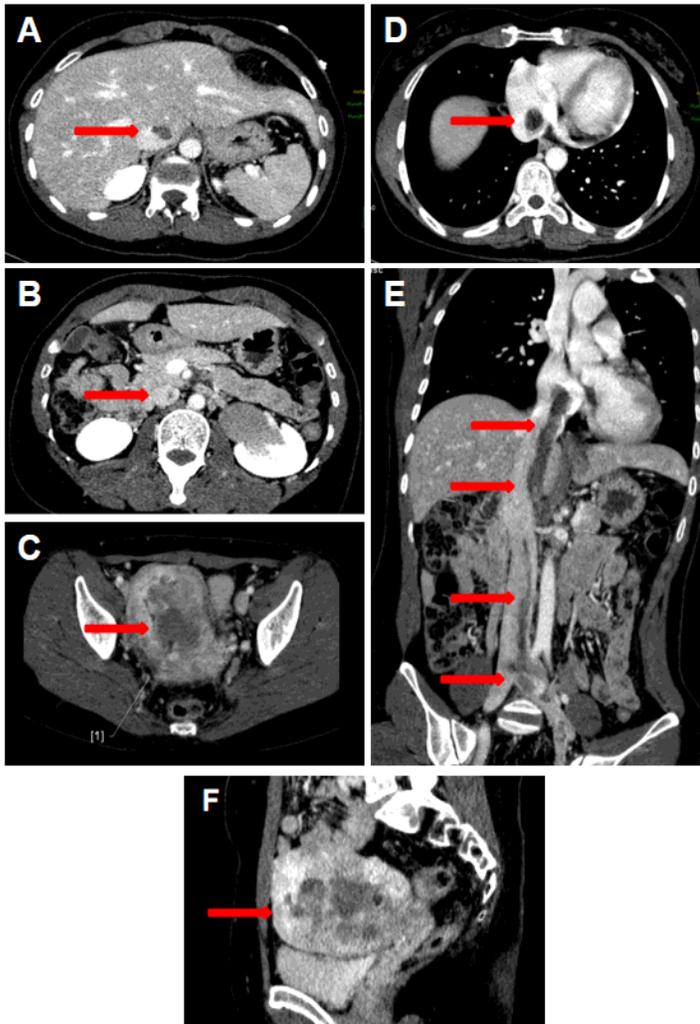


Figure 1
Imaging by the superior and inferior vena cava CTA of the an enlarged uterine as well as low density image in the venous system and the right atrium. The red arrow indicates the mass.

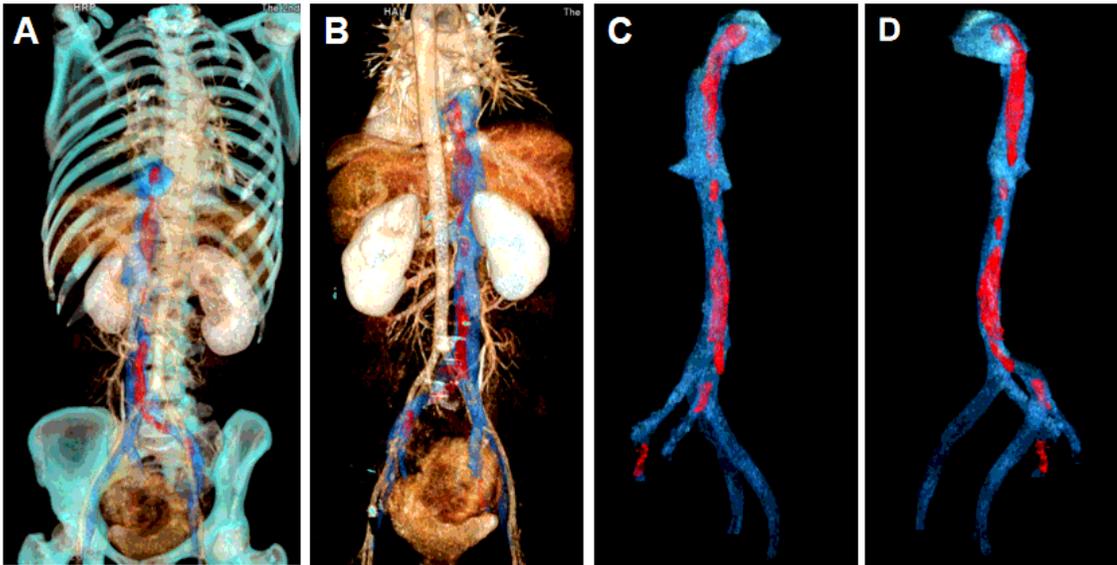


Figure 2
 The three-dimensional computed tomography reconstruction of the whole path of the mass originating from the uterine and invaded into the venous system and eventually to the right atrium. A. Positive view B. Back view C.D. Endovascular imaging of the tumor. Red represents the mass. Blue represents the blood vessel.

