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Twin pregnancy in a partial septate uterus and the contribution of magnetic resonance imaging. A case report and brief literature review

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Summary

Congenital malformations of the female genital tract are defined as deviations from normal anatomy. They are due to embryological maldevelopment of the Müllerian or paramesonephric ducts. Septate uterus is the most common of all Müllerian duct defects. A rare case of a viable twin pregnancy in separate components of a septate uterus is reported. The diagnosis of uterine malformation was made before pregnancy. The viability of the twin gestation was detected and confirmed by transabdominal and transvaginal sonography. Moreover, the location of the fetuses was confirmed by a magnetic resonance imaging (MRI). A specific schedule of prenatal screening was provided. Two healthy neonates were delivered by a cesarean section in the 34th week of gestation. The importance of MRI and management of this high-risk pregnancy is discussed.

Key words: Twin pregnancy; Uterine malformations; Septate uterus; Magnetic resonance imaging (MRI).

Introduction

The fallopian tube, uterus, cervix, and the upper two-thirds of the vagina of the female reproductive tract are derived from a pair of Müllerian ducts. Normal development of the Müllerian ducts includes three phases: organogenesis, fusion, and septal resorption [2]. The exact mechanisms of these phases are unknown [3]. A wide variety of malformations can occur when this system is disrupted. The uterine malformations are the most common [4, 5].

A complete or partial septate uterus is the most common (34-48%) type of uterine malformation [6, 7]. It is a result of the septal resorption process failure. Septate is defined as the uterus with normal outline and an internal indentation at the fundal midline exceeding 50% of the uterine wall thickness. This indentation is called septum and is composed of poorly vascularized fibromuscular tissue. It could divide partly (partial septum) or completely (complete septum) the uterine cavity including in some cases the cervix and/or the vagina. In a partial septate uterus, the septum partly divides the uterine cavity above the level of the internal cervical os [8].

The most common symptoms of a septate uterus are dyspareunia, dysmenorrhea, and primary or secondary infertility. This Müllerian tract defect is also associated with poor reproductive outcome and obstetrical complications including abortions, premature labour, malpresentations, retained placenta, fetal intrauterine growth retardation, antepartum hemorrhage, and increased cesarean section rates [9-11]. The fetal survival rates have been reported to be 6-28% [12]. Adverse effects are even higher in twin pregnancies, where data is limited to case reports. In a recent retrospective cohort study of twin pregnancies, uterine malformations were found to be associated with an increased risk of cerclage, preterm birth, and lower birth weights, but not fetal growth restriction [13].

Case Report

A 29-year-old woman in her third pregnancy was referred to the present department in the 12th week of gestation of a twin pregnancy. In her obstetrical history, a delivery of a dead fetus in the 26th week of gestation (in year 2001) and a full term cesarean section delivery of a healthy male neonate were mentioned. Her general medical history was not remarkable except for the administration of thyroxin tablets, because of hypothyroidism, heterozygous thalassaemia, and deficiency of glutamate-6 pyruvate dehydrogenase (G6PD) enzyme. A transvaginal ultrasound scan was performed and revealed a diamniotic, dichorionic twin pregnancy. Both fetuses showed normal cardiac activities and the crown-rump lengths (CRL) corresponded to the gestational age.

Ultrasonography performed at the 16th week of pregnancy, detected each fetus in separate compartments of a partial septate uterus. Both fetuses were of similar size and the measurements corresponded to the gestational age. Placentas were fundal and anterior and no evidence of congenital anomaly was detectable in either of fetuses. This unusual twin pregnancy was confirmed by MRI. The MRI examination revealed a smooth fundus with a uterine septum ending two mm above the internal cervical os, and common cervical canal. The outer uterine contour appeared nor-

