

CLINICAL CASE

A PARTICULAR CASE OF DOUBLE UTERUS

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Abstract

Uterine congenital malformations of the uterus appear due to the fusion and/or resorption defects of the Mullerian paramesonephric ducts, throughout the organogenesis. One of the relative frequent malformations is the double uterus, in which there are two uterine cavities and two cervixes which enter the vagina (with or without a longitudinal median septum). The overall frequency of uterine malformations is difficult to appreciate because of the asymptomatic evolution in most cases. In 25% of the cases, congenital malformations of the uterus are accompanied by abnormalities of the excretory system. We present the case of a 27-year-old patient taken out to 7 weeks of amenorrhea. Transvaginal ultrasound confirmed the diagnosis of double uterus with gestational bag in the right uterine cavity. The renal ultrasound performed confirmed the caliceal malformation. Labor triggers in week 35, complicated with acute fetal distress, thus an emergency caesarean section was decided. A segmental-arched hysterotomy is performed along with a breech-presentation delivery; the fetus masculine with G=2600g, L=48cm, APGAR 6. The evolution of the patient and of the fetus was favorable, they were discharged after 14 days postpartum. This case required a complex approach, the evaluation protocol of the patient being an individualized one, in order to minimize the risks.

Keywords: *uterine congenital malformations, infertility and double uterus***Introduction**

Uterine congenital malformations of the uterus appear due to the fusion and/or resorption defects of the Mullerian paramesonephric ducts, throughout the organogenesis [1-4]. Uterine duplications are caused by the lack of paramesonephric ducts fusion. Sometimes, the uterus is completely halved (bicornuate uterus), in the least severe form this one being indented only on the median line (arcuate uterus).

One of the relative frequent malformations is the double uterus, in which there are two uterine cavities and two cervixes which enter the vagina (with or without a longitudinal median septum).

Cases of fully or partially atresia of one of the paramesonephric ducts are characterized by the lack of specific hemicavities or the presence of an endometrial sensitive hormone and the menstrual blood accumulates at this level. Frequently, the body uterine atresia associates with the atresia of the cervix. Uterine congenital malformations may be accompanied by vagina development anomalies, but they are more rare. Over time, according to the embryological development, there have been proposed several classifications of the congenital uterine malformations

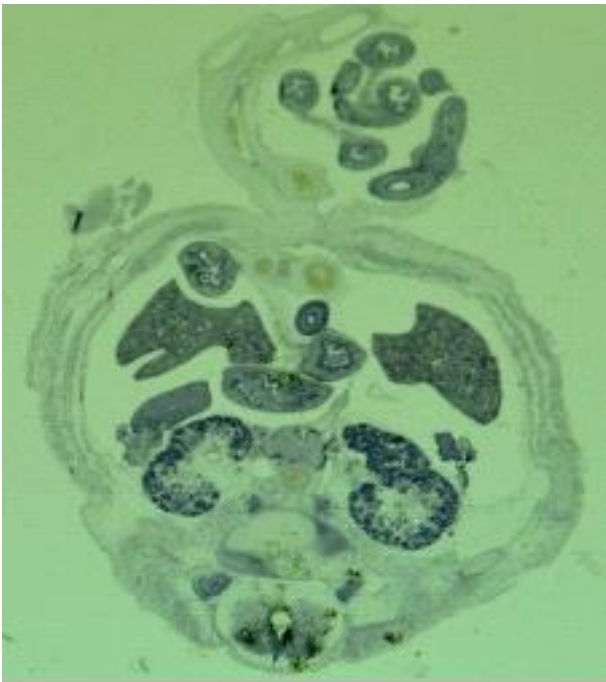


Figure 1 – Microscopic view of fetus

Musset and Belaish Classification (1964) divides the congenital uterine malformations into 4 classes:

- Aplasia/ agenesis of the mullerian ducts
- Complete bilateral (incompatible with life because it is associated with the bilateral renal agenesis)

- Incomplete bilateral (Mayer Rokitansky-Huster-Hauser syndrome)
 - Complete unilateral (real unicornuate uterus)
 - Incomplete unilateral (pseudounicornuate uterus)
- Fusion anomalies of the mullerian ducts
- Didelphic uterus
- Bicornuate uterus
- Resorption anomalies of the mullerian ducts:
 - Septate uterus
 - Arcuate uterus
 - Hypoplasia of the mullerian ducts (uterine hypoplasia)

