

Unicornuate Uterus with Non-Communicating Horn Diagnosis and Management

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Abstract

We present a case of unicornuate uterus with a rudimentary horn of non-communicating variety in a young unmarried girl. The case is described because of rarity and difficulties in the diagnosis and management of such congenital malformation.

Key Words: Mullerian duct anomalies (MDA), Rudimentary uterine horn, hematometra, miscarriage, unicornuate uterus. **Abbreviations:** USG (Ultra sonography) MRI (Magnetic Resonance Imaging) HSG (hysterosalpingography)

INTRODUCTION

Female genital organs develop from three origins; Ovaries from germ cells, uterus, fallopian tubes and upper 2/3 of vagina arises from Mullerian (paramesonephric) duct and lower 1/3 of vagina from sinovaginal bulb. Mullerian duct anomalies (MDAs) are embryological consequence of non development, nonfusion or non resorption of the mullerian ducts¹ resulting in absent uterus and vagina, unicornuate uterus, didelphys, bicornuate uterus, Septate, Arcuate and Diethylstilbestrol exposed T-shaped uterus.^{2,3} MDAs occur with an incidence estimated to be 4.3%⁴. Accurate diagnosis of the various subtypes is of great importance as MDAs are often associated with a vast variety of obstetric and gynecological problems⁵. A unicornuate uterus occurs approximately with a frequency of 1 in 4000 in healthy fertile population. It is a type of mullerian duct abnormality, which is smaller than a normal uterus and 65% of women have a rudimentary horn; which may or may not be connected with the rest of the uterus and vagina⁶. The diagnosis of unicornuate uterus is usually delayed, as it remains asymptomatic until adolescence and its initial symptoms are variable. It is often diagnosed after the appearance of gynecological complications, such as severe spasmodic dysmenorrhoea, haematometra, endometriosis, infertility, ectopic pregnancy, dyspareunia and recurrent miscarriage^{7,8}. Sometimes it is associated with other obstetric complications like pre term labor, intra uterine growth retardation, intra peritoneal hemorrhage and uterine rupture⁹. Various

imaging modalities have been used in the diagnosis and evaluation of MDAs, including hysterosalpingography, ultrasound, and magnetic Resonance imaging (MRI). Although HSG and ultrasound (USG) may suggest a Mullerian duct anomaly, further confirmation by MRI, hysteroscopy and laparoscopy is required¹⁰.

CASE REPORT

Miss K, D/O Mr. A of age 14 years unmarried student of 9th class presented with painful menstruation. Pain was of mild intensity initially, but increased on third and fourth day of menstrual cycle. It was spasmodic in nature and relieved with injectable analgesia. It was followed by continuous lower abdominal pain of moderate intensity. Patient could not perform her routine activities without analgesia. Her past medical history remained insignificant except that she had been taking injectable analgesia off and on for last 8 months. She was not allergic to any medicine. She belonged to lower middle class and her family history was unremarkable. Her G.P.E revealed moderately pale looking girl with pulse 88 beats per min., blood pressure 120 /70 mmHg, Temp 98° F, Respiratory rate 20/min. She was having no jaundice and edema. Accessible lymph nodes were not palpable, abdomen was soft non tender revealing no mass. Rectal examination elicited tenderness and mass on left side of pelvis the size of which could not be assessed due to tenderness. Baseline investigations Blood group B+ve, Hb 9.0 gm/dl. Rest of investigations were normal A

transabdominal ultrasound scan was done which disclosed a bicornuate uterus and absence of left kidney which was confirmed on IVU. Tumor marker screen was also within normal range. MRI revealed uterus Didelphus with haematometra and haematocolpos on left side while right side was normal. A provisional diagnosis of MDA was made. She was counseled and prepared for laparotomy which was performed under general anesthesia. During operative procedure, Unicornuate uterus with non-communicating left horn was recognized. Blind ended horn was found to be distended with chocolate colored fluid. While right horn was normal. It was also noted that patient had stage II endometriosis. The rudimentary horn was fused to the unicornuate uterus by a thick band of tissue. The blood supply was identified within this band. The left tube, ovarian ligament, and round ligament were clamped cut and transfixed. Similarly second and third pedicles were lamped, cut and transfixed. The rudimentary horn was removed without much difficulty. The endometriotic spots were ablated. Post operative recovery was uneventful and patient was discharged on fifth day. Patient was alright after six months of follow up.

Figure-1
Removal of Horn

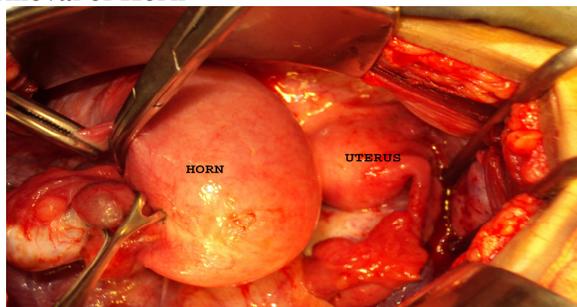


Figure-2
Removal of horn after separation of thick Fusion with unicornuate uterus

