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Uterus Didelphys: A Rare Müllerian Anomaly Presents with Recurrent Miscarriages – A Short Review

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ABSTRACT

The uterus didelphys is a rare congenital Müllerian duct (Paramesonephric duct) fusion anomaly. The complete non-fusion of the paired Müllerian ducts generate this problem. This is characterised by the presence of two separate uteri, two cervices, and frequently a longitudinal vaginal septum. The majority of patients are asymptomatic but some patients may have gynaecological and obstetric problems. Uterus didelphys is diagnosed during evaluation for recurrent pregnancy loss. It is interesting to note the clinical presentation, diagnostic challenges and management options.

Keywords

Uterus didelphys, Müllerian anomaly, Congenital uterine anomaly, Recurrent miscarriages, Preterm delivery, Double cervix.

Background

The word uterus didelphys came from ancient Greek words *di-* two and *delphus* is uterus.

The uterus didelphys affects about 0.3% of the population. The incidence is 1 in 3000 women. It is much lesser than other non fusion anomalies such as arcuate uterus, septate uterus, and bicornuate uterus [1].

Uterus didelphys is an uncommon congenital Müllerian duct (Paramesonephric duct) anomaly due to the non-fusion of Mullerian duct. Uterus didelphys represents a Class III anomaly according to the American Society for Reproductive Medicine (ASRM) [2].

This results in complete duplication of the uterus, cervix, and sometimes vagina. The patients may be asymptomatic or associated with reproductive and obstetric complications.

Generally, the patients present with 3 or 4 mid trimester recurrent miscarriages in the past. They may also present with heavy menstrual periods and pelvic pain. With these complains they are diagnosed to have uterus didelphys. They may also have fibroids in both the uterus and suffers from heavy and painful menstrual periods.

They have no difficulty in conception. Their pregnancies occur spontaneously. There will be no history of infertility. The menstrual cycles are regular with mild dysmenorrhoea. There is no history of dyspareunia, chronic pelvic pain, or urinary symptoms.

The patient may not have any significant medical or surgical history. They do not have history of diabetes, hypertension, or thrombophilia. The family history will be unremarkable.

On general examination, the vital signs are stable. Abdominal examination was normal. The speculum examination reveals two separate cervices. There is no vaginal septum.

The 1st miscarriage shall be at the period of gestation of 20 weeks. The ultrasound examination at that time will identify uterine didelphys. The MRI examination conforms two uterus and two

cervix. There may be or may not be a vaginal septum.

The patient is counselled regarding the nature of the anomaly and its reproductive implications. As surgical unification is not recommended for uterus didelphys, conservative management is advised. Resection of the vaginal septum is not indicated since most of the patients are asymptomatic.

The 2nd miscarriage will be in 1-2 years at the period of gestation of 18-20 weeks.

The 3rd miscarriage will be again after few months at the period of gestation of 18-20 weeks.

The 4th miscarriage will be again at the period of gestation of 18-20 weeks.

The following investigations are done in recurrent miscarriages

a) Transvaginal examination for the cervical length. The cervical length will be normal.
b) Genetic investigations including parental karyotyping and chromosomal microarray of products of conception. All will be normal
c) Thyroid profile, diabetic screening, luteal phase progesterone and PCOS evaluation- All will be normal.
d) Immunological / thrombophilia screening test on two occasions 14 weeks apart done. Lupus anticoagulant, Anticardiolipin antibodies (IgG, IgM) and Anti-β2 glycoprotein I antibodies- All will be normal.
e) Inherited thrombophilia: Factor V Leiden mutation, Prothrombin gene mutation, Protein C, Protein S and Antithrombin III deficiencies screening

Usually after the 4th miscarriage, the patients not want to go through any pregnancy.and emotional agony

Most of the couple adopt a child

Discussion

During embryogenesis the fusion of two paramesonephric ducts (also called as Müllerian ducts) occurs between the 12th and 16th weeks of gestation. Uterus didelphys is the embryogenetic non fusion of the Müllerian ducts. This results in two separate uterus and two separate cervix. There may be two vagina in some cases [3].

