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CASE REPORT

Endometrial Carcinoma Arising in Didelphic Uterus Accompanied by Right Renal Agenesis: A Case Report

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MÜLLERIAN duct anomalies (MDA) are abnormalities occurring in the müllerian duct due to abnormal development of the uterus, cervix and vagina. Reported prevalence of this malformation in general population was 4%-5%. But real figure may be greater because of unawareness of these diseases due to its asymptomatic nature.¹

Bicornuate and didelphic uteri are two common forms of MDA, with the incidence of 25% and 11%, respectively,² and the latter is usually associated with an absent, hypoplastic or duplicated kidney on the side of obstruction. However, endometrial carcinomas arising in didelphic uterus are rarely reported according to our literature searches from both PubMed and domestic publication databases. Here we report an unusual case with endometrial carci-

noma in right horn of didelphic uterus accompanied by ipsilateral renal agenesis. The combination of müllerian anomaly and malignant tumor posed a great challenge to clinicians in terms of diagnosis and treatment.

CASE DESCRIPTION

A forty-four-year-old, middleweight (height 162 cm, weight 51 kg) woman with a five-month history of paroxysmal vaginal drainage was referred to the department of Gynecological Oncology, Zhongnan Hospital, Wuhan University (Wuhan, China) in pursuit of a definite diagnosis and thorough treatment. She had once Gravidia 1 in the left cornu of her uterus, Para1 through cesarean delivery, and still had regular menstrual periods and capacity. Before admitted to hospital, she took antibiotic agents by herself but there was not any effect. She did not smoke or drink alcoholic beverages. History of medical diseases, previous surgeries, and oral contraceptives were

denied. There was no family history of breast cancer or any other malignancy either.

On admission, gynecologic examination showed one vagina and two cervixes out flowing thin serous secretions without stench. Pelvic two-dimensional ultrasound revealed didelphic uterus. The thickness of the two cornu endometria was 9 mm (left) and 14 mm (right), respectively. The endometrium of the right one was 10 mm thick three months earlier. Ultrasound imaging indicated neoplasms in the right uterine cavity and no abnormality in the left one. Abdominal computed tomography (CT) imaging found absence of right kidney. Endometrial biopsy of the lesion in the right uterus was performed and revealed endometrioid adenocarcinoma histologically. Based on the results of examinations above, the final diagnosis was established as endometrioid adenocarcinoma arising in didelphic uterus accompanied with right renal agenesis, which is clinical rare.

Preoperative laboratory tests revealed normal blood indexes and liver and kidney functions. Given the situation, total abdominal hysterectomy, bilateral salpingo-oophorectomy and staging (bilateral and pelvic lymph node dissection, pelvic washings) were scheduled and performed. Gross examination showed a bicornuate uterus with two cervixes, bilateral fallopian tubes and ovaries. The left cornu measured 6.0 cm×3.5 cm×2.0 cm and the right one 6.5 cm×5.0 cm×2.0 cm (Fig. 1A). On uterine sagittal slice, multiple irregular

polypoidal masses occupied the whole uterine cavity of the right horn, additionally, of the left horn, there was a small mass seen on the top of the isthmus uteri (Fig. 1B), which was not detected preoperatively in ultrasound. Rapid intraoperative pathological diagnosis of the uterus indicated malignant neoplasm of endometrium (right) and benign polyps (left).

Postoperative histopathological outcomes of multiple punch sampling further demonstrated a well-differentiated endometrioid adenocarcinoma with superficial myometrium invasion. No metastasis of lymph node, and no invasion of the ligament and blood vessels occurred. The vaginal cutting margin or upper part of cervix was all free from tumor cells. Immunohistochemical stainings for estrogen receptor, progesterone receptor and P53 were all positive. The positive expression rates of Ki-67 in endometrial cancer tissue were 60%. No malignant cell was found in the pelvic washings. The disease was consistent with FIGO stage IA (Figs. 1C and 1D).

Although close eyes were kept on the patient for postoperative care, uroschisis did occur two days after the surgery. The reason was considered to be our early pulling out of the catheter which was inserted preoperatively since the patient's function of automatic micturition was decreased then. We reinserted the catheter and urinary retention was alleviated. The patient recovered from surgery three weeks later and took adjuvant therapy subsequently.

