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Case Report

Didelphys uterus: an interesting case report of pregnancy in a rare Mullerian anomaly

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ABSTRACT

Mullerian duct anomalies (MDAs) are congenital defects of the female genital system that arise from abnormal embryological development of the Mullerian ducts. A didelphys uterus, also known as a double uterus, is one of the least common amongst the MDAs. This report discussed a case of pregnancy with uterus didelphys. This patient was a 27-year-old primigravida with 34.2-week gestation with spontaneous conception who presented with decreased fetal movements. On examination patient had a non-communicating, thick vaginal septum extending from the introitus to the cervix was seen and two cervices one on each side of the septum were located. The patient underwent emergency lower segment caesarean section in view of foetal distress and doppler changes. Intra-operatively, evidence of didelphys uteri was seen. Intraoperative and post-operative period was uneventful. A fetus of 1790 gram was delivered, with APGAR 9/10. There were no renal anomalies on subsequent ultrasonography.

Keywords: Mullerian duct anomalies, Didelphys uterus, Congenital defects

INTRODUCTION

MDAs are female genital system congenital malformations caused by aberrant Mullerian duct embryological development. Failure of development, fusion, canalization or reabsorption, which generally happens between 6 and 22 weeks in utero, causes several defects. According to most estimates, these anomalies affect between 0.5 and 5.0 percent of the general population.[1-4]

With a mean incidence of 35%, septate uterus is the most prevalent uterine defect, followed by bicornuate uterus (25%), and arcuate uterus (20%).[4] Women with mostly secondary infertility may experience delayed natural conception as a result of uterine abnormalities.[4]

It is generally accepted that having a uterine anomaly is associated with poorer pregnancy outcomes such as increased chances of spontaneous abortion, preterm labour, caesarean delivery due to abnormal presentation and positions and decreased live births as compared to a normal uterus.[1-5] The severity of these results, however, varies depending on the type of uterine anomaly and other obstetric variables such as multiple pregnancy, amniotic fluid index, placenta placement, gestational hypertension and diabetes.

The pregnancy outcome of patients with untreated bicornuate and septate uterus is also poor, with a term delivery rate of only 40%.[4] Unicornuate and didelphys uterus have a term delivery rate of 45 percent, and patients with untreated bicornuate and septate uterus have a term delivery rate of only 40 percent. With term delivery rates of 65 percent, an arcuate uterus is associated with a somewhat better but still compromised pregnancy outcome.[4]

The most recent classification system for the different types of Mullerian duct abnormalities is by American Society of Reproductive Medicine in 2021 (Figure 1).[17]

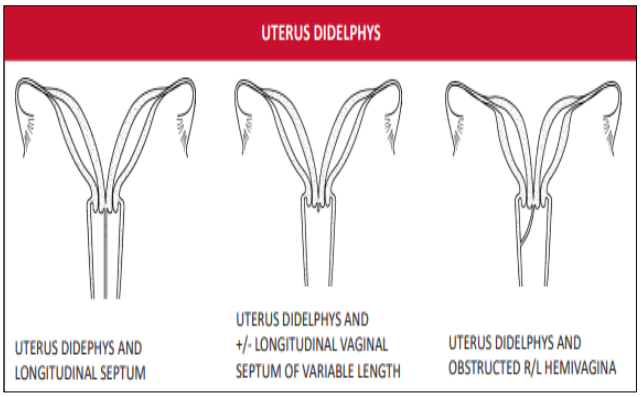

Figure 1: Uterus didelphys and its classification.

In this case report, we discussed a rare case of didelphys uterus with pregnancy.

CASE REPORT

This patient was a 27-year-old primigravida with 34.2-week gestation from Mumbai who presented to our emergency room. The patient was referred from a peripheral hospital for decreased fetal movements and doppler changes on USG.

Patient was primigravida, conceived spontaneously and registered with a general gynaecologist at 24 weeks of gestation. Patient had uncomplicated antenatal care from 24 weeks and did not give any history of signs of threatened abortion or signs of threatened preterm labour. Her routine ANC investigations and vital signs remained within normal limits and fetal ultrasounds (1st one done at 24 weeks) also remained within normal limits. Foetus showed appropriate growth. The patient was referred from a peripheral hospital for decreased fetal movements and Doppler changes on USG.

At 34.2-week gestation, ultrasonography was done which showed a single live intrauterine gestation of 33.5 weeks with cephalic presentation, posterior placenta. Estimated fetal weight was 2310 gm. Liquor was adequate. Two loops of cord around fetal neck were noted. On middle cerebral artery doppler, C-reactive protein (CPR) was less than 1, suggestive of brain sparing effect.

