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Case Report

Bicornuate bicollis didelphic uterus with renal anomaly: pregnancy risks and complications

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Abstract

Congenital Müllerian duct anomalies affect the female genital tract, including the fallopian tubes, uterus, cervix, and/or upper vagina. These anomalies occur due to agenesis or failed fusion of the paramesonephric ducts or failed resorption of the uterine septum during fetal development. The incidence of Müllerian duct anomalies ranges from 0.5% to 5.0% [1].Bicornuate uterus accounts for approximately one-fourth of these anomalies.

Keywords: ASRM,OHVIRA syndrome, Mullerian anomaly, vaginal septum ,Herlyn-Werner-Wunderlich syndrome

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1. Introduction

Congenital Müllerian duct anomalies affect the female genital tract, including the fallopian tubes, uterus, cervix, and/or upper vagina. These anomalies occur due to agenesis or failed fusion of the paramesonephric ducts or failed resorption of the uterine septum during fetal development. The incidence of Müllerian duct anomalies ranges from 0.5% to 5.0%.[1] Bicornuate uterus accounts for approximately one-fourth of these anomalies, while didelphic uterus, or "double uterus," is one of the rarest, representing only 8% of cases.[2] Genetic factors, hormonal imbalances, or environmental influences during embryonic development can contribute to Müllerian anomalies. A rare congenital anomaly also seen where a ureter inserts into a blind-ending hemivagina, associated with an atrophic kidney.

Anomaly arises congenitally due to abnormal migration of the ureteric bud during its insertion into the urinary bladder.[3]

Symptoms vary depending on the type of anomaly, and may include infertility, recurrent miscarriages, painful periods, preterm labor, delivery complications, and fetal malpresentation,Recurrent UTI.[4] Research has also linked congenital Müllerian duct anomalies to an increased risk of renal and urinary tract abnormalities.[5]

According to a review by Saravelos et al., the prevalence of congenital uterine anomalies is 6.7% in the general population, 16.7% in women with recurrent miscarriage, 7.3% in infertile women[6] Incomplete resorption of uterine or vaginal septa can lead to rudimentary and obstructed uterine horns or obstructive hemivagina and ipsilateral renal anomaly, also known as OHVIRA or Herlyn-Werner-Wunderlich syndrome.[7,8]

The exact incidence of transverse vaginal septum is unknown, but reported cases range from 1 in 2,100 to 1 in 72,000, making it one of the rarest female genital tract anomalies. The etiology of this condition is unclear, although most cases are believed to result from sex-limited autosomal recessive transmission.[9-10] In this case report, we describe a nulliparous patient diagnosed with a rare

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Müllerian congenital anomaly, classified as U3BC2V3 according to the ESHRE/ESGE classification.[11]12 This anomaly consists of a double cervix, transverse vaginal septum, complete bicorporeal uterus, and right renal agenesis.

Ultrasound,MRI,CT SCAN assesses the uterine shape, size, and configuration, helping diagnose bicornuate didelphic bicollis uterus.Renal Anomaly Detection: MRI can detect associated renal anomalies, such as ectopic kidneys, renal agenesis, or horseshoe kidney.

MRI and CT scans play important roles in evaluating bicornuate didelphic bicollis uterus with renal anomaly. MRI is the preferred imaging modality due to its higher soft-tissue resolution, less radiation exposure, and multi-planar imaging capabilities. CT scans can gives additional information about renal defect and ureteral evaluation

The American Society for Reproductive Medicine (ASRM) has revised the old AFS (1988) classification, introducing the ASRM Müllerian Anomalies Classification (ASRM MAC 2021). This updated classification expands on the original AFS classification, dividing anomalies into nine distinct groups. However, even with this revision, the ASRM MAC 2021 may not encompass all known anomalies, given the vast number of possible variations.[12]

2. Case Report

A was a 24 years old lady who presented at Gynaecology outpatient clinic on account of inability to get pregnant for 9 years.There was no fever, no urinary or gastrointestinal symptom. She attained menarche at the age of 13 years and had a normal menstrual history with no complaints of excessive bleeding, dyspareunia, or vaginal discharge. Routine hematological laboratory tests were normal. Hysterosalp ingography showed uterine cavity appear small,no other structures noted.

The ultrasound examination of the pelvis confirmed bicornuate uterus with rudimentary left uterine horn. The general abdominal ultrasonography revealed absence of the Right kidney (Right renal agenesis): MRI of pelvis and abdomen shows uterus bicornuate with 2 separate cavity divided till the cervical level with left horn small rudimentary hypoplastic and cervix is also divided into 2 cavity suggestive of bicornuate bicollis, right kidney is not seen in right renal fossa and left kidney shows compensatory hypertrophy.Both ovary appears normal.