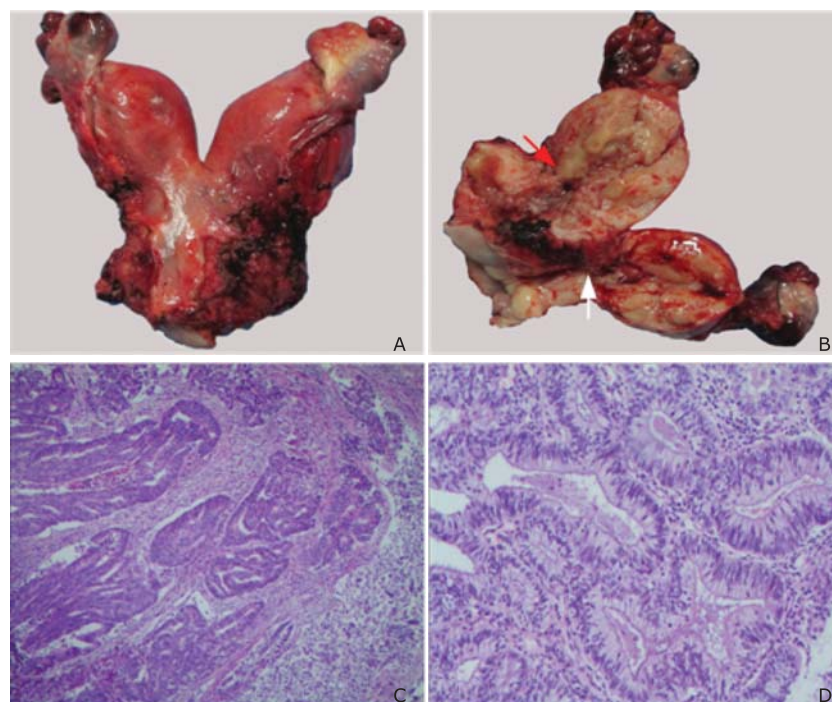


Figure 1. Photograph of the external surface of the bicornuate uterus (A). Photograph of the bivalved specimen of didelphic uterus: endometrial carcinoma in right horn (red arrow) and benign polyps in left uterine horns (white arrow) (B). Photomicrograph of the endometrioid-type component of the endometrial carcinoma (C, HE staining, ×40). Photomicrograph of the endometrioid-type adenocarcinoma (D, HE staining, ×200).

DISCUSSION

Several techniques are available for the evaluation of uterine malformations including the widely used pelvic ultrasound, CT, colposcopy and hysteroscopy. Although these modalities have been proved to be appropriate and effective to detect urogenital malformations,³ accurate diagnosis still could be a dilemma, as most patients with vaginal drainage usually menstruate regularly and prefer anti-inflammatory therapies. In such cases, a combination of physical examination and ultrasound is usually essential to confirm diagnosis. Meanwhile, pathological biopsy serves as gold standard to make a definite diagnosis. Moreover, application of CT as well as magnetic resonance imaging has been well established and the accuracies were reported as high as 100% in the diagnosis of uterine malformations.^{4, 5} To the best of our knowledge, fractional dilatation and curettage are the most common and valuable methods to diagnose endometrial carcinoma. Considering that the patient had bicornuate uterus, it seems reasonable to assume that there was half chance of obtaining a benign biopsy result which actually means missing the cancerous lesion. Accordingly, utilizing multi-means to locate the endometrial cancer in didelphic uterus is necessary to reduce the risk of delayed and/or inadequate diagnosis.⁶

Currently, the treatment for endometrial cancers with didelphic uterus is still the same as those with normal uterus. However, in our direct experience, management of this complex congenital anomaly is of great challenge and requires careful anatomic consideration, since the enlarged cervical canal connects closely with adjacent organs, making the operation more difficult to perform. Therefore, surgeons should be careful to avoid damaging ureters, bladder and rectum when separating conjunctive tissues, particularly for those with only one nephric duct.

Specialized nursing care affects both early and late outcomes of patients after surgery, so more attention should be given to prevent from postoperative complications. The patient in this report developed uroschisis in two days after the removal of catheter, causing the adverse recovery of functions of bladder. The reason may lie in an underestimation of the complexity of such congenital urinary malformations and early pulling out the catheter. A lesson we learned in the management of this case is that more comprehensive assessment should have been made to prevent complications, such as uroschisis in this case. However,

the mechanisms underlying the postoperative bladder function recovery in urogenital malformation, as well as their effects on uroschisis, have not yet been fully elucidated. Further investigations are necessary to clarify the influence of urinary organs agenesis on the postoperative outcomes of uroschisis.

Intriguingly, a research with long-term follow-up suggested that didelphic uterus was not associated with a higher incidence of endometriosis or genital neoplasm, whereas the structural malformations of uterus may increase the risk of developing endometrial carcinoma.⁷ Presumably this is because of the higher amount of the hormone receptors in two endometrial cavities than that in one, and the poor sensitivity to hormonal change in bicornuate uterus.

The case we report here raises issues of diagnosis, therapy and biopsy that need to be addressed today and helps better understanding of this uncommon disease in the future.

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