

CASE REPORT OF HERLYN-WERNER-WUNDERLICH (OHVIRA) SYNDROME: MRI FINDINGS AND RADIOLOGICAL GUIDE

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ABSTRACT

Herlyn-Werner-Wunderlich (HWW) syndrome, a rare Müllerian duct anomaly (MDA) consists of didelphic uterus (MDA III), hemivaginal septum and ipsilateral renal agenesis (Mesonephric anomaly). This is given the name of OHVIRA syndrome comprising of obstructed hemivagina and ipsilateral renal anomaly. Patients are usually diagnosed at puberty after menarche. Diagnosis is made solely on the basis of suspicion due to nonspecific symptoms and also because of seldom encounter of Mullerian duct anomalies (MDA) clinically. If the history, examination and imaging findings correlate, existence of this syndrome is raised. The role of imaging is to help detect, diagnose and distinguish surgically correctable forms of Müllerian duct anomalies from inoperable forms. We report a case of a 16-year old girl with this condition who was diagnosed as uterus didelphys with unilateral hematocolpos and ipsilateral renal agenesis on the basis of sonography and confirmed by MRI. MRI proved to be of great help in correct diagnosis avoiding surgical interventions.

Patient was treated with drainage of hematocolpos and single stage vaginoplasty and followed up subsequently.

Case Report

A 16-years old girl presented with severe abdominal pain; she also had irregular menstruation with severe dysmenorrhea and she reported that since menarche. The pain worsened immediately prior to menstruation and improved with menstrual flow. Her physical examination was unremarkable except tenderness in lower abdomen. Gynaecological examination did not reveal any abnormality of external genitalia.

Ultrasound of abdomen and pelvis showed two divergent uterine horns with distended cavity and wide opened cervix. Right kidney was not seen in right flank. On IVP it was confirmed as agenesis of right kidney (Fig. 1 & 2).

MRI was advised which revealed a uterine-vaginal malformation consisting of didelphys uterus with double cervix, of which the right vagina was obstructed. A large fluid collection seen in vagina with wide opened right cervix. Right uterine cavity was



Figure 1: U/S imaging shows didelphy uterus with two divergent uterine horns

also distended. The fluid characteristics were suggestive of methemoglobin. All these findings were consistent with didelphys uterus with obstructed right hemivagina resulting in hematocolpos (Fig. 3 & 4).

Both ovaries were normal in size, shape and echotexture. Included images of the abdomen demonstrate agenesis of right kidney in abdomen and pelvis with compensatory hypertrophy of left kidney.

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Figure 2: Right renal agenesis on IVP examination

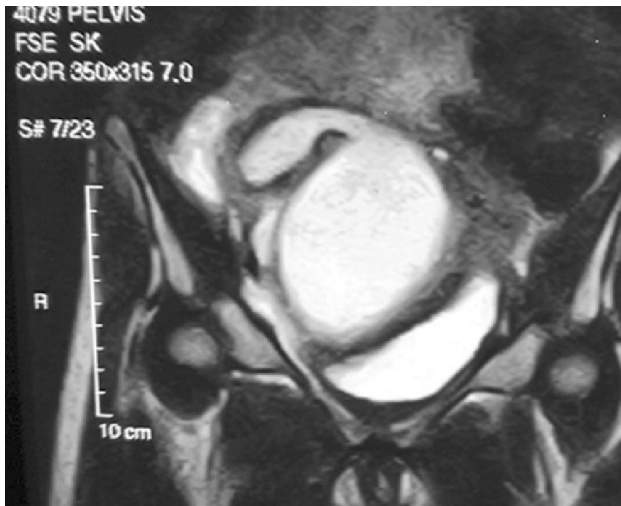


Figure 3: Coronal MRI T2 image shows didelphys uterus with double cervix and obstructed right vagina. High T2 signal intensity fluid seen in right hemivagina extending into right cervix and right endometrial cavity suggestive of hematocolpos. The fluid is displacing bladder inferiorly and left uterine cavity has been pushed posteriorly

Patient was treated with drainage of hematocolpos and single stage vaginoplasty and followed up subsequently.

