Mullerian Anomalies a Variant of Uterus Didelphys

Pallavi Misra*

Abstract
The case report is of a patient who initially presented as a case of primary infertility and upon work up and investigation was found to be a case of double uterus or uterus didelphys. Uterus didelphys or double uterus results from failure of the midline fusion of the two Mullerian ducts. Mullerian anomalies arise due to defects in fusion of the two Mullerian ducts during intrauterine life. Most cases go unreported as they are asymptomatic and do not cause any menstrual, coital or reproductive problems. The odd cases presenting thus end up getting investigated thoroughly and are unforgettable learning experiences worth reporting and sharing. The case below is reported for its rarity and as testimony to the fact that despite all advances in our understanding of human embryology, some questions still remain unanswered.

Introduction
Mullerian anomalies result from defective fusion or absorption of the Mullerian ducts during embryonic life. Their incidence is difficult to estimate as majority of these anomalies are minor or insignificant and do not affect menstrual and reproductive function and thus remain asymptomatic. Patients with symptomatic anomalies usually have symptoms of obstruction or reproductive failure. Diagnostic methods for evaluation of the exact nature of the anomalies have evolved immensely over the past years. The advent of ultrasound, hysteroscopy and magnetic resonance imaging has facilitated rapid and accurate diagnosis. The increased availability and utilization of these diagnostic techniques yields complete information about the anomaly under investigation and has resulted in higher reported incidence of various anomalies.

Case History
The patient - Mrs. VM - a 26 year old housewife presented in the out patient clinic for investigation of primary infertility. She had been married for 08 yrs and had been living with her husband since then. She gave history of regular coitus two to three times a week with no dyspareunia. Her menstrual history was normal. She had attained menarche at the age of 13 years and had regular cycles since then with moderate flow for three to four days and no dysmenorrhoea. There was no history of any addictions or drug intake and no significant past medical or surgical history.

Her husband was 30 years old and working as a motor mechanic. He was a non smoker and non-alcoholic and had no addictions. There was no positive medical or surgical history in his case too.

The patient gave history of being investigated for primary infertility on two occasions in the past, however, no reports were available from the first instance and only films of the hysterosalpingogram from the second.

Examination
The general and systemic examination was normal. Her vitals were normal and there was no pallor. There was no thyroid swelling or galactorrhoea and no evidence of any signs suggestive of hyperandrogenism.

Gynaecological examination
Per abdomen (P/A) examination was normal.

Per speculum (P/S) examination revealed a complete longitudinal vaginal septum with a cervix visualised in each of the two vaginal cavities (Figs. 1

*Consultant Gynaecologist, Kandivali (East), Mumbai – 400 101.
Per vaginum (P/V) examination was performed through both the vaginal cavities and revealed a single uterus which was anteverted, normal in size, freely mobile and both the fornices were free.

**Investigations**

The Hysterosalpingogram (HSG) plates available from one of the previous investigations revealed the presence of a septate uterus with two horns. The fallopian tubes were not visualised. There was no spill of contrast into the peritoneal cavity. It was thus decided not to repeat the investigation (Figs. 3 and 4).

The other tests (haemogram, urine analysis, X-ray chest and biochemistry) were normal and were performed with a view to and prepare the patient for examination under anaesthesia (EUA) with diagnostic laparoscopy and dilatation and curettage (D and C).

Husband's semen analysis (HSA) showed evidence of oligoasthenospermia. A urological examination was requested in view of the above and was reported as normal. Ultrasoundography (USG) of the pelvis of the patient (trans abdominal and trans vaginal) confirmed presence of a vaginal septum and a septate uterus with visualization of two separate horns of the uterus. Rest of the pelvic anatomy was unremarkable (Figs. 5 and 6).

USG - Kidney Ureters and Bladder was normal.

**Provisional diagnosis** - Uterus Didelphys with bicollis with bicolpos and patient was posted for EUA with diagnostic laparoscopy and D and C under general anaesthesia.

EUA revealed a complete longitudinal vaginal septum with a cervix in each vaginal cavity. The left cervix was not communicating with the uterus while the right one was patent. The upper cervical length was one and half inches. The per vaginum examination was performed through both vaginal cavities and a single uterus was found to be present. The uterus was anteverted, normal sized, freely mobile and both fornices were free.

**Laparoscopy** revealed presence of a single uterus which appeared normal in shape and size. The tubes and ovaries were unremarkable. A corpus luteum was visualised in the right ovary (Fig. 7).