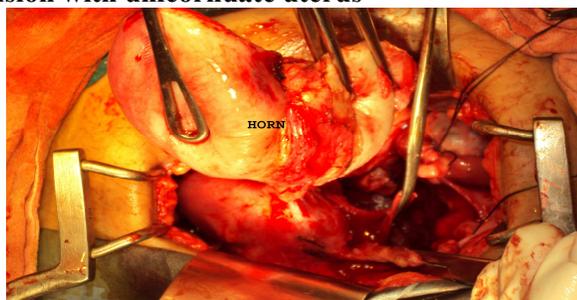


Figure-3
Complete removal of Horn

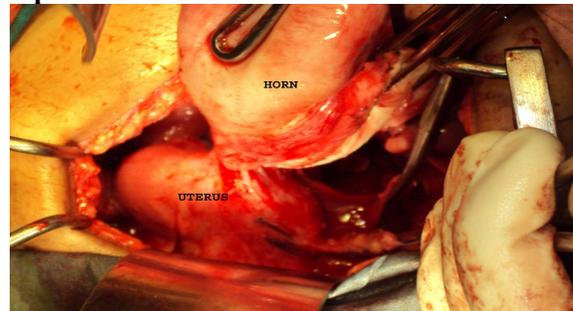
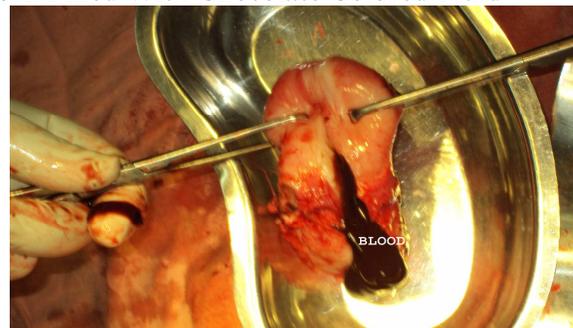


Figure-4
Horn Filled With Chocolate Colored Fluid



DISCUSSION

A unicornuate uterus with rudimentary horn occurs as a result of failure of complete development of one of the müllerian ducts and incomplete fusion with the contra lateral side. Unicornuate uterus may have either communicating or non-communicating rudimentary horn, absent rudimentary horn or a rudimentary horn with no cavity³. One of the horns which has functioning endometrium and non communicating variety leads to onset of cyclic pain and enlargement of that horn. More than a few functional noncommunicating horns present during or after the third decade of life with acute obstetric uterine rupture¹¹. Different obstetric presentations associated with unicornuate uteri are miscarriage in 37%, preterm birth in 16%, and term birth in only 45%. cases⁶. If pregnancy occurs in rudimentary horn, the most often outcome of pregnancy is rupture in second trimester in 90% of cases¹². Whenever the patient with suspicion of rudimentary horn is seen it is advisable to remove the rudimentary horn and its tube in order to avoid the high number of ectopic pregnancy and life threatening torrential intraperitoneal hemorrhage^{9,13}. Similarly it

was concluded by Nirmala Duhan et al¹⁴ that rudimentary uterine horns whether they occur in association with a patent uterovaginal tract or in isolation, should be excised. By removing horn further it may result in reduction or prevention of dysmenorrhoea and endometriosis¹⁵. Uterine anomalies can be suspected or screened on history, physical examination, hysterosalpingography and ultrasonographic evaluation. But diagnosis by these means is only in 14% of cases before these become symptomatic¹¹. In assessing a unicornuate uterus with HSG, blocked or non-communicating rudimentary horns will not be detected on film¹⁶. In this case it was not diagnosed accurately by USG and MRI. In experienced hands, both MRI and 3D-ultrasound are satisfactory investigations for MD. As the high sensitivity and specificity of 3D USG makes it a best means for identifying women with MDAs but studies that compare MRI with 3D USG for the diagnosis and classification of MDAs are deficient¹⁷. However MRI is helpful in diagnosis of MDA and proved to be 100 % in one study carried out on four patients prior to laparoscopy or laprotomy⁵. Additional diagnostic procedures are combined hysteroscopy and laparoscopy which are set aside for women in whom therapeutic intervention is expected to be undertaken¹⁸. Mesonephric duct giving rise to development of renal system is closely related to the reproductive tract and urinary tract abnormalities are frequently associated with MDA, most commonly a unicornuate uterus¹⁸. That is why we explored renal system and it was found that left kidney was absent in the above case.

In review of literature renal tract anomalies appeared to be 15-40%^{11,19,20} including ectopic kidney, renal agenesis, double renal pelvis, horseshoe kidneys and unilateral medullary sponge kidney¹⁹. MDA is also associated with skeletal anomalies like congenital fusion or absence of vertebra and it is seen in of 12-50% cases. An association of müllerian duct aplasia, renal aplasia and cervicothoracic somite dysplasia called MURCS has also been documented.²¹

In the mentioned patient no skeletal abnormality was noted. As unicornuate uterus with rudimentary horn is of grave consequences, a serious notice of symptomatology of patients must be taken. In our case patient was teenage and presented with severe dysmenorrhoea and pain lower abdomen. We suspected MDA and tried to confirm the subtype. MRI revealed it to be a uterus didelphys. Keeping in mind

the severity of clinical findings, counseling of patient was done. Laparotomy is preferred instead of laparoscopy, we confirmed diagnosis and treated anomaly successfully at the same time. Falcon et al²² described that there are two anatomical varieties which exist in regard to attachment of rudimentary horn with unicornuate uterus; with either thick connective tissue band or with complete fusion without any external marks of separation. Removal of horn in later case is not easy situation the same we faced in our case. Diagnosis and removal of horn by laparoscopy would be another option but to limited extent.

CONCLUSION

We should not over look symptoms like severe dysmenorrhoea in young girls and must have high index of suspicion for müllerian duct abnormalities especially unicornuate uterus with rudimentary horn so that surgery may be opted prior to any misfortune. Furthermore, urinary tract must always be explored for any abnormality.

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