Low-grade Endometrial Stromal Sarcoma with Intracaval or Intracardiac Extension: A Retrospective Study of 8 Cases

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Abstract

Purpose: This study aimed to improve the knowledge of low-grade endometrial stromal sarcoma (LG-ESS) with intracaval or intracardiac extension and tried to identify the potential risk factors and optimal treatment method influencing prognosis.

Methods: We performed a retrospective review of eight LG-ESS patients with intracaval or intracardiac extension who underwent treatment at Peking Union Medical College Hospital between 2012 and 2020.

Results: The median age at diagnosis was 44 years, ranging from 28 to 56 years. Abnormal uterine bleeding was the most common intimal symptom (3/8), followed by low back discomfort (2/8), edema of the lower limbs (2/8), abdominal pain (1/8), and dyspnea (1/8). All patients underwent resection of the intravascular and extravascular portions of the tumor. Two patients were in stage IIIC, and 6 were in stage IVB. After surgery, 4 patients received adjuvant radiotherapy, of whom 3 also received letrozole. One patient was treated with letrozole alone, and 1 patient received medroxyprogesterone. The average follow-up time was 34.5 months, ranging from 6 to 98 months. No patients died or relapsed during the follow-up period.

Conclusions: LG-ESS with intracaval or intracardiac extension is an uncommon type of tumor which is easily misdiagnosed and can only be diagnosed by histological evaluation after surgery. Complete tumoral excision followed by adjuvant therapy may benefit patient survival time. Long-term follow-up is essential due to the high rate of late recurrence.

Introduction

Endometrial stromal sarcoma (ESS) is an uncommon tumor of the uterus. The incidence of ESS is approximately 1–2/1000000 women per year, accounting for approximately 10% of all uterine sarcomas and 0.2% of uterine malignancies(1). ESS can be separated into low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS) based on mitotic activity(2). LG-ESS tumors have fewer than 10 mitotic figures per high-power field. LG-ESS is a slow-growing neoplasia with a favorable prognosis, and the overall 5-year survival rate is between 80 and 100%. However, it has the characteristic of late recurrence, and approximately 37–60% of patients show late tumor recurrence(3). Usually, lesions in cases of intrauterine, intracaval, or intracardiac extension are highly uncommon and have rarely been reported in previous studies(2). Owing to the atypical presentation, patients with this disease are susceptible to misdiagnosis, especially with intracaval leiomyomatosis. Given the rarity of this disease, the clinicopathologic characteristics and prognosis are not clear.

In this study, we retrospectively investigated the clinicopathologic characteristics and outcomes of LG-ESS patients with intracaval or intracardiac extension and aimed to improve knowledge of this disease. We also tried to identify the potential risk factors and optimal treatment methods influencing prognosis.

Method

Patient population

We retrospectively analyzed LG-ESS patients with cardiovascular, inferior vena cava (IVC) or iliac vessel involvement between January 2012 and December 2020. The inclusion criteria were as follows: (1) patients who were diagnosed with metastatic endometrial stromal sarcoma with iliac veins, IVC or cardiac involvement confirmed by histopathology at Peking Union Medical College Hospital (PUMCH); patients with complete information; and (3) patients without other malignant tumors. During this period, 8 patients were included in our study. Clinical features, manifestations, related examinations, and surgical details were recorded. This study was approved by the Ethics Committee of PUMCH. The need for written informed consent was waived due to the retrospective nature of the study, and all personal information was deidentified to protect patient privacy.

Treatment

All patients underwent gynecological and cardiovascular evaluations preoperatively, postoperatively, and during long-term follow-up. Of all 8 patients, 5 underwent hysterolapingo-oophorectomy as well as resection of intracaval or intracardiac tumors. Resection of intracaval or intracardiac tumors was performed in the remaining cases because they had received hysterectomy before the surgery due to leiomyoma. Radiotherapy, hormonal therapy (HT) or other treatments were also performed after surgery.

Follow-up

After treatment, all patients were regularly followed up for a prolonged period. The follow-up evaluations included assessment of treatment outcomes, tumor markers and abdominopelvic ultrasound. Computerized tomography venography (CT) or computerized tomography venography (CTV) was also performed if necessary.

Results

Demographics and clinical manifestations

The clinicopathologic characteristics of the patients are summarized in Table 1. Among the 8 patients, the median age at diagnosis was 44 years, ranging from 28 to 56 years. Two patients had natural menopause, and 2 had surgical menopause. Abnormal uterine bleeding was the most common intimal symptom (3/8). Other clinical presentations, including abdominal pain (1/8), low back discomfort (2/8), edema of the lower limbs (2/8) and dyspnea (1/8), were also observed. Tumor markers were normal in most patients (7/8). Evaluation of prolactin was found in only one patient. CT or CTV of the chest,

abdomen, and pelvis showed masses in the iliac vessel in patients (Fig. 1). Meanwhile, 6 of them extended toward the inferior vena cava, and 2 extended toward the right atrium and ventricle.

