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An interesting case report on Herlyn-Werner-Wunderlich syndrome with hematoclopos and communicating blind duplex ureters

Preeti Lewis *, Ashish Zarariya, R. G. Daver, Shraddha Choudhary and Karan Kale
Dept. of OBGY, Grant Govt Medical College, Mumbai, India
*Corresponding author: Preeti Lewis

ABSTRACT

Herlyn-Werner-Wunderlich syndrome represents a complex female genital malformation with uterus didelphys, unilateral low vaginal obstruction, and ipsilateral renal agenesis, all being secondary to mesonephric duct-induced müllerian anomalies. Clinically, this often presents in postpubertal adolescent or adult women where hematometrocolpos produces a more pronounced mass effect and pain on the side of the obstructed hemivagina. Mullerian duct anomalies have an incidence of 2-3%, while **OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL ANOMALY [OHVIRA]** has an incidence of **0.1-3.5%** of all such anomalies.[1,2] We hereby report a similar case which had in addition, duplication of the ipsilateral ureter with ectopic opening in the urethra and communication with hematocolpos making it even more rare.

Keywords: hematocolpos, hemivagina, didelphic uterus,Obstucted hemivagina and ipsilateral renal agenisis [OHVIRA]

INTRODUCTION

A spectrum of uterine fusion anomalies can occur during fetal development. Lateral fusion defects are the most common type of mullerian duct anomalies which can range from symmetric or asymmetric to obstructed or non-obstructed fusion anomalies.

Mullerian duct anomalies have an incidence of **2-3%**, while **OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL ANOMALY [OHVIRA]** has an incidence of **0.1-3.5%** of all such anomalies.[1,2]

OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL ANOMALY, also known as Herlyn –Werner – Wunderlich Syndrome, is a **rare syndrome with only a few case** reported. Herlyn-Werner-Wunderlich syndrome comprises of a complex female genital malformation with uterus didelphys, unilateral low vaginal obstruction, and ipsilateral renal agenesis, secondary to mesonephric duct-induced mullerian anomalies.

Symptoms usually present, usually after menarche, when haematocolpos develops during menstruation, resulting in dysmenorrhoea and a pelvic mass. Ultrasonography, typically demonstrates a pelvic fluid collection, which can simulate other disease process such as –Cystic ovarian malignancy, Pelvic abscess, Ovarian cyst, Pyosalpinx, Hydrosalpinx; thus confounding the diagnosis. MRI is confirmatory.

We hereby report a similar rare case which had in addition, **duplication of the ipsilateral ureter with ectopic opening in the hemivagina and urethra .**

CASE REPORT

12 year old, unmarried girl came with severe cyclical pain in abdomen right sided spasmodic during menses since 6 months with menarche 1.5 yrs back. She was apparently alright for one year, after which she started developing cyclical right sided dysmenorrhoea.

She had no bowel or bladder symptoms.

Her previous menstrual cycles were regular of moderate flow but painful since 5 months which was relieved by medication and on walking.

Her past medical or surgical and family history was insignificant.

***On examination*:** she was short statured , height was **136.5cm** with **Polydactyly** with well developed secondary sexual characters. Vitals were stable

Per Abdomen no palpable mass felt.

On local Examination, hymen was intact.

On per rectal examination, hematocolpos present on the right side.

ULTRASONOGRAPHY : It revealed uterus didelphys, left horn of uterus measure 6.1x2.7x1.7cm, and endometrial thickness of left horn was 5mm.Right horn of uterus measures 6.5x3x2.7 cm. Small fluid noted in endometrial cavity. Right fallopian tube is dilated tortuous and filled with internal echoes, measuring 13x 2.2 cm . A large fluid collection with dense internal echoes measuring approximately 9.4x4.0 cm was noted in right vagina.The restricting membrane (transverse vaginal septum/ hymen) was 7.0 mms in thickness. Bilateral ovaries normal. Right kidney is not visualized. Left kidney hypertrophied. **Impression** was- uterus didelphys with hematocolpos, hematometra, and hematosalpinx on right side with right renal agenesis.

