ORIGINAL RESEARCH PAPER

Obstetrics and Gynaecology

# VARIANT OF HERLYN WERNER WUNDERLICH SYNDROME – A RARE MULLERIAN DUCT ANAMOLY

**KEY WORDS:** Mullerian Duct anomalies, Uterus Didelphys, Hematometra

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**ABSTRACT**

**Introduction-**The incidence of Mullerian duct anomalies is 0.5-5% , of which uterus didelphys/double uterus is rarest of all anomalies. Knowledge on them is important as they have great impact on women's quality of life.
**Case Report-**A 15 year old unmarried girl admitted in the Departement of OBGY , NRI hospital,guntur with pain abdomen of one month duration. Her cycles are regular with severe dysmenorrhoea. USG and MRI pelvis revealed Two uterine horns with two endometrial cavities, two cervical canals with obstructed left horn with hematometra of size 16.5*5.5cm with hematotrachelos, hematosalphinx with absent left kidney. Laparotomy was done and drianage of hematometra , excision of left sided cervix, uterine horn and the fallopian tube with fimbrial cyst was done. Postoperatively, patient was relieved of her symptoms.
**Discussion –** Mullerian duct anomalies occur due to abnormal embryological development of Mullerian duct. They often associated with renal anomalies. They present with amenorrhoea,severe dysmenorrhoea and palpable mass due to underlying hematotrachelometra. Association of uterine didelphys with renal agenesis and obstructed hemi vagina is called Herlyn-Werner-Wunderlich syndrome, and our case is variant of it.
**Conclusion –** Early and accurate diagnosis is vital because untreated cases may develop retrograde tubal reflux , endometriosis, impaired fertility and obstetric complications in later life.

## INTRODUCTION :

- Mullerian duct abnormalities occur due to failure of development , fusion, canalization or reabsorption of Mullerian ducts which normally occurs between 6 and 22 weeks in utero.[1,2]
- Uterine didelphys is the rarest of all and usually asymptomatic. If symptomatic ,present with dysmenorrhoea , mass per abdomen due hemato metrocolpos and associated with renal anamolies like renal agenesis, high riding aortic bifurcation, inferior vena cava duplication, malposition of ovaries.
- Association of uterine didelphys with renal agenesis and obstructed hemi vagina is called Herlyn-Werner-Wunderlich syndrome .

- **CASE REPORT**
- **:**A 15 year old unmarried girl admitted with pain abdomen of one month duration with mass in suprapubic region more towards left side.Attained menarche at 13yrs of age and had regular cycles with severe dysme norrhoea. Investigated outside where ultrasonography of abdomen and pelvis showed non-visualisation of left kidney with possibility of left horn of bicornuate uterus with hematometra and hematosalpinx or ?left ovarian cyst. In NRI hospital, she underwent MRI pelvis which revealed Two uterine horns with two endometrial cavities, two cervical canals with obstructed left horn with hematotrachelos, hematometra (16.5*5.5cm) , heamtosalpinx with absent left kidney.

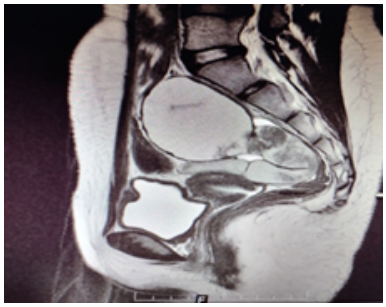
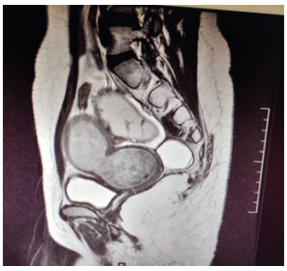


**Figure 1 : Normal sized right uterus with cervix and vagina**



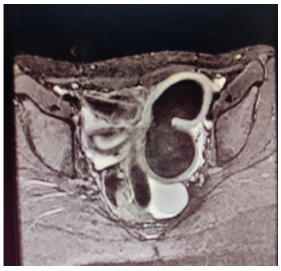


**Figure 2 & 3: Left hematotrachleos, hematometra and hematosalpinx**

- Case posted for Examination under Anesthesia where ,on abdominal mass of 18weeks size , on P/S: a single vagina and single cervix were seen. Uterine sound passed through cervical os and uterocervical length of 2.5 inches noted.
- P/V – mass of 18week size appreciated anteriorly in left fornix- cystic mass felt-propably left hematosalpinx or ovarian cyst, another small mass felt posteriorly and right to 18 week mass probably uterus.
- Case, then proceeded to laparotomy ,Intraoperatively we found a big cystic and smooth surfaced mass (18 week size) on opening the peritoneal cavity. A long coiled and distended structure attached to this mass, on lifting it revealed small uterus on the right side along with right fallopian tube and right ovary.
- Then, 18week mass traced down which is left uterus, cervix, fallopian tube is attached to it and was distended .
- Left hematosalpinx of size 15*3 cm noted and fimbrial cyst of size 10*7 cm noted which is adherent to bowel and omentum all over its surface and same released.
- After confirming that there was no communication to the vagina from this swelling, we gave small stab incision over the lower part of swelling and clotted blood drained. Lowest portion of the mass was attached to the right sided uterine cervix. Then releasing the adhesions ,we were able to remove entire length of left-sided uterus with cervix and attached fallopian tube . Left side ovary was normal.