Table 1
Clinicopathologic characteristics of the LG-ESS patients with intracaval or intracardiac extension

No.	Age	Initial symptom	Tumor marker	Tumor Location	Op. approach	FIGO Stage	Histopathology	Adjuvant therapy	Follow-up (m)	Outcome
1	46	None	Normal	IVC	Thrombectomy + Resection of pelvic mass	IVB	LG-ESS	MPA	36	NED
2	56	Abdominal pain	Normal	LVIC	TH + BSO + Thrombectomy + Resection of pelvic mass	IIIC	LG-ESS	Radiotherapy	21	LOST
3	51	Low back discomfort	NA	LVIC	BSO + Thrombectomy	IIIC	LG-ESS	Radiotherapy + Letrozole	42	NED
4	28	Abnormal uterine bleeding/Lower Limb Edema	Normal	IVC	TH + BSO + Thrombectomy	IVB	LG-ESS	Radiotherapy + Letrozole	40	AWD
5	43	Abnormal uterine bleeding	Normal	RV/RA/IVC/RVIC/RVII	TH + BSO + Thrombectomy + Resection of pelvic mass	IVB	LG-ESS	No	20	NED
6	40	Lower Limb Edema	Normal	IVC/BVIC/BVII	TH + BSO + Thrombectomy	IVB	LG-ESS	Radiotherapy + Letrozole	13	NED
7	46	Abnormal uterine bleeding/Low back discomfort	PRL 34	RV/IVC/RIV	TH + BSO + Thrombectomy	IVB	LG-ESS	Letrozole	6	NED
8	41	Lower Limb Edema	Normal	IVC/RVIC	Thrombectomy + Resection of pelvic mass	IVB	LG-ESS	No	98	NED

IVC, inferior vena cava; VIC, venae iliac communis; RV, right ventricle; RA: right atrium, VII: venae iliac interna; TH + BSO, transabdominal hysterosalpingo-oophorectomy; LG-ESS, low-grade endometrial stromal sarcoma; MPA, medroxyprogesterone; NED, no evidence of disease; AWD, alive with disease.

Histopathological results

According to the FIGO staging system, 2 patients were in stage IIIC, and 6 were in stage IVB. Two patients had heterologous elements, of which had leiomyoma differentiation. No pathological evidence of lymph node metastasis was found. For patients who underwent immunohistochemical staining, the positivity rate of estrogen receptor (ER) was 87.5% (7/8), that of the progesterone receptor (PR) was 87.5% (7/8), that of CD10 was 87.5% (7/8), that of desmin was 50% (4/8), that of CD34 was 100% (6/6), and that of SMA was 71.4% (5/7). No one showed positivity for P53 (0/5). The median value of the Ki 67 index was 5% (1–10%).

Treatment regimens

All the patients underwent surgery. Five of them underwent hysterosalpingo-oophorectomy as well as resection of intracaval or intracardiac tumors with or without resection of the pelvic mass. Resection of intracaval or intracardiac tumors was performed in the remaining cases because they had received hysterectomy before the surgery for other reasons. One of them also underwent resection of the residual ovaries. During the surgery, we could see the thrombus extending from the internal and common iliac veins toward the inferior vena cava, some of them even to the right ventricle (Fig. 2). Total hysterectomy with bilateral salpingo-oophorectomy, resection of intracaval or intracardiac tumors, and pelvic mass resection were performed without any residual tumor in most women (7/8). However, a pelvic lesion located in the uterorectal fossa remained in one patient. Abnormal uterine bleeding, edema and other clinical manifestations were improved in all patients postoperatively. After surgery, four patients received adjuvant radiotherapy, and 3 of them received letrozole maintenance treatment. In addition, 1 patient was treated with letrozole alone, and 1 patient received medroxyprogesterone.

Follow-up and outcomes

Patients were followed regularly during our study period, with a mean follow-up time of 34.5 months, ranging from 6 to 98 months. At the last follow-up, 2 patients continued to take letrozole for maintenance treatment, and the other 2 had stopped on their own. During the follow-up period, 1 patient was lost to follow-up after 21 months without signs of disease recurrence. Among the remaining 7 patients, all showed no evidence of disease. One patient was alive with disease because she underwent partial resection of the tumor, and no progression or new metastatic lesion was diagnosed. No patients died or relapsed during the follow-up period.