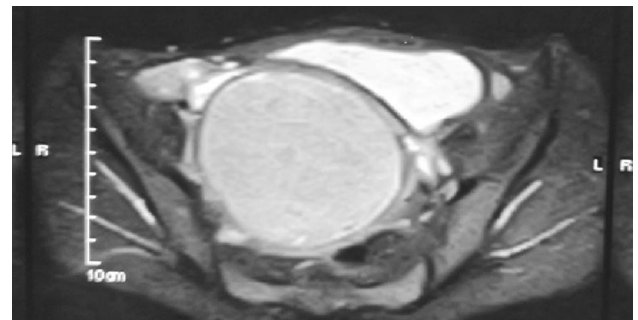


Figure 4: Axial MRI T2 image shows collection in right vagina extending into right cervix

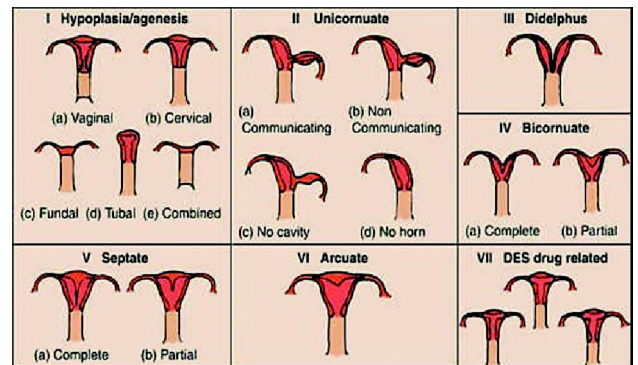


Figure 5: represents diagrammatic representation of types of Mullerian duct anomalies¹⁵

Discussion

Close relationship between the urinary and reproductive system during embryogenesis explains the coexistence of urinary tract and reproductive abnormalities. HWW syndrome is a rare malformation involving both Mullerian (female genital tract) and Wolffian (urinary) ducts. The arrest occurs at 8th week of gestation affecting both mullerian and metanephric ducts.

Embryology: Transformation of müllerian ducts into the segments of the female reproductive tract requires completion of 3 phases of development as follows:

- **Organogenesis:** Failure of this phase results in abnormalities like uterine agenesis or hypoplasia (bilateral) or unicornuate uterus (unilateral).
- **Fusion:** There are two types, lateral and vertical. Failure of lateral fusion (lower segments of paired mullerian ducts fuse to form uterus, cervix and upper vagina) results in anomalies such as bicornuate or didelphys uterus.

Incomplete vertical fusion (lower one third vagina fuses with upper two thirds) results in an imperforate hymen.

- Septal resorption: Nonresorption is the cause of septate uterus.

Müllerian duct anomalies are categorized most commonly into 7 classes according to the American Fertility Society (AFS) Classification Scheme^{1,2} as follows:

Class I: (hypoplasia/agenesis) includes uterine/cervical agenesis or hypoplasia. The most common form is the Mayer-Rokitansky-Kuster-Hauser syndrome, which is combined agenesis of the uterus, cervix, and upper portion of the vagina.^{3,4,5}

Class II: (unicornuate uterus) is the result of complete, or almost complete, arrest of development of müllerian duct with communicating or non-communicating horns containing endometrium or no endometrial cavity in contralateral horn. Incidence is 6-25%.

Class III: (didelphys uterus) results from complete nonfusion of both müllerian ducts with incidence of 5-11%.

Class IV : (bicornuate uterus) results from partial or complete nonfusion of the müllerian ducts upto or not extending to internal os. It is the next commonest type having incidence of 10-39%.

Class V: (septate uterus) is complete or incomplete division of uterus extending to the internal cervical os. It is the commonest anomaly, incidence of 34-55%

Class VI: (arcuate uterus) has a single uterine cavity with a convex or flat uterine fundus or may demonstrate a small fundal cleft or impression (>1.5 cm). It has an incidence of 7%.

Class VII: (T-shaped uterus) is seen in women treated with diethylstilbestrol (DES), an estrogen analogue prescribed to prevent miscarriage.

Herlyn-Werner-Wunderlich (HWW) syndrome is cha-

racterized by a triad of type III Müllerian duct anomaly, obstructed hemivagina and mesonephric duct anomalies. Most often the latter manifests as renal agenesis.⁶ (Fig. 5) represents the diagrammatic representation of types of Müllerian duct anomalies and schematic diagram shows that HWW is a type III anomaly.

Irrespective of imaging modality, the typical findings are of:

- duplication of the uterus, cervix and vagina
- unilateral haematocolpos or haematometocolpos
- absent kidney on the same side as the uterovaginal obstruction

Patients present with recurrent severe dysmenorrhea, chronic pelvic pain, excessive foul smelling mucopurulent discharge, spotting and intermenstrual bleeding. A palpable abdominal, pelvic or vaginal mass (mucocolpos or pyocolpos) may also be present.