The classification from 2013, suggested by Conuta ESHRE/ESGE Working Group divides the uterine anomalies in 7 classes [8,9]:

- U0- normal uterus;
- U1-dysmorphic uterus;
- U2- septate uterus;
- U3-bicorporeal uterus;
- U4- hemi uterus (with or without rudimentary cavity);
- U5- aplastic uterus;
- U6- unclassified malformations;

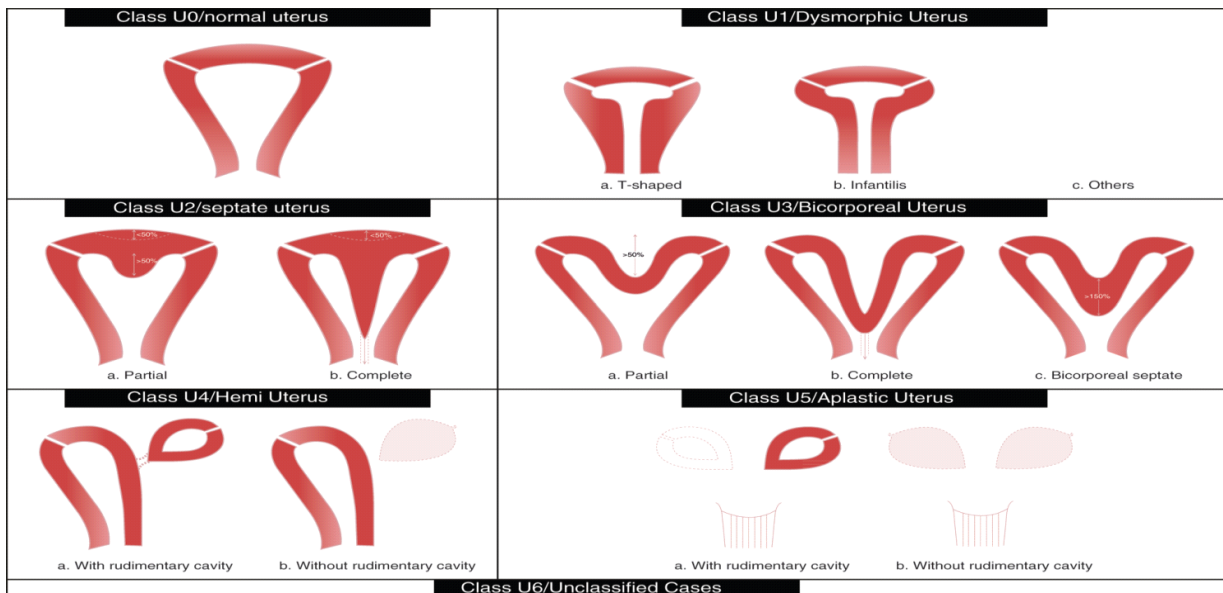


Figure 2 - Conuta ESHRE/ESGE Working Group Classification (source: https://embryology.med.unsw.edu.au/embryology/index.php/File:Uterine_anomalies_ESHRE-ESGE_classification.jpg)

ESHRE/ESGE classification of uterine anomalies: schematic representation (Class U2: internal indentation >50% of the uterine wall thickness and external contour straight or with

indentation <50%; Class U3: external indentation >50% of the uterine wall thickness; Class U3b: width of the fundal indentation at

the midline >150% of the uterine wall thickness).

The overall frequency of uterine malformations is difficult to appreciate because of the asymptomatic evolution in most cases [10]. In 2011 Chan et al. concluded that the prevalence of uterine malformations is 5.5% in general population, 8% in infertile patients and 13 % in those with a history of miscarriages [11,12]. The incidence of the congenital uterine malformations within couples with reproductive disorders was 6.8%. In any case, due to the unspecific and asymptomatic evolution of the malformation, the incidence can be higher [13].

There have been commonly reported in patients with congenital uterine malformations complications such as infertility, miscarriage, premature birth, abnormal placental insertions or labor dystocia in situations of pregnancy that evolved to the term [14-19]. With common mesodermal origin, from the Muller paramesonephric duct, genital development is closely related to the urinary system - in 25% of the cases, congenital malformations of the uterus are accompanied by abnormalities of the excretory system.

In terms of management of pregnant women with congenital uterine malformations, diagnosis and the type of the uterine malformation is established after clinical examination and imaging (pelvic ultrasound – conventional and 3D reconstruction, MRI, hysterosalpingography, hysterosonography, hysteroscopy) [20]. Likewise, for any patient diagnosed with congenital uterine anomalies an exam of the reno-urinary system (kidney, urography, ecography) [21] must be assessed.