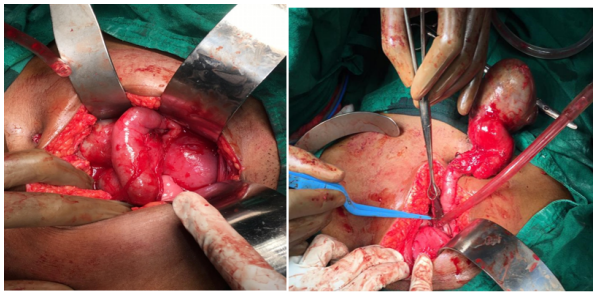


**Fig: left side uterus with left hematosalphnix with paratubal cyst.**

- Excision of left cervix, uterinehorn, fallopian tube and fimbrial cyst done(left hemi hysterectomy).
- Right and Left ovaries appeared normal and retained.
- Right cervix, uterine horn, fallopian tube appeared normal and retained .
- Hemostasis secured well. Patient was relieved of symptoms postoperatively.

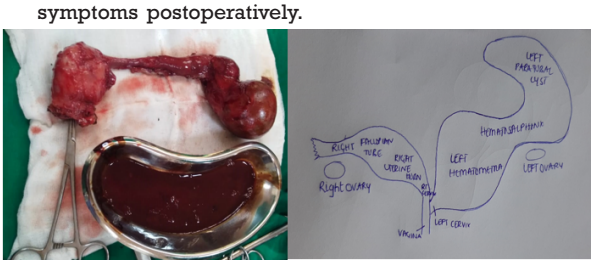



**Fig: Excised left-sided cervix ,uterus ,hematosalpinx para-tubal cyst with drained hematometra in the kidney tray.**

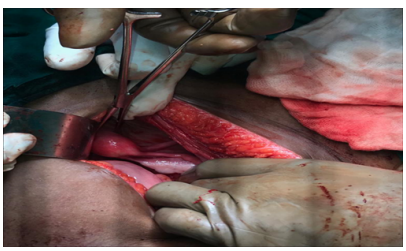
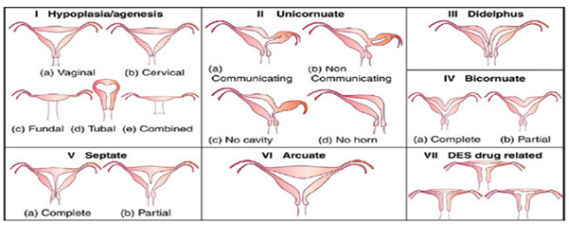


**Fig: Right-sided normal uterus,fallopian tube and ovary.**

- Histopathology report: endocervix – chronic non-specific endocervicitis with nabothian cysts ,proliferative endometrium , normal myometrium, with hematosalpinx with mild salphingitis. Left fimbrial cyst showed infected para-tubal cyst with an organized thrombus in the lumen ,eosinophilic proteinaceous material and cholesterol clefts.

## DISCUSSION :

Both the fallopian tubes,uterus ,cervix and upper 2/3rd of vagina develops from Mullerian duct and lower 1/3rd of vagina develop from urogenital sinus. In our case each mullerian duct developed into fallopian tube,uterus,cervix separately. But the vagina developed and communicated with right side cervix..Incidence is believed to be between 0.5 and 5.0%.[3,4] Mullerian duct anomalies [MDA] are divided into six groups on the basis of the Buttram and Gibbons system.[5]




American Fertility Society (AFS) Classification of Müllerian Anomalies (1988) :

- Class I: Müllerian agenesis/Hypoplasia—segmental,
- Class II: Unicornuate uterus.
- Class III: Didelphys uterus.
- Class IV: Bicornuate uterus.
- Class V: Septate uterus
- Class VI: Arcuate uterus,
- Class VII: Diethylstilbestrol (DES)-related abnormality.

## CONCLUSION:

Even though patient attained menarche with regular menstrual cycles , presenting with mass per abdomen and dysmenorrhoea ,Mullerian duct anomalies are still possible. Early & accurate diagnosis is vital as untreated cases may develop retrograde tubal reflux ,endometriosis, impaired fertility and obstetric complications in later life.

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