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**Fig. 1**: Shows the longitudinal vaginal septum as seen on per speculum examination

**Fig. 2**: Shows the two vaginal cavities with cervices visualized in each of them.

**Fig. 3**: Shows the uterus as seen on USG in longitudinal and transverse sections.

**Fig. 4**: Shows the two horns of the uterus as visualised on USG.

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Chromopertubation was attempted but was not possible through the left cervix. The same was successfully performed through the right cervix. There was moderate resistance to the passage of dye with only bilateral staining of tubes but no free spill into the peritoneal cavity.

D and C was not possible through the left cervix. It was performed through the right cervix and revealed a single uterine cavity with moderate endometrium obtained on curettage which was sent for histopathology examination (HPE).

The HPE report revealed endometrium in late secretory phase.

Final Diagnosis - Incomplete uterus didelphys with bicollis with bicolpos.

Treatment - Patient was advised surgical correction with In vitro fertilisation with embryo transfer but could not proceed with the same in view of the financial implications of the treatment.

Discussion

Uterus didelphys is a Class III category of malformation as per the American Fertility Society’s Classification of uterine malformations. In such cases the mullerian tract fails to fuse along all or most of its length. There maybe complete duplication of the vagina, cervix and uterus and the two halves maybe divided by a ligament of connective tissue (Fig. 8).
Embryology

In a female foetus, the uterus starts out as two small tubes - the mullerian ducts. As the development occurs, the tubes normally join to create one larger, hollow organ — the uterus. Sometimes, however, the tubes do not join completely. Instead, each one develops into a separate cavity. Lack of fusion of the two mullerian ducts results in duplication of corpus and cervix. This condition is called Double Uterus or Uterus Didelphys.

Each cavity in a double uterus often leads to its own cervix. Some women with a double uterus also have a duplicate or divided vagina. A vaginal septum is present in about 75 per cent of patients with uterus didelphys. Double uterus is rare — and sometimes not even diagnosed as these patients usually have no difficulties with menstruation and coitus.

According to one estimate, double uterus occurs in 2 per cent to 4 per cent of women who have normal pregnancies. The percentage may be higher in women with a history of miscarriage or premature birth.

It is not known as to what exactly causes a double uterus. Congenital anomalies of the mullerian ducts are frequently associated with abnormalities in the urinary tract, which suggests that something may influence the development of these related tubes before birth. The renal abnormality is usually present on the same side as the mullerian defect. Normal vaginal development is subsequent to the canalization and fusion of the two Mullerian ducts once vacuolization has occurred, but not their union, hence, a duplication of different degree will be the consequence.

Investigation and Diagnosis

A double uterus is often diagnosed during a routine pelvic exam when the doctor observes a vaginal septum, double cervix or an unusual shaped uterus. It may also be a finding in cases presenting for investigation of primary infertility or after repeated reproductive failures. If the doctor suspects an abnormality, he or she may recommend any of the following tests to help confirm the diagnosis.

- Magnetic resonance imaging (MRI).
- Ultrasound.
- Hysterosalpingography.
- Hysteroscopy.
- Laparoscopy.

Treatment for this anomaly is basically surgical and is needed only if a double uterus causes symptoms or complications, such as pelvic pain or repeated miscarriages. The reproductive outcome for patients with uterus didelphys is similar to that of patients with unicornean uteri. In view of the acceptable rates of foetal salvage and technical difficulties of uteroplasty, surgical correction is rarely performed. The various surgical modalities that have been suggested for this anomaly include a modified Strassmann procedure and cervical circlage. Strassmann’s unification procedure is the operation of choice although hysteroscopic metroplasty with resection of the dividing wall and preservation of both the cervices is also a possibility.

Prognosis

Many women with a double uterus have normal sex lives, pregnancies and deliveries. In fact, the more complete the duplication, the lesser the chances of likely complications. Sometimes a double uterus leads to infertility or miscarriage. A double uterus may also cause premature birth or malpresentations of the baby.

References
1. Kenneth J Ryan, Ross R Berkowitz, Robert L
China’s Barefoot Doctor: Past, Present, and Future

In 1968, the programme of barefoot doctors was introduced by the journal Red Flag as a national policy focussed on quickly training paramedics to meet rural needs. Most barefoot doctors, who graduated from secondary school education, practised after training at the country or community hospital for 3-6 months.

Despite a low level of service in terms of technique and medical instruments the barefoot doctor programme effectively reduced costs and provided timely treatment to the rural people.

WHO regarded China’s barefoot doctor system as a successful example of solving shortages of medical service in developing countries.

In January, 1985, the title of barefoot doctor was cancelled by the Ministry of Health, and some of them either became village doctors or were lost to other professions. As private practitioners, however, village doctors focused on treatment of diseases with economic benefits in mind, such that the public health of the village was a low priority.

The new system differs from the old: it is organized and financially supported by the Government. The local government pays 10 Renminbi per year for everyone covered by the new system, and the fund covers the costs of serious diseases.