In terms of reproduction, the septated uterus has the worst prognosis (the highest risk of miscarriage in the first trimester). Patients with double uterus, although they have a much higher incidence of obstetric complications, have a good reproductive prognosis, especially if a reconstruction of the uterine cavity is performed (special attention should be given to restoring optimal vasculature, whose damage can alter the functionality of the body). There is no standard tracking protocol for patients diagnosed with congenital uterine malformations during pregnancy, management being individualized.

Case presentation

We present the case of a 27-year-old patient without a history of heredo-colateral antecedents. From obstetric antecedents we mention three miscarriages in the first trimester of the pregnancy. From the previous pathological medical antecedents we mention: pyelocaliceal malformation (duplication of the bilateral pelvis), hereditary thrombophilia with factor V Leyden gene mutation. No antecedents of exposure in utero to DES (diethylstilbestrol).

The patient was taken out to 7 weeks of amenorrhea - clinical examination was performed, Pap test, TORCH screening and no other modifications were observed. Transvaginal ultrasound confirms the diagnosis of double uterus with gestational bag in the right uterine cavity (Figure 3).



Figure 3 – Transvaginal ultrasound

The renal ultrasound performed confirms the caliceal malformation. Clinical evaluation is recommended. Prophylactic tocolysis treatment and antiplatelet therapy are prescribed. The ultrasound reveals no fetal malformations and the development of the fetus is confirmed in the right uterine cavity (week 12). At week 15, the patient is hospitalized for metrorrhagia and the administration of tocolytic therapy and progesterin begins. At week 23, normal fetal morphology is confirmed.



Figure 4 – Ultrasound image

At week 29, the patient is hospitalized for uterine contractions, an increased risk of premature birth exists, but evolution is favorable under the tocolytic treatment (musculotropic and beta-sympathomimetic drugs) and anticoagulant (low molecular weight heparin). Labor triggers in week 35, complicated with acute fetal distress. The decision of extracting the fetus by emergency caesarean section is taken.

Pfannenstiel incision was performed. Intraoperatively the double uterus with gestation bag in the right uterine cavity is revealed. Its longest axis was disposed longitudinally, the lower segment was very thin and in tension. A segmental-arched hysterotomy is performed along with a breech-presentation delivery; the fetus masculine with G=2600g, L=48cm, APGAR 6

The hysterorraphy was done with great difficulty due to the heavy bleeding from the transected level of the hysterotomy. We used double layer suture for the uterus using with slowly absorbable thread separated. Also in the course of fetus extraction, a rupture was propagated to the lateral edge of the hemiuterus, until the nearby of the ligament of the uterus insertion. No other pathological modifications were detected during the inspection of the peritoneal cavity. The parietorraphy was performed in anatomical layers. The skin suture was performed intradermally. 10 days postoperatively the wire from the skin was suppressed. The evolution of the patient and the fetus was favorable, they were discharged after 14 days postpartum (hospitalization was prolonged due to the need of neonatal intensive care of the newborn).

Discussions

This case required a complex approach imposed by the uncertainty of the three miscarriages in the patient's obstetric history- because at the first three pregnancies no anticoagulant treatment was administered, we can not say whether recurrent abortions were due to coagulation abnormalities or uterine malformations. Our patient presented the most common obstetric complications- unorganized uterine contractions during pregnancy, being

hospitalized repeatedly with a diagnosis of threat of miscarriage/premature birth. Labor was prematurely triggered in the week 35 of pregnancy and it was complicated by the acute fetal distress, requiring emergency caesarean extraction of the fetus. It proved to be a typical congenital anomaly of double uterus by associating the reno-urinary malformations and complications but also due to the complications that appeared during the pregnancy. The evaluation protocol of the patient is individualized; in order to minimize the risks an evaluation at each 3 weeks was imposed.

Conclusions

We consider that this case reported is important because we presented the pre-, peri- and postnatal conduct of a pregnant woman with double uterus, a malformation with low incidence and guarded prognosis, to whom the right management was applied, with favorable results, following the administration of a specific treatment. The peculiarity of the case derives from the fact that unlike standard therapy, the pregnant woman needed specific prophylactic therapy, imposed by the associated pathologies. It also proved to be a particular case of malformation due to the premature birth in the week 35.

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