



RESEARCH ARTICLE

AN ODDBALL EXAMPLE OF MULLERIAN DYSGENESIS: A RARE CASE REPORT

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ABSTRACT

Mullerian ducts are responsible for the formation of female reproductive system. Any deviation in the process leads to abnormal presentation. Proper examination, investigation and correct diagnosis of the mullerian anomaly leads to its correct management and good outcomes. In this case there was presence of vaginal and cervical septum with single uterine cavity and it was against the law of resorption of mullerian ducts. Hystero-lapqroscoy was done and after proper identification septum was resected.

INTRODUCTION

Mullerian ducts form the base of development of genitourinary system in female. In female due to absence of expression of SRY gene and anti-mullerian hormone, regression of wolffian duct occurs and differentiation of mullerian ducts continue. There are three stages of development: During first stage superior aspect of each mullerian duct develops into the left and right fallopian tubes, while the more caudal portions of each duct develop into left and right uterus, cervix and upper 2/3rd of vagina. The lower 1/3rd of vagina develops from the urogenital sinus and the ovaries develop from gonadal ridge independent of mullerian ducts. During the second stage there is midline fusion of the separate left and right uterus, cervix and upper vagina. During the third stage of development, there is resorption of midline fused segments which starts caudally and progresses cranially.¹

CASE REPORT

A 28 year old hindu female came to gynae OPD with complaint of spontaneous loss of pregnancy .She had two spontaneous abortions at period of 2 month and 3 month amenorrhoea respectively. On USG, uterus was retroverted with two separate uterine cornu in fundal region suggestive of bicornuate uterus or sub septate uterus.

After excluding all other factors of infertility, diagnostic hysteron-laparoscopy was performed. On per speculum examination there was a vertical septum extending from anterior vaginal to posterior vaginal wall and two separate cervical os and two vaginal canals were visualized and were healthy looking. On per vaginal examination,² cervical os felt, mid position, uterus retroverted, retroflexed normal size freely mobile and bilateral fornices free. On diagnostic laparoscopy uterus and bilateral adenexa were found normal in size, shape and appearance. Bilateral chromo-pertubation test was positive. Hysteroscope was introduced in one cervical os and dilator in other cervical os and both were found in same single uterine cavity. Uterine cavity was spacious, endometrium was healthy and bilateral ostia were visualized. So decision of vaginal septum resection was taken and done in usual way to reach cervical os. As single uterine cavity was already confirmed so cervical septum resection also done to create a single cervical os. Patient is advised to conceive after 6 months followed by elective cervical cerclage in next pregnancy.

DISCUSSION

Patient with mullerian dysgenesis are known to have higher incidence of infertility, repeated first trimester spontaneous abortions, fetal intrauterine growth retardation, fetal malposition, preterm labour and retained placenta.²



Picture showing vaginal septum being resected with cervical septum(double cervix appearance)



Picture showing single cervix after cervical septum resection

Mullerian duct anomalies are those structural anomalies which are caused by errors in mullerian duct development during embryonic morphogenesis. Factors that are responsible for such behavior are genetics, and maternal exposure to teratogens.^{3,4}

Malformations of the mullerian ducts in fetuses can result in exhibition of extragenital anomalies such as urological anomalies that include unilateral renal agenesis, horseshoe kidney or malformation of collecting ducts. The present case according to the European Society of Human Reproduction and Embryology (ESHRE) Classification belongs to U0C2V1 and is an exception of the unidirectional developmental theory⁵ which suggests that resorption of septum progresses caudally to cranially. A systemic clinical examination, 3D USG, MRI and diagnostic hystero laparoscopy combined allows correct evaluation of such patients. Surgical correction of the anomaly with close antepartum surveillance and choosing appropriate mode of delivery results in optimal maternal and fetal outcomes.

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