Case Report

Endometrial stromal sarcoma metastatic to the inferior vena cava: a case report and literature review

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Background: Endometrial stromal sarcoma (ESS) is a uterine stromal tumor with a very low incidence, accounting for 10–15% of all uterine stromal malignancies and 0.2% of all uterine malignancies. The most common extrauterine location of ESS is the ovary, and it is relatively rare outside the uterus. Although most recurrences occur within the pelvis, distant metastases can occur.

Case Description: We report a rare case of low-grade ESS (LG-ESS) metastatic to the inferior vena cava (IVC) which is difficult to distinguish from leiomyoma clinically. A 56-year-old woman attended outpatient complaining right thigh pain. She underwent a surgery of hysterectomy and bilateral adnexectomy 12 years ago. Abdominal contrast-enhanced computed tomography (CT) demonstrated that the vaginal stump was thick, with peripheral multiple nodular shadow. Soft tissue shadow in the right pelvic cavity. Thickening and enhancement of soft tissue shadow were observed in the peripheral blood vessels of the vaginal stump, the right internal iliac vein and the external iliac vein to the IVC of the liver segment. Malignancy (recurrence or metastasis) were considered. After multidisciplinary consultant, a preoperative diagnosis of leiomyomatosis of the IVC was made and surgical treatment was performed. Surgeons performed laparotomy, resection of tumor in IVC, right common iliac vein, right external iliac vein, right internal iliac vein and left common iliac vein. Post-operative pathology of dissected tumor demonstrated LG-ESS. The source may be the ovarian venous stump left after surgery 12 years ago. After a gynecological consultant, chemotherapy is recommended and is currently under follow-up.

Conclusions: We report a rare case of LG-ESS metastatic to the IVC, which was probably a lesion derived from the ovarian venous stump remaining after surgery 12 years ago.

Keywords: Endometrial stromal sarcoma (ESS); inferior vena cava (IVC); chemotherapy; case report

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Introduction

Endometrial stromal sarcoma (ESS) is a uterine stromal tumor with a very low incidence (1), accounting for 10–15% of all uterine stromal malignancies and 0.2% of all uterine malignancies (2,3). After initial appearance it can recur or metastasize many years later. However, the most common extrauterine location of ESS is the ovary, and it is relatively rare outside the uterus (4). Extrauterine pelvic extension of low-grade ESS (LG-ESS) is frequently associated with endometriosis (5). We report a rare case of LG-ESS metastatic to the inferior vena cava (IVC). The source may be the ovarian venous stump left after surgery 12 years ago. After a gynecological consult, chemotherapy is recommended and is currently under follow-up. We present the following case in accordance with the CARE reporting checklist (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-317/rc).

Case presentation

A 56-year-old woman presented in outpatient with right thigh pain. She underwent a surgery of hysterectomy and

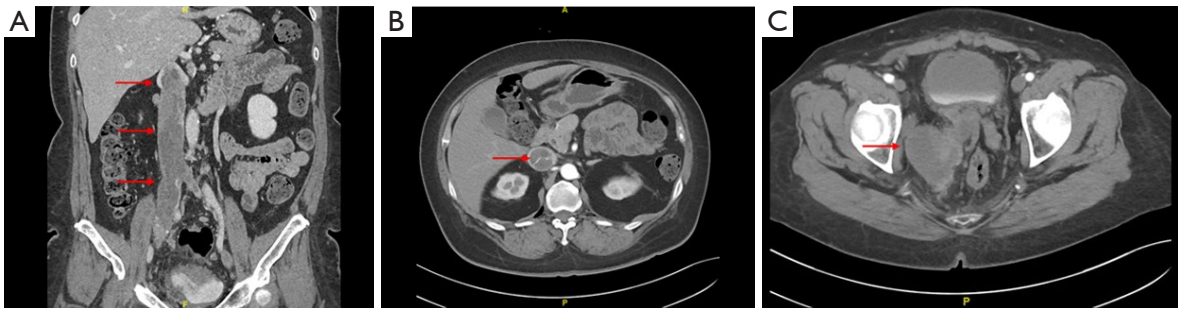

Figure 1 Abdominal contrast-enhanced CT. As indicated by the red arrows, the IVC and iliac vein lumens are dilated with irregular enhanced soft tissue (A,B). The vaginal stump thickened, soft tissue shadow in the right pelvic cavity (C). CT, computed tomography; IVC, inferior vena cava.

