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CASE REPORT

U2bC1V2: A Rare Mullerian Abnormality—Complete Uterine Septum, Double Cervix and Complete Vertical Vaginal Septum

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Introduction

The abnormalities of Mullerian development are rare and have incidence of 1–6%. With advances in diagnostic techniques, such cases are diagnosed with more accuracy with a combination of 3D Ultrasound, MRI, Laparoscopy and Hysteroscopy.

We present a rare Mullerian duct abnormality where the woman had complete uterine septum, double cervix and complete longitudinal vaginal septum. According to American Fertility Society (AFS), such cases fall into unclassified group as only handful of such cases have been reported. However, the European Society of Human Reproduction and Embryology (ESHRE) classifies such anomalies. Since it is a rare and difficult case, the management needs to be customized for each patient's requirements. We discuss our case in detail.

Case History

A 28-year-old woman, married for 9 months presented to the OPD. Her chief complaint was inability to consummate her marriage. On Per Speculum examination she was found to have two vaginas and two cervices. Ultrasound examination revealed two uterine cavities raising suspicion of uterus didelphyus. The vaginal septum was complete. Hence it was decided to do a vaginal septal resection and planned for hysteroscopy and laparoscopy for making a definitive diagnosis.

Patient was given general Anaesthesia after consent. A 5 mm port was put in umbilicus and laparoscope was introduced. We could visualize only single uterus with no dip on fundus. The fundus was smooth. Both ovaries looked normal. Right tube had mild hydrosalphinx.

The vaginal septum was held on top and bottom by long clamps and cauterised. The septum was completely divided till both cervices were seen together and then hysteroscopy was carried out. The hysteroscope was first introduced in right and then left cavity. The cervical canals were separate indicating that the uterine septum was complete. Each cavity had a single ostia which had polyps around it. The cavity was same on either side but looked smaller than a single uterine cavity. On Chromopertubation the dye spilled out from each tube. Hence diagnosis of complete uterine septum with double cervix was made.

On discharge, the couple was counselled about the findings and risk of miscarriages, early preterm delivery, cervical incompetence and poor reproductive outcome with the uterine septum. They were offered resection of uterine septum. The couple agreed to discuss and revert to their decision.

The couple followed up after 3 months and confirmed consummation of their marriage as well as no problems in sexual intercourse. They decided on resection of uterine septum. To check thickness of septum MRI was advised. The MRI showed two uterine cavities separated by thick septum of 4–6 mm thickness Single fundus and two cervices were visualised. It was planned to do surgery in few weeks.

Discussion

The complexity of female reproductive organs is well established. The development involves Mesonephros, Wolffian and Mullerian ducts which form the reproductive

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organs. Abnormal fusion including absence of fusion can lead to variety of defects. It is believed that three processes play a role in formation of uterus and vagina embryologically. The fusion occurs laterally, vertically and then reabsorption helps. If these processes fail it leads to abnormalities. Either fusion doesn't happen, or canalization fails which leads to formation of uterus didelphyus or complete septum. The theories to explain the formation of woman's reproductive tract are many and American Fertility Society (AFS) made a good attempt to explain that development of these organs is in one direction from bottom to the top. However, if we see our case and assume that cervical duplication happened due to failure of fusion of Mullerian duct on two sides, it's not possible to have single, smooth fundus. This unique Mullerian anomaly was first described by McBean and Bumstead.

Therefore, alternative theories should be considered. One hypothesis by Musset et al. was that the ducts fuse and reabsorb at level of isthmus and proceed in north and south direction simultaneously.

AFS classification has not classified this anomaly but according to ESHRE 2013, we can classify it as U2bC1V2. It becomes easy to understand the anomaly this way.

Diagnosis is confirmed by combining diagnostic modalities like 3D ultrasound [1], MRI and hysterolaparoscopy.

