Uterus didelphys complicated with endometrial carcinoma

A case report of uterus didelphys with endometrial carcinoma

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Abstract

**Rationale:** The incidence of uterine malformations is low (4%–7%). Currently, the National Comprehensive Cancer Network clinical practice guidelines in oncology recommend minimally invasive surgery for early endometrial cancer. Minimally invasive surgery for the treatment of uterus didelphys with endometrial cancer is rare due to the large size of the uterus. To date, only 2 such patients have been reported to have undergone laparoscopy. Whether such patients can be treated with minimally invasive surgery needs to be further explored.

**Patient concerns:** A 40-year-old woman with uterine didelphys was hospitalized for menorrhagia in the past 2 months.

**Diagnosis:** Endometrial adenocarcinoma was found in both the uterus and cervix using fractional dilation and curettage.

**Interventions:** The patient underwent laparoscopic surgery. Postoperative adjuvant radiotherapy and chemotherapy were administered.

**Outcomes:** There was no sign of recurrence during routine follow-up.

**Lessons:** The use of a uterine manipulator to lift either side of the uterus could help to expose the narrow ipsilateral para-uterine field. It is difficult to remove the uterus entirely through the vagina, making it necessary to select appropriate cases wherein screening is performed to check if the vagina is loose, and the uterus is of appropriate size. Minimally invasive surgery may be feasible for suitable patients.

**Abbreviation:** EC = endometrial carcinoma.

**Keywords:** endometrial cancer, laparoscopy, uterine didelphys

1. Introduction

The overall incidence of uterine anomalies is low and the clinical diagnosis of a specific malformation is often difficult and confusing.\textsuperscript{1} Uterine didelphys may often be misdiagnosed as bicornate/mediastinal uterus. Uterine didelphys occur when the bilateral Müller tubes completely fail to become proximal and fuse with each other and instead continue to develop individually, thus forming 2 uterine cavities, 2 cervixes, and often 2 vaginas separated by the longitudinal diaphragm and with normal menstrual flow. A bicornuate uterus occurs when the fusion of 2 Müllerian structures fails, resulting in 2 uterine horns and only 1 cervix. The mediastinal uterus is the result of insufficient reabsorption of the septum after fusion of Muller’s structure.

In developed countries, the most common malignant tumor of the female reproductive system is endometrial carcinoma (EC). In China, its incidence is second only to cervical cancer but...
shows a significant upward trend. Randomized controlled trials and meta-analyses have shown that compared to laparotomy, laparoscopy, a minimally invasive treatment, offers the benefits of a lower incision infection rate, lower blood transfusion rate, lower venous thrombosis incidence, shorter hospital stay, higher quality of life, and no difference in tumor-related prognosis. Therefore, laparoscopy is the primary recommendation of the National Comprehensive Cancer Network clinical practice guidelines in oncology for early endometrial cancer.\(^{[2–6]}\) Uterine didelphys complicated by EC are rare. Furthermore, enlargement of uterine didelphys makes the use of minimally invasive surgery even more challenging. To date, only 2 cases involving uterine didelphys have been reported. Here, we report a case in which laparoscopy was successfully performed for uterine didelphys complicated by EC.

### 2. Case report

A 40-year-old woman was hospitalized for menorrhagia over the previous 2 months. She did not have any other diseases and had no particular family history. The patient was married and had a childbearing history of G0P0L0A0. Computed tomography showed uterine didelphys, thickened endometrium in the uterine and cervical canals, mild-to-moderate enhancement in the right endometrium, and small lymph nodes in the abdominal cavity and retroperitoneum (Fig. 1). Gynecological examination revealed a normal vulva and urethral orifice, an unobstructed vagina, and smooth mucosa. A double cervix was observed above the vagina and both cervices had a diameter of approximately 2 cm. The pathology of fractional dilation and curettage was as follows: left uterine cavity, moderately to highly differentiated endometrioid carcinoma; right uterine cavity, moderately differentiated endometrioid carcinoma; left cervical canal, well-differentiated endometrioid carcinoma; and right cervical canal, moderately to highly differentiated endometrioid carcinoma. Two diagnoses were established, EC and uterine didelphys.