MRI: It showed, two separate endometrial cavities with two separate cervices and two vaginal cavities suggestive of uterine didelphys. The endometrial thickness was 6mm on both sides. Hyper intense collection measuring 10x5x4.5 cm distending right hemivagina and cervix with an obstructing low transverse vaginal septum measuring 3mm in thickness suggestive of hematocolpos [Figure 1,2]. Left hemivagina not dilated. A large tubular tortuous structure (max diameter=1.8cm) containing reflux blood products was seen communicating with lateral wall of obstructed right hemivagina and extending into the right half of pelvis in right adenexa, right para rectal and paravesical spaces. This represented an ectopic dilated remnant of right ureter. Another linear blind ending tubular structure (max diameter=0.4 cm) containing reflux blood products was seen, opening ectopically into the urethra, where it was obstructed. It showed a long ectopic intramural course in posterior wall of bladder with localized dilatation. Just proximal to its opening into urethra it was seen to communicate with ? right ureteric remnant. It extended proximally in the course of ureter into lower abdomen antero-medial to left iliac vessels most likely representing an ectopic remnant of a duplex right ureter. Right kidney was not visualized suggestive of congenital agenesis . Compensatory hypertrophy of left kidney noted. Final diagnosis of **uterine didelphys with obstructed right hemivagina, ipsilateral renal agenesi-s (OHVIRA syndrome) and possible remnant duplex right ureter.**

Cystoscopy s/o Non-visualized Right Ureter[Figure 3].

Congenital heart disease was ruled out by **2-D Echo**.

Vaginal drainage of haematocolpos was done under spinal anaesthesia. The transverse septum of approximately 4-5 mm, was incised and marsupalization of margins were done[Figure 4,5]. Patient referred to urologist for further management.

DISCUSSION

Retrospective studies of similar cases have shown that the mean age of presentation is about 15 years.[3]

Pelvic pain is the most common presenting symptom (90%) followed by abdominal mass (40%). Patients can present at a later age with foul smelling vaginal discharge due to pyocolpos.[4]

Some present with pelvic pain, worsening dysmenorrhoea and pelvic mass.[5]

Some occasionally present with urgency, frequency or vaginal discharge. [6]

A didelphic uterus is due to an embryologic arrest occurring during the 8th week of gestation, which ultimately affects mullerian and metanephric duct.[7] If one of the wollfian duct is absent, the kidney and the ureter on ipsilateral side will fail to fuse in midline. This process may occur completely or incompletely. If failure to fuse is complete then a uterus didelphys is formed. Mullerian duct on the side, lacking the wollfian duct displaces itself laterally and cannot come into direct contact with the urogenital sinus in the centre, resulting in the blind sac, imperforate or obstructed vagina. However the distal part of the vagina , developing from the urogenital sinus is not affected.[7]

MRI is helpful in characterizing the didelphic uterus and vaginal septum for pre-operative planning to prevent complication such as endometriosis or adhesion from chronic infection with subsequent infertility.

In a case review series of 15 patients, those patients who wanted pregnancy following vaginoplasty and/or hemi hysterectomy demonstrated successful pregnancy rates similar to females with didelphic uteri.[8]

Cetinkya et al. have discussed the use of hysteroscopy to excise the vaginal septum and preserve hymenal integrity.[9]

Those cases where an ectopic ureter inserts into the obstructed hemivagina, laparoscopic surgical ligation or surgical removal is advocated to prevent recurrent fluid accumulation in the obstructed hemivagina.[6] It could be argued that surgical excision might be better as there have been reported cases of malignant transformation occurring in the ectopic ureter.[6]

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FIGURES:

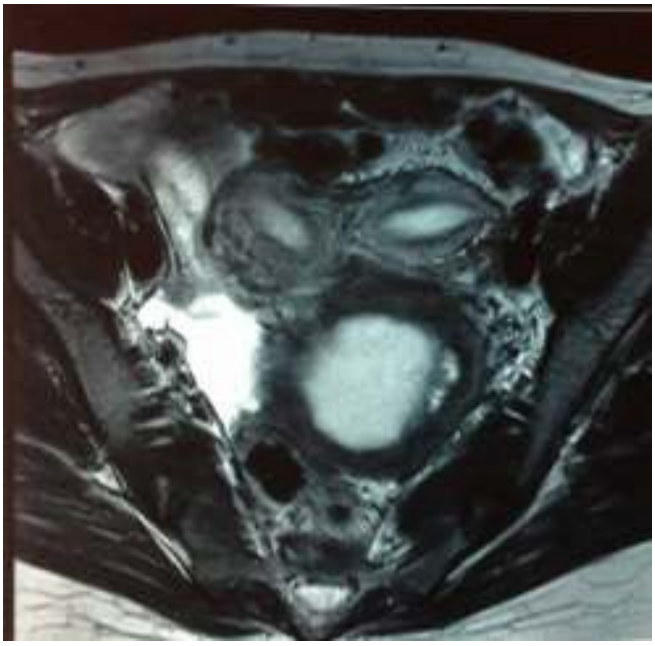

Figure 1: Magnetic Resonance Imaging showing didelphic uterus with hematocolpos

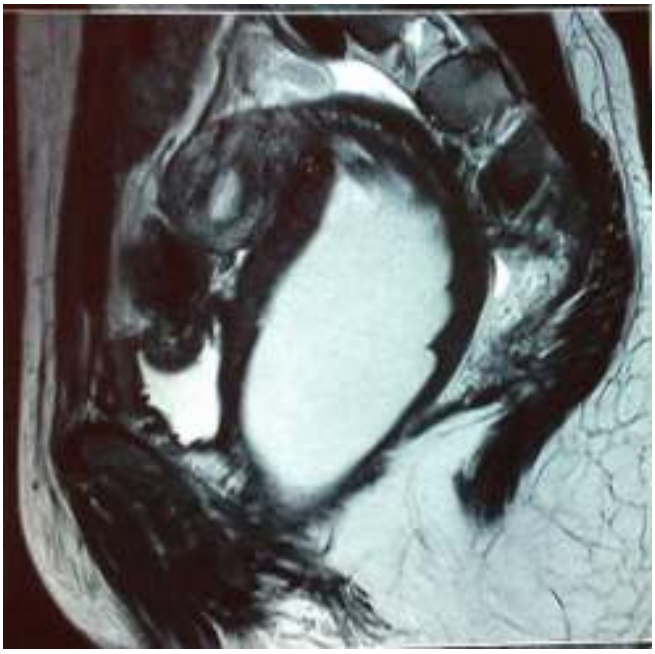

Figure 2: Sagittal veiw of hematocolpos on Magnetic Resonance Imaging