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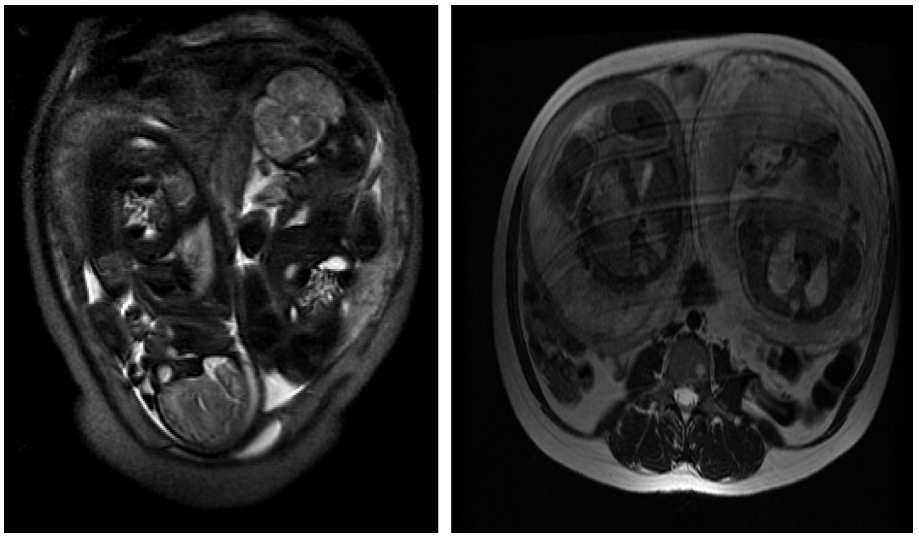

Figure 1. — A) T2-weighted abdominal image (coronal plane) showing simultaneous 34-week pregnancy in each horn of a uterus didelphys in a 29-year-old woman. B) T2-weighted abdominal image (axial plane) showing simultaneous 34-week pregnancy in each horn of a uterus didelphys in the same patient.

mal. Both uterine hemi-cavities were normal in appearance and each fetus were in these separate hemi-cavities (Figures 1A, B). Maternal urinary tract ultrasound examination was normal. Serial ultrasound examinations taken twice a month demonstrated normal fetal development. The pregnancy course was uneventful until the 33rd week of gestation. In the end of the 33rd week, the pregnancy was complicated by the occurrence of preterm contractions confirmed by cardiotocography monitoring which showed normal fetal heart rate with an increased myometrial activity. The pregnant woman was treated with bed rest, oral magnesium administration, and i.m injection of betamethasone in order to enhance fetal lung maturation. At 33+4 weeks gestation, premature preterm rupture of membranes was noted and regular uterine contractions began. An emergency cesarean section was performed. Both fetuses were delivered successfully: the first one was a female in a breech presentation with a birth weight of 2,150 grams and the second was a male fetus presenting in vertex position with a birth weight of 2,465 grams. Both newborns had normal Apgar scores (8/10). Their postnatal course was uneventful. The two placentas, each in its respective hemi-cavity, were easily delivered. The uterine septum was clearly identified during cesarean section. Two distinct uterine cavities were separated by approximately seven mm(lower and midportion] to 18 mm (fundal portion) of intervening fibrous tissue and one cervical canal were noted. The uterus contracted well after the delivery of the twins. The postpartum course was uneventful. The patient was discharged on the eighth postoperative day in a stable condition.

Discussion

The accurate incidence of uterine malformations in the general population and among infertile women is not well known. This is due to the fact that many of these malformations are asymptomatic and remain undiagnosed. Moreover, the diagnostic methods used are inaccurate and there is not a uniform system of classification [15]. Therefore, the incidence of uterine congenital anomalies is approximately estimated and reported to be 3.2% among infertile women, while of 0.001-10% in general population [16, 17]. It has also been reported that the incidence of Müllerian anomalies is significantly higher in infertile patients (6.3%) ($p < 0.05$), in comparison to fertile (3.8%) and sterile (2.4%) women [15].

The etiology of Müllerian anomalies has not yet been fully clarified. The karyotypes are normal (46 XX) in 92% of the women with Müllerian anomalies and abnormal (sex chromosome mosaicism) in 8% of these women. Polygenic and multifactorial causes have been proposed [18]. Hypoxia during pregnancy, medications such as methotrexate or diethylstilbestrol (DES), ionizing radiation, and viral infections may contribute to Müllerian malformations [19, 20]. Among the drugs, thalidomide and DES lead to malformations of the uterine cavity. DES, a nonsteroidal estrogen, was widely used in the 1950s, principally in the United States, in the treatment of obstetrical conditions, like miscarriage and preeclampsia, resulting in uterine malformations (especially of the uterine cavity) in the daughters of the women who were treated with DES [21]. A recent study incriminated the deficiency in the antiapoptotic protein Bcl 2 for the persistence of the intrauterine septum [18].