2.1. On local pelvic examination

1. **Per abdominal examination-** Abdomen soft, non-tender, umbilicus centrally placed, no dilated veins and scar mark, seen.
2. **Pervaginal examination –** External genitalia appear normal but a small hole seen just below the urethral meatus, Anormal 6 week size uterus with bilateral fornix free and clear and no abnormal discharge was seen.
3. **Per speculum examination-** Shows 2cervix with few nabothian cyts was present, no abnormal discharge present.

External genitalia appear normal but a small hole seen just below the Urethral meatus. Per speculum examination shows 2 cervix with few nabothian cyst was present. Per vaginal examination shows a normal 6 week size uterus with bilateral fornix free and clear and no abnormal discharge was present.Per abdominal examination- abdomen soft,non tender,umblicus centraly placed,no dilated veins and scar mark.

2.1. Operative procedure

In light of the findings, a diagnostic hysteroscopy and laparoscopic surgery followed by vaginal septal resection performed. The diagnostic hysteroscopy, at first glance, cervix opening negotiated with difficulty, fibrosis noted at internal os ,fibrosed part cut with hysteroscopic scissor, tabular cavity seen,pale endometrium and tubal ostia are not seen.(**Figure 1**,**Figure 2**)

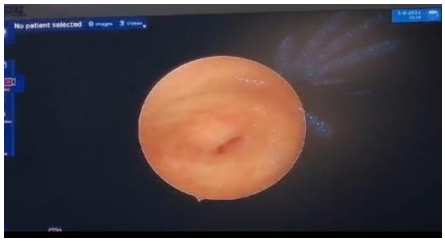

Figure 1: Shows cervical opening and fibrosis noted at internal os

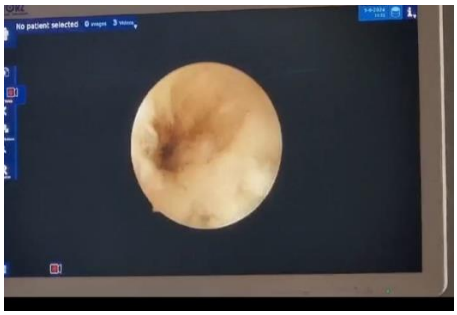

Figure 2: Shows cervical opening and fibrosis noted at internal os.

In Vaginoscopy 1 cervix opening noted with vagina appear normal.

The diagnostic laparoscopy revealed heart shaped uterus with left rudimentary horn, both tubes and ovaries seen normal(**Figure 3**)

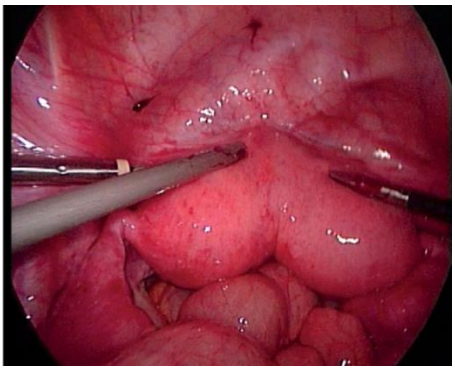

Figure 3: Heart shaped uterus

Perspeculum examination shows a small opening noted inferior to Urethral meatus,opening dilated by using hegar dilators, hysteroscope inserted into this opening,opening shows another cervix where endometrium appeared normal and tubal ostia seen.(**Figure 4**,**Figure 5**,**Figure 6**)

Chromopertubation test was positive in right side, vaginal septal resection was done.(**Figure 7**,**Figure 8**,**Figure 9**)

Figure 4: shows small opening inferior to Urethral meatus.

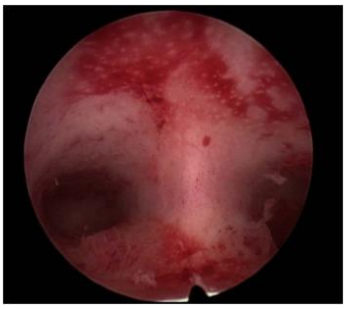

Figure 5: Another cervix shows normal endometrium and ostia.

Figure 6: Another cervix shows normal endometrium and ostia.