On March 23, 2018, surgery was performed under general anesthesia. Fractional dilation and curettage before surgery indicated that both the uterus and cervical canal showed cancer involvement; therefore, modified radical hysterectomy was performed. The patient underwent laparoscopic modified radical hysterectomy, double adnexal resection, pelvic lymphadenectomy, and abdominal para-aortic lymphadenectomy (Fig. 2).

Postoperative gross pathology findings were as follows (Fig. 3): the volume of the right uterus was \(10.5 \times 8 \times 4.5 \text{ cm}\). The area in the uterine cavity showing grayish-red erosion was approximately \(6 \times 4 \text{ cm}\). The volume of the left uterus was \(9 \times 7 \times 4 \text{ cm}\), and the rough endometrial surface area was grayish-white or grayish-red, measuring approximately \(4 \times 2 \text{ cm}\). Routine pathological diagnosis was as follows: the right uterus showed a differentiated endometrioid adenocarcinoma, which invaded the superficial myometrium and involved the internal orifice of the cervical canal. The left uterus showed differentiated endometrioid adenocarcinoma invading the superficial myometrium but not the internal orifice of the cervical canal. The cervix showed bilateral, chronic mucosal inflammation. No lymph node metastasis was observed. The immunohistochemical status was 70% estrogen receptor (+) and 70% progesterone receptor (+).

A postoperative pathological diagnosis of stage I and G2 endometrial adenocarcinomas was made. Postoperatively, paclitaxel + carboplatin combination therapy was administered for 4 cycles simultaneously with afterloading radiotherapy. The patient was routinely followed up by telephone in December 2021. She underwent computed tomography examination in 2020, wherein she showed no signs of tumor recurrence or obvious complications.

The findings during the operation were as follows: The space between the uterine isthmus/cervix and the posterior wall of the bladder was dense and unclear. The uterus and bladder were separated to expose the cystocervical space mainly via sharp separation. Since both uterine cavities are large and occupy the majority of the pelvic operation field, it is difficult to lift the bladder and expose the para-uterine space. The left uterus was lifted to expose and resect the left para-uterine tissue and the right uterus was lifted to expose and remove the right uterine tissue. This effectively solved the problem of limited visual field exposure. After resection of the bilateral pelvic funnel ligaments, only the left uterus became ischemic and discolored when severing the left uterine artery. This suggests that unilateral uterine arteries supply only the uterus along their corresponding sides. After complete resection of both uteri, removal of uterine didelphys through the vagina was challenging. Thus, we deflected the uterus and first pulled out the left uterine horn of the left uterus, removed the small uterus on the left, and successfully removed the larger uterus on the right.

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**Figure 1.** CT findings of the uterus. (A) CT shows bilateral enlargement of the uterine body. (B) Bilateral uterine fusion and adhesion to the bladder wall. (C) Bilateral cervix and bilateral uterine arteries. CT = computed tomography.
3. Discussion

The incidence of uterine malformations is low (4%–7%),[9,10] and there is no evidence that uterine malformations lead to an increase in the incidence of EC. As fractional dilation and curettage revealed endometrioid adenocarcinoma in both uteri, complete resection of both uteri was performed. We reviewed published articles on uterine malformations complicated by endometrial cancer in PubMed from January 1, 1990 to June 30,
2021, and screened the articles on double uterine malformations. We found only 18 cases of uterine didelphys complicated by malignant tumors, including 15 cases of EC, 2 cases of carcinosarcoma, and 1 case of sarcoma. The incidence of bilateral uterine involvement was even lower, with only 3 cases reported in the literature, including 1 case of sarcoma. In our patient, diagnostic curettage revealed involvement of both the uterine cavity and cervix; therefore, modified radical hysterectomy was chosen. For uterine didelphys, diagnostic curettage should avoid missing 1 side of the diseased uterus, which may result in missed diagnosis. Patients with uterine didelphys often present with double vaginas, and special attention should be paid to identifying exceptions. In patients with bilateral cervical involvement, cervical tube curettage should be performed separately to avoid missing lesions.