At 34.2-week gestation, the patient presented to our hospital with decreased fetal movements and doppler changes on ultrasonography. On physical examination, patient was normotensive and afebrile. On per abdomen examination, uterus was 32-week size, fetal heart rate was present and abdomen relaxed. On per vaginal examination, a non-communicating, thick vaginal septum extending from the introitus to the cervix was seen and two cervices one on each of the septum were located. However, patient and her husband were not aware of the patient's condition until that day. The patient did not report having dyspareunia, dysmenorrhea or chronic abdominal pain in the past (Figure 2).

The fetal heart tracing showed a fetal heart rate of 170 per minute at baseline, decreased variability, with accelerations and late decelerations. At the end of this work-up, the patient was admitted for doppler changes of brain sparing effect and fetal distress. The patient underwent emergency lower segment caesarean section under spinal anaesthesia in view of foetal distress.

Intraoperatively, evidence of didelphys uterus seen, where two uteri with one fallopian tube and one ovary each on one side with two cervices. The pregnancy was continued in right uterus. The foetus was in oblique lie. Baby delivered by vertex presentation. A female foetus of 1790 gram was delivered with APGAR 9/10. The uterus was closed in double layers. Haemostasis was achieved at blood pressure 110/80 mmHg. Abdomen was closed in layers. The surgery was uneventful. The puerperium was also uneventful. There were no renal anomalies on subsequent USG (Figure 3).

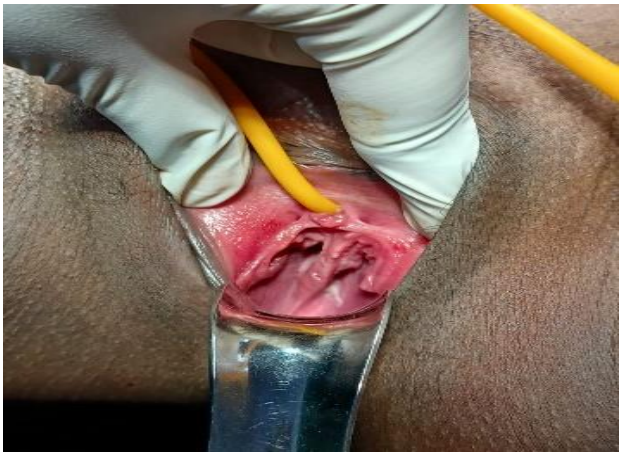

Figure 2: Vulval examination showing non-communicating, thick vaginal septum extending from the introitus to the cervix.

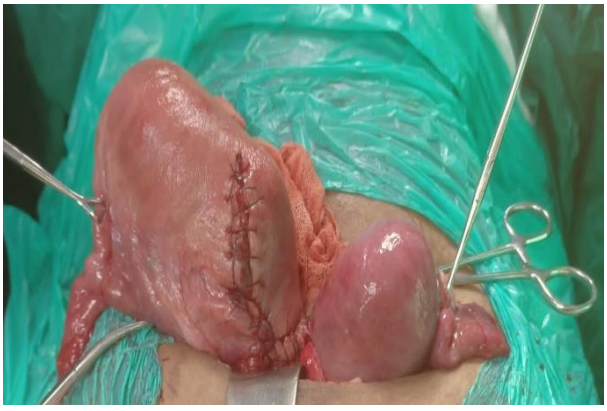

Figure 3: Intraoperative evidence of didelphys uterus.

DISCUSSION

A didelphys uterus is an extremely rare MDA compared to other anomalies, according to the Buttram and Gibbons classification of 1978. For diagnosis, very invasive methods such as hysteroscopy, hysterosalpingography or laparoscopy have been used.[10] The first sort of imaging that was routinely done was a 2D ultrasound. It was, however, insufficient for diagnosis because it cannot consistently distinguish between kinds of abnormalities. The diagnostic method was 3D/4D ultrasonography, which was non-invasive and allowed assessment of both the endometrial cavity and the uterine fundus in coronal view. In the diagnosis of MDAs, magnetic resonance imaging is as accurate as other invasive modalities. Its non-invasive and it can also detect linked urinary tract disorders at the same time. Despite this, subjectivity, slight changes in morphology and shifting classification systems make it difficult to discriminate between these distinct anomalies on imaging modalities.[4]

