

## Surgical management of complex atypical endometrial hyperplasia in a woman with rare genitourinary anomalies: unicornuate uterus with rudimentary horn, ipsilateral ectopic ovary and pelvic kidney

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**SUMMARY:** Surgical management of complex atypical endometrial hyperplasia in a woman with rare genitourinary anomalies: unicornuate uterus with rudimentary horn, ipsilateral ectopic ovary and pelvic kidney

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*Aim.* According to the so far published literature, only one case of endometrial cancer in a patient with unicornuate uterus has been reported. This is a case report study, presenting a rare case of complex atypical endometrial hyperplasia in a woman with unicornuate uterus and multiple genitourinary anomalies.

*Case report.* A 43-year old G1P1 woman presented with episodes of menorrhagia and anemia. She had previous surgical history of laparoscopy due to infertility, in which she was diagnosed

with unicornuate uterus with a rudimentary left uterine horn and ipsilateral ectopic ovary in the anatomic place of the left kidney. Dilatation and curettage was performed. Histology showed complex atypical endometrial hyperplasia. The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy, in an extremely interesting operation due to the multiple genitourinary anomalies. The uterus with a 6-centimeter uterine myoma and the adnexae were removed en block. Great effort was put into dissecting the left fallopian tube which arised from the cervix and via the rudimentary horn led to the left ectopic ovary that was located at the left kidneys' anatomic space. The patient recovered well and final histology was negative for malignancy.

*Discussion.* All necessary imaging examinations have to be scheduled prior to surgical intervention in order to give valuable anatomic information in cases of women diagnosed with Mullerian abnormalities.

KEY WORDS: Endometrial hyperplasia - Unicornuate uterus - Rudimentary horn - Ectopic ovary - Pelvic kidney.

### Introduction

Congenital anomalies of the female genital tract are defined as deviations from the normal anatomy arising from embryological maldevelopment of the Mullerian or paramesonephric ducts (1). The prevalence of uterine congenital malformations in the general population is estimated between 4-7% (2-4). In populations with obstetrical history of miscarriage, infertility or recurrent miscarriage the prevalence of uterine anatomic anomalies is even higher (2, 3). However, it is true that the incidence of Mul-

lerian anomalies seems underestimated as they remain clinically silent.

The clinical manifestation of Mullerian duct anomalies includes women with a. either obstructive symptoms (primary amenorrhea and pain or progressive dysmenorrhea), or b. without symptoms (non-obstructive reproductive tract anomalies) who have often been referred to a gynecologist due to infertility, miscarriage or recurrent miscarriage. Also, the association between Mullerian duct anomalies and premature delivery or breech presentation is well documented.

Mullerian anomalies have not been considered as a significant risk factor for the development of gynecological cancer. The association between endometrial cancer and uterine congenital anomalies is extremely weak, with only 25 cases reported during the past 26 years (1). According to the so far pub-

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lished literature, only one case of endometrial cancer in a patient with unicornuate uterus has been reported (5). This is a case report study, presenting a rare case of complex atypical endometrial hyperplasia in a woman with unicornuate uterus and multiple genitourinary anomalies that made surgical intervention extremely interesting.

## Case report

A 43-year old G1P1 woman presented with episodes of menometrorrhagia and anemia. She had previous surgical history of laparoscopy due to infertility, performed by the same surgical team, in which she was diagnosed with unicornuate uterus with a rudimentary left uterine horn and ipsilateral ectopic ovary in the anatomic place of the left kidney. Later magnetic resonance imaging (MRI) and cystography, also revealed left ectopic pelvic kidney. After in vitro fertilization (IVF) she achieved twin pregnancy with delivery at term via caesarean section, eight years before.

Dilatation and curettage were performed due to menometrorrhagia. Histology showed complex atypical endometrial hyperplasia. The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. The uterus with a 6-centimeter uterine myoma and the adnexae were removed en block. Great effort was put into dissecting the left fallopian tube which arised from the cervix and via the rudimentary horn led to the left ectopic ovary that was located at the left kidneys' anatomic space (Figure 1). The procedure was performed after surgical incision of the left parametrium, with special interest in protecting the left ectopic pelvic kidney and its artery from iatrogenic trauma.

The patient recovered well and final histology was negative for malignancy.

## Discussion

Endometrial cancer is considered as the most common neoplastic disease of the female genital tract in developed countries. Approximately 75% of



Figure 1 - Surgical specimen. A: cervix, B: unicornuate uterus, C: uterine myoma, D: right ovary, E: left fallopian tube, F: rudimentary horn, G: left ovary.

patients with endometrial cancer are diagnosed in early stages (FIGO stages I or II), and 5-year overall survival is estimated at 74-91% (1, 6, 7). On the other hand, the incidence of endometrial cancer and complex atypical endometrial hyperplasia in women with congenital abnormalities of the female genital tract is extremely low, as only few cases have been reported during the last decades.

This epidemiologic observation has led to the investigation of potential differences in the expression of the disease between women diagnosed with Mullerian abnormalities in comparison with patients having a normal uterus. It seems that, compared to the endometrial cancer of a normal uterus, their epidemiologic characteristics, risk factors and histological parameters, are similar (1).

Anomalies of the urinary tract are the most common abnormality associated with congenital malformations of the female reproductive tract, as the development of normal Mullerian ducts occurs in association with the normal development of the mesonephric ducts. The incidence of associated genital anomalies in women with renal anatomic abnormalities is estimated between 25-89% (8). The commonest abnormality of the urinary tract is renal agenesis (31.8%) and most frequent in association with didelphys uterus (9).

In our case, the presence of left ectopic pelvic kidney that was already known after MRI and cystography made the operation challenging. Surgical incision of the left parametrium space was performed prior to the hysterectomy, in order to protect the left ectopic kidney and its artery from iatrogenic trauma. It is characteristic that the left renal artery supplied the left ovary that was located at the left kidneys' space via the left ovarian artery. As a result, great effort was put into appropriate ligation of the left ovarian artery without restriction of the blood supply of the left pelvic kidney through the left renal artery. Additionally, the left ureter and left fallopian tube were carefully dissected. The left fallopian tube arised from the cervix and - via the rudimentary horn - led to the left ectopic ovary that was located at the left kidneys' anatomic space.

## Conclusions

Though rare, gynecological malignancies and atypical endometrial hyperplasia can be observed in women with congenital abnormalities. All necessary imaging examinations have to be scheduled prior to surgical intervention in order to give valuable anatomic information. Detailed imaging of the urinary tract is needed in cases of Mullerian abnormalities, especially before major operations.

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### Conflicts of interest

All Authors declare no conflicts of interest.

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