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Pregnancy in Mullerian anomaly – A diagnostic dilemma

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ABSTRACT

Aim: To report rare case of pregnancy in Mullerian Anomaly of the uterus and to understand the concept of precise diagnosis for rare cases.**Materials and Methods:** This is a case report at CIMS, Lucknow of patient who came to our hospital at first trimester with history of recurrent abortions and preterm delivery. She was diagnosed to have cervical incompetence initially until her abdomen was opened for LSCS when rare Mullerian Anomaly was seen to be the cause of previous obstetric history.**Results:** Rare congenital anomaly was reported and the bad obstetric outcomes were known to be the symptoms of the rare cause.**Conclusions:** We need to broad our diagnosis with the fact that rare cases can also be the cause of worst conditions or some initial primary conditions.This is an Open Access (OA) journal, and articles are distributed under the terms of the [Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License](https://creativecommons.org/licenses/by-nc-sa/4.0/), which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.For reprints contact: reprint@ipinnovative.com

1. Introduction

Among the normal fertile women the incidence of Müllerian defects varies currently from 7% to 8% and more than 25% among women with recurrent miscarriage. However, in 5% of general population clear uterine malformations are observed in 2% to 3% of fertile women, in 3% of infertile women and in 5% to 10% of patients with recurrent miscarriage.¹

Congenital uterine anomalies (CUAs) may lead to symptoms such as pelvic pain, prolonged abnormal bleeding, recurrent pregnancy loss, or preterm birth and thus may be identified in patients, including adolescents, who present with these disorders. Some Congenital uterine anomalies may be suspected because of associated findings on physical examination, such as a longitudinal vaginal septum. Others may be detected when imaging studies are

performed to evaluate patients with infertility.

The classification and clinical manifestations of major congenital anomalies of the uterus (septate, unicornuate, bicornuate, and didelphys uterus), along with their potential associated cervical and vaginal anomalies, could be understood. The treatment of these anomalies should be known by the obstetricians.

Cervical agenesis or hypoplasia, congenital vaginal abnormalities, and diethylstilbestrol (DES)-related anomalies should also be understood by the obstetricians for a clear diagnosis.

One of the rare Mullerian anomaly is unicornuate uterus with an incidence of 0.06%.² A unicornuate uterus development is from partial or complete lack of development of one Mullerian duct between the 7th and 8th week of gestation.

Unicornuate uterus is rare congenital anomalies of uterus. In this only 1 Mullerian duct is present. Because of

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Fig. 1: Mullerian Anomaly - Bicornuate uterus

1 Mullerian duct female has $\frac{1}{2}$ of uterus, 1 fallopian tube, $\frac{1}{2}$ cervix, and $\frac{1}{2}$ upper vagina. There can be complete agenesis, on communicating or non-communicating rudimentary horn on other side. Mullerian duct also known as paramesonephric duct are embryological structures for developing urogenital system. The presence of any maternal uterine anomaly is associated with an increased risk of preterm birth, placenta previa, breech presentation, preterm premature rupture of membranes, cesarean section, placental abruption and intrauterine growth retardation (IUGR).³

A unicornuate uterus is a type II WHO classification with unilateral hypoplasia or agenesis that can be further classified into communicating, non-communicating cavity and no horn.⁴ In 83% of cases, the rudimentary horn is non-communicating and often associated with ectopic pregnancies.⁵ Pregnancy in non-communicating rudimentary horn is still possible. This can be seen by transperineal migration of sperm or fertilized ovum. In approximately 1 out of 76,000 pregnancies this could happen. The risk of uterine rupture occurs mostly in second trimester which could be approx 50 to 90%.⁶

The clinical presentation of women with a unicornuate uterus is variable owing to the heterogeneity of the four morphological variants. It has been reported previously that in most cases, there is a functional non-communicating rudimentary horn, there can be non-functional rudimentary horn.⁷ Severe dysmenorrhea and pregnancy and obstetric

complications are common in women with functional rudimentary horn because of presence of endometrium which is also functional⁸ and they are also at risk of developing chronic pelvic pain and endometriosis at a young age.^{9–11} Unicornuate uterus is often associated with renal anomalies, occurring in 40% of cases,¹² with renal agenesis being the most common,¹³ but our patient had no such abnormalities.