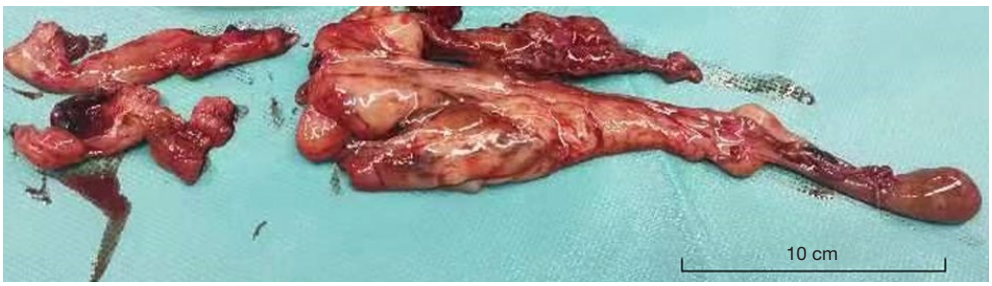

Figure 2 Specimen of resected tumor in IVC, right and left common iliac vein, right external and internal iliac vein. IVC, inferior vena cava.

bilateral adnexectomy 12 years ago. Denying a family history of genetic disease. Laboratory findings were almost normal. A computed tomography (CT) of the lumbar was performed at the local hospital and showed that the diameter of the IVC and its branches was thickened, the density in the lumen was uneven, high-density shadows were visible. Then she was transferred to our hospital for surgery. Abdominal contrast-enhanced CT demonstrated that the vaginal stump was thick, with peripheral multiple nodular shadow. Soft tissue shadow in the right pelvic cavity. Thickening and enhancement of soft tissue shadow were observed in the peripheral blood vessels of the vaginal stump, the right internal iliac vein and the external iliac vein to the IVC of the liver segment. Malignancy (recurrence or metastasis) were considered (*Figure 1*). The patient's vital signs were normal. Physical examination showed that the abdomen was soft and there was no abdominal pain. Mild swelling of the left leg and tortuous dilated veins could be seen on the lateral side of the left leg. Artery pulsation of both lower limbs were palpable and her skin temperature was normal. Laboratory results were not significant abnormalities, and tumor biomarkers such as carcinoembryonic antigen (CEA) did not increase.

Although our hospital's detailed examination did not yield a clear diagnosis, after multidisciplinary consultation, IVC leiomyomatosis was considered and surgical treatment was performed. Surgeons performed laparotomy, resection of tumor in IVC, right common iliac vein, right external iliac vein, right internal iliac vein and left common iliac vein and disconnection of the right internal iliac vein was performed (*Figure 2*). The specimens were sent for histopathology. Postoperative pathology was LG-ESS.

The findings in the specimen did not support endometriosis. No lymph node metastasis was seen. Postoperative investigations confirmed stage IV (6). On histopathological examination, the tumor of IVC was mesenchymal tumor, with abundant interstitial blood vessels, mitotic 0–2/10 HPF. The tumor cells were positive for CD10, SMA, P16, ER, PR, scattered slightly positive for CyclinD1, but negative for Desmin, CD34, CD117, Dog-1, STAT6, S-100 and HMB45 on immunostaining. Ki-67 index was about 8% (*Figure 3*). Combined with immunophenotype and clinical history, a definitive diagnosis of LG-ESS metastatic to the IVC was made. After