HWW syndrome is an uncommon but often treatable cause of infertility. Patients with müllerian such anomalies are known to have a higher incidence of impaired fertility and obstetric complications (repeated first-trimester spontaneous abortions, fetal intrauterine growth retardation, fetal malposition, preterm labor, and retained placenta) later in life.

Based on evidence from the patient history and physical examination, additional imaging workup is required. Imaging techniques involve mainly ultrasound, hysterosalpingography, intravenous urography, CT and MRI. Laparoscopy is the gold standard for the evaluation of female reproductive tract anomaly. In some lesions, the surgical approach is altered based on imaging findings.^{7,8}

Ultrasound: unilateral renal agenesis is diagnosed prenatally. Female fetuses and neonates should be screened for genital malformations. Uterine anomaly is better visualized in neonatal period when uterus is still under maternal hormonal stimulation with characteristic myometrium and echogenic endometrium. However, follow-up ultrasound scanning of these asymptomatic patients is advised till puberty. First examination ordered is pelvic ultrasound. Abdominal and pelvic imaging at puberty allows corr-

ect diagnosis by showing uterovaginal duplication, hematocolpos or hematometocolpos and ipsilateral renal agenesis. Presence of hematocolpos causes difficulty in visualizing vaginal septum rendering it inconclusive for differentiating between it and other associated anomalies.⁹ In spite being the first imaging modality chosen it has limitations as it is operator dependent. Overlying bowel gas can confound transabdominal imaging. Transvaginal imaging, although superior to the transabdominal approach, may not always be possible, as in patients with vaginal septa. Image resolution is a limiting factor.

Hysterosalpingography (HSG): Performed under fluoroscopy, allows evaluation of the uterine cavity and tubal patency. Anomalies may be suggested but positive findings often are nonspecific for precise diagnosis as overlap may exist with subsequent Mullerian duct anomalies.¹⁰

Intravenous urography confirms renal agenesis and replaced by pelvic MRI.¹¹

Computed tomography though gives information about congenital anomalies, it is not routinely performed due to the use of ionizing radiation.

MRI: Is the criterion standard for imaging uterine anomalies due to high contrast resolution images of the uterine anatomical structure. In addition, it can help evaluate the urinary tract for concomitant anomalies and replace intravenous urography. Most types of uterine anomalies can be diagnosed confidently using pelvic MRI.¹²

Two separate normal-sized uteri with preserved endometrial and myometrial width and two cervixes are seen. Obstructed vagina is best identified in between uterine cavities. MRI thus has an added advantage of better visualization of pelvis showing normal ovaries.

MRI has consistently demonstrated high sensitivity and specificity for evaluation of uterine anomalies. For patients requiring surgery, MRI demonstrated 100% sensitivity and specificity, compared to 67% sensitivity and 100% specificity for EVS. For nonsurgical lesions, both MRI and EVS had 100% sensitivity and specificity. Data suggests very low false-negative and false-positive rates.¹³

Diagnostic laparoscopy: Is an invasive and expensive diagnostic method compared to MRI. Though the

gold standard, it could be reserved when the diagnosis is not clear after imaging or when MRI is not available and not performed as a routine procedure.

However, laparoscopy can be also therapeutic in some selected cases such as: drainage of hematocolpos/hematometocolpos, septectomy, or marsupialization of the blind hemivagina. It is considered very helpful in order to reduce pelvic pain and lower the risk of infection and of further hematometra.

Vaginal septectomy with marsupialization and drainage of hematocolpos/hematometocolpos to provide relief of pain and further complications is a preferred surgical approach with long term outcome. Salpingectomy for pyosalpinx is done if needed. Hemihysterectomy with or without salpingectomy is rarely indicated. Subsequent successful pregnancy in the obstructed uterus has been reported.¹⁴

If surgery is not an immediate option for patients with HWW syndrome, menstrual suppression with combined oral contraceptive pills is advised to prevent further accumulation of hematocolpos and further hematometra.

Conclusion

Genetic counseling, possible sex limited autosomal dominant inheritance, with no recurrence in female siblings unless mother is affected but a 50% risk for female offspring is predicted. Prenatal diagnosis should be aimed. Correct diagnosis though difficult due to absence of specific findings on examination and nonspecific symptoms is the key to good outcome of these patients.

Diagnosis requires lower abdominal MRI extending to upper abdomen to check for ipsilateral kidney. Prompt treatment is then necessary to relieve acute symptoms, preserve normal fertility and prevent several medical complications.

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