A unicornuate uterus or unicornis unicollis is a type of Mullerian Duct Anomaly (class II). It is characterized by a banana-shaped uterus usually draining into a single fallopian tube.¹⁴ It can be classified into the following types according to the American Fertility Society-¹⁵

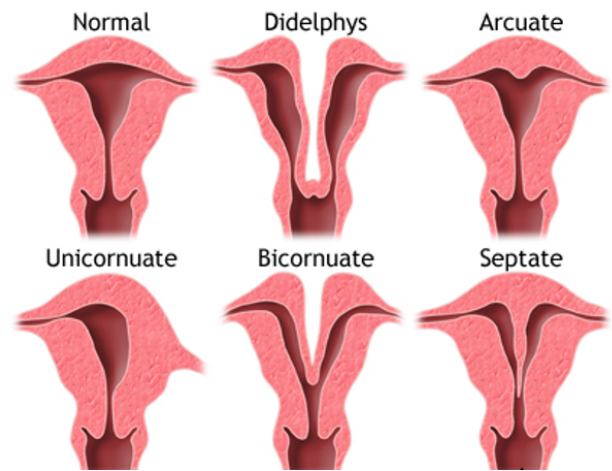


Fig. 2: Types of Mullerian Anomaly¹⁶

Type a: with rudimentary horn.

a1: horn contains endometrium.

a1a: communicating contralateral rudimentary horn contains endometrium **a1b:** non-communicating contralateral rudimentary horn contains endometrium **a2:** contralateral horn has no endometrial cavity.

Type b: no horn

2. Who classification

1. Class I- Agenesis of uterus
2. Class II- Unicornuate uterus
3. Class III- Uterine didelphy
4. Class IV- Bicornuate uterus
5. Class V- Septate uterus
6. Class VI- Arcuate uterus
7. Class VII- DES.

Unicornuate uterus is major cause for spontaneous abortion, preterm labour & malpresentation. This is considered under high risk pregnancy which make challenging for women to have favourable pregnancy. In women already diagnosed with uterus anomaly antenatal monitoring should be aggressive.

The septate uterus is the most common uterine anomaly among the other mullerian anomalies in the general population and is also associated with the poorest reproductive outcomes. It is composed of fibro muscular tissue, because of which the septum has a decreased blood supply, which may lead to poor implantation, resulting in early pregnancy loss. Later in gestation, the septum may compromise the available space for growth, leading to pregnancy loss, malpresentation, or preterm birth.

Studies suggest that resection of the uterine septum results in higher delivery rates than in women without treatment. A septum incision by hysteroscopy, or metroplasty, is the treatment of choice for women with a septate uterus. A large meta-analysis found in a study conducted that hysteroscopic resection of the uterine septum increased live birth rates from 6.1% before surgery to 83.2% after metroplasty. The benefits of hysteroscopic metroplasty have not yet been assessed by a prospective randomised trial.¹⁷ Other congenital abnormalities, such as bicornuate and unicornuate uterus, are more frequently associated with later trimester losses or preterm delivery than recurrent abortions.

One such case of unicornuate uterus is described in following report.



Fig. 3: Closure of Unicornuate uterus after delivery of fetus

3. Aim

To report a rare case of unicornuate uterus with history of recurrent abortion and preterm labour.

4. Materials and Methods

This is a case report at Career institute of medical science and hospital Lucknow of 33yr old female, home maker by occupation socio-economically lower class. She came to OPD at 16wks of gestation for the first time in current pregnancy. She had a history of recurrent abortions and 1 preterm delivery 1.5 yrs back at 7 months of gestation of which baby did not survive. Her previous 4 abortions were spontaneous. Her obstetrics score was G6P1L0A4. Her duration of marriage was 4yrs.

