# Endometrioid adenocarcinoma arising in uteri with incomplete fusion of Mullerian ducts. Report of three cases

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**Summary**

The clinicopathological findings of three cases of endometrial adenocarcinoma arising in uteri with developmental anomalies are described.

*Key words:* Uterus; Didelphys; Bicornis; Adenocarcinoma; Polyps.

## Introduction

The most common developmental abnormality of the uterus results from failure of fusion of a portion or of the entire length of the lower Mullerian ducts.

The incidence of congenital malformations of the upper part of the female genital system in less than 0.2% of the female population. In a selective series such as a hysterosalpingography study, 8.6% of individuals were discovered to have a uterine malformation [1, 2].

If a uterine malformation is identified in childhood it may be part of a polyphenotypic syndrome with genitourinary tract anomalies [3]. In women without chromosomal abnormalities the most usual malformations consist of incomplete fusion of the Mullerian ducts [4-7].

Etiologic factors include fetal exposure to diethylstilbestrole or thalidomide, environmental factors and excessive hormonal stimulation during the 10-12th week of fetal development [4-7].

Usually these malformations are not recognized until the reproductive years, when problems of fertility, abortions, fetal deformities, abnormal birth presentations or uterine perforation arise as adverse consequences of these malformations [2].

A number of classifications have been proposed for mullerian anomalies, [8, 9] but the most simple is one presented by Buttram and Gibbons (Table 1).

In the case of didelphys uterus the two structures may not be of equal size. Development of a neoplasm in one of the uterine endometrial cavities is an extremely rare event and few reports are encountered in the literature [10-15].

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Table 1. — *Classification of Mullerian fusion anomalies (Buttran V. C., Gibbons E. E., 1979).*

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|---|---|
| **Class I** | Segmented Mullerian agenesis or hypoplasia which is classed as a. Vaginal, b. Cervical, c. Fundal, d. Tubal and e. Combined. |
| **Class II** | Unicornuate uterus with a rudimentary horn and communicating endometrial cavity, non-communicative, uterine cavity, no uterine cavity. Unicornuate uterus without rudimentary horn. |
| **Class III** | Uterus didelphys. Bicornuate uterus complete, partial or arcuate. |
| **Class IV** | Bicornuate uterus, complete to the internal os, partial or arcuate. |
| **Class V** | Septate uterus, with a complete or incomplete septurm. |
| **Class VI** | Uterus with internal minimal changes. |

## Case Report

Six cases with incomplete fusion of the Mullerian ducts were examined in our laboratory during a 15-year period. Three of these cases presented with an adenocarcinoma in one of the uterine cavities.

*Case 1*

The patient was a 58-year-old woman with a history of persistent metrorrhagia, ten years after menopause. She had no children. Pap smear and diagnostic curettage were negative for malignancy. A total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Pathological examination showed a didelphys uterus composed of two cervices 2.8 cm each in length, a right uterine body measuring 6 x 4 x 3 cm, and a left uterine body measuring 4 x 3 cm. The right uterine cavity was lined by friable, whitish neoplastic tissue extending deeply into the myometrium. Both uteri contained multiple fibroids 0.5-2 cm in diameter.

Histological examination showed an endometrioid adenocarcinoma focally papillary, poorly differentiated. No normal endometrial mucosa was preserved.

The tumor infiltrated the entire thickness of the myometrium. There was no involvement of the surface, cervix or adnexa. The left uterine endometrial cavity showed atrophic endometrium with focal ulceration and acute inflammatory changes, probably post-curettage.

Both cervices had inflammatory changes and squamous metaplasia of endocervical mucosa. The adnexae were atrophic. The patient received adjuvant chemo- and radiotherapy and was well one year after the diagnosis. No follow-up information is available.

*Case 2*

A 49-year-old woman with metrorrhagia was referred to our Clinic. The Pap-test and diagnostic curettage were negative for malignancy. Because of multiple leiomyomas, the patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Pathological examination revealed a bicornuate uterus consisting of a cervix measuring 2.8 cm in length and two endometrial cavities measuring 2 x 3.5 cm (right) and 1.5 x 2.5 cm (left). In the latter a polyp was identified measuring 1.5 cm in diameter. The myometrium contained four leiomyomas 1.3-10 cm in diameter. The adnexae were grossly unremarkable.

The endometrium of the right endometrial cavity was proliferative. The endometrium of the left endometrial cavity was hyperplastic. The endometrial polyp was adenomatous with foci of atypical complex hyperplasia and focal development of well-differentiated adenocarcinoma. There was no involvement of the underlying myometrium.

The patient had no further treatment and she was well at the 2-year follow-up.

*Case 3*

A patient, 76 years of age, presented with metrorrhagia. Because of the suspicion of traumatic rupture of the uterus during a diagnostic D&C she underwent total abdominal hysterectomy. Pathological examination revealed a bicornuate uterus with a double uterine cavity measuring 2 x 4.5 cm (right) and 1.5 x 4 cm (left).

The longest cavity was involved by a well-differentiated endometrioid adenocarcinoma that extended to the cervix and infiltrated the entire thickness of the uterine wall. The other endometrial cavity was affected by an adenomatous polyp with focal infiltration by adenocarcinoma.

No adjuvant therapy was given to the patient and she was well six months after the diagnosis.

## Conclusion

In our cases adenocarcinoma developed in one cavity of a didelphys uterus (Class III malformation) and in two bicornuate uteri (Class IV malformation).

In one case the tumor extended into both uterine cavities and in two cases, an endometrial polyp was involved.

Although a rare event, neoplastic change may occur in uteri where complete or incomplete fusion exists and proper clinical and laboratory diagnostic evaluation of the patient is essential for the correct therapeutic procedure [14].

In all cases of incomplete fusion of the upper genital tract a thorough investigation is mandatory to exclude the possibility of missing a cancer arising focally in only one of the two endometrial cavities.

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