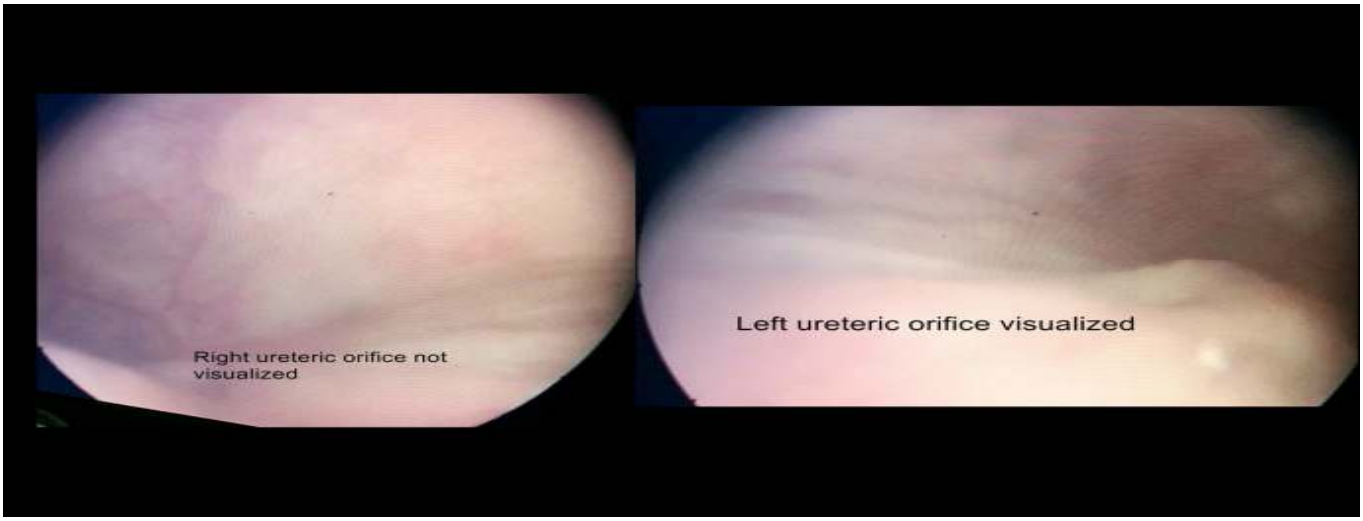

Figure 3: Cystoscopy. showing right ureteric orifice

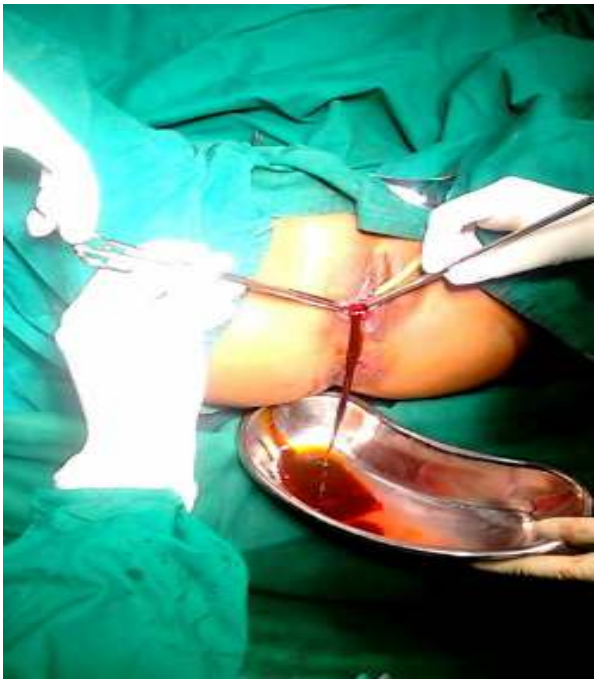

Figure 4: Drainage of hematocolpos of blind hemivagina

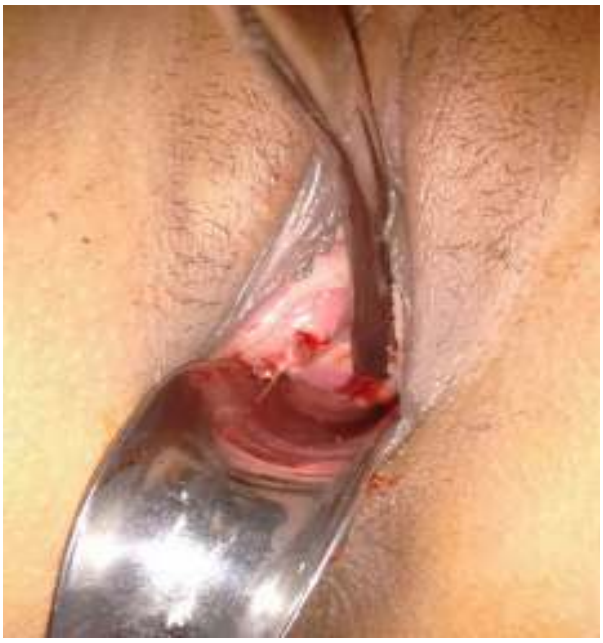

Figure 5: Post operative image