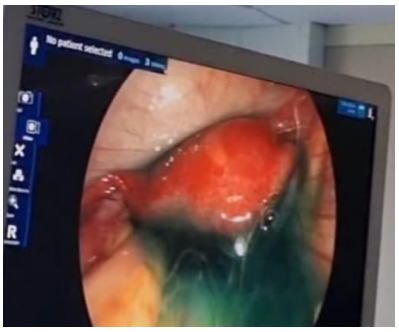

Figure 7: shows chromopertubation test positive in right side.

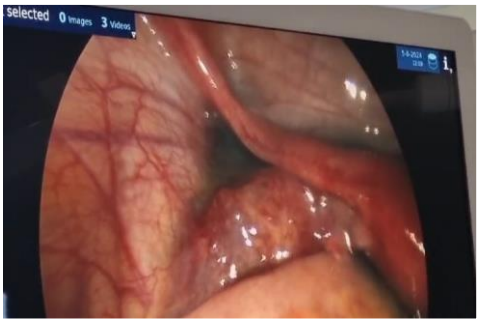

Figure 8: shows negative chromopertubation test in left side.

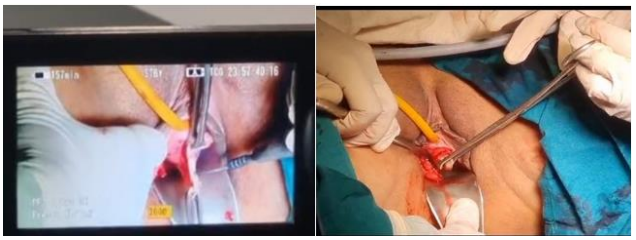

Figure 9: vaginal septal resection.

Clinical significance of this case can causes Emotional and psychological disturbance and affects the quality of life This condition can increase the risk of infertility, miscarriage, preterm labor, placenta abnormalities, intrauterine growth restriction. Abnormal uterine and renal anomaly can also increase the risk of recurrent urinary tract infections,chronic kidney injury.

3. Conclusion

The impact of common congenital anomalies of the female genital tract is hugely variable. Some Mullerian anomalies are easily diagnosed, but others have unusual presentations that make diagnosis and therapy difficult. A good knowledge of basic embryology is important for understanding the path-ogenesis and clinical features of these anomalies. All gyne-cologists should be aware of these conditions and their possible clinical presentations.

Multidisciplinary team work, regular follow-up, strict monitoring, proper treatment ,emotional support and care helpful for achieving successful pregnancy.

This type of incidence is noted to be rare in our tertiary care teaching college and hospital that it is the first case of its kind reported after a long time.

4. Discussion

We report the case of a Nulliparous patient with bicornuate bicollis uterine anatomy and unilateral renal agenesis.Bicornuate uterus is a common Mullerian duct anomaly and can be accompanied with a single cervix (unicollis) or a double cervix (bicollis) depending on the extent of the duplication.[8] Diferentiating bicornuate bicollis uterus from didelphic uterus can be challenging, as the anatomy of these anomalies is similar. Te key diference between these anomalies is that a didelphic uterus has two widely spaced and completely separate uterine cavities. By comparison, bicornuate anatomy demonstrates some degree of fusion between the two uterine horns, although the septum can extend to the level of the cervix to yield two cervices some cases.The bicornuate uterus is a more severe Whereas anomaly than septate uterus". in septate uterus, there is a soft tissue septum usually from the fundus downwards but not reaching the cervical region, partially dividing the uterine cavity into two, in bicornuate uterus there is complete separation of the two cornua of the uterine body which can either involve the cervix (bicollis) or spare it (unicollis)".[13]

Abnormal uterine anatomy has been well-documented and studied, and complex distal mesonephric congenital anomalies including cases of unilateral renal agenesis and ipsilateral cervicovaginal atresia or an ipsilateral blind hemivagina have been described.[14] Similarly, there have been cases with communicating bicornuate bicollis uterine anatomy associated with atretic blind hemivagina and ipsi-lateral renal agenesis.[14] Thus, in patients presenting with bicornuate uterine anatomy and unilateral renal agenesis, it is reasonable to suspect anomalies of this nature. This patient did not have evidence of any of the aforementioned cervical and/or vaginal fndings. Rather, the patient had apparently noncommunicating uterine horns with respective cervices; a true bicornuate bicollis anatomy with unilateral renal agenesis. This anatomy has not been documented in the literature and represents a very rare anomaly. Magnetic resonance imaging and three-dimensional computed tomography angiography are useful tools for diagnosing complex mullerian anomalies, and operative laparoscopy may be an appropriate alternative treatment for these cases.[15]

5. Source of Funding

None.

6. Conflict of Interest

None.

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