The use of laparoscopy, because of its minimal invasiveness, is advantageous in EC and offers a prognosis comparable to that of laparotomy. It is also associated with lesser trauma, faster recovery, and fewer intestinal adhesions, making it superior to open surgery. Our patient was satisfied with the minimally invasive surgery and had a short postoperative recovery period. After a thorough literature review, we found that only 1 case each of robotic and laparoscopic surgeries has been reported. This may be attributed to the uterine didelphys being generally large and thus difficult to remove through the vagina. Despite the challenge of the enlarged uterus, we pulled it out completely through the vagina. This proves that minimally invasive surgery can be performed after screening to determine whether the vaginal fornix is loose. In our case, there were some complications with laparoscopy: both uteri were large, the field of vision was narrow and difficult to expose, and lifting the uterus was inconvenient. However, a uterine manipulator was used to lift 1 side of the uterus and move it toward the opposite side to expose and remove ipsilateral para-uterine tissue. This could effectively solve the problem of exposure difficulties in the bilateral para-uterine fields. Recently, we performed laparoscopic hysterectomy in another patient with uterine didelphys and only 1 kidney. The patient was married and had a childbearing history of G1P1L1A0. This patient had adenomyosis in both uteri that was complicated by endometriotic cysts in both ovaries. The patient experienced severe dysmenorrhea and required surgery to relieve her symptoms. During the surgery, the visual field was as difficult to expose as in the previous patient. We used the above-mentioned method to solve this problem, which was using uterine manipulator to lift 1 side of the uterus to expose the ipsilateral para-uterine field.

During the surgery, we found that 1 side of the uterine artery was supplied to only 1 side of the uterus in this patient with uterine didelphys. In young patients wishing to retain reproductive function, if only 1 side of the uterus is affected by endometrial cancer, it may be possible to cutoff the artery supplying the affected side and resect the affected side of the uterus. Jiao et al reported a 28-year-old infertile woman with uterine didelphys who tried to preserve the uterus even after the failure of megestrol treatment. The patient underwent transabdominal surgery to remove the left uterus and fallopian tube, thereby preserving the contralateral uterus and appendages. This relationship was also verified in another patient with uterine didelphys. This preliminarily confirms that the embryonic stage of the uterine didelphys is formed separately, and that it is feasible to retain 1 side of the uterus and uterine artery.

Minimally invasive surgery has the benefits of reduced trauma and a faster postoperative recovery. To check if patients with uterine malformations are eligible for minimally invasive surgery, we suggest that the following conditions should be met: the vaginal fornix should be loose and the uterus should be of appropriate size. Understanding the different anatomical characteristics and careful cooperation of the entire team are also warranted.

Author contributions

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References

[1] Szpera-Goździejewicz A, Gruca-Stryjak K, Bęborowicz GH, Ropacka-Lesiak M. Uterine arteriovenous malformation - diagnosis and management. Ginekol Pol 2018;89:276–9.
[2] Scalici J, Laughlin BR, Finan MA, Wang B, Rocconi RP. The trend towards minimally invasive surgery (MIS) for endometrial cancer: an ACS-NSQIP evaluation of surgical outcomes. Gynecol Oncol 2015;136:512–5.
[3] Janda M, Gebski V, Davies LC, et al. Effect of total laparoscopic hysterectomy vs total abdominal hysterectomy on disease-free survival among women with stage I endometrial cancer: a randomized clinical trial. JAMA 2017;317:1224–33.
[4] Fader AN, Weise RM, Sinno AK, et al. Utilization of minimally invasive surgery in endometrial cancer care: a quality and cost disparity. Obstet Gynecol 2016;127:91–100.
[5] Galaal K, Donkers H, Bryant A, Lopes AD. Laparoscopy versus laparotomy for the management of early stage endometrial cancer. Cochrane Database Syst Rev 2018;10:CD006655.
[6] Kornblith AB, Huang HQ, Walker JL, Spirtos NM, Rotmensh J, Cellia D. Quality of life of patients with endometrial cancer undergoing laparoscopic international federation of gynecology and obstetrics staging compared with laparotomy: a Gynecologic Oncology Group study. J Clin Oncol 2009;27:5337–42.
[7] Walker JL, Piedmonte MR, Spirtos NM, et al. Laparoscopy compared with laparotomy for comprehensive surgical staging of uterine cancer: Gynecologic Oncology Group Study LAP2. J Clin Oncol 2009;27:5331–6.
[8] Mannschreck D, Matsumo RK, Moriarty JP, et al. Disparities in surgical care among women with endometrial cancer. Obstet Gynecol 2016;128:526–34.
[9] Gao J, Zhang J, Tian W, et al. Endometrial cancer with congenital uterine anomalies: 3 case reports and a literature review. Cancer Biol Ther 2017;18:123–31.
[10] Lei C, Huang M, Li N, An J, Xiong S, Xu Y. Intensity-modulated radiotherapy combined with intracavitary brachytherapy for locally advanced cervical cancer with uterus didelphys. Gynecol Oncol Rep 2021;36:100724.
[11] Martínez-Beltrán M, Gíménez J, Acín P. Uterus didelphys with septate cervix and unilateral endometrial carcinoma: a case report. J Genit Syst Disor 2012;1:
[12] Vázquez Vicente D, Di Fiore HA, Garcia-Foncillas J, Plaza Atranz J. Endometrial adencarcinoma in one horn of a didelphys uterus with vaginal duplication. BMJ Case Rep 2014;2014:bcr2013203280.
[13] Vanichchantkul A, Huang KG, Hsu CC. Endometrial carcinoma arising in one horn of a didelphys uterus. Taiwan J Obstet Gynecol 2020;59:162–4.
[14] Tsukahara Y, Fukumatsu Y, Tomita K, Shiozawa T, Inuma H, Fukuta T. Endometrial carcinoma arising from a double uterus. Gynecol Obstet Invest 1990;29:311–2.
[15] Molpus KL, Puleno JG, Williams AM, Bernal KL, Remmenga SW. Endometrial adenocarcinoma within a single horn of a didelphys uterus: a report of 2 cases. J Reprod Med 2004;49:123–5.
[16] Suprasert P, Khunamornpong S. Carcinosarcoma arising in uterine didelphys after tamoxifen therapy for breast cancer: a case report. J Med Assoc Thai 2010;93:608–12.

