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Didelphys Uterus - A Case Report and Review of the Literature

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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 MDAs. This report discusses a case of didelphys uterus that successfully conceived.

Keyword: MDA, OPD, UTERUS

Introduction

Mullerian duct anomalies (MDAs) are congenital defects of the female genital system that arise from abnormal embryological development of the Mullerian ducts. These abnormalities can include failure of development, fusion, canalization, or reabsorption, which normally occurs between 6 and 22weeks in utero. Most sources estimate an incidence of these abnormalities to be from0.5 to5.0%inthe general population [1–4]. Septate uterus is the commonest uterine anomaly with a mean incidence of ~35% followed by bicornuate uterus (~25%) and arcuate uterus (~20%) [4]. Uterine anomalies may have a part in the delayed natural conception of women with mainly secondary infertility [4]. It is generally accepted that having a uterine anomaly is associated with poorer pregnancy outcomes such as increased chances of spontaneous abortion, premature labor, cesarean delivery due to breech presentation, and decreased live births, compared to a normal uterus [1–5]. However, the degree of these outcomes varies among different types of uterine anomalies. Uni cornuate and didelphys uterus have term delivery rates of ~45%, and the pregnancy outcome of patients with untreated bicornuate and septate uterus is also poor with term delivery rates of only ~40% [4]. Arcuate uterus is associated with a slightly better but still impaired pregnancy outcome with term delivery rates of ~65% [4]. Most women with a didelphys uterus are asymptomatic, but some present with dyspareunia or dysmenorrhea in the presence of a varying degree of longitudinal vaginal septum. Rarely, genital neoplasms, hematocolpos/hematometrocolpos, and renal anomalies are reported in association with didelphys uterus. Despite some of these complications, there are many cases of women with a didelphys uterus that did not exhibit any reproductive or gestational challenges. When classifying these anomalies solely based on abnormal development, four major types are apparent. 2. Presentation of Case This patient is 22year old Female Primigravida with two months Amenorrhea Presented in OPD with complain of

bleeding per Vaginum, Sudden in onset, not associated with Pain, No history of trauma. On PV Examination- Non communicating thick Vertical Vaginal Septum felt with two Vaginal cavity and two cervices. To confirm P/V Findings USG Advised & done which showed bicornuate uterus with Missed aboration of approximate 8 weeks. For furthers evaluation of other associated congenital anomalies, CT scan of whole abdomen advised – no renal anomalies or other congenital anomalies seen, except didelphys uterus.

- Dilatation & Evacuation done after explaining the condition of patient to attendants with due verbal & Written Consort along with all necessary investigation done.
- Post D & C patient condition was stable
- Patient was discharged after six hour of observation post op.

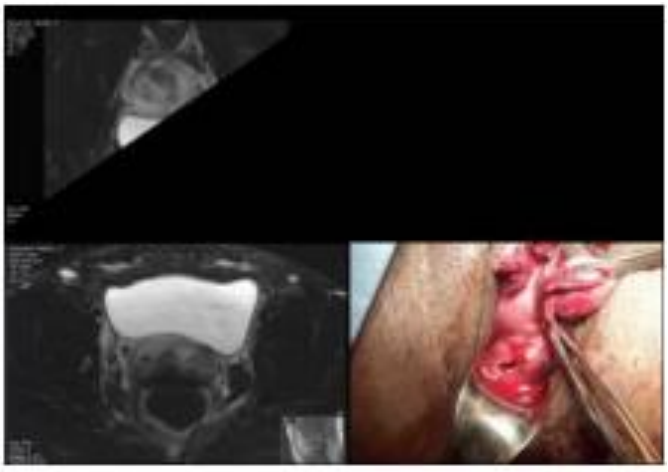

Figure 1

Discussion A didelphys uterus remains a very rare Mullerian duct anomaly in comparison to other anomalies described in the Buttram and Gibbons classification. Most of the data on the clinical significance and outcomes of this uterine anomaly are based on small retrospective, observational, or case studies. The results of these studies are mixed, not only due to the types of studies, but also due to the very low incidence of the anomaly in the population and the fact that more research has been directed to the more common malformations: arcuate, septate, bicornuate. Figure 4: MRI of abdomen and pelvis with contrast: Series # 5, T2 axial FS: one cervix on the right and one cervix on the left, 2 separate cervices. Figure 5: MRI of abdomen and pelvis with contrast: Series # 4 coronal FS (Fast): right uterus and left bulky postpartum uterus. Most women with a didelphys uterus are asymptomatic but may present with dyspareunia or dysmenorrhea in the presence of a thick, sometimes obstructing, vaginal septum. This obstructing vaginal septum can lead to hematocolpos/hematometrocolpos and thus present as chronic abdominal pain as well. Rarely, genital neoplasms and endometriosis are reported in association with cases of didelphys uterus [1, 2, 14]. The fertility of women with untreated didelphys uterus has been shown by some sources to be better than those with other Mullerian duct abnormalities but still less than women with normal uterine anatomy. There is also an increased risk of spontaneous abortion, fetal growth retardation, and prematurity with an estimated 45% (or lower) chance of carrying a pregnancy to term in comparison to a normal uterus, which is similar to that of a uni cornuate uterus. This indicates poor reproductive performance, but still not as poor as a septate or bicornuate uterus which are more common amongst the MDAs [1, 2, 5, 15]. The body of literature on didelphys uterus, although limited, generally shows that the anomaly may lead to better pregnancy outcomes in comparison to the other anomalies; however, there are also studies that demonstrate the contrary. For example, Acien's prospective observational study [5] of the reproductive outcome of women with different uterine anomalies in