Chang et al. also found 5 patients with exact similar anomaly. Their clinical checkup and radiological and surgical workup helped in diagnosis. The main complaint was dyspareunia and patient was diagnosed to have complete uterine septum with two cervix and vertical vaginal septum (Figs. 1, 2).

Such an anomaly can have outcomes like mid-trimester abortions, preterm labor, malpresentation and difficulty in conceiving. Vaginal septal resection is easy and should be done. However, whether to remove uterine septum or not has to decided individually.

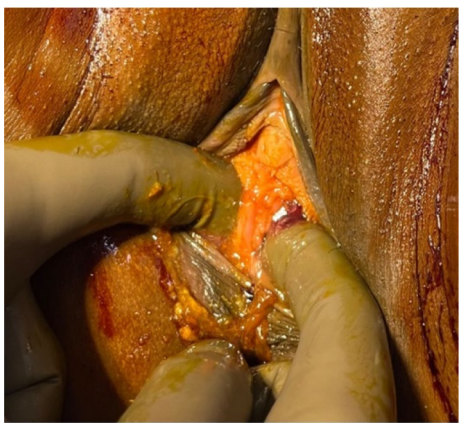

Fig. 1 Complete vaginal septum

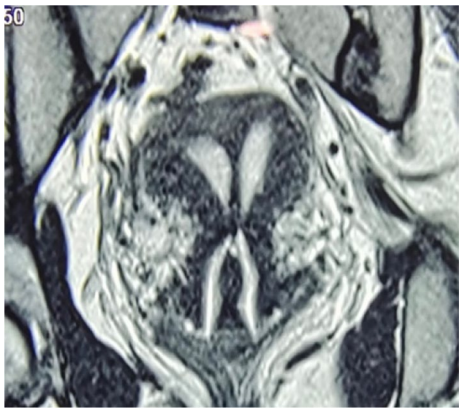

Fig. 2 MRI showing two uterine cavity with single fundus and complete uterine septum. Two cervices can be seen

Philip Patton et al. studied sixteen women presenting with either pregnancy loss in parous women or dyspareunia in nullipara. All women had the same anomaly that we are discussing. Preoperative pregnancy loss was 81%. They used both hysteroscopic and transabdominal route to remove the uterine septum. They found live birth rate of 75% post-surgery. Their conclusion was uterus didelphys was the most common misdiagnosis of this condition. The chances of miscarriage, obstetric complications and dyspareunia are commonest complications and hence it is advised to go ahead with surgery.

Alka Vijay [2] et al. recommend to go ahead with the hysteroscopic removal of the septum under guidance of transabdominal ultrasound. However, Arpita De et al. [3] were of the opinion to leave the uterine septum as it is and try for spontaneous pregnancy.

There are controversies around which treatment to choose. The approach which relieves the symptoms and preserves fertility has to be chosen. Removing Vaginal septum is simple and should be done. However, there are different opinions about whether to remove uterine septum or unify the cervices. Many people advocate hysteroscopic removal of the uterine septum to improve the outcomes of pregnancy [4]. Metal probes, plastic dilators or Foleys catheters have been used for hysteroscopic incision. Many don't advocate cervix unification since it can lead to cervical incompetence or bleeding during pregnancy. The chances of cervical dystocia also increase. Each couple has to counselled and tailor-made approach should be taken.

Conclusion

The correct diagnosis for such an anomaly is essential to plan for management. A combined approach of physical examination, 3D ultrasound, MRI and hysterolaproscopy

makes sure that there is no mistake in diagnosis. The commonest error is misdiagnosing it as uterus Didelphyus (as we had initially thought). Further management depends on symptoms. Vaginal septum resection is universally advocated. However uterine septal resection can be discussed with couple, the risk and benefits discussed and then individualized management should be done.

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Declarations

Conflict of interest The authors declare that there is no conflict of interest.

Ethical Approval All procedures performed on our patients were in accordance with the ethical standards of the institutional and /or national research committee.

Informed Consent The patient and her spouse have agreed for this publication.

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