[17] Kunos C, Woods C, Colussi VC, Abdul-Karim FW, Waggoner S. Low-dose-rate brachytherapy for treatment of uterine didelphys malignancy. J Clin Oncol 2011;29:e104–6.

[18] Bhalla R, Evans H, Berger L, Crow J, Deheragoda M, Taper Y. A uterus didelphys bicornis, with endometrial cancer in both uteruses. J Obstet Gynaecol 2005;25:823–5.

[19] Kosinski A, Dini M. Endometrial cancer in a double uterus. A report of two cases. J Reprod Med 1994;39:926–7.

[20] Holub Z, Shomani A. Uterine reduplication, unilateral ureteral and renal aplasia syndrome associated with endometrial cancer: a case report. Eur J Gynaecol Oncol 1998;19:573–6.

[21] Kondi-Pafiti A, Spanidou-Carvouni H, Dimopoulou C, Kontogianni CI. Endometrioid adenocarcinoma arising in uteri with incomplete fusion of Mullerian ducts. Report of three cases. Eur J Gynaecol Oncol 2003;24:83–4.

[22] Chen CY, Yen MS, Yang MJ, Wu YC. Uterus didelphys with adenocarcinoma in the right cavity diagnosed by 2-dimensional sonography and magnetic resonance imaging. J Ultrasound Med 2008;27:1802–3.

[23] Fanfani F, Fagotti A, Restaino G, Guerriero M, Scambia G. Endometrial cancer arising in both horns of didelphys uterus in a Down’s syndrome woman. Gynecol Oncol 2006;101:537–9.

[24] Iavazzo C, Kokka F, Sahdev A, Singh N, Reynolds K. Uterine carcinosarcoma in a patient with didelphys uterus. Case Rep Obst Gynecol 2013;2013:401962.

[25] Yu J, Shang J, Wen H, Xu Y. Fertility preservation in patients with uterus didelphys and endometrial carcinoma: a case report. BMC Womens Health 2021;21:319.

[26] Kobayashi M, Kobayashi H, Nakayama S, Adachi H. Robot-assisted laparoscopic hysterectomy for endometrial cancer in a patient with Herlyn-Werner-Wunderlich syndrome. BMJ Case Rep 2021;14:e240001.

[27] Sassine D, Moufarrij S, Hodgson A, et al. Case report: Sentinel lymph node mapping of endometrial carcinoma occurring in uterine didelphys. Gynecol Oncol Rep 2021;36:100769.