As the foetus develops, the Mullerian ducts fuse together to form the uterus, fallopian tubes, cervix, and the upper section of the vagina. A double Mullerian system means a fusion failure, but for unknown reasons. The abnormality can affect the vagina and kidneys, and from time to time, the skeleton [4].

The actual aetiology is unknown. The genetic factors may play a role. They may have family history of reproductive anomalies. The autoimmune disorders involve this connective tissue disorders may be a contributory factor.

Symptoms of uterus didelphys

The majority of the women are asymptomatic.

The common symptoms are:

a) Heavy menstrual periods: Two uterus and two endometrium can lead to heavy menstrual period. Two shedding may last longer and heavier.
Two menstrual periods can occur in one cycle. Both uterus shed their lining may be at different time.
b) Pelvic pain: They may experience pelvic discomfort and pain during menstrual period.
c) Reproductive problem: They may have difficulty on conceiving, problem in carrying the pregnancy until term.

A specific association of uterus didelphys (double uterus), unilateral hematocolpos (inadequate draining of menstrual blood) and ipsilateral renal agenesis (having only one kidney) has been described [5].

Diagnosis

They may not have any symptoms at all. Heavy menstrual periods and dysmenorrhoea are also experienced by normal women. However recurrent miscarriages and preterm labour may arise suspicion.

On speculum examination two cervices can be seen. Some patients may have vaginal septum and some may not have septum.

Imaging tests

Imaging tests are invaluable make a diagnosis of this condition

a) Ultrasound examination: 2 D and 3 D transvaginal ultrasound examination will identify two separate uterus and two separate cervix
b) MRI: Magnetic resonance image can identify two uterus, two cervix and often two vagina clearly.
c) HSG: Radiopaque substances is sent through both cervix and X Ray identifies two separate uterus and double cervix
d) Sono-hysterogram- In this procedure, thin catheters are inserted into both uterine cavities. Saline is infused into both uterus. Then transvaginal ultrasound scan is done to get the images of the cavity as the fluid goes through the cervix and uterus.

Differential diagnosis

The following conditions may mimic uterus didelphys. We should carefully exclude these anomalies to make a precise diagnosis.

a) Bicornuate Uterus: This results from the partial fusion of Mullerian duct. The uterus is heart shape in imaging
b) Septate Uterus: The uterus divided by a fibrous or muscular septum.
c) Unicornuate Uterus: In this condition only one Müllerian duct develops, resulting in a single, underdeveloped uterus.

Complications

a) Infertility: Usually the people with uterus didelphys have no problem in conception but rarely some may have difficulty in conceiving.
b) Uterus didelphys have high chance of late miscarriages and preterm birth due to accommodation problem
c) The incidence of ectopic pregnancy is relatively high in these patients.
d) Incidents of breech presentation are relatively high compared to normal population. The baby has limited space to move freely. It is difficult for the foetus to get the cephalic presentation.
e) The incidence of Caesarean section also slightly high.

A number of twin gestations have occurred where each uterus carried its pregnancy separately. There have only been about 100 cases worldwide of a woman with a double uterus being pregnant in both wombs at the same time [6].

The majority of patients with uterus didelphys can go through normal pregnancy and successfully deliver term babies.

Heinonen studied 26 women with uterus didelphys. In this study 18 patients 69.2%.delivered term babies and 8 women 30.8% had preterm delivery. The breech presentation was in 43% and Caesarean section was performed in 82% of the cases [7].

Pregnancy outcomes should be carefully monitored since problems can quickly arise. Caesarean is often recommended [8].

Conclusion

Uterus didelphys is often asymptomatic and unnoticed. Some women have menstrual problems and obstetric complications. Accurate diagnosis using imaging modalities is essential for appropriate counselling and management, particularly in women with infertility or adverse obstetric outcomes.

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