Discussion

LG-ESS is a rare uterine tumor that commonly spreads to the vagina, pelvis, and peritoneal cavity(4). In this study, we reviewed the demographics, clinicopathological characteristics, treatments and outcomes of 8 LG-ESS patients with intracaval or intracardiac extension, constituting the largest case series collected to date.

Intracaval or intracardiac extension of LG-ESS has rarely been reported, so the pathophysiology is still unclear. Based on previous studies reported in the literature, we hypothesize that the possible underlying mechanisms are as follows: 1) ESS exhibited hematogenous spread to the lung and thereafter to the pulmonary veins and direct extension from the pulmonary veins into the heart(5, 6). At this point, intracardiac and lung metastases could be detected without concurrent abdominal or caval metastasis. 2) Tumor thrombi directly spread through the venous system, stretching through the IVC from the iliac vein to the heart(7–10). Most cases reported, including ours, spread in this way. LG-ESS typically develops in premenopausal and perimenopausal women, with a mean age of 46 (range 18–83) years(11). Older age was thought to be an independent risk factor for OS in some studies(4, 12). In our study, the median age was 44 years, ranging from 28 to 56 years. Most of them were premenopausal or perimenopausal, similar to previous study results.

The clinical presentation of these lesions depends on the localization of the tumor. The typical clinical presentation in more than 90% of cases is abnormal uterine bleeding, which also presents with dysmenorrhea and pelvic pain. Patients with extrauterine spread tend to present with progressive dyspnea, abdominal pain, and lower limb edema. The diagnosis is usually confirmed by imaging modalities such as echocardiography or CT(13). In our study, abnormal uterine bleeding was the most common intimal symptom, and other clinical presentations, including abdominal pain, low back discomfort, edema of the lower limbs and dyspnea, were also observed. Due to the atypical presentation, this tumor is susceptible to misdiagnosis before tumor resection(14). In view of tumor invasion into the major cardiovascular channels, clinical behaviors are similar to those of intravenous leiomyomatosis (IVL), a very rare smooth muscle tumor. IVL is characterized by worm-like extensions into the uterine veins or into other pelvic veins. Tumor growth involving the inferior vena cava in more than 10% of patients with IVL and even cardiac involvement is occasionally seen. Indeed, all cases in our series had a preoperative diagnosis of IVL at the initial treatment. We suggest that the spread pattern of our cases may resemble IVL. The histological diagnosis of LG-ESS was confirmed both in primary tumors and in metastatic sites in all cases. In addition, tumor infiltrations to adjacent organs were observed in our series, while such invasion is seldom seen in IVL.

Many investigators have agreed that imaging examination should be regarded as a diagnostic technique for the preoperative assessment of the extent of neoplastic caval thrombus. Generally, uneven density or low density filling defects are seen in the iliac vein, IVC or heart. In our case, CT or CTV delineated the extent of the thrombus and visualized any residual caval flow (Fig. 1). Thus, imaging could help to guide the scope of surgery. However, differentiation between LG-ESS and IVL according to their enhancement of the characteristics upon imaging modalities is unreliable, and there is no suitable tumor marker for this disease(15). An accurate diagnosis can only be made utilizing histological evaluation(16). Immunohistochemical detection of markers such as CD10, SMA, and desmin can have very good auxiliary use(17). In our research, the positivity rate of CD10 was 87.5%, that of desmin was 50%, and that of SMA was 71.4%, showing value in differentiating ESS from uterine smooth muscle tumors. Therefore, a high suspicion index is required not only to predict the disease but also to monitor recurrence. In addition, a multidisciplinary team consisting of a cardiac surgeon, radiologist and oncologist should be involved(18).

Given the rarity of this kind of disease, the optimal treatment for this condition is still a matter of debate. Surgery represents the most important procedure for the management of patients with ESS. In cases with intracaval or intracardiac extension, the disease condition is particularly complicated and associated with high mortality and postoperative recurrence rates. Herein, surgery for such tumors requires the participation of clinicians from various disciplines. The role of lymphadenectomy in the surgical management of ESS patients is not conclusive. Lymph node involvement among these patients was uncommon, and no differences were found either in recurrence-free survival or in overall survival after performing lymphadenectomy(19–21). In our cohort, all the patients underwent surgery without lymphadenectomy under the direction of gynecologic, vascular and cardiac surgeons. All patients recovered well after surgery, and no recurrence was found after treatment during the follow-up period. Complete resection of the intravascular and extravascular portions of the tumor is mandatory to relieve symptoms and prevent recurrence(22). Whenever possible, radical tumor resection should be attempted. Considering the longer surgical duration and higher risk of massive hemorrhage in a one-stage procedure, two-stage operations were also recommended (23).