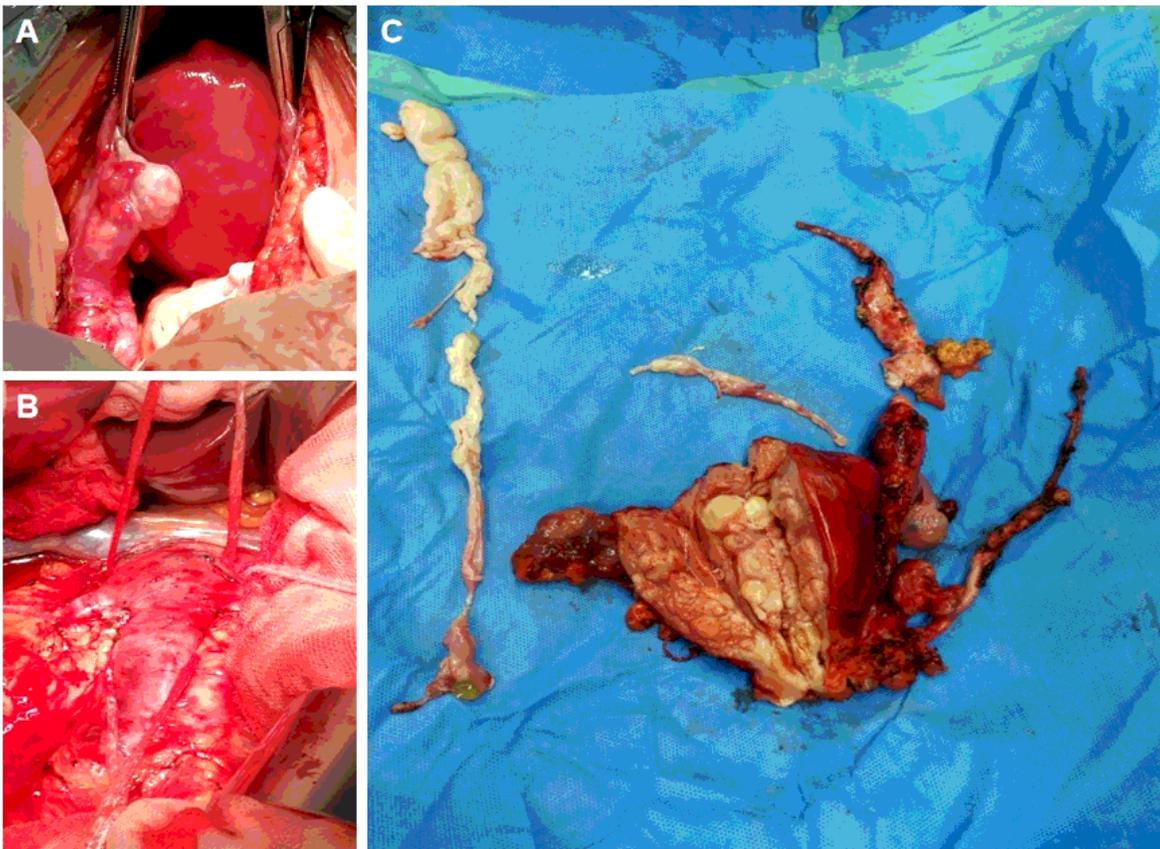


Figure 3
 Imaging of the operation and gross specimen.