comparison to a normal uterus found the rate of term delivery for a didelphys uterus significantly lower than the normal uterus group but the rate was not as 4 Case Reports in Obstetrics and Gynecology low as that of the bicornuate group and septate group [5]. Grimbizis et al. also confirmed this conclusion in a review on the clinical implications of uterine malformations [4]. Another study by Ludmir et al. also found, with high-risk obstetric intervention, more pregnancies from a didelphys uterus reached term and fetal survival rate was higher in comparison to the bicornuate and septate group [16] On the other hand, a large retrospective longitudinal study of 3181 patients by Raga et al. demonstrated poor reproductive performance in women with didelphys uteri with a higher rate of pre term delivery, spontaneous abortion, and the lowest chance of having a term delivery than the other MDAs[3]. In addition, a long term retrospective follow-up of 49 women with didelphys uterus found no impairment with fertility and decreased rate of spontaneous abortion; however the rate of prematurity was increased in comparison to other known studies on septate and bicornuate uteri [2] . The association between having a Mullerian duct anomaly and fertility is debatable. The review by Grimbizis demonstrated the incidence of MDAs in infertile patients (3.4%) similar to that of the general population and/or fertile women (4.3%), which they concluded demonstrated that MDAs may not have a negative impact on fertility [4]. To go further, there are reported cases of women with didelphys uteri pregnant with twins or triplets demonstrating the ability to conceive and support the healthy growth of a fetus in either one of the uteri [17–20]. In contrast, the large retrospective study done by Raga et al. found the incidence of Mullerian duct anomalies to be significantly higher in infertile women than in fertile women, suggesting a link between infertility and the MDA [3]. A retrospective study on fertility and obstetric outcome done by Zhang et al. in China demonstrated that women with a didelphys uterus more frequently required infertility treatments than with other anomalies to conceive [21]. Certain procedures may be undertaken to increase fertility, decrease chances of prematurity, and improve the quality of life. Surgical correction of a didelphys uterus (metroplasty) is not usually indicated and the literature on women with didelphys uterus who underwent metroplasty is very limited. With that said, metroplasty would only be considered on a case-by-case basis after all other ways in which reproductive performance could be improved are exhausted [4, 5, 22]. Observational studies cite women with septate or bicornuate uteri with a history of repeated abortions and infertility demonstrating improvement in reproductive and gestational outcome after metroplasty [4]. Longitudinal vaginal septum excision is considered if the woman is symptomatic, complaining of dyspareunia or pain from hematometrocolpos due to obstruction. Some septa can be easily displaced to the side to facilitate vaginal birth and others may be thick and inelastic, increasing the risks of vaginal dystocia and thus requiring excision. A didelphys uterus is not an indication for cesarean delivery and thus vaginal delivery should be considered first [23–25]. Finally, cervical incompetence is not usually associated with didelphys uterus and thus cerclage is not routinely used unless there is a history of cervical incompetence or premature dilation is found on exam during early second trimester [2, 5, 16]. A didelphys uterus has been shown in many case reports to occur as a part of a syndrome, more specifically called, Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemi vagina and

ipsilateral renal anomaly (OHVIRA). It is a very rare congenital anomaly of the urogenital tract involving Mullerian ducts and Wolffian structures, and it is characterized by the triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis [26]. This condition can cause hematometrocolpos or hematocolpos on the side of the obstructed hemivagina which produces amass effect with subsequent lower abdominal pain [8, 27, 28]. Most cases present after menarche as intense lower abdominal pain and/or a protruding mass over the vaginal introitus [8, 27, 28]. Sudden, intense vaginal pain has been documented as a rare presenting symptom as well [28]. A preliminary pelvic Ultrasound is done followed by an MRI to confirm the diagnosis. One case report identified this syndrome in a newborn who was diagnosed with renal agenesis in utero and born with a protruding vaginal mass and a hydrocolpos was found on imaging [29]. Although this condition is extremely rare, it is important for a physician, especially an ER physician, to keep it in mind when a postpubertal female presents with sudden lower abdominal pain and all other causes have been ruled out [8, 27, 28].

Conclusion The didelphys uterus is a very rare Mullerian duct anomaly with varying reproductive and gestational outcomes in comparison to other more common abnormalities. The ability to conceive remains a debatable issue as well. There is insufficient data on surgical correction (metroplasty); therefore, it is not usually indicated; however, excision of the vaginal septum may be required if the women is symptomatic. Didelphys uterus is not an indication for cesarean delivery unless the vaginal septum is thick and inelastic resulting in an increased risk for vaginal dystocia. Cervical incompetence has not been shown to occur in conjunction with the didelphys uterus. Lastly, when a didelphys uterus is diagnosed, renal anomalies should also be investigated to rule out Herlyn-Werner-Wunderlich (HWW) syndrome. Overall, the literature available on the didelphys uterus is quite limited at the present time. Therefore, more studies are needed in order to better determine the reproductive and gestational outcomes, so that clinicians can adequately advise and care for their patients.

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