

A PRESENTING OF UTERUS DIDELPHYS: A CASE REPORT

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ABSTRACT: *This is a case of double uterus reported by a student in Chennai, India. The patient is a 18year old female from Malaysia of an Indian descent who is of normal health condition. Uterus didelphys (sometimes also uterus didelphys) represents a uterine malformation where the uterus is present as a paired organ as the embryo genetic fusion of the mullerian ducts failed to occur. As a result there is a double uterus with two separate cervix, and often a double vagina as well. Each uterus has a single horn linked to the ipsilateral fallopian tube that faces its ovary. The uterus is formed during embryogenesis by the fusion of the two paramesonephric ducts (also called mullerian ducts). This process usually fuses the two mullerian ducts into a single uterine body but fails to take place in these affected women who maintain their double mullerian systems. A didelphic uterus will have a double cervix and is usually associated with a double vagina. Causes for the failure to fuse are not known. Associated defects may affect the vagina, the renal system, and less commonly, the skeleton. The condition is less common than these other uterine malformations: arcuate uterus, septate uterus, and bicornuate uterus. It has been estimated to occur in 1/3,000 women. The findings of this case study is that the double uterus condition is not abnormal and if the patient suffers acute pain then the patient will need to seek medical help and may in extreme cases need corrective surgery. This is evidence that the double uterus condition is not abnormal and that it can be managed if detected. In most of the cases unless a pelvis scan is done this will not be detected.*

KEYWORDS: *Uterus didelphys, embryogenic, ipsilateral fallopian tube, paramesonephric*

1. INTRODUCTION

Uterus didelphys (sometimes also uterus didelphys) represents a uterine malformation where the uterus is present as a paired organ as the embryo genetic fusion of the mullerian ducts failed to occur. As a result there is a double uterus with two separate cervix, and often a double vagina as well. (Madureira et al., 2006)Each uterus has a single horn linked to the ipsilateral fallopian tube that faces its ovary. The uterus is formed during embryogenesis by the fusion of the two paramesonephric ducts (also called mullerian ducts).(Kim et al., 2007) This process usually fuses the two mullerian ducts into a single uterine body but fails to take place in these affected women who maintain their double mullerian systems. A didelphic uterus will have a double cervix and is usually associated with a double vagina. Causes for the failure to fuse are not known. Associated defects may affect the vagina, the renal system, and less commonly, the skeleton.(Golan et al., 1989)The condition is less common than these other uterine malformations: arcuate uterus, septate uterus, and bicornuate uterus. It has been estimated to occur in 1/3,000 women. Women with the condition may be asymptomatic and unaware of having a double uterus.

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However, a study by Heinonen showed that certain conditions are more common. (Burgis, 2001) In his study of 26 women with a double uterus gynecological complaints included dysmenorrhea and dyspareunia. All patients displayed a double vagina. The fetal survival rate in 18 patients who delivered was 67.5%. Breech presentation was present in 43% and premature delivery common (21%) (García González et al., 2009; Jindal et al., 2009). A pelvic examination will typically reveal a double vagina and a double cervix. Investigations are usually prompted on the basis of such findings as well as when reproductive problems are encountered. Not all cases of uterus didelphys involve duplication of the cervix and vagina. Helpful techniques to investigate the uterine structure are transvaginal ultrasonography and sonohysterography, hysterosalpingography, MRI, and hysteroscopy. (Olpin and Heilbrun, 2009) More recently 3-D ultrasonography has been advocated as an excellent non-invasive method to evaluate uterine malformations. Uterus didelphys is often confused with a complete uterine septum. Often more than one method of investigation is necessary to accurately diagnose the condition. Correct diagnosis is crucial as treatment for these two conditions is very different. (Kaufman and Lam, 2008; Rock and Breech, 1997) Whereas most doctors recommend removal of a uterine septum, they generally concur that it is better not to operate on a uterus didelphys. In either case, a highly qualified reproductive endocrinologist should be consulted. Patients with a double uterus may need special attention during pregnancy as premature birth and malpresentation are common. Cesarean section was performed in 82% of patients reported by Heinonen. (Asha and Manila, 2008) Uterus didelphys, in certain studies, has also been found associated with higher rate of infertility, spontaneous abortion, intrauterine growth retardation, and postpartum bleed. A number of twin gestations have occurred where each uterus carried its pregnancy separately. (Acién, 1993) A recent example occurred on February 26, 2009, when Sarah Reinfelder of Sault Ste. Marie, Michigan delivered two healthy, although seven weeks premature, infants by cesarean section at Marquette General Hospital. It is possible that the deliveries occur at different times, thus the delivery interval could be days or even weeks. A UK woman with a double uterus gave birth to triplets in 2006. Hannah Kersey, of Northam in Devon, gave birth to a pair of identical twins from an egg that implanted into one womb and then divided, and to an infant from a single egg that implanted into the other womb. This was the first known birth of viable triplets in a woman with a double uterus. (Chandler et al., 2009; Madureira et al., 2006) It is estimated that the possibility of such a birth is about 1 in 25 million. A triplet pregnancy in a woman with uterus didelphys was reported from Israel in 1981; one baby died in utero, and of the remaining babies, one was delivered at 27 weeks gestation and the other 72 days later.