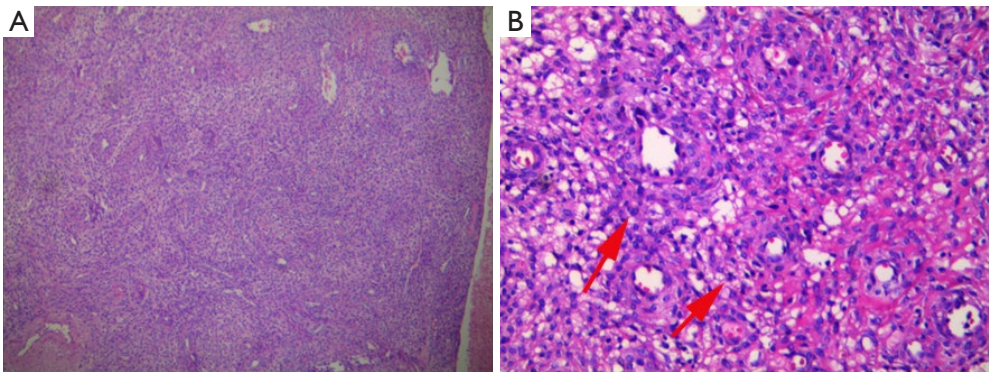

Figure 3 Pathological analysis of the surgical specimen. (A) HE, ×100. (B) HE, ×200. As indicated by the red arrows, the proliferation of endometrial stromal cells surrounded spiral arteriolar vessels, the nucleus of tumor cells are round or spindle with less atypia.

carefully inquired her previous history, she suffered from LG-ESS 12 years ago. The diagnosis was LG-ESS with extrauterine extension into the bilateral ovarian venous vessels, leiomyomas and adenomyosis. Hormonal treatment was performed for 2 years. She recovered well, then she was not followed up. Therefore, we believed that the disease might have originated from the stump of the ovarian vein left after surgery 12 years ago. The patient improved and was discharged from the hospital on the 22nd postoperative day without obvious postoperative complications. Further to this, after gynecology consultation, she was referred for chemotherapy due to the incomplete removal of tumor. She felt the surgery was successful and decided to continue her gynecological treatment. And now she is on follow-up.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

We reported a case of ESS which metastasized to the IVC and the disease is difficult to distinguish from leiomyoma clinically. According to the current WHO classification, uterine sarcomas is divided into the following types: endometrial stromal nodule (ESN), LG-ESS, high-grade endometrial stromal sarcoma (HG-ESS), undifferentiated uterine sarcoma (UUS) (7). LG-ESS is a slow-growing indolent disease that mainly affects perimenopausal women, especially women aged 40–55 years, and sometimes occurs in elderly or younger women (8,9). Although most recurrences occur within the pelvis, distant metastases can occur. According to previous reports, the main extrauterine sites of ESS include the ovaries, bowel wall, peritoneum, sigmoid colon, pelvis, urinary bladder and vagina (8,10-12). After initial appearance it can recur or metastasize many years later and the risk of recurrence is 10–20% (1). The primary treatment for LG-ESS is total hysterectomy and bilateral adnexectomy, which can achieve an increased cure and long-term survival (13,14). The benefits of lymphadenectomy and tumor debulking are unclear (13,15). It is commonly associated with obesity, diabetes, younger age at menarche, estrogen intake, pelvic radiation and endometriosis (12,16,17). Microscopically, LG-ESS resembles the proliferative phase of endometrial stroma featuring small cells with oval to spindle nuclei ordered in sheet surrounded by spiral arteriole like vessels. The immunohistochemistry is positive for CD10, wt-1, vimentin, actins, interferon induced transmembrane protein 1 (IFITM1), estrogen, androgen, and progesterone receptors (18). It is difficult to make a diagnosis before operation if it develops at unusual sites. If the patient has a history of ESS and considers ESS recurrence, the biopsy and immunostaining may be useful for diagnosis. The risk of recurrence of LG-ESS is 10–20%, characterized by late recurrence over 10–30 years (9). Some studies report that ESS is a hormone-dependent malignancy and postoperatively hormonal treatment in form of progesterone, gonadotropin-releasing hormone (GnRH) analogues and aromatase inhibitors may have activity as maintenance therapy in recurrent disease. However, the benefits are unclear because the small number of patients

(13,19). Postoperative radiotherapy in patients with LG-ESS only provides locoregional control, but long-term side effects of pelvic irradiation need to be considered carefully (13).

Conclusions

We report a rare case of LG-ESS metastatic to the IVC, which was probably a lesion derived from the ovarian venous stump remaining after surgery 12 years ago. After gynecology consultant, she was referred for chemotherapy and currently under follow-up.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-317/rc

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-317/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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