In addition to surgery, hormone therapy, radiotherapy and chemotherapy are important adjuvant treatments for LG-ESS, but their effects need to be identified and discussed. Hormonal therapy with progestins, aromatase inhibitors, and gonadotropin-releasing hormone analogs has been considered an effective adjuvant treatment, especially for recurrent tumors, and their use should be guided by the hormone receptor status of the tumor concerned(24, 25). On the one hand, aromatase inhibitors are associated with longer RFS in patients with advanced LGESS, which is better tolerated than progestins and can be primary HT for LG-ESS(26). ER and PR were found in 87.5% of our cases; therefore, letrozole was used in 4 patients. However, the optimal dose, regimen and duration are not well established(27). On the other hand, a recent meta-analysis concluded that HT could only reduce the risk of recurrence in patients with FIGO stage I-II but without conferring any benefit in terms of overall survival(28). Moreover, adjuvant radiotherapy alone appears to improve local control, but the impact on survival is unclear(29). Adjuvant chemotherapy with anthracyclines and ifosfamide may be beneficial in patients with advanced or recurrent hormone-unresponsive tumors(30). Nevertheless, most studies have failed to show a clear survival benefit from adjuvant radiotherapy or chemotherapy, and no standardized or validated guidelines for adjuvant radiotherapy and chemotherapy exist as of yet(31). In our report, four patients received adjuvant radiotherapy, and 3 of them received letrozole maintenance treatment. In addition, one patient was treated with letrozole alone, and one patient received medroxyprogesterone. No patients have relapsed to date. The best combination, providing the best long-term survival, may be complete surgical resection and postoperative adjuvant therapy. However, we still need to expand the sample size for further study to investigate the optimal adjuvant treatment.

The 5-year survival rate for ESS is 54–100% at stage I and 30% at stage II, followed by a dramatic decrease to 11% at stages III and IV(2). However, patients in our study seemed to have a relatively good prognosis even in stage III or IV compared with previous studies. Most patients underwent radical resection of the tumor though a multidisciplinary team and received adjuvant therapy, which may prolong the overall survival time. In addition, the age in our patients was relatively young, and the oldest patient was only 56 years old, which may be associated with a better prognosis. According to a previous study, the risk of recurrence in LG-ESS is 10–20%, and 37–60% of patients show late tumor recurrence(3). Approximately 40% of recurrences were limited to the pelvis, and 60%

occurred as distant metastases or as a combination of all forms of dissemination(32). The median period until the appearance of recurrence is 5.4–9.3 years in stages I and II and only 9 months in stages III and IV(32). Although the patients in our research were in an advanced stage, no patients showed a sign of recurrence during the follow-up period. The possible reason may be that most patients were followed up within 5 years, while late recurrence after more than 10–30 years are characteristic of this disease(11). Thus, long-term follow-up is crucial because of the high rate of late recurrence.

In conclusion, LG-ESS with intracaval or intracardiac extension is an uncommon type of tumor. Due to the lack of effective detection methods, it can easily be misdiagnosed as IVL, and accurate diagnosis can only be made by means of histological evaluation postoperatively. Complete tumoral excision followed by adjuvant hormone therapy provides relatively long-term survival in most patients. Multidisciplinary treatment and regular follow-up for 10 or 20 years after initial diagnosis are necessary due to the high rate of late recurrence.

Declarations

Funding

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Conflict of interest

The authors declare that they have no conflict of interest.

Ethical approval

All procedures performed in the study were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

This research study was conducted retrospectively according to medical record data obtained during hospitalization. We consulted extensively with the IRB of Peking Union Medical College Hospital (PUMCH) who determined that our study did not require informed consent.

Availability of data and material

The dataset supporting this article is included within the article, and additional data are available from the corresponding author upon request.

Author Contribution

Conceptualization: Wang J; Data curation: Chen J, and Shi X; Formal analysis: Chen J and Wang J; Software: Chen J; Writing – original draft: Chen J, and Wang J; Writing – review and editing: Cao D, Yang J, Shen K, and Huang H.

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Figures

Figure 1

Computed tomogram of the chest, abdomen, and pelvis. A large mass extending from the pelvis to the inferior vena cava.

Figure 2

Gross specimens of the resected tumor thrombus. (a) Thrombus extending from the right internal and common iliac veins to the inferior vena cava and the mass in the right ventricle. (b) Uterus, bilateral ovaries and tubes, and the tumor extending from the right uterine vein and common iliac vein to the inferior vena cava.