Despite these challenges, Grimbizis et al found that the septate uterus was the most prevalent, accounting for 35% of all uterine abnormalities, followed by bicornuate at 25%, arcuate at 20%, unicornuate at 9.6% and complete agenesis at 3%. Didelphys uterus was shown to be the second least frequent MDA, accounting for 8.3% of all MDAs.[4] Chavan et al found 36.6 percent septate uterus, 23.3 percent arcuate uterus, 20 percent uterine didelphys, 13.3 percent unicornuate uterus and 6.6 percent uterus bicornuate in another study from India.[17]

The Mullerian ducts had completely failed to merge, resulting in two uterine chambers and two cervices in a didelphys uterus. There was also a longitudinal vaginal septum, which can be thin and easily moved or thick and inelastic. A standard speculum exam, when observation of anatomical abnormalities demanded further study, usually led to initial suspicion of the disorder, followed by diagnosis. Furthermore, because Mullerian ducts frequently form in combination with Wolffian ducts, kidney abnormalities may be discovered alongside uterine abnormalities.[1,2]

The majority of women with a didelphys uterus were asymptomatic, but a thick, occasionally blocking vaginal septum can cause dyspareunia or dysmenorrhea. This obstructive vaginal septum can cause hematocolpos/ hematometrocolpos, which can cause chronic abdominal pain.[1,2,7]

Untreated didelphys uterus had a greater fertility and pregnancy rate than other Mullerian anomalies, but it was less than women with a normal uterus. There was a higher risk of spontaneous abortion, foetal growth retardation, and preterm as well as a decreased likelihood of carrying a pregnancy to term (45 percent). The results were equivalent to unicornuate uterus; however, they were poor when compared to the more common MDAs of septate and bicornuate uterus.[1,2,5,8]

In a prospective observational study of the reproductive outcome of women with various uterine anomalies compared to a normal uterus, Acién et al discovered that the rate of term delivery for a didelphys uterus was significantly lower than the normal uterus group, but not as low as the bicornuate and septate groups.[5] In a review of the clinical implications of uterine abnormalities, Grimbizis et al validated this conclusion.[4] Ludmir et al observed that when high-risk obstetric intervention was used, more pregnancies from a didelphys uterus reached term and the foetal survival rate was higher than in the bicornuate and septate groups.[9]

Metroplasty and longitudinal vaginal septum excision are two procedures that can be used to promote fertility, reduce the risk of preterm, and improve overall quality of life. Surgical repair of a didelphys uterus is usually not recommended unless all other options for improving fertility have been exhausted.[4,5,10] If the lady is symptomatic, with dyspareunia or pain from hematometrocolpos due to blockage, longitudinal vaginal septum excision may be explored. Some septa are thin and elastic, allowing for vaginal birth, whereas others are thick and inelastic, increasing the likelihood of vaginal dystocia and necessitating excision. The presence of a didelphys uterus is not a reason for a caesarean section, thus vaginal birth should be considered first.[11-13] Finally, because didelphys uterus is rarely associated with cervical incompetence, cerclage is rarely utilised unless there is a history of cervical incompetence or premature dilation is discovered on inspection during the early second trimester.[2,5,9]

Didelphys uterus has only been reported in a few case reports as part of the Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA). The triad of didelphys uterus, blocked hemivagina, and ipsilateral renal agenesis characterises this unusual foetal abnormality involving Mullerian ducts and Wolffian structures.[14] This syndrome can result in hematocolpos or hematometrocolpos on the side of an obstructed hemivagina, resulting in a mass effect and lower abdominal pain.[6,15,16]

CONCLUSION

In compared to other more typical Mullerian duct anomalies, the didelphys uterus is a highly unusual Mullerian duct aberration with varied reproductive and gestational consequences. The ability to conceive is also a contentious topic. Surgical correction (metroplasty) is not normally recommended due to a lack of data; nevertheless, if the women are symptomatic, excision of the vaginal septum may be required. The presence of a Didelphys uterus is not a reason for a caesarean section unless the vaginal septum is thick and inelastic, increasing the risk of vaginal dystocia. Cervical incompetence hasn't been linked to the didelphys uterus in any studies. Finally, if a didelphys uterus is seen, renal abnormalities should be

looked at to rule out the Herlyn-Werner-Wunderlich (HWW) syndrome.

Overall, there is a scarcity of literature on the didelphys uterus at the moment. As a result, further research is needed to better understand the reproductive and gestational outcomes so that doctors may provide appropriate advice and treatment to their patients.

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