2. CASE PRESENTATION

This patient is an 18-year-old female of Indian descent. She has been suffering from lower abdomen pain during her menstrual cycle. This patient has been having her regular menstrual cycle every month but with heavy flow. Moreover, this patient has always been having severe abdomen pain when she reached puberty and since then she has consulted a General Practitioner and that respective person told the patient that it's normal to have pains during the menstrual cycle. But the pains got stronger and severe every time she had her menstrual cycle and the patient would fall sick such as have fever, vomit and diarrhea. So, she decided to consult a doctor in India as she is studying in India too. The doctor decided to do a pelvic scan and when the report came back the diagnosis was found to be a bicornuate uterus. Then the doctor explained to the patient about what the diagnosis was and said that it could be treated and she need not worry. Therefore, the patient is on medications such as T. Trapic and Martifur.

Figure 1:



Figure 2:

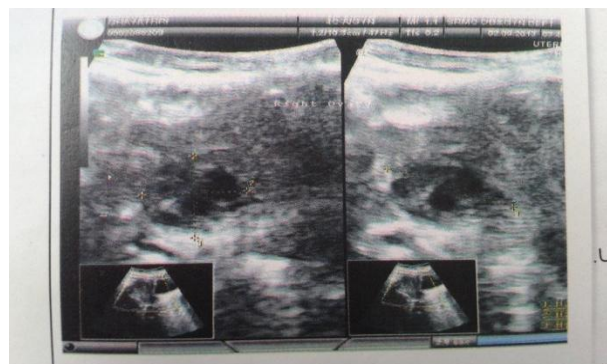


Figure 3:



Bicornuate Uterus

3. DISCUSSION

There is a high association between mullerian duct and renal anomalies such as unilateral agenesis. Routine imaging for infertility workup often detects the anomaly. Uterus didelphys with obstructed hemivagina is an obstructive mullerian anomaly caused by the lateral nonfusion of the mullerian ducts with asymmetrical obstruction. (Buttram and Gibbons, 1979) Resection of the vaginal septum is the treatment of choice for obstructed hemivagina for symptom relief and preservation of reproductive capabilities. Hysteroscopic resection of the septum under transabdominal guidance may also be carried out so as to preserve the hymenal integrity in young females. Didelphic uterus have the highest association with

transverse vaginal septum.(Fertility and sterility, 1988; Woelfer et al., 2001)Renal agenesis most commonly occurs in association with uterine didelphys than with any other type of mullerian anomaly.

The reported incidence of renal anomalies in this group is 20%. OHVIRA syndrome represents obstructed hemivagina with ipsilateral renal anomaly and is classically associated with uterus didelphys. Obstructed unilateral vagina in patients with uterus didelphys is frequently associated with ipsilateral renal and ureter agenesis.(Raga et al., 1996)The role of imaging is to help detect, diagnose and distinguish surgically correctable forms of mullerian malformations from inoperable forms Abdominal or transvaginal ultrasound is cheap and noninvasive. Three-dimensional (3D) has higher sensitivity and specificity to evaluate malformations. Computed tomography (CT) has a limited role in the evaluation of the female pelvis, whereas magnetic resonance imaging (MRI) is highly sensitive. The vaginal septum is difficult to visualize on ultrasound and is best shown on MRI.Uterus didelphys has been associated with higher rate of infertility, spontaneous abortion, intrauterine growth retardation, preterm labor and postpartum bleed.(Takagi et al., 2003)Patients may present with severe dysmenorrhoea, lower abdominal pain, paravaginal mass, excessive foul smelling mucopurulent discharge and intermenstrual bleeding depending on the existence of uterine or vaginal communications. In patients presenting with palpable abdominal, pelvic or vaginal mass (mucocolpos or pyocolpos), mullerian duct anomalies must be excluded. The diagnosis of this rare mullerian anomaly, also known as Herlyn-Werner-Wunderlich Syndrome (HWWS) should be suspected in women who have ipsilateral renal agenesis with a pelvic mass.(Martínez-Beltrán et al., 2012)++

4. CONCLUSION

There is a high association between mullerian duct and renal anomalies such as unilateral agenesis. Patients most often present for medical attention because of infertility , repeated pregnancy loss or severe abdomen pain with heavy flow during menstruation.(Raga et al., 1997) Uterus didelphys with obstructed hemivagina is an obstructive mullerian anomaly caused by the lateral nonfusion of the mullerian ducts with asymmetrical obstruction.(Karaca et al., 2015) Renal agenesis most commonly occurs in association with uterine didelphys than with any other type of mullerian anomaly.

ACKNOWLEDGEMENT

The authors of this study acknowledge the institute, for their help towards collecting all the patient case records and other datas in relevance to the current study.

CONFLICT OF INTEREST

The authors declare that there was no conflict of interest.

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