Three systems have been proposed for the classification of female genital tract anomalies. The American Society of Reproductive Medicine (ASRM) classification system, the embryological–clinical classification system of genito-urinary malformations, and the Vagina, Cervix, Uterus, Adnexae and associated Malformations system based on the Tumor, Nodes, Metastases principle in oncology. Recently, The European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE), have established a common working group under the name (CONgenital UTerine Anomalies) (CONUTA), in order to develop a new classification

system [8].

Currently, many diagnostic tools have been used in order to diagnose these congenital malformations. Hysterosalpingography (HSG) allows satisfying assessment of the uterine cavity and tubal patency, but enables assessment of the external uterine contour. Ultrasound is a very useful, quick, and economical tool, with no exposure of the patient to radiation. However, the image may not be satisfying and diagnosis may not be simple in large patients. Three-dimensional (3D) ultrasound has been shown to be more accurate than two-dimensional ultrasound and equal or better than MRI at assessing Müllerian duct anomalies (MDA) [22]. 3D ultrasound has the potential of becoming the imaging standard for MDA [22]. However, MRI is currently considered to be the best imaging modality for MDA. MRI offers a visualization of both the internal and the external uterine anatomy, with no radiation exposure. Moreover, MRI has been shown to have excellent agreement with the clinical diagnosis of the subtypes of MDA [23].

As mentioned above, of all uterine anomalies, the septate uterus is associated with the poorest reproductive outcome and the highest incidence of obstetrical complications, with fetal survival rates between 6-28 % [12]. Furthermore, it has been reported that the septum surface area is correlated with the incidence of complications. This is because greater surface area is associated with septal implantation, leading possibility to abnormal placentation or uncoordinated contractility of the septal musculature [24]. However, a septate uterus does not seem affect fertility and is not considered as an infertility factor [25]. Heinonen *et al*. have reported that pregnancy in the septate uterus can progress without any surgical treatment [26]. It has been reported that the pregnancy outcome depends on the capability of the uterus cavity to expand in proportion to their variation from the normal [27]. Consequently, pregnancies in women with Müllerian defects may even result in an absolutely normal obstetric outcome [28].

As long as twin pregnancies in women with Müllerian defects are concerned, they seem to be very rare. Twin pregnancies with each fetus placed in separate compartments of a septate uterus are managed as high-risk pregnancies and should be offered meticulous prenatal care. [29] Although a normal pregnancy course is possible, most of the uterine abnormalities are associated with a considerably lower percentage of viable babies, particularly in twin pregnancies [30-32]. Moreover a twin pregnancy with a fetus in each hemi-cavity of a septate uterus has been reported to have a worse prognosis concerning fetal survival rate than a pregnancy in a uterus bicornuate, didelphys or arcuatus [33]. In addition, the incidence of breech presentation and prematurity are much higher in this group of pregnant women [34]. In the present case, after a short time administration of tocolytic treatment, both fetuses were delivered by a cesarean section in the 34th week of gestation.

It is worth mentioning that early detection of uterine anomalies is of great importance. As mentioned above, these pregnancies run increased risk of obstetrical complications. Although sonography and especially transvaginal sonography have been reported to be useful and reliable in identifying uterine anomalies in the very early stages of pregnancy, in many cases [35-38] the diagnostic value of the MRI is undeniable. MRI demonstrates fibroid tissue, with a high sensitivity and specificity in distinguishing these anomalies [39]. The specificity of MRI ranges from 96-100% for diagnosing Müllerian malformations [40]. In cases of a septate uterus, the septum is recognized as an entity of different signal intensity, according to its composition. Fibrous septa are seen as low-intensity signals on T2-weighted images and muscular septa as intermediate-intensity T2 signals [41]. MRI may demonstrate the exact uterine abnormality [42]. In the present case, MRI helped in the exact recognition of the uterus septum, and fetuses' and placenta placements. Accordingly, a successful pregnancy management, delivery, and the optimal outcome of a viable twin pregnancy succeeded.

Although, in most cases of twin gestation in septate uterus the outcome is reported to be poor, the present patient managed to reach the 34th week of gestation without complications and had a delivery of two healthy neonates by cesarean section. It is notable that a previous normal delivery does not exclude the presence of uterine malformations. The present authors believe that early detection of uterine malformations with the use of transvaginal sonography and proper follow-up by MRI should be of great importance in the management of these high-risk pregnancies in the future. Unfortunately, these high-risk pregnancies lack management guidelines, resulting many times in management dilemmas among obstetricians. Consequently, they would like to underline the need for management guidelines of pregnancies complicated by MDA, because even though they are rare, they are precious.

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