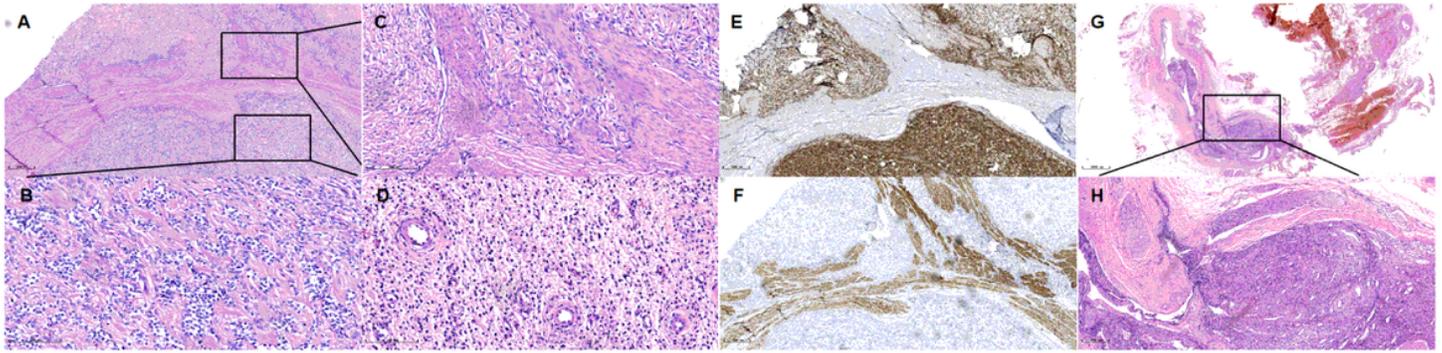


Figure 4

Histopathology and immunohistochemistry. A. Photomicrograph shows a small and uniform population of round to oval cells with little - moderate eosinophilic cytoplasm, nuclear mitotic activity 0-1/HPF(H&E,orig.x50)B. At higher power tumor cells show smooth muscle differentiation (H&E,orig.x200); C. The infiltrating growth of tumor cells and dividing the surrounding smooth muscle tissue(H&E,orig.x200); D. Spiral arterial differentiation (H&E,orig.x200); E. Immunohistochemistry shows tumor cells were reactive to CD10 (H&E,orig.x50); F. Immunohistochemistry shows tumor cells were inactive to SMA (H&E,orig.x50); G: Tumor tissue is seen in the venous wall (H&E,orig.x10);H: tumor tissue partially infiltrated the vein.

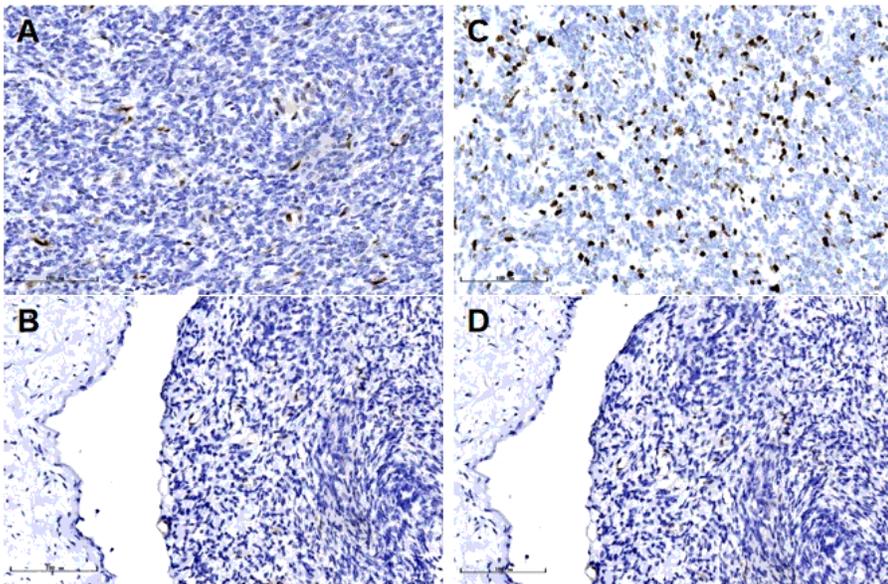


Figure 5

Immunohistochemistry of cyclin D1 and Ki-67. A. cyclin D1(5%) staining in the primary uterine foci (H&E,orig.x200); B. cyclin D1(<1%) staining in metastatic lesions (H&E,orig.x200); C. Ki-67(20%) staining in the primary uterine foci (H&E,orig.x200); D. Ki-67(2%) staining in metastatic lesions(H&E,orig.x200).

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- [checklist.pdf](#)