In the current pregnancy major precautions and aggressive monitoring was done. Her antenatal investigations were done. She was immunized with tetanus toxoid at 18 weeks of gestation & Mc-Donald Cerclage was done at same time. Her Targetted imaging for fetal anomalies (TIFFA) scan was done and no congenital anomaly in fetus was reported. To prevent preterm delivery- from her first visit in OPD she was started on Progesterone, Isoxspurine, Anafortan. Close monitoring was done of the patient, and she was asked bed rest keeping in mind her bad obstetric history. There was no history of illness in the past, there was no significant family history.

At 29 weeks of gestation, she complained of pain in her abdomen so was admitted & injection progesterone and isoxspurine were given. She was discharged on Day 10 of her admission, when she was symptomatically better.

She was admitted and planned for emergency LSCS at 36 weeks of gestation when she complained of decreased fetal movement and pain abdomen. Her USG done the same day was suggestive of oligohydramnios (AFI-5cm). Her cerclage was opened. On opening the abdomen by pfennestiel incision and following the same standard steps for lower segment cesarean section all answers to her bad obstetric history were now known. She was diagnosed with a Unicornuate uterus with non-communicating rudimentary horn. Her incision was increased in J manner to deliver the baby. No other complications were seen. Bleeding was within normal limit blood loss was about 1000 ml.

A baby girl weighing 2.2kg was delivered and cried immediately after birth. APGAR Scores 6 and 9 at 1 and 5 minutes, respectively. Antibiotics, IV fluid, and analgesics were given to the mother after shifting to the intensive care unit (ICU). Breastfeeding started after 4 hours. Foley's was removed on Postoperative day 3. Her lab investigation post-delivery turned out normal. Her dressing was done on 4th and 8th day resulted in healthy stitch line. No induration was present.

The patient was discharged on advice for contraception, and exclusive breastfeeding for the newborn. The baby was advised to follow immunization chart for timely immunization.

Medication such as iron, calcium, protein powder, and vitamins were prescribed.

5. Results

With broad diagnosis once could avoid the risks in pregnancy. Mullerian anomaly of unicornuate uterus was found to be the cause of recurrent abortions and preterm labor.

6. Discussion

Knowing anatomy and embryology very important in congenital anomalies of uterus. Most of women are asymptomatic but some may experience menstrual irregularities, dysmenorrhea, (especially in unicornuate uterus with outline endometrium in rudimentary horn).

Women with anomalies uterus have less changes of normal pregnancy as it does not provide suitable physiological environment. It gives rise to obstetric complication such as infertility, ectopic pregnancy, spontaneous abortion.

Usually with such history of current case one usually thinks of cervical incompetence rather than any other cause. But the need to elaborate out D/Ds for such history is very important.

7. Conclusion

In this case report after repeated bad obstetric history of the patient it was seen that the patient had unicornuate uterus which led to such obstetrical outcomes. Patients who have a unicornuate uterus with rudimentary horn have an increased incidence of gynecologic problems at menarche or later in their life, they present with symptoms such as dysmenorrhea and chronic pelvic pain.⁶ This is why patients with such symptoms should be screened with 2D or 3D ultrasonography at an early age. However, the ultrasonography diagnosis of Mullerian anomalies could be missed at times. The reproductive outcome of women with a unicornuate uterus is very poor, but a successful pregnancy is possible. Excision of the rudimentary horn should be undertaken during the non-pregnant state laparoscopically, and it would be necessary to screen such pregnancies for the development of intrauterine growth retardation, which is very common in mullerian anomalies of the uterus. There is a need for increased awareness of this rare anomaly and a high index of suspicion, especially in developing countries where the possibility of early detection is low. There can be chances of rupture in pregnancy in mullerian anomalies, and that could be life-threatening.

Uterine malformation can have significant effect on outcome of pregnancy. High prevalence of abortion of PTLs and fetus malposition are associated with uterine malformation.

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None.

